# Pediatric Anesthesiology Review

## Clinical Cases for Self-Assessment

Robert S. Holzman
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*Third Edition*

Springer

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# Preface

This text is designed for those who would become consultants in pediatric anesthesia. It is based on a curriculum developed since 1992 in our department to illustrate the breadth and depth of the practice of pediatric anesthesia. Weekly meetings are held with our fellows and many of our faculty who are or who have been associate examiners of the American Board of Anesthesiology. The program is an integral part of the didactic series in the Department of Anesthesiology, Perioperative and Pain Medicine at Boston Children's Hospital.

An ability to explain *why* various data are required before or during the care of a patient or *why* a certain anesthesia care plan was chosen was critical to us in our philosophy of the course, and we have tried to preserve that ideal during the crafting of this text. Although the interactive aspect of a dialog between examiner and examinee cannot be effectively recreated through a textbook, the reader is encouraged—strongly so—to use this book in creative ways to try to mimic the spontaneity achievable through conversation. First of all, a "buddy" system is advisable. Secondly, a small handheld recorder is extremely useful when using the questions as prompts; the contemplative reader will listen critically to the responses he or she has offered and then hopefully improve as the recording continues. Using materiality as the best endpoint for adequate answers, the discerning reader should attempt to answer the question to the satisfaction of an imaginary partner—whether a parent, a surgeon, a pediatrician, or another anesthesiology colleague calling for help. With practice and introspection, it is amazing how similar, rather than different, the answers are to these diverse audiences.

This third edition has the same purpose as our previous endeavors—to accompany the reader's journey in attaining proficiency, expertise, and, finally, mastery in the consultative practice of pediatric anesthesiology. The formatting of the book is designed to encourage the reader's free flow of ideas. One should begin with looking at both facing pages, then progress to covering the answers on the right side, and eventually cover the questions on the left. In this very simple, programmed text manner, practice at generating the appropriate breadth and depth of answers, and then questions, can be encouraged.

The written examinations, seen at the beginning of the text as a baseline in pediatric medicine, are primarily knowledge-based, reflecting factual medical information necessary for the subspecialty practice of pediatric anesthesiology.

With this basic guidance, the reader is encouraged to be creative throughout this book, to use imagination and a fund of knowledge in bringing yourself "into the operating room" and managing the patient in an expert fashion—one that would, in the eyes of peers as well as patients and their families, merit the awarding of "consultant in pediatric anesthesiology."

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# Part I
# Pediatric Medicine for Pediatric Anesthesiologists

# Chapter 1
# General Pediatrics


**Thomas J. Mancuso**



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## Questions

1. Which of the following are considered risk factors for the development of tuberculosis (TB) in children?
   1. HIV infection
   2. Exposure to an infectious adult
   3. Malnutrition
   4. Passive exposure to cigarette smoke
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

2. Tuberculosis infection may involve which of the following organs/systems?
   1. The lungs
   2. The bones
   3. The CNS
   4. The kidneys
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

3. What percentage of immunocompetent adults infected with tuberculosis will develop active disease during their lives?
   A. 100%
   B. 50%
   C. 25%
   D. 5%
   E. 2%

4. Which of the following factors may affect the accuracy of the Mantoux test (the intradermal injection of 5 TU of PPD in 0.1 ml diluent)?
   1. Concurrent penicillin treatment
   2. The presence of other infections
   3. Presence of fever >38 °C
   4. Prior BCG vaccination
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

## Answers

1. A. 1, 2, 3

TB in the USA is unfortunately becoming more common in adults as well as in children. It is an important cause of mortality worldwide. In the USA, infection of children often results from exposure to an untreated individual with active disease. Reservoirs of TB include people with HIV/AIDS, the homeless, patients living in overcrowded conditions, and new immigrants.

2. E. All of the above

TB infection can involve most organ systems. It most commonly affects the lungs. Superficial lymph node infection is a common manifestation of TB infection. The major cause of death in children from TB is meningitis. Cerebrospinal fluid (CSF) findings in TB meningitis include a predominance of lymphocytes in low numbers (50–500 cells/μl), low glucose, and elevated protein. The TB organism is seen in less than 50 % of cases, and CSF cultures become positive only after several weeks. Miliary tuberculosis, so called because the small lesions found throughout the body resemble millet seeds, is due to blood-borne spread of the organism.

3. D. 5%

Predisposing factors for the development of serious disease in patients infected with TB include young age, pregnancy, and decreased vigor of the immune response (HIV/AIDS, poor nutrition, steroid treatment).

4. E. All of the above

Patients previously immunized by bacille Calmette-Guérin (BCG) will show a positive PPD. The BCG vaccine, derived from a mycobacterium related to TB, activates cell-mediated immunity. Since the many vaccines derived from strains of the bacterium differ from one another in antigenicity, the immune response to the vaccines is quite variable.

5. Group A beta-hemolytic streptococci cause:
   1. Scarlet fever tonsillitis
   2. Impetigo
   3. Erysipelas
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

6. Which one of the following is true regarding the nonsuppurative complications of group A beta-hemolytic infections?
   A. Rheumatic fever may develop after tonsillitis.
   B. Neither nephritis nor rheumatic fever develops after impetigo.
   C. Rheumatic fever is caused by the same strains of the organism as nephritis.
   D. Nephritis develops only after scarlet fever rashes.

7. *Helicobacter pylori* (*H. pylori*):
   1. Has been cultured from children with hypertrophic pyloric stenosis
   2. Has been implicated as a cause of chronic abdominal pain in children
   3. Generally causes watery, but not bloody, diarrhea
   4. Is considered a contributing factor in the pathogenesis of peptic ulcer
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

8. Colitis due to infection with toxigenic *Clostridium difficile*:
   1. Is due to overgrowth of the organism after antibiotic therapy
   2. Is characterized by watery, often bloody, diarrhea
   3. Is due to the toxins produced by *C. difficile*
   4. Can also be caused by ingestion of preformed toxin found in poorly refrigerated food
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

5. E. All of the above

This gram-positive organism, also called *S. pyogenes*, can be divided into over 60 subtypes based on surface protein, such as the M proteins. Impetigo is most common in younger children and tonsillitis/pharyngitis in school-aged children.

6. A. Rheumatic fever may develop after tonsillitis.

Many serologic types of group A *Streptococcus* infecting the throat can be associated with rheumatic fever. Nephritis, in contrast, is related to a limited number of types and may occur following skin infections, while rheumatic fever only follows pharyngitis.

7. C. 2, 4

Infection with this bacterium is associated with ulcer disease, acute gastritis, and chronic abdominal pain. *H. pylori* is responsible for at least 50 % of duodenal and gastric ulcers in adults, but it is the cause of a lower percentage of ulcers in children. In some patients with chronic abdominal pain, eradication of *H. pylori* has been associated with diminution of the pain.

8. A. 1, 2, 3

Antibiotic-associated diarrhea is due to toxins produced by *C. difficile*. Overgrowth of the bacteria occurs when antibiotic treatment suppresses normal flora in the GI tract. Symptoms continue for 7–10 days after stopping the antibiotic therapy. In more severe cases, IV and/or enteral vancomycin therapy may be needed. Food poisoning is caused by ingestion of *C. perfringens* capable of forming spores. Botulism is a form of food poisoning caused by ingestion of the neurotoxin made by *C. botulinum*.

9. Regarding the clinical manifestations of bacterial meningitis beyond the neonatal period:
   1. Focal neurologic signs are seen in 10–15% of cases.
   2. If seizures occur, it is very likely that the child will be left with a permanent seizure disorder.
   3. Fever need not be present.
   4. Photophobia, due to inflammation of the optic nerve, may lead to permanently impaired vision.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

10. Regarding the prognosis of bacterial meningitis beyond the neonatal period:
    1. Some degree of hearing loss is seen in approximately 10% of survivors.
    2. Neurologic abnormalities seen shortly after the onset of meningitis may resolve over time.
    3. The mortality rate is 1–5%.
    4. Brain abscesses are commonly seen during the course of antibiotic therapy.
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

11. The hemolytic uremic syndrome:
    1. Typically has a prodrome of 3–5 days of diarrhea
    2. May include neurologic dysfunction such as seizures or coma in its presentation
    3. May include hypertension as part of its presentation
    4. Generally is treated with supportive care (careful fluid and electrolyte management, dialysis, and transfusion as needed)
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

9. B. 1, 3

Fever is often part of the presentation of bacterial meningitis in children. Lethargy, vomiting, and decreased level of consciousness may also be part of the presentation.

10. A. 1, 2, 3

The worst prognosis is seen in younger children with higher bacterial counts in the CSF. Cerebral or spinal cord infarction, another unusual complication seen in children with bacterial meningitis, can be diagnosed by CT.

11. E. All of the above

HUS is primarily a disease of young children. HUS is characterized by hemolytic anemia, thrombocytopenia, and renal dysfunction. Prognosis for survival is very good, and long-term morbidity such as hypertension and mild azotemia is seen in <10 % of cases. Many causes and associations have been noted. The syndrome can be seen as a result of a toxin-producing *E. coli*, following a prodrome of diarrhea. Treatment is mainly supportive, with careful fluid and electrolyte management.

12. Children with the hemolytic uremic syndrome (HUS):
    1. May have had infection with *E. coli*, *Shigella*, or *Salmonella*
    2. Have anemia, thrombocytopenia, and low WBC counts due to bone marrow failure
    3. Are generally younger than 5 years of age
    4. Are best treated with IV immunoglobulin
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

13. In children with a temperature greater than 39 ° C without a source for the fever:
    1. Bacteremia will likely occur in 1–5% of cases.
    2. Bacteremia, if it occurs, will most often be due to *Streptococcus pneumoniae*.
    3. The risk for occult bacteremia is greatest among those younger than 24 months.
    4. Almost all of the children who have bacteremia will develop purulent complications.
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

14. Which of the following are seen relatively often in children with immunodeficiencies?
    1. Growth failure
    2. Chronic diarrhea
    3. Skin rashes
    4. Recurrent or chronic infections
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

15. Scabies:
    1. Is characterized by beefy red skin with satellite lesions
    2. Has 1–2 mm red papules which may be excoriated or crusted
    3. Is caused by contact with an allergen
    4. Is a pruritic rash, particularly at night
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

12. B. .1, 3

Treatment of children with HUS is supportive. The low red cell and platelet counts are due to hemolysis and increased destruction, respectively. The hemoglobin at presentation may be as low as 2 g/dl and platelet count <100,000/mm$^3$.

13. A. 1, 2, 3

Children who present with fever without a source often have viral illnesses, but in children <36 months of age, a WBC count with differential may help identify those with a much greater likelihood of bacteremia.

14. E. All of the above

Immunodeficiencies can be primary or secondary. Primary immunodeficiencies can involve defects in B cells, complement, T cells, or neutrophils. Secondary immunodeficiencies can result from malnutrition, viral infections, metabolic disorders (diabetes mellitus, sickle cell disease, uremia) or malignancies, and cancer chemotherapy.

15. C. 2, 4

Scabies is an intensely pruritic rash, and its preferred sites are interdigital spaces, wrists, elbows, and ankles. Other common rashes seen in infants and children include *Candida albicans*, which commonly complicates diaper dermatitis (which does not have the same beefy red appearance and satellite lesions), and tinea corporis, which is well described by its common name, ringworm.

16. Urticaria (hives) in children may be associated with:
    1. Airway edema
    2. Contact with a food or chemical
    3. Exposure to cold
    A. Exercise1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

17. Urticaria:
    1. Is an evanescent rash consisting of red-pink wheals
    2. May be treated with PO diphenhydramine
    3. Is commonly associated with beta-streptococcal infections
    4. Is especially common in children with abnormalities in T-cell function
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

18. The first teeth to erupt, the lower central incisors, do so at the age of:
    A. 4 months
    B. 7 months
    C. 12 months
    D. 15 months

19. Which of the following is the most common form of child maltreatment?
    A. Physical abuse
    B. Neglect
    C. Sexual abuse
    D. Emotional abuse

16. E. All of the above

Urticaria is characterized by a localized or generalized erythematous, raised rash with lesions of various sizes.

17. A. 1, 2, 3

Up to 20 % of the general population experience urticaria at some point in their lives. Angioedema is a different lesion involving deeper skin layers or submucosa that involves the periorbital and perioral areas, lips, tongue, respiratory tract, hands, feet, and GI tract.

Hereditary angioedema (HAE) is a different condition, transmitted as an autosomal dominant trait. HAE results from partial deficiency of C1 esterase, an enzyme that inhibits the first part of the complement system. This deficiency allows activation of the complement system with resultant symptoms such as angioedema. This edema, without urticaria, can involve the airway.

18. B. 7 months

Deciduous teeth erupt as follows:

6–7 months: upper (first) and lower incisors
7–9 months: upper and lower (first) lateral incisors
16–18 months: bicuspids
12–14 months: first molars
20–24 months: second molars

Permanent teeth begin erupting at 6–7 years of age with incisors first and then molars, followed by bicuspids.

19. B. Neglect

Each year in the USA, there are approximately one million confirmed cases of abuse or neglect of children. The true incidence of abuse and neglect is almost certainly much greater than the one million confirmed reports, however. Physicians are required by law in all states to report all cases of suspected child abuse. Cultural and geographic norms vary greatly, but a working definition of abuse is parental (or guardian) behavior that damages the normal physical and psychological development of a child.

20. Sudden infant death syndrome (SIDS) has been associated with:
    1. Inadequate nutrition
    2. Recent immunization
    3. Maternal smoking
    4. Concurrent upper respiratory infection
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

21. SIDS:
    1. Is the most common cause of death in the first 2 weeks of life
    2. Accounts for 35% of post-perinatal deaths/year in the USA
    3. Occurs with the same frequency in all ethnic groups
    4. Has no pathognomonic markers at autopsy
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

22. A brief resolved unexplained event (BRUE):
    1. Would have previously been called a near-miss SIDS event or apparent life-threatening event (ALTE)
    2. Is more likely to occur following immunizations
    3. May present with pallor, cyanosis, limpness, and apnea
    4. Would be much more likely to occur in firstborn children
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

23. Which of the following conditions are often associated with BRUEs?
    1. Gastroesophageal reflux (GER)
    2. Acute upper respiratory infections (URI)
    3. Seizures
    4. Failure to thrive (FTT)
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

20. B. 1, 3

SIDS occurs almost exclusively in the second through fifth months of life with the peak in the midpoint of that time period. The incidence does not differ much in various seasons or in different climates.

21. E. All of the above

The diagnosis is often one of exclusion. The incidence appears to be stable. In some cases of SIDS, there may have been suffocation by an adult, but this is difficult to prove.

22. B. 1, 3

Although infants who suffer an ALTE requiring intervention may seem to have a slightly higher chance of dying from SIDS, most infants who do succumb to SIDS have not had a prior ALTE.

23. A. 1, 2, 3

While these conditions are seen with higher frequency in infants who have suffered an ALTE, they are not seen more often in infants who have succumbed to SIDS. The pathologic hallmark of SIDS is that there is no pathognomonic finding for SIDS.

24. In children with obstructive sleep apnea syndrome (OSAS), also called sleep-disordered breathing:
    1. The physical exam during wakefulness may be entirely normal.
    2. There is anatomical narrowing of the upper airway.
    3. There is abnormal neuromuscular control of upper airway patency.
    4. The complications which may develop include FTT, hyperactivity, and poor school performance.
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

25. Myelomeningocele:
    1. Is the most common severe form of neural tube defect
    2. Occurs less often in children of mothers who took supplemental folate in the periconceptional time period
    3. May be located anywhere along the neuraxis
    4. Is associated with a Chiari type II defect in 80% of cases
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

26. Tetanus immunization is usually done in combination with other immunizing agents (DTP, Td, DT). Active immunization with tetanus toxoid:
    1. Provides 10 years of immunity
    2. Is given with pertussis in children only until 7 years of age
    3. Is unnecessary in persons with superficial clean wounds who have received their last tetanus toxoid within the past 10 years
    4. Should be given to persons with more serious and dirty/animal wounds if their most recent tetanus toxoid dose was given more than 5 years ago
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

24. E. All of the above

In addition to the problems mentioned, nighttime hypoxemia with resultant pulmonary hypertension and cor pulmonale can develop in children with sleep-disordered breathing. Sleep studies are used to confirm the diagnosis. A history of nighttime snoring is not sufficient to diagnose sleep-disordered breathing.

25. E. All of the above

The caudal neuropore closes by the fourth to fifth week of gestation. Failure of this closure to occur leads to the development of a variety of congenital anomalies including spina bifida occulta, spina bifida cystica, meningocele, and myelomeningocele. Spina bifida occulta is seen in 10 % of the population and generally causes no symptoms. Spina bifida cystica, a saclike lesion associated with unfused vertebrae, is seen in 0.1 % of people. Myelomeningocele is seen in approximately 0.1 % of live births. The location within the cord determines the clinical picture of this condition. Affected children undergo repair within 1–2 days of life and commonly ventriculoperitoneal shunt placement shortly thereafter. The problems (orthopedic, urological, gastrointestinal) persist throughout life. Most children with myelomeningocele have normal intellect.

26. E. All of the above

Tetanus is fortunately very rare in the USA. The bacterium *Clostridium tetani* produces two toxins, but only one, tetanospasmin, produces disease. It is a very potent neurotoxin. Generalized tetanus, the most common presentation, involves trismus, nuchal rigidity, difficulty swallowing, as well as headache. Subsequently, affected individuals develop generalized, uncoordinated muscle spasms. These muscle spasms can lead to fractures, dysphagia, and even respiratory failure.

27. Which of the following statements are true regarding current vaccines given to children?
    1. Paralytic polio is very rarely (1 in 2.6 million) caused by oral polio vaccine (OPV) in either vaccine recipients or contacts.
    2. Although measles vaccine may cause fever in 15% of recipients, more serious side effects are exceedingly rare.
    3. Mumps vaccine may rarely cause orchitis.
    4. Local reactions may occur in up to 25% of recipients of *Haemophilus influenzae* type B vaccine.

    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

28. Therapy for suspected tetanus infection includes:
    1. Penicillin G to kill the *C. tetani*
    2. Tetanus immune globulin (TIG) to neutralize circulating toxin before it binds to neuronal membranes
    3. Active immunization with tetanus toxoid
    4. Dialysis to remove toxin if the patient deteriorates, developing more and more severe muscle spasms

    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

29. Regarding pertussis infection in the USA:
    1. Mortality is highest among infants.
    2. The attack rate of approximately 1 per 1,000,000 is due to high vaccination rate.
    3. Approximately 50% of reported cases are in children <1 year of age.
    4. It is extremely contagious among nonimmunized children.

    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

27. E. All of the above

28. A. 1, 2, 3

Treatment involves inactivation of the circulating toxin, treatment of the infection to stop toxin production, and supportive care as needed. If there is significant tissue necrosis, IV antibiotics will not reach therapeutic levels, and these wounds must be debrided. In very severe cases, amputation should be considered.

Tetanus is caused by an exotoxin produced by *C. tetani*. TIG has no effect on toxin that has already bound to neural tissue and does not cross the blood-brain barrier.

29. E. All of the above

Herd immunity ("community immunity" – "when the vaccination of a portion of the population (or herd) provides protection to unvaccinated individuals") keeps the incidence of the illness low, protecting those infants who are not fully immunized.

30. The clinical manifestations of pertussis include:
    1. Severe paroxysms of coughing, particularly at night
    2. A characteristic inspiratory sound (whoop) between coughing spells
    3. A calm appearance between coughing spells
    4. Normal temperature throughout the illness
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

31. Complications of pertussis infection include:
    1. Bronchopleural fistula
    2. Seizures and mild, transient encephalitis
    3. Coagulopathy
    4. Pneumonia
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

32. Regarding reactions to pertussis immunization:
    1. Temperature elevations >38 °C are seen in approximately 50% of vaccine recipients.
    2. Seizures occur in approximately 1 of 2000 vaccine recipients.
    3. Reactions seem more common and perhaps more severe in children who are older than 7 years when vaccinated.
    4. Evidence for pertussis vaccine encephalopathy, autism, or SIDS following the vaccine has not been found.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

33. Regarding vaccination against polio:
    1. The inactivated polio vaccine (IPV) is contraindicated in immunocompromised children.
    2. Lifelong, but type-specific, immunity is conferred by recognized infections.
    3. Paralytic polio has never been seen in a contact of a recipient of OPV.
    4. OPV and BP are trivalent and provide immunity to three virus types.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

30. A. 1, 2, 3

Morbidity and mortality of infants are due to the severe paroxysms of coughing. The infant with these severe coughing spells cannot feed and may aspirate during attempted feeds. Temperature elevations to 40 °C are part of the illness. WBC counts in pertussis may be so high that the diagnosis of acute lymphoblastic leukemia (ALL) may be considered.

31. C. 2, 4

Pneumonia is often due to bacterial superinfection, not the *B. pertussis* organism itself. Treatment is generally empiric since the infectious organism may not be recovered from the child.

32. E. All of the above

The vaccine has been suspected as an etiologic agent in autism, various forms of encephalopathy, or developmental delay, but a causative link has never been proved despite numerous reviews of databases both in the USA and the UK.

33. C. 2, 4

Inactivated polio vaccine is one of the several inactivated virus vaccines. The others given in childhood are hepatitis A virus (HAV) and influenza. Other types of vaccines in use are made up of immunogenic components of the organism such as pertussis, *Haemophilus influenzae* type B (HIB), and *Streptococcus pneumoniae*. Attenuated live virus vaccines in use include measles, mumps, rubella, and varicella.

34. Influenza vaccine is recommended for:
    1. Children with diabetes
    2. Children with asthma
    3. Children with seizures
    4. All children below the age of 3 years

    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

35. In which of the following groups are accidents **not** the leading cause of death?

    A. 10–14-year-old males
    B. 10–14-year-old females
    C. 14–19-year-old males
    D. 14–19-year-old females
    E. None of the above
    F. All of the above

34. A. 1, 2, 3

The vaccine is recommended for children with medical conditions that may lead them to suffer a more severe form of influenza should they contract the illness. Influenza is passed from person to person via the respiratory route. Infection with influenza is associated with considerable morbidity and mortality. After infection, there is a 2–3-day incubation period prior to the onset of symptoms. Generally in adults and older children, the onset is sudden, with high fevers, headache, myalgias, and chills. This lasts for several days followed by a 2–4-week period of more prominent respiratory symptoms including a prominent dry cough. In young children, influenza infection presents in a manner similar to other viral respiratory illnesses, with fever, cough, coryza, and fussiness. Serious morbidity in otherwise well individuals is due to bacterial respiratory superinfections.

35. E. None of the above

The cause for mortality in children varies by age. The four most common causes are:

| | |
|---|---|
| Birth to 1 year of age | |
| Perinatal factors | 50% |
| Congenital anomalies | 20% |
| Infections | 4% |
| Cardiac disease | 3% |
| 1–4 years of age | |
| Injuries | 40% |
| Congenital anomalies | 13% |
| Infections | 8% |
| Cancer | 8% |
| 5–14 years of age | |
| Injuries | 55% |
| Cancer | 15% |
| Cardiac disease | 4% |
| Congenital anomalies | 4% |
| 15–25 years of age | |
| Injuries | 75% |
| Cancer | 6% |
| Cardiac disease | 4% |
| Infections | 2% |

36. Which of the following milestones are appropriate for a 6-month-old with normal development?
    1. Able to feed her-/himself
    2. Able to sit unsupported
    3. Speaks single syllables or imitates speech sounds
    4. Beginning walking

    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

37. Which of the following milestones are appropriate for a 12-month-old with normal development?
    1. Beginning walking
    2. Wave bye-bye
    3. Have a one- to three-word vocabulary
    4. Play ball with parent

    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

36. A. 1, 2, 3

Another important developmental milestone for anesthesiologists and other medical professionals to note is the beginning of stranger anxiety. Most 6–8-month-old infants will easily go to a smiling stranger, but at around 8–9 months, most will be quite fearful of people with whom they are not very familiar.

37. E. All of the above

At this age, most language will be intelligible only to the parents or close family members. Phrases and sentences that strangers will understand will not be articulated until 2–4 years of age. A solid understanding of growth and development is a crucial part of caring for children and an invaluable aid in establishing rapport with these young people.

## Suggested Readings

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# Chapter 2
# Newborn Medicine


**Thomas J. Mancuso**



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## Questions

1. In the neonatal period (day 0–28 of life), mortality is higher than any other period in infancy and childhood. Regarding neonatal mortality, the following is true:
   1. It is inversely correlated with birth weight with most deaths occurring in neonates with birth weights <1.5 kg.
   2. It is most commonly due to prematurity and its complications.
   3. Most neonatal deaths occur in the first week of life.
   4. The high neonatal mortality in African-American babies is due to the higher rate of premature births in this group.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

2. Regarding apnea of prematurity:
   1. It occurs in nearly all infants born weighing <1000 g.
   2. It usually resolves by 36–37 weeks postconceptual age (PCA).
   3. It is treated with theophylline or caffeine.
   4. Infants with this problem require home monitoring until 60 weeks PCA.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

3. Which of the following are associated with poor fetal growth and therefore SGA births?
   1. Reduced uteroplacental blood flow
   2. Intrauterine infection
   3. Chromosomal abnormalities
   4. Poor maternal nutrition
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

## Answers

1. All of the above

Low birth weight, which is distinct from preterm birth (see definitions), occurs in approximately 7% of live births in the USA. Mortality of low birth weight infants is higher than mortality of normal birth weight infants by approximately the following:

Moderately low birth weight (MLBW 1501–2500 g) 40 times increased, very low birth weight (VLBW 1000–1500 g) 200 times increased, and extremely low birth weight (ELBW <1000 g) 600 times increased.

Mortality for low birth weight infants has decreased with improvements in newborn care. Common causes for mortality in the newborn are different for term and preterm newborns.

Term: congenital anomalies, birth asphyxia, infection, and meconium aspiration syndrome

Preterm: respiratory distress syndrome (RDS), intraventricular hemorrhage (IVH), infection, and necrotizing enterocolitis (NEC)

The LBW (<2500 g) rate in the USA has increased from 6.6% to 7.5% from 1981 to 1997. The USA still lags behind many industrialized countries in neonatal mortality, while the rate of teen pregnancy exceeds that of many industrialized countries.

2. A. 1, 2, 3

Apnea is defined as cessation of airflow into the lungs for a specified period of time, usually 1–20 s. Once the known potential causes for apnea have been ruled out, the diagnosis of apnea of prematurity can be made. Infants with apnea of prematurity may be discharged home without monitoring provided they have had 7–10 days free of apneic spells. The incidence of SIDS does increase with decreasing birth weight, but apnea of prematurity is not an independent risk factor for SIDS.

3. E. All of the above

Intrauterine growth restriction can be considered a final common pathway for a myriad of influences on the fetus including genetic factors and environmental influences. The intrauterine environment is determined by uterine blood flow, placental function, and placental and umbilical circulation. Maternal factors that affect birth weight include maternal weight gain, maternal age, and medical conditions such as hypertension or diabetes mellitus.

4. What maintenance fluid would you order for a 2 kg, 2-week-old who will be NPO for 6 h?
   1. D5 0.2 NS at 8 mL/h
   2. D10 0.45 NS at 10 mL/h
   3. D5 LR at 10 mL/h
   4. D5 0.45 NS at 12 mL/h

5. Which of the following is (are) true regarding maintenance fluids, electrolytes, and glucose administration to the newborn after the first week of life?
   1. Approximately 100–125 mL/kg/day of water will replace urine output and insensible losses.
   2. Glucose utilization, 6–10 mg/kg/min, can be supplied with D10 given at 100 mL/kg/day.
   3. Excessive sodium losses, due to renal tubular immaturity, must be replaced with 0.9% NS.
   4. Preterm newborns require less fluid than term infants because of their decreased urine output.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

6. Newborns have difficulty maintaining temperature because:
   1. They have a large surface area relative to their weight.
   2. Their increased tone leads to excessive heat loss.
   3. Shivering thermogenesis is limited.
   4. Brown fat is a poor insulator.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

4. D. D5 0.2 NS at 8 mL/

Water administration to term older infants and children is related to caloric expenditure in the following manner on a 1 mL/cal basis:

0–10 kg: 100 cal/kg/day divided by 24 h/day = 4 mL/kg/h.
10–20 kg: 50 cal/kg/day divided by 24 h/day = 2/mL/kg/h.
20 kg: 20 cal/kg/day divided by 24 h/day = 1 mL/kg/h.

Sodium requirements are in the neighborhood of 2–3 meq/kg/day. 0.2–0.45% NS is adequate for sodium replenishment for children up to 45 kg.

Fluid requirements for the newborn change dramatically in the first few days of life. For DOL #1, the fluid needed by the newborn is 60–80 mL/kg/day, gradually increasing to 100–140 mL/kg/day over the subsequent several days. D10 provides sufficient glucose to the newborn.

5. A. 1, 2, 3

The newborn has higher insensible fluid losses than older children. Transdermal evaporative losses are affected by the ambient temperature, while respiratory evaporative losses are affected by the humidity. Maintenance glucose requirements can be met with the administration of 6–8 mg/kg/min. D5 at 100 mL/kg/day provides 5 g/kg/day or 5000 mg/kg/day of glucose or 3.5 mg/kg/min (5000 mg/kg/day × 1 day/1440 min/day = 3.5 mg/kg/min). D10 given at 100 mL/kg/day will provide 6.7 mg/kg/min of glucose. Normal newborns lose little sodium in the first few days of life, often receiving only D10W during the first 24 h of life. Preterm newborns require more fluid because of increased transdermal losses.

6. B. 1, 3

Surface area/weight in a newborn is three times that of an adult. Newborns lose heat at a rate approximately four times that of adults. Nonshivering thermogenesis, which occurs in the brown fat, is a neonatal response to cold. In nonshivering thermogenesis, fat is oxidized and oxygen consumption is increased.

7. The neutral thermal environment for a 10-day-old 1.5 kg infant lying on a warm mattress in a draft-free room of moderate humidity:
   1. Is a room temperature of 34–35 ° C
   2. Is the environment at which the baby will be actively warmed
   3. Is the environment at which $O_2$ consumption is lowest
   4. Includes warming lights
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

8. The Apgar score:
   1. Has a 0–10 scale
   2. Is a useful guide to interventions needed in neonatal resuscitation
   3. Can be used to estimate the likelihood of neonatal acidosis
   4. Was developed in the 1950s by Virginia Apgar, an anesthesiologist
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

9. The Apgar score includes all of the following, which are scored 0–2, except:
   1. Heart rate
   2. Presence of gag reflex
   3. Respiratory effort
   4. Tone
   5. Reflex irritability
   6. Color
      A. 1
      B. 2
      C. 3
      D. 4
      E. 5
      F. 6

7. B. 1, 3

The neutral thermal environment is one with the ambient temperature in which the newborn loses the least amount of heat while maintaining normal body temperature. A neutral thermal environment is one in which the infant neither gains nor loses heat. The newborn loses heat by four means:

Convection to the cooler surrounding air

Conduction to the cooler surfaces which contact the newborn's skin

Radiation to nearby solid objects

Evaporation from moist skin and lungs

Newborns respond to ambient temperature below the neutral thermal environment with increased oxygen consumption to produce heat. The increased oxygen consumption response is limited, however, and once this occurs, the temperature of the newborn begins to fall.

8. E. All of the above

This score is of value in assessment of the newborn at birth and the effectiveness of any resuscitation efforts. Apgar scores at 1 and 5 min correlate poorly with longer-term neurologic outcome. The American Academy of Pediatrics and American College of Obstetrics and Gynecology emphasize using the Apgar score only as a tool in evaluating the condition of the newborn at the time of birth.

9. F

The Apgar score range is 0–10. Term newborns without congenital anomalies with a normal cardiopulmonary adaptation to extrauterine life should have a score of 8–9. Newborns with a score of 0–3 require resuscitation. Most cases of low Apgar scores are due to inadequate ventilation, not to cardiac causes.

In her original work (Apgar, V. *Current Research in Anesthesia and Analgesia* 1953:32;260), Dr. Virginia Apgar demonstrated that the score could differentiate between infants born to mothers who had general anesthesia and infants born to mothers who had spinal anesthesia.

10. A newborn whose Apgar score was 2 at 1 min has been intubated and is being adequately and appropriately ventilated. The heart rate is now 60/min. The next intervention should be:
    1. Volume expansion with 10 cc/kg isotonic fluid
    2. Correction of acidosis with $NaHCO_3$, 1 meq/kg, slowly
    3. Observation and active warming in the special care nursery
    4. Closed cardiac massage
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

11. Intraventricular hemorrhage in preterm infants has been associated with:
    1. Acidosis
    2. Hypoxemia
    3. Cerebral blood flow alterations
    4. Germinal matrix hyperplasia
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

12. Possible consequences of germinal matrix hyperplasia (GMH)/intraventricular hemorrhage (IVH) include:
    1. A normal neurologic exam after grade I IVH
    2. Posthemorrhagic hydrocephalus (PHH)
    3. Motor and cognitive deficits in 50% of infants with grade IV IVH
    4. Hydrocephalus in virtually all infants with grade III–IV IVH
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

10. D. 4

The goals of neonatal resuscitation are to prevent morbidity and mortality of hypoxic-ischemic damage and to reestablish spontaneous respiratory effort and cardiac output. Although the 1 min Apgar score is useful in evaluation of the newborn, there are occasions when intervention should be immediate. Please review resuscitation of the newborn in one of the references.

11. A. 1, 2, 3

Immature vessels in the gelatinous subependymal germinal matrix of preterm newborns are subject to various forces predisposing the preterm to intraventricular hemorrhage (IVH). Contributory factors include prematurity, respiratory distress syndrome (RDS), pneumothorax, hypotension, hypertension, and increased venous pressure. Most IVH occurs within the first week of life and can present with seizures, apnea, cardiovascular instability, and acidosis. The risk for IVH decreases with increasing gestational age. In many surveys, approximately one-half of infants with birth weights <1500 g have imaging evidence of IVH.

12. E. All of the above

The incidence of IVH increases with decreasing birth weight: 60–70% of 500–750 g. Infants and 10–20% of 1000–1500 g infants have IVH. There are four grades defined by ultrasound (done through the anterior fontanelle):

Grade I: bleeding in the germinal matrix

Grade II: blood in the ventricle filling <50% of the ventricle

Grade III: >50% of the ventricle filled with blood

Grade IV: grade III + intraparenchymal blood

Marked clinical deterioration (apnea, seizures, metabolic acidosis, decreased tone) accompanies the occurrence of the IVH, usually within the first week of life. Neurological sequelae are more severe in newborns with the more severe grades of IVH.

13. The initial laboratory evaluation of a healthy neonate with a normal perinatal history who has a brief seizure and who is now clinically stable should include:
    1. Measurement of electrolytes, $Ca^{++}$, and glucose
    2. Neuroimaging
    3. An EEG
    4. A lumbar puncture
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

14. Regarding neonatal respiratory distress syndrome (RDS):
    1. It is rare in infants born after 30 weeks of gestation.
    2. It is due to surfactant deficiency.
    3. Lung compliance is decreased in infants with RDS.
    4. It is associated with the premature closure of the PDA (patent ductus arteriosus).
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

15. Which of the following are features of RDS?
    1. Grunting
    2. Nasal flaring
    3. Air bronchograms on CXR
    4. Central cyanosis with peripheral plethora
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

16. Therapies for RDS include:
    1. Distending airway pressure
    2. Administration of sodium bicarbonate
    3. Surfactant administration
    4. Hypertonic fluid administration
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

13. E. All of the above

The most common cause of seizures in the newborn is hypoxic-ischemic encephalopathy. Other causes include infectious, metabolic, hemorrhagic (see above), and structural abnormalities. Seizure types in the newborn include:

Myoclonic, involving the extremities

Focal, often involving the facial muscles

Subtle, involving chewing, blinking, and respiratory alterations including apnea and multifocal clonic seizures

14. A. 1, 2, 3

RDS occurs in approximately 75% of infants born at <28 weeks of gestation and in about 5% of those born after 37 weeks. Increased incidence (controlling for gestational age) is seen in infants of diabetic mothers, multi-fetal pregnancies, and cesarean delivery. Preterm white males have the highest incidence. Surfactant deficiency leads to higher surface tension within the alveoli, development of atelectasis, and a decreased FRC leading to hypoxemia.

15. A. 1, 2, 3

Rapid, shallow breathing, indicative of poor compliance, is seen within minutes of birth in RDS. The natural course is one of progressive cyanosis and dyspnea. Newborns with RDS exhibit nasal flaring, grunting (in an effort to develop end-expiratory distending airway pressure), and tachypnea. Affected and untreated infants may develop mixed acidosis, hypotension, temperature instability, and apnea.

16. B. 1, 3

Impaired gas exchange in the lung is the basic pathophysiology requiring treatment. Warm humidified oxygen should be given to maintain $SpO_2$ >90%. If this is not accomplished with an $FiO_2$ of 60%, CPAP via nasal prongs should be started. At this point, administration of exogenous surfactant via endotracheal tube should also be considered, and assisted mechanical ventilation may be needed. Surfactant administration should be started within the first 24 h of life and may be repeated every 6–12 h for up to two to four doses depending upon the clinical situation.

17. Transient tachypnea of the newborn (TTN):
    1. Is primarily seen in prematures born between 30 and 34 weeks of gestation
    2. Can progress to chronic lung disease if untreated
    3. Resolves within 24–48 h
    4. Has a CXR identical to that seen with RDS

18. The ductus arteriosus:
    1. Has right to left blood flow in the normal fetus
    2. Closes in the postnatal period as a result of higher oxygen tension in the blood
    3. If open in the preterm, may lead to congestive heart failure
    4. If open in the newborn, causes a characteristic harsh diastolic murmur
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

19. The diagnosis of PDA is supported by:
    1. The presence of a shadow at the aortic knob on CXR
    2. The presence of diminished peripheral pulses due to excessive pulmonary blood flow
    3. The presence of pulsus paradoxus
    4. The findings of bounding pulses, tachypnea, and a systolic murmur

20. Which of the following maternal/perinatal factors is (are) often associated with congenital heart disease?
    1. The presence of a chromosomal abnormality
    2. Maternal rubella infection
    3. Maternal alcohol abuse during pregnancy
    4. Maternal cocaine use during pregnancy
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

17. C

TTN is seen in newborns following an uneventful term vaginal or cesarean delivery. The infants may have a minimal oxygen requirement. TTN resolves within 2–3 days. It is thought to be due to delayed absorption of fetal lung fluid. CXR will show prominent pulmonary vascular markings, fluid lines in the fissures, and over-aeration.

18. A. 1, 2, 3

In the fetus, RV output is 66% of the combined ventricular output, and the ductus arteriosus carries 90% of that RV output to the descending aorta, with 10% going to the lungs. In the normal newborn, the patent ductus arteriosus (PDA) may have a continuous murmur, often described as machinelike. In newborns, a large PDA may present with bounding pulses, cardiomegaly, and other signs of CHF. Bounding peripheral pulses are the result of increased LV stroke volume due to the increased LV volume load and diastolic runoff due to the low diastolic pressure. A small PDA may be asymptomatic.

19. D. The findings of bounding pulses, tachypneaand a systolic murmur

The CXR in a newborn with a large PDA will show increased pulmonary vascular markings and possibly cardiomegaly. The echo will show an enlarged left atrium, picked up by an abnormal LA/Ao ratio. The ductus can often be seen with 2D echo. The LA is enlarged due to the R to L shunt through the PDA. Spontaneous closure of the PDA beyond infancy is rare. The risk of endarteritis is such that all PDAs should be closed either surgically or via catheter closure.

20. A. 1, 2, 3

Infants born to mothers who abused cocaine have many problems, but an increased incidence of congenital heart disease is not one of them. Problems these children do have as a result of intrapartum cocaine exposure include spontaneous abortion, preterm birth, IUGR, microcephalus, abnormal EEG, poor expressive language and verbal comprehension, and later behavioral problems.

21. In persistent pulmonary hypertension of the newborn (PPHN):
    1. Pulmonary blood flow is decreased.
    2. There is systemic hypoxemia.
    3. Blood flow through the PDA is right to left.
    4. The systemic vascular resistance is much lower than it was during fetal life.
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

22. At birth, the right ventricle:
    A. Is hypoplastic
    B. Is approximately as thick-walled as the left ventricle
    C. Has much thicker walls than the left ventricle
    D. Has poor contractility until PVR decreases

23. Which of the following congenital heart defects is the most common in full-term newborns?
    A. Coarctation of the aorta
    B. Tetralogy of Fallot
    C. Patent ductus arteriosus
    D. Ventricular septal defect
    E. Hypoplastic left heart syndrome

24. Hypoglycemia is seen in the following neonates:
    1. SGA newborns
    2. Infants with polycythemia/hyperviscosity
    3. Preterm newborns
    4. Infants with Beckwith-Wiedemann syndrome (macroglossia, visceromegaly, omphalocele)
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

21. A. 1, 2, 3

PPHN may occur in term and postterm infants after birth asphyxia, meconium aspiration, group B streptococcal sepsis, or polycythemia. The normal decline in pulmonary vascular resistance (PVR) that usually occurs after birth does not occur. Excessively high PVR leads to a return to a fetal pattern of circulation, with increased right to left flow through the PDA from the RV and markedly diminished pulmonary blood flow.

Labile hypoxemia, out of proportion to CXR findings, is seen. Hypoxemia, hypercarbia, and acidosis worsen the degree of pulmonary vasoconstriction. A transthoracic echocardiogram can confirm the diagnosis and rule out other causes of profound hypoxemia such as congenital heart disease.

22. B. Is approximately as thick-walled as the left ventricle

During fetal life, the RV delivers approximately 90% of its output to the systemic circulation via the open ductus arteriosus and 10% to the very high-resistance pulmonary circulation. The ECG of a newborn shows prominent right-sided forces with right axis deviation and large R waves. The upright T waves in the precordial leads seen at birth often revert to negative within a few days after birth.

23. D. Ventricular septal defect

Ventricular septal defects (VSD) comprise approximately 25% of all congenital cardiac lesions, exclusive of PDA in preterms, bicuspid aortic valves, and peripheral pulmonic stenosis. The majority are of the membranous type, located posteroinferiorly, anterior to the septal leaflet of the tricuspid valve. The severity of the VSD can be characterized by the ratio of pulmonary to systemic flow ($Q_p/Q_s$). An infant with a ventricular septal defect with a $Q_p/Q_s$ >2:1 will exhibit clinical signs and symptoms of congestive heart failure (CHF) such as effortless tachypnea, diaphoresis, and poor feeding (the equivalent of “exercise intolerance” in the newborn).

24. E. All of the above

There are four groups of newborns at risk for hypoglycemia: infants of diabetic mothers, IUGR newborns, very immature and/or ill newborns, and newborns with metabolic/genetic disorders such as galactosemia, glycogen storage diseases, etc.

25. Hypoglycemia in the term neonate:
    1. Is diagnosed only by the presence of signs and symptoms and not a specific number
    2. Should only be treated if it occurs after the first 3–4 h of life
    3. Is very rarely seen in large, term infants
    4. Is commonly defined as a glucose of <45 g%
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

26. Symptoms and signs of hypoglycemia in the neonate include:
    1. Tremors or seizures
    2. Apnea
    3. Lethargy
    4. Poor feeding
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

27. In the treatment of glucose of <30 mg% in a newborn under anesthesia in the OR, an IV bolus of 200–300 mg/kg glucose (2–3 mL/kg of D10) is given, followed by:
    1. 4 mL/kg/h of D10
    2. D5.2 NS at maintenance
    3. 6–8 mg/kg/min glucose
    4. Glucagon 0.3 mg/kg IM up to a maximum of 1.0 mg
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

28. Regarding hemoglobin in the newborn:
    1. The mean venous hemoglobin in term infants is 18 g/dL.
    2. The physiologic anemia in preterm infants lasts longer and has a lower nadir than that seen in full-term infants.
    3. Hemoglobin concentration increases during the first few days of life as plasma volume decreases.
    4. RBC survival is normal (120 days) in term infants.
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

25. D. 4

The incidence of hypoglycemia varies with the definition used, the population studied, and the method of measurement. In term infants, a glucose of less than 35 mg% requires intervention, while symptomatic infants with glucose measurements >40 mg% also may be treated. Preterm newborns are not more tolerant of low glucose than full-term newborns. Term infants and preterm newborns are equally at risk for severe neurodevelopmental sequelae if left with a low serum glucose.

26. E. All of the above

In the newborn, hypoglycemia may present with neurologic (apnea, seizures, lethargy, coma) or sympathomimetic (pallor, palpitations, diaphoresis) symptoms. The brain in a newborn uses glucose at a rate of approximately 20 mg/min or 4–5 mg/100 g brain/min. The rate of glucose utilization of 5–7 mg/kg/min for a 3.5 kg newborn leads to an overall rate of glucose utilization of 17–24 mg/min.

27. B. 1, 3

Treating hypoglycemia with larger amounts of glucose than 200–300 mg/kg results in rebound hypoglycemia. If the hypoglycemic newborn is seizing, 400 mg/kg may be given. The infusion is begun following the bolus and the glucose level is closely followed afterward. The prognosis of asymptomatic hypoglycemia is generally quite good. If hypoglycemia is accompanied by seizures, it is associated with abnormal intellectual development.

28. A. 1, 2, 3

Hemoglobin levels in very low birth weight (VLBW) infants are 1–2 g lower than those of term infants.

29. The physiologic anemia (expected drop in hemoglobin) of infancy:
    1. Is due to decreased erythropoiesis in the oxygen-rich postnatal environment
    2. Occurs more rapidly and has a lower nadir in preterm infants compared to term infants
    3. Occurs at 10–12 weeks of age in term infants
    4. Has its nadir at 9–10 g/dL in term infants
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

30. Neonatal polycythemia:
    1. Is seen in infants of diabetic mothers
    2. Is diagnosed with a venous HCT >65%
    3. Is treated with partial exchange transfusion in symptomatic infants
    4. Can lead to development of seizures, CNS damage, or necrotizing enterocolitis (NEC)
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

31. Polycythemia in the neonate (a venous HCT >65% on two separate specimens):
    1. Is commonly idiopathic
    2. Occurs in infants of diabetic mothers
    3. Is associated with prolonged labor and fetal distress
    4. Occurs in newborns with intrauterine growth restriction
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

32. Polycythemia in the neonate should be treated:
    1. In all infants whose venous HCT is >65%
    2. With simple phlebotomy to reduce the HCT to <60%
    3. With exchange transfusion to reduce the HCT to <45%
    4. With partial exchange transfusion in all symptomatic infants whose venous HCT is >65% on two separate specimens
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

29. E. All of the above

The anemia of prematurity occurs at 1–3 months and may present with pallor, apnea, poor weight gain, tachypnea, and tachycardia. As the total hemoglobin concentration drops, the concentration of fetal hemoglobin decreases; the newborn makes more HbA, a hemoglobin that releases oxygen more readily than fetal Hb. The P50 of fetal hemoglobin is a $PaO_2$ of 19 mmHg, while that in the adult, with no fetal Hb, is a PaO2 of 32 mmHg.

30. E. All of the above

With increases in hematocrit from 40% to 60%, blood viscosity changes very little. With increases above 65%, blood viscosity increases rapidly. The incidence of polycythemia is increased in babies born at altitude, postmature vs. term infants, SGA babies, infants after delayed clamping of the umbilical cord, and infants of diabetic mothers.

31. A. 1, 2, 3

Clinical manifestations of polycythemia include lethargy, tachypnea, respiratory distress, hypoglycemia, and thrombocytopenia. Infants may appear ruddy or plethoric. Severe complications also may occur such as seizures, necrotizing enterocolitis (NEC), and pulmonary hypertension (PPHN). Although studies are not conclusive, it appears that long-term sequelae such as neurodevelopmental abnormalities can be prevented by treatment of affected infants with partial exchange transfusion.

32. D. 4

The goal of the partial exchange transfusion is to reduce the hematocrit to <50%. The long-term prognosis of polycythemia is unclear. Some adverse outcomes reported include problems with speech and fine motor control and perhaps lower IQ scores. Partial exchange transfusion, when performed through an umbilical vein, is associated with an increased incidence of NEC.

33. "Physiologic" hyperbilirubinemia in the healthy term newborn:
    1. Usually does not exceed 8–9 mg/dL of unconjugated (indirect) bilirubin
    2. Is seen only in breastfed infants
    3. Can be partly accounted for by the low levels of glucuronyl transferase in the newborn
    4. Is diagnosed with a bilirubin level > 15 mg/dL within the first week of life
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

34. Factors which are important in the decision to institute phototherapy treatment for unconjugated hyperbilirubinemia include:
    1. The neonate's gestational age
    2. The neonate's chronological age
    3. The presence of other illnesses such as sepsis or respiratory distress
    4. The neonate's hemoglobin concentration
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

35. Bilirubin results from hemolysis. Causes of hemolysis in the newborn associated with hyperbilirubinemia include:
    1. Cephalohematoma
    2. Rh or ABO incompatibility
    3. Circulating bacterial endotoxin from group B *Streptococcus*
    4. Sickle cell trait
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

36. Bilirubin toxicity:
    1. May be seen in term neonates whose bilirubin levels exceed 25 mg/dL
    2. Need not be seen in term infants whose bilirubin exceeds 30 mg/dL
    3. May be seen in preterm infants weighing <1500 g whose bilirubin level exceeds 15 mg/dL
    4. Results from damage to the basal ganglia and cranial nerve nuclei
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

33. B. 1, 3

Jaundice is observed in approximately 60% of term and 80% of preterm infants during the first week of life. The color results from accumulation of unconjugated (indirect-reacting) bilirubin in the skin. "Physiologic jaundice" appears on day 2 or 3 of life, but jaundice appearing at this time may also represent a more severe form. Clinical jaundice and indirect hyperbilirubinemia are reduced upon exposure of the skin to visible light in the blue (420–470 nm) range. Conventional phototherapy is applied continuously, and the baby should be turned to expose the maximum amount of skin. The eyes should be covered. Complications of phototherapy include loose stools, rashes, and dehydration. Exchange transfusion is another more definitive but also more invasive procedure to lower bilirubin.

34. A. 1, 2, 3

There are many algorithms for the use of phototherapy. In general, phototherapy for unconjugated hyperbilirubinemia has begun at lower bilirubin concentrations in younger, smaller, and sicker infants and infants in whom the rate of rise of unconjugated bilirubin is more rapid.

35. A. 1, 2, 3

The causes include factors which increase the amount of bilirubin presented to the liver for conjugation (hemolysis, infection, shortened red blood cell life span) or factors that decrease the liver's ability to conjugate the bilirubin (liver immaturity, enzyme deficiency, prematurity, hypothyroidism).

36. E. All of the above

Kernicterus is the neurologic syndrome resulting from deposition of unconjugated bilirubin in brain cells. The relationship between serum bilirubin levels and kernicterus in healthy term infants is uncertain. The less mature the infant, the greater the susceptibility to kernicterus. Suggested maximum unconjugated bilirubin levels (in mg/dL) in relatively healthy preterms are:

<1000 g: 12–13
1000–1250: 12–14
1250–1500: 14–16
1500–2000: 16–20

37. The clinical signs of bilirubin toxicity include:
    1. Lethargy
    2. High-pitched cry
    3. Rigidity
    4. Choreoathetosis
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

38. A mother with type O+ blood delivers a 35-week, 2600 g infant with type A+ blood. She is breastfeeding. On day 2 of life, the infant's indirect bilirubin is 12 mg/dL. Management includes:
    1. Cessation of breastfeeding for 2–3 days
    2. Coombs test, Hb, RBC morphology, and indices
    3. Partial exchange transfusion
    4. Observation with daily bilirubin measurements
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

39. Group B streptococcal sepsis in the newborn:
    1. May occur early, within the first 72 h after birth, primarily with bacteremia
    2. May occur later, between 10 and 30 days of age often including meningitis
    3. Is fatal in 10–15% of cases
    4. Will be less likely by treatment of women colonized with the bacteria with appropriate antibiotics during labor
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

40. Congenital rubella infections are characterized by:
    1. Various congenital cardiac defects
    2. Cataracts
    3. Intrauterine growth retardation
    4. Brain calcifications
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

37. E. All of the above

More long-term neurologic problems associated with kernicterus include mental retardation, choreoathetosis, spastic diplegia, and deafness. The incidence of kernicterus at autopsy in hyperbilirubinemic preterm newborns ranges from 2% to 16%.

38. C. 2, 4

Evaluation of a well newborn with clinical jaundice involves a search for the etiology before deciding that the cause is "physiologic." While it is true that breastfed infants have higher bilirubin measurements than comparable formula-fed infants, breastfeeding is rarely held. Overall, approximately 7% of term infants have bilirubin levels >13 mg%, while less than 3% have levels >15 mg.

39. E. All of the above

Sepsis in the newborn may present with a variety of signs and symptoms including apnea, tachypnea, temperature instability, metabolic acidosis, hypoxemia, or DIC. Initial empirical treatment of infants suspected of having systemic bacterial infection usually consists of an aminoglycoside and ampicillin.

40. A. 1, 2, 3

Congenital rubella affects virtually all organ systems. IUGR is the most common manifestation. Other findings include developmental delay, anemia, blueberry muffin skin lesions, structural cardiac defects (PDA, PA stenosis), hearing loss, microphthalmia, cataracts, and meningoencephalitis. Brain calcifications are seen in children with congenital toxoplasmosis or congenital cytomegalovirus infection and two other parts of the TORCH (toxoplasmosis, others, rubella, cytomegalovirus, herpes), acronym of congenital infections.

41. A newborn with a vesicular rash, retinopathy, and meningoencephalitis likely has:
    1. Group B streptococcal infection
    2. Congenital rubella infection
    3. Congenital herpes simplex virus infection
    4. Chlamydia infection

42. Which of the following are risk factors for the development of BPD or chronic lung disease (CLD) of infancy?
    1. Lower gestational age
    2. Prolonged mechanical ventilation and oxygen therapy
    3. Male gender
    4. Exchange transfusion
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

43. Bronchopulmonary dysplasia (BPD) or chronic lung disease (CLD) of infancy:
    1. Is only seen in infants who suffered severe RDS
    2. Is caused by oxygen toxicity
    3. Is characterized by hypoxia and hypercarbia
    4. Is seen as often in ex-full-term infants as in ex-preterm newborns
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

44. IM vitamin K is given to newborns:
    1. To make up for the relative deficiency of vitamin K in breast milk because newborns have inadequate stores of vitamin K
    2. Because the newborn lacks sufficient bacterial flora to produce vitamin K
    3. To prevent hemorrhagic disease of the newborn due to lack of vitamin K-dependent coagulation factors
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

41. C. Congenital herpes simplex virus infection

Most cases of neonatal herpes occur due to infection during delivery with most cases manifesting themselves in the first month of life. One-third of infected infants will never have a skin lesion, while symptoms of encephalitis (lethargy, seizures, poor tone) occur in 50–80%. Newborns with postnatal infection also often have keratoconjunctivitis. Acyclovir is the mainstay of treatment for HSV. Newborns with intrauterine infection may also present with microcephaly.

42. A. 1, 2, 3

Chronic lung disease results from injury to the newborn lungs from mechanical ventilation and oxygen therapy. It is defined as an oxygen requirement in an infant beyond 36 weeks of postconceptual age. Uncomplicated RDS begins to improve in the third or fourth day, while infants developing CLD show X-ray and clinical worsening. Most affected infants recover by 6–12 months, but some may have respiratory symptoms throughout childhood. Right-sided heart failure may be seen in severely affected infants.

43. B. 1, 3

Treatment of CLD includes nutritional support, fluid restriction, maintenance of adequate oxygenation, and vigorous treatment of infection. Recovery is dependent on growth of healthy new lung tissue. Medications often used to treat these children are diuretics, bronchodilators, and dexamethasone. Infants with CLD often exhibit growth failure, psychomotor retardation, nephrolithiasis (from long-term diuretic therapy and TPN), osteopenia, and subglottic stenosis (from long-term/multiple intubations).

44. E. All of the above

A moderate decrease in some coagulation factors (II, VII, IX, X) occurs in all newborns between the second and third day of life. These gradually return to normal by the tenth day of life. Hemorrhagic disease of the newborn is characterized by GI, nasal, intracranial, or post-circumcision bleeding. Vitamin K administration prevents the fall in vitamin K-dependent factors in term infants but is not effective in all preterm newborns.

45. Which of the following are characteristics of human milk?
    1. It has a casein/whey ratio of 1:4.
    2. It meets all the nutritional needs of infants for only the first 1–2 months of life.
    3. It contains lactose.
    4. Its iron content is adequate for the first year of life.
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

46. Instillation of 1% silver nitrate into the conjunctival sac of newborns shortly after birth:
    1. Is an effective strategy for preventing gonococcal ophthalmia neonatorum
    2. Will not prevent chlamydia conjunctivitis
    3. Can be replaced by instillation of 1% tetracycline ophthalmic ointment
    4. Should not be considered adequate treatment of ophthalmia neonatorum
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

45. B. 1, 3

There are several advantages to breastfeeding: allergy to cow's milk is avoided, human milk contains antibodies, it is free of contaminating bacteria, it contains macrophages and lactoferrin, and it supplies many important nutrients to the infant. Supplements of iron and vitamin D should be started at 4–6 months. If the water supply is not adequately fluoridated, the infant should receive this as a supplement as well.

46. E. All of the above

Other routines of newborn care include warming and drying to help conserve heat; treatment of the umbilical cord with triple dye, bacitracin, or another bactericidal agent; and screening for various diseases (these are state-specific).

# Chapter 3
# Respiratory System


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## Questions

1. Respiratory syncytial virus (RSV) :
   1. Is the second most important lower respiratory tract pathogen in early childhood
   2. Causes infected cells to form characteristic syncytia
   3. Confers lifelong immunity after one infection
   4. Infects well over one million children annually
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

2. Which of the following is part of the clinical presentation of RSV bronchiolitis?
   1. It is commonly seen in children less than 2 years of age.
   2. Young infants with the illness may have lethargy and apnea.
   3. Respiratory distress (caused by small airway obstruction).
   4. Wheezes, rales, and rhonchi all may be heard on auscultation of the lungs.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

3. Respiratory syncytial virus (RSV) can cause:
   1. An upper respiratory illness
   2. Bronchiolitis
   3. Otitis media
   4. Pneumonia
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

## Answers

1. C. 2, 4

RSV is the most important respiratory tract pathogen in childhood. It is the major cause of bronchiolitis and pneumonia in children less than 1 year of age, although placentally transmitted antibody may offer protection for the first 4–6 weeks of life. RSV is a medium-sized RNA virus that produces characteristic syncytial cytopathology. The occurrence of outbreaks each fall and winter and the very high incidence in the first year of life are characteristics not seen with other respiratory viruses.

2. E. All of the above

Infants and children infected with RSV first present with rhinorrhea and then cough accompanied by audible and auscultatory wheezing. There is intermittent fever, and the clear rhinorrhea persists throughout the illness. Hospitalized infants with RSV have normal CXRs only about 10% of the time.

3. E. All of the above

RSV most typically causes coryza and pharyngitis, often with fever. In 10–40% of infected children, there is lower respiratory tract involvement (pneumonia, bronchiolitis). RSV infection is usually an outpatient illness. Generally, 1–3% of infected infants are hospitalized.

4. Infection with RSV:
   1. Is very common among infants
   2. Often leads to more serious respiratory distress in infants aged 2–6 months
   3. Occurs in epidemics annually during the months of November through April
   4. Confers lifelong immunity to the RSV virus
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

5. The pathologic changes brought about by RSV infection include:
   A. Necrosis of the respiratory epithelium
   B. Edema of the submucosa
   C. Destruction of cilia
   D. Small airway obstruction by edema and necrotic cells
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

6. Infection with RSV leads to more severe respiratory distress in:
   A. Ex-preterm newborns
   B. Infants with seizure disorders
   C. Children with congenital heart disease
   D. Infants with sickle cell trait
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

7. Treatments for RSV bronchiolitis include:
   A. Amoxicillin
   B. Ribavirin
   C. Racemic epinephrine
   D. Oxygen
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only

4. A. 1, 2, 3

Annual epidemics of RSV occur during the 4–5 months of the winter. It is estimated that up to 50% of susceptible infants undergo infection during each epidemic. Infection is almost universal by the second birthday. Reinfection occurs at a rate of 10–20% per epidemic throughout childhood with higher rates in day care settings.

5. E. All of the above

The pathology seen in the lung includes necrosis of the respiratory epithelium, mucus secretion, and edema of the submucosa. These changes lead to mucus plugging of the small airways with distal hyperinflation or atelectasis.

6. B. 1, 3

Infection of immunocompromised infants with RSV often results in more severe disease. RSV infection in the first few weeks following bone marrow or solid organ transplant can be as high as 50%. Children for whom immunoprophylaxis is considered useful are ex-preterm newborns with BPD or CLD and ex-preterm newborns discharged from hospital during RSV season.

7. C. 2, 4

Most hospitalized infants are hypoxemic, requiring humidified oxygen therapy. A trial of inhaled bronchodilators is often undertaken and continued if the clinical status of the child improves.

Antibiotics are not useful in uncomplicated RSV bronchiolitis. They may be indicated if a consolidated pneumonia develops, however. Ribavirin has been shown to have a modest effect on the course of RSV pneumonia, but hospital stay and mortality have not been reduced. Long-term effects are unknown. It is currently recommended only for high-risk infants with RSV such as those with CLD, congenital heart disease, or immunodeficiency.

Administration of palivizumab (Synagis®), a monoclonal antibody against RSV or RSV-IVIG, high-titer antibody against RSV, is recommended for protecting high-risk infants from serious complications of RSV. It has been shown to reduce total hospital days in this population.

8. All of the above true statements regarding the prognosis for infants with RSV bronchiolitis include:
   1. Infants who develop the illness are more likely to have recurrent wheezing later in life.
   2. Approximately 1–2% of infants hospitalized with this illness die.
   3. Two to 5% of hospitalized infants with this illness develop respiratory failure.
   4. Anti-RSV antibody administration will dramatically decrease the severity of the illness.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

9. The differential diagnosis of wheezing in children during the first year of life includes:
   1. Bronchiolitis (RSV)
   2. Ataxia-telangiectasia with pulmonary involvement
   3. Gastroesophageal reflux (GER)
   4. Cystic fibrosis
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

10. Asthma, a chronic disease of reversible airway obstruction:
    1. Is characterized by episodes of recurrent wheezing and coughing
    2. Only rarely has an allergic basis in children
    3. Often begins before the sixth birthday
    4. Is decreasing in prevalence and severity
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

8. A. 1, 2, 3

Administration of Palivizumab (Synagis®), a monoclonal antibody against RSV or RSV-IVIG, high-titer antibody against RSV, is recommended for protecting high-risk infants from serious complications of RSV. It has been shown to reduce total hospital days in this population.

9. E. All of the above

Wheezing is a manifestation of obstruction in the lower respiratory tract in children. There are many etiologies:

Acute wheezing: asthma (intrinsic, exercise, anxiety, or cold induced), infection, airway foreign body, aspiration of GI, oral secretions

Chronic: asthma (as above), tracheo- or bronchomalacia, airway compression (various vascular compressions, enlarged lymph nodes, tumors), bronchitis, cystic fibrosis, sequelae of RDS (chronic lung disease or bronchopulmonary dysplasia)

10. A. 1, 2, 3

Asthma is the most frequent admitting diagnosis in children's hospitals. Before puberty, males are affected twice as often as females. Thereafter, the incidence is equal. Thirty percent of children who will later be diagnosed as asthmatics are symptomatic by 1 year of age, and 80% present by the fourth birthday. Although up to 50% of asthmatic children are nearly symptom-free by 20 years of age, resolution is rare in children with steroid-dependent disease.

11. Airway narrowing in asthma is due to:
    1. Thickened basement membranes
    2. Edema of the small airways
    3. Mucus secretion
    4. Increased airway smooth muscle tone
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

12. Causes of wheezing in asthmatic children include:
    1. Viral respiratory infections such as RSV infection
    2. Tobacco smoke
    3. Aspirin
    4. Animal dander
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

13. The changes in the small and large airways that occur in asthma lead to:
    1. Increased airway resistance, especially noticeable during exhalation
    2. Hypercarbia resulting from decreased respiratory drive
    3. Ventilation-perfusion (V/Q) mismatch due to nonuniform airway involvement
    4. Increased specific compliance due to much lower resting lung volumes
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

14. Pathophysiologic alterations seen in asthmatic children include:
    1. Nonuniform small airway obstruction
    2. V/Q mismatch
    3. Decreased lung compliance as a result of hyperinflation
    4. Atelectasis
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

11. E. All of the above

The airway obstruction in asthma is due to bronchoconstriction, mucus hypersecretion, mucosal edema, cellular infiltration, and also desquamation of epithelial and inflammatory cells within the airways.

12. E. All of the above

Wheezing is a complex process involving autonomic, immunologic, infectious, endocrine, and psychological factors. In children with extrinsic or allergic asthma, wheezing results from exposure to environmental factors, and these patients have increased IgE against the implicated allergens. Children with intrinsic asthma do not have such antibodies. Viral infections are the most important infectious triggers of asthma (see RSV). Emotional factors may trigger wheezing, and children with this chronic disease may suffer emotional consequences from the illness.

13. B. 1, 3

$PaCO_2$ is generally low early in asthma attacks, rising as the obstruction worsens. $PaO_2$ is often low during an acute exacerbation and may remain so for several days after the worst of the attack is over. Reversible airway obstruction is a hallmark of asthma, with PEFR and $FEV_1$ increasing at least 10% following bronchodilator administration.

14. E. All of the above

CXR abnormalities often seen in children during acute exacerbations of asthma include hyperinflation, atelectasis, infiltrates, and pneumomediastinum. PEFR and $FEV_1$ are decreased, often by more than 15%. ABG abnormalities are described above.

15. Treatment of acute exacerbations of asthma includes:
    1. CPAP
    2. Steroids
    3. Cromolyn
    4. Beta-agonists
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

16. Regarding the use of theophylline as a treatment for asthma:
    1. The medication has a narrow therapeutic range.
    2. It inhibits phosphodiesterase and is an adenosine receptor antagonist.
    3. It is effective orally and intravenously.
    4. Side effects include sleep disturbances, nausea, vomiting, and headaches.
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

17. Which of the following are common side effects of nebulized albuterol?
    1. Nausea and vomiting
    2. Jitteriness, sleep disturbances
    3. Suppression of adrenal secretion
    4. Tachycardia
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

18. Complications of asthma seen in children with asthma include:
    1. Pneumothorax
    2. Pneumonia
    3. Pneumomediastinum
    4. Sudden death
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

15. C. 2, 4

Therapy of acute asthma is aimed at lessening bronchoconstriction and reducing inflammation. Oxygen is administered by mask or nasal prongs. Bronchodilation is achieved with various inhaled medications such as beta-2 agonists (albuterol) and/or cholinergic antagonists (ipratropium bromide). Systemic corticosteroids are often given for a short course. CPAP will likely worsen air trapping and is avoided. Cromolyn is useful for prophylaxis, especially with exercise-induced asthma. Cromolyn is a maintenance medication with little use during acute exacerbations.

16. E. All of the above

Theophylline may be given orally as a sustained release preparation for children with moderately severe asthma as an alternative to inhaled steroids or cromolyn. It also may be used IV in the treatment of acute severe asthma. The therapeutic range is 10–20 mg%. Toxicity may be seen with serum levels of 25–30 mg%.

17. C. 2, 4

Other treatments for asthma include:

Ipratropium: a cholinergic antagonist that may cause tachycardia and abdominal pain.
Cromolyn: an inhaled powder, which may cause coughing especially when first used. It is used as a preventive measure in asthma, not a treatment of acute exacerbations.

Albuterol: the jitteriness from albuterol usually occurs with excessive use of either the PO or inhaled forms.

18. E. All of the above

Death from childhood asthma is rare, but mortality rates have been increasing. Mortality rates are several times higher in African-American children than in white children.

19. Clinical manifestations of cystic fibrosis include:
    1. Productive cough and recurrent respiratory infections
    2. Hemoptysis, pneumothorax, and atelectasis
    3. Maldigestion due to exocrine pancreatic insufficiency
    4. Diabetes insipidus
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

20. Cystic fibrosis, the major cause of severe chronic lung disease in children:
    1. Occurs in 1:3000 white and 1:17,000 black live births
    2. Is characterized by thickened secretions
    3. Primarily involves the pulmonary and gastrointestinal systems
    4. Is inherited as an autosomal dominant trait
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

21. Treatments that patients with cystic fibrosis (CF) might receive include:
    1. Pancreatic enzyme replacement, high calorie diets, and fat-soluble vitamin supplements
    2. Antibiotics to control progression of pulmonary infections
    3. Bronchodilator and anti-inflammatory agents
    4. Oxygen
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

19. A. 1, 2, 3

CF is characterized by obstruction and infection of the airways and malabsorption of many important nutrients. After 10 years of age, 85% of children with cystic fibrosis will develop diabetes mellitus. People with CF have varying degrees of the following respiratory tract problems: failure to clear mucus secretions, dehydrated mucus secretions, and chronic infection in the respiratory tract. The rate of progression of lung disease is the chief determinant of morbidity and mortality. The first lung pathology is bronchiolitis, followed later by bronchiectasis. Interstitial disease is not a regular feature although eventually fibrosis does develop. The paranasal sinuses are filled with secretions, and the epithelial lining is hyperplastic and hypertrophic. The nasal mucosa is edematous and develops polyps.

20. A. 1, 2, 3

The CF gene is most common in Northern and Central Europeans. It codes for a protein called the cystic fibrosis transmembrane conductance regulator (CFTR) that is expressed largely in epithelial cells of the airways, GI tract, sweat glands, and GU system. The most prevalent mutation of CFTR is the deletion of a single phenylalanine residue at amino acid 508 (F508del). Fifty percent of CF patients with Northern European ancestry are homozygous for F508del.

21. E. All of the above

Antibiotics, given PO, IV, and via inhalation, are used to control the progression of lung infection. Steroids are used to treat allergic pulmonary aspergillosis. Anti-inflammatory agents may slow the progression of lung disease.

22. Croup, a clinical syndrome of barking cough, hoarseness, and inspiratory stridor, has several causes, including respiratory viruses. Characteristics of croup include:
    1. The illness lasts for 4–6 days.
    2. There is a characteristic CXR finding called the pencil (or steeple) sign indicative of subglottic tracheal narrowing.
    3. Treatment with inhaled racemic epinephrine (0.5 cc of a 2.25% solution) temporarily improves the stridor.
    4. Dexamethasone, 0.3–0.5 mg/kg, is a treatment for the illness.
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

23. Clinical characteristics of croup (laryngotracheobronchitis) include:
    1. Mild temperature elevation, rarely reaching 39 °C
    2. The presence of a URI (upper respiratory infection) for 1–3 days prior to the onset of stridor
    3. A peak incidence during the ages of 18 months to 3 years
    4. A typical barking cough
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

24. Acute epiglottitis presents differently than viral croup in the following way(s):
    1. The course of epiglottitis is much more rapid and fulminating.
    2. The temperature elevation in epiglottitis is greater.
    3. The age range of children with epiglottitis is older.
    4. Very often other family members of children with epiglottitis have been ill with URI symptoms.
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

22. E. All of the above

Croup is the most common form of acute upper airway obstruction and is most commonly caused by a virus. Symptoms are characteristically worse at night. Most children with croup progress to stridor and slight dyspnea and then begin to recover. Agitation and crying, with associated more rapid respiratory rate and turbulent airflow, worsen the situation. Children with croup prefer to sit upright.

23. E. All of the above

Older children are generally not seriously ill. Other family members may have a mild respiratory illness. The nighttime worsening may recur for several consecutive days before the illness resolves.

24. A. 1, 2, 3

Epiglottitis is usually seen in children aged 2–7 years, while croup is more often seen in younger children. Epiglottitis is caused by bacteria, while croup a virus. Other family members are not acutely ill with respiratory viruses as is the case with croup. Epiglottitis is a severe bacterial infection associated with high fever, rapidly progressing airway obstruction, and dyspnea.

25. Aspirated airway foreign bodies:
    1. Can usually be seen on either a PA or lateral CXR
    2. Most often occur in 2–4-year-old children
    3. Are usually first noted during an acute URI when the child has more severe symptoms than usual
    4. May not be noted until sometime after the aspiration episode
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

26. Bacterial tracheitis, a cause of upper airway obstruction that occurs as a superinfection of viral laryngotracheitis:
    1. Is often caused by coagulase+ staph or *Haemophilus influenzae*
    2. Is diagnosed with airway endoscopy
    3. Is regularly treated with endotracheal intubation and IV antibiotics
    4. Is seen only in the teenage years
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

27. Regarding acute otitis media (AOM) in children:
    1. Both bacteria and viruses are known causative agents.
    2. Meningitis is a possible complication of untreated bacterial AOM.
    3. It is generally treated with PO antibiotics.
    4. Infants less than 1 month of age with AOM should be thoroughly evaluated for systemic infection.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

25. C. 2, 4

Most airway foreign bodies are not radio-opaque and many are actually food, often peanuts. Although up to 50% of cases of airway foreign body aspiration come to medical attention soon after the aspiration event and the parents give a history of a specific choking episode, a substantial minority of cases are discovered in the evaluation of a child with recurrent wheezing for several months.

26. A. 1, 2, 3

Bacterial tracheitis is one of the laryngotracheal respiratory tract infections affecting children. The others are croup, viral laryngotracheobronchitis, and epiglottitis. Bacterial tracheitis is acute in onset, affects children 4–5 years of age, and is associated with a cough and harsh stridor. Treatment of affected children often involves IV antibiotics, hospitalization, and intubation.

27. E. All of the above

AOM is a very common childhood infection. Management strategies vary. The etiologic agent in a particular case is rarely identified. The tympanic membrane in AOM is red, often bulging, and immobile, and the normal landmarks are not seen. With repeated episodes of AOM or with chronic serous OM, pediatricians often refer their patients to an ORL specialist for myringotomy and tube placement. Untreated AOM can develop into acute mastoiditis, which can destroy the mastoid air cells.

28. Children with acute sinusitis:
    1. Have URI symptoms (nasal discharge and cough) that persist for more than 10 days
    2. May have persistent daytime cough
    3. May have facial pain and swelling in association with their URI
    4. May complain of headache
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

29. Regarding URIs in children:
    1. Mucociliary dysfunction can persist for weeks after recovery from the URI.
    2. Viral URIs predispose children to bacterial infections such as pneumonia, sinusitis, or otitis media.
    3. Nasal discharge, initially watery, becomes mucopurulent after 5–7 days.
    4. Young infants may develop fever to 38 °C or 39 °C with uncomplicated URIs.
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

30. Children with allergic rhinitis:
    1. Often also have conjunctivitis
    2. Have pale edematous nasal membranes
    3. May have nasal polyps
    4. Will have fewer and lessened symptoms during exercise
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

28. E. All of the above

Sinusitis often accompanies the common cold or even allergic rhinitis. The maxillary, ethmoid, and sphenoid sinuses are present at birth, and the frontal sinuses develop at around the first birthday. These sinuses gradually become air filled over the first several years of life. Affected children have persistent purulent nasal drainage, nighttime cough, and facial tenderness and pain. Treatment is with PO antibiotics unless extension from the sinuses is considered a possibility.

29. E. All of the above

The common cold or viral URI is a frequent problem in children. These occur most often in the winter months, from early fall through late springs. Toddlers and young school-aged children can have up to six to nine colds/year. The number per year decreases, with most adults reporting one to three URIs/year.

30. A. 1, 2, 3

The differential diagnosis of rhinitis in children includes sinusitis, viral URI, nasal foreign body, and allergic rhinitis. Children with allergic rhinitis do not have fever but often have allergic "shiners," nasal polyps, and pale edematous nasal mucosa.

31. Regarding URIs in children:
    1. The incidence is highest between the ages of 6 and 8 years.
    2. School-aged children normally experience one to two colds/year.
    3. Boys have more URIs than girls.
    4. Among children aged 1–4 years, those in day care have fewer URIs than those cared for only at home.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

32. Which of the following causes pharyngitis most often?
    A. Mycoplasma pneumoniae
    B. Rhinovirus
    C. Adenovirus
    D. *Haemophilus influenzae*, untypeable
    E. Beta-hemolytic strep

33. Which of the following are considered etiologic agents for the common cold in children?
    1. Parainfluenza viruses
    2. Group B beta-hemolytic streptococci
    3. Respiratory syncytial virus
    4. *Haemophilus influenzae* type B
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

34. Influenza viruses:
    1. Can cause primary pneumonia
    2. Cause epidemic respiratory infections
    3. Are spread from person to person via the respiratory route
    4. Confer lifelong immunity to all strains after one symptomatic infection
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

31. B. 1, 3

A viral URI typically lasts only 7 days with a minority lasting up to 2 weeks. As the URI resolves, the nasal secretions change from clear and thin to a thicker and yellow-green consistency.

32. C. Adenovirus

In children less than 2 years of age, the cause for pharyngitis is usually viral, whereas in children over 5 years of age group, a strep is the most common causative agent. Making the diagnosis of strep pharyngitis is important because appropriate antibiotic treatment will prevent rheumatic fever as well as minimize the chance of local suppurative complications such as abscess formation. Diagnosis is with rapid antigen detection or throat culture. Antibiotic treatment does not, however, prevent the development of poststreptococcal glomerulonephritis.

33. B. 1, 3

The common cold in children is caused by a variety of viruses. It is not a bacterial infection and thus not treatable with antibiotics. Bacterial complications of viral URIs include sinusitis, otitis media, and pneumonia. There is no therapy for the common cold save symptomatic measures, and most over-the-counter medications sold for URI treatment have not been shown to be effective in reducing the symptoms.

34. A. 1, 2, 3

Influenza A viruses are responsible for epidemics. These epidemics follow a shift in one of the major antigens, neuraminidase, or hemagglutinin. The clinical picture in young children is milder than that seen in adults. Young children may exhibit bronchitis, laryngotracheitis, and/or mild upper respiratory tract symptoms. Older children and adults have high fever of abrupt onset, myalgias, chills, and cough. The cough and congestion may last for 2 weeks.

# Chapter 4
# Cardiology


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## Questions

1. Paroxysmal supraventricular tachycardia:
   1. Can cause low output congestive heart failure
   2. Can be treated with IV adenosine
   3. Can be prevented with PO digoxin
   4. Should be initially treated with verapamil
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

2. The ECG of a newborn or infant:
   1. Will show increased right ventricular forces
   2. Will show a prominent R wave in V1
   3. Will show inverted T waves in lead V1
   4. Will show right bundle branch block pattern until 5 years of age
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

3. The most common cardiac lesions, exclusive of PDAs in premies, are (choose 2):
   A. ASD
   B. Tetralogy of Fallot (TOF)
   C. Hypoplastic left heart syndrome
   D. VSD

4. Coarctation of the aorta (CoA), a relatively large VSD:
   1. Will often present with dyspnea, feeding difficulty, and poor growth
   2. Presents at 1–3 months of age as pulmonary vascular resistance (PVR) decreases
   3. Will cause a $Q_p$:$Q_s$ > 2:1
   4. Will present with a harsh holosystolic murmur and a loud P2
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

## Answers

1. A. 1, 2, 3

SVT is a tachydysrhythmia that originates from above the bifurcation of the bundle of HIS. The onset is usually paroxysmal, hence paroxysmal supraventricular tachycardia (PSVT). In children, the rate is generally >230 beats/min, and in infants the rate can exceed 300 beats/min. The QRS is narrow, differing little from the QRS seen during normal sinus rhythm. Predisposing factors for the development of PSVT are the preexcitation syndromes such as Wolf-Parkinson-White and congenital heart disease and sympathomimetic medications such as atropine or glycopyrrolate.

2. A. 1, 2, 3

At birth, the RV and LV walls are of approximately equal thickness; thus, the infant has relative RV hypertrophy with prominence of right and anterior forces.

3. A. ASD and D. VSD

Atrial septal defects (ASDs) come in four varieties: patent foramen ovale (PFO), secundum (at the fossa ovalis) ASDs, coronary sinus defects (absence of wall separating the coronary sinus and LA), and sinus venosus defects (immediately below the SVC opening). Sinus venosus defects are associated with partial anomalous pulmonary venous return.

4. E. All of the above

VSD is the most common congenital cardiac malformation in children with an incidence of approximately 2–3:1,000 live births. VSDs are often not apparent in the newborn because the relatively high pulmonary vascular resistance limits the right to left flow through the defect. Congestive heart failure (CHF) becomes clinically apparent as the infant grows and PVR decreases. The severity of the shunt is characterized by the $Q_p$:$Q_s$ ratio (pulmonary to systemic blood flow). With a Qp:Qs >2, signs and symptoms of CHF are seen. CHF in infants and newborns presents with poor feeding, diaphoresis with feeding, effortless tachypnea, lethargy, and FTT. The typical murmur of a VSD is holosystolic, harsh, and best heard along the left sternal border.

5. Regarding ventricular septal defects (VSDs) :
   1. They occur with an overall incidence of 3–4/1000 live births.
   2. They undergo spontaneous closure in approximately 25% of cases.
   3. Complications of repair are rare and include acquired complete heart block.
   4. Most VSDs occur in association with other congenital anomalies.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

6. Regarding ASDs:
   1. They can be divided into primum, secundum, and sinus venosus based on location and etiology.
   2. Secundum ASDs are the most common type.
   3. Secundum ASDs often are asymptomatic in childhood.
   4. There may be associated partial anomalous venous return with an ASD.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

7. Which of the following organisms are associated with sepsis occurring after cardiac surgery?
   A. *Haemophilus influenzae* type b
   B. *Neisseria gonorrhoeae*
   C. *Staphylococci*
   D. Enteric gram-negative rods

8. The so-called innocent murmur of childhood:
   1. May be heard in up to 30% of children at some point in their lives
   2. Is best heard in a localized area along the left lower sternal border
   3. Is a short, vibratory ejection-type murmur
   4. Is generally heard in children between the ages of 3 and 7 years
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

5. A. 1, 2, 3

VSDs are classified by location in the septum. Membranous or perimembranous defects are the most common. These are located below the aortic valve and adjacent to the tricuspid valve. Other types of VSD include AV canal-type, subpulmonary, conoventricular, and muscular VSDs. Many VSDs undergo spontaneous closure with larger defects less likely to do so. Currently, repair is undertaken in infancy. The risk of heart block in the postoperative period is related to the size and location of the patch used to repair the defect.

6. E. All of the above

Isolated secundum ASDs, which represent 80% of all ASDs, generally do not present in infancy. Children with this defect generally are in sinus rhythm. Primum ASDs are often associated with a cleft mitral valve. Sinus venosus and coronary sinus ASDs are actually defects in the embryologic sinus venosus. The murmur noted in children with relatively large ASDs is a pulmonary flow murmur with associated fixed splitting of S2. Patients with unrepaired ASDs often do relatively well into their 20s when progressive cyanosis and dyspnea develop.

7. C. Staphylococci

8. E. All of the above

The history is unremarkable in these children since they have no cardiac disease. Murmurs are more frequently heard in children during febrile episodes. The cause of the murmur is unknown. Some speculate that it is heard only in childhood because the relatively thin chest wall of young children transmits extra-cardiac sounds more easily.

9. Congestive heart failure in childhood may present:
    1. With chronic cough
    2. As failure to thrive
    3. With respiratory distress during feedings
    4. With the child's complaint that it is difficult to keep up with peers during play
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

10. Tetralogy of Fallot:
    1. Is the most common cyanotic congenital cardiac lesion presenting after 2 weeks of age
    2. Includes pulmonary stenosis, VSD, overriding aorta, and RVH
    3. May have infundibular and valvar pulmonary stenosis
    4. Is inherited as an autosomal dominant
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

11. The so-called tet spells:
    1. Occur in infants with unrepaired tetralogy of Fallot
    2. Are the result of increased tone in the infundibulum of the RV outflow tract
    3. Result in intense cyanosis and diminution of the systolic ejection murmur as pulmonary blood flow dramatically decreases
    4. Can be treated with IV fluid administration and/or increasing SVR
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

12. The most common congenital cardiac defect presenting with cyanosis in the newborn is:
    A. Transposition of the great arteries (TGA)
    B. Patent ductus arteriosus (PDA)
    C. Hypoplastic left heart syndrome (HLHS)
    D. Tetralogy of Fallot (TOF)
    E. Truncus arteriosus

9. E. All of the above

CHF presents in infancy with tachycardia, tachypnea, poor feeding, and failure to thrive. In older children, decreased exercise tolerance is noted, as in adults. Recurrent respiratory infections are common, although rales are not heard until later in the course of CHF.

10. A. 1, 2, 3

TOF is found in approximately 6–10% of infants with cyanotic congenital heart disease. TOF has no known inheritance pattern, but it is found in association with a number of syndromes such as Goldenhar (oculo-auriculo-vertebral hypoplasia), VACTERL (vertebral anomalies, esophageal atresia with tracheoesophageal fistula, radial dysplasia, renal anomalies, imperforate anus, cardiac defects), CHARGE association (choanal atresia, heart defects, deafness, genital hypoplasia in males, coloboma), and Klippel-Feil syndrome (short neck, limited neck motion, low occipital hairline).

11. E. All of the above

With repair of TOF now routinely performed in the neonate and infant, "tet spells" are rare.

12. A. Transposition of the great arteries (TGA)

Without mixing of the two parallel circulations, the newborn with TGA cannot survive.

The presentation of TGA is affected by the presence of other anomalies such as a VSD, left ventricular outflow tract obstruction (LVOTO), or sub-pulmonic stenosis. Newborns with intact ventricular septa rely on the presence of a PFO or PDA to mix oxygenated and deoxygenated blood.

13. A newborn with isolated TGA may present with:
    1. Cyanosis
    2. Tachypnea without dyspnea or respiratory distress
    3. Normal peripheral pulses
    4. An ECG showing right ventricular hypertrophy, indistinguishable from that of a newborn with a normal heart
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

14. Which of the following may be done to newborns with TGA to improve arterial SpO2?
    1. A Rashkind-Miller procedure
    2. Administration of milrinone
    3. Prostaglandin E1 administration
    4. Dilation of the ductus arteriosus in the catheterization lab
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

15. Regarding the ductus arteriosus:
    1. More than 90% of fetal RV output passes through the ductus.
    2. If it remains open postnatally, it has flow through it both during systole and diastole.
    3. It closes functionally during the first day of life in most term infants.
    4. Spontaneous closure of a persistently open PDA is unlikely after the age of 6 months.
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

16. Endocardial cushions contribute to which of the following cardiac structures?
    1. The lower part of the atrial septum
    2. The part of the ventricular septum where the AV valves insert
    3. Tissue that forms part of the mitral and tricuspid valves
    4. Part of the intraventricular conduction system
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

13. E. All of the above

In newborns with TGA and IVS (intact ventricular septum), there often is no murmur present. A CXR may be normal, but approximately 30 % shows an "egg on a string" pattern of the cardiothymic shadow.

14. B. 1, 3

Maneuvers that enhance mixing of the parallel circulations improve the situation in newborns with TGA. Prostaglandins keep the PDA open, and the Rashkind procedure involves creation of an atrial septostomy using a specially designed catheter.

15. E. All of the above

In full-term infants, persistent patent ductus arteriosus accounts for approximately 10% of congenital heart disease. PDAs are much more common in the premature newborn (see question/answer 18 and 19 in newborn medicine). Commonly, the PDA is picked up when a murmur is heard in an asymptomatic child who is being examined for another reason. The typical murmur is continuous (machinelike) and heard best in the midclavicular line between the first and second interspace.

16. A. 1, 2, 3

AV canal defects, also called endocardial cushion defects, involve a primum ASD, defects in one or more of the AV valves, and also a defect in the ventricular septum.

17. Common atrioventricular canal defects (CAVC):
    1. Result in communication between all four cardiac chambers
    2. Have abnormal mitral and/or tricuspid valves
    3. Often present in a manner similar to large VSDs
    4. Predispose the child to the early development of pulmonary vascular obstructive disease (PVOD)
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

18. In total anomalous pulmonary venous return (TAPVR):
    1. It is often divided into several types based upon the site of pulmonary venous drainage.
    2. All the pulmonary veins drain into the systemic venous system, not the left atrium.
    3. There often is pulmonary venous obstruction.
    4. There is an ASD or PFO allowing a right to left shunt to compensate for the left to right shunt resulting from the TAPVR.
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

19. Regarding the locations of the pulmonary venous drainage in TAPVR:
    1. Supracardiac, the most common, involves drainage into an anomalous vein which eventually empties into the SVC.
    2. With intracardiac TAPVR, venous drainage is direct into the RA or coronary sinus.
    3. With infracardiac TAPVR, a vein passes through the diaphragm.
    4. Mixed TAPVR, a combination of the other types, is the least common.
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

17. E. All of the above

AV canal defects have variable anatomy. The atrial and ventricular septa and AV valves are affected in many different ways. Down syndrome is frequently associated with CAVC defects. Two-dimensional echo demonstrates absence of the atrial septum, while pulsed Doppler echo demonstrates the presence of AV valve regurgitation.

18. E. All of the above

TAPVR is classified into four types. In decreasing order of frequency, they are supracardiac, cardiac, infracardiac, and mixed. Obstruction of the anomalous venous drainage leading to pulmonary congestion may occur at any point along the anomalous venous pathway. Obstruction almost always occurs in the infracardiac type.

19. E. All of the above

The presence and degree of venous obstruction and the degree of intra-atrial mixing determine the severity of clinical symptoms. Infants with obstruction in the anomalous venous connections develop cyanosis and respiratory distress early in life. Infants without obstruction and a nonrestrictive inter-atrial communication may have only minimal symptoms during the first year of life.

20. Coarctation of the aorta (CoA):
    1. Is a congenital narrowing of the aorta near the insertion of the ductus arteriosus
    2. Is commonly associated with VSD or hypoplastic left heart syndrome (HLHS)
    3. May cause reduced lower body perfusion
    4. Produces aortic insufficiency
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

21. Newborns and infants presenting with coarctation of the aorta:
    1. Generally have more severe coarctation
    2. May have metabolic acidosis as a result of poor lower body perfusion
    3. May present with signs of LV failure
    4. Have an extensive network of collaterals
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

22. Children with coarctation of the aorta:
    1. Have had time to develop collateral flow through intercostal and other arteries
    2. Usually require cardiopulmonary bypass for surgical repair
    3. May have systemic hypertension
    4. Are often managed medically until adulthood
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

23. The hypoplastic left heart syndrome can include:
    1. Hypoplasia of the LV and RA
    2. Mitral atresia
    3. Coronary, carotid, and subclavian flow via retrograde filling of a small ascending aorta from the ductus
    4. RV hypertrophy
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

20. A. 1, 2, 3

The aortic narrowing seen in CoA is located in the descending thoracic aorta just across from the insertion of the ductus arteriosus. This anomaly has a 2:1 male predominance. The major problem associated with CoA is increased LV afterload. For CoA to become clinically significant, the aortic diameter must be decreased by at least 50%. Pulses and measured blood pressure in the lower extremities are diminished compared to the upper extremities.

21. E. All of the above

Newborns with CoA may appear well initially, but cardiac failure and respiratory distress quickly develop as the ductus closes. Prostaglandin administration may improve the situation as the dilated ductus allows improved lower extremity and renal perfusion.

22. B. 1, 3

Children with isolated CoA often have no specific complaints. With a careful history, the child may report leg cramps. The coarctation may be discovered during an evaluation of systemic hypertension. The ECG may show no changes or LVH by voltage criteria may be seen. The pathognomonic CXR finding of rib notching, due to rib erosion by the enlarged collateral vessels, is rarely seen in children younger than 5–6 years of age.

23. A. 1, 2, 3

HLHS is seen in 3–4:10,000 live births. There is a spectrum of anomalies in this left-sided obstructive lesion. The LV and ascending aorta are underdeveloped. The mitral valve is often involved, exhibiting stenosis, hypoplasia, or atresia. The RV provides both pulmonary and systemic flow in HLHS. There is a L to R shunting of pulmonary venous return at the atrial level and a R to L shunting of RV output at the PDA, with the ascending aorta and its vessels (carotids, subclavian, and coronaries) perfused retrograde via flow from the PDA.

24. What happens to newborns with HLHS when the PDA closes?
    1. There is improved systemic blood pressure.
    2. There is reduced coronary flow.
    3. There is increased systemic flow.
    4. There is decreased systemic flow.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

25. In HLHS, systemic flow is affected by:
    1. SVR
    2. PaO2
    3. PVR
    4. pH
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

26. Which of the following CHD diagnoses is matched with the past or present appropriate surgical procedure?
    1. TOF-BT shunt
    2. TGA switch
    3. HLHS Stage I – Glenn-Fontan
    4. VSD-PA band
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

24. C. 2, 4

The PDA allows the RV output to flow to the pulmonary circuit and the systemic circuit and retrograde into the ascending aorta. Once the diagnosis of HLHS is made, prostaglandin should be infused to keep the PDA open and the newborn monitored for the development of metabolic acidosis.

25. B. 1, 3

The ratio of systemic to pulmonary vascular resistances is crucial in determining flows into these vascular beds. Excessive pulmonary flow will lead to underperfusion of the body. Hyperventilation and oxygen administration may increase SpO2 but lead to systemic hypoperfusion and the development of metabolic acidosis. Chromosomal abnormalities have been reported in 11% of infants with HLHS, and autopsies have revealed neurologic abnormalities in 29% of these patients.

26. E. All of the above

Palliation of TOF is no longer done as a routine, but if done, the goal is to achieve an increase in pulmonary blood flow. A Blalock-Taussig shunt (B-T shunt) diverts subclavian artery flow to the pulmonary circulation, either using a Gortex graft or by an end-to-side anastomosis of the subclavian artery to the PA.

Transposition of the great arteries (TGA) is treated surgically with a so-called switch operation in which the PA and aorta are moved to the appropriate ventricular outflow tract. It is very important to know the coronary arterial anatomy beforehand. The coronaries are removed from the aortic root along with a small area of surrounding tissue and moved to the newly "switched" aorta.

Hypoplastic left heart syndrome accounts for 1% of all CHD. There is a small LV, mitral valve, aortic valve, and aortic arch. A stage I procedure is done in the newborn period. Systemic flow is carried by the PDA, and coronary flow is retrograde in the small aortic arch. The stage I procedure involves creation of a neoaorta from the hypoplastic aortic arch, main PA, and homograft. A large ASD is created and pulmonary blood flow is via a modified (graft material) B-T shunt.

Ventricular septal defects often become clinically apparent in the third month of life, when PVR decreases substantially and the higher left ventricular pressures divert more and more blood to the lower pressure right ventricle, leading to CHF. The degree of shunt is characterized by the ratio of systemic to pulmonary flow ($Q_p$:$Q_s$).

27. The Fontan operation, also called total cavopulmonary connection:
    1. Directs systemic venous return to the PA
    2. Is the surgical procedure for many patients with single ventricle physiology
    3. Is generally performed at 1–2 years of age
    4. Is generally preceded by a bidirectional Glenn procedure

    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

A 2:1 shunt or greater is associated with CHF. Currently, most VSDs are repaired primarily in the OR or with a device in the cardiac catheterization lab. A PA band was used previously as a temporary means to increase resistance to pulmonary flow, thus decreasing the shunt.

27. E. All of the above

The Fontan procedure involves a so-called passive flow of systemic venous return into the pulmonary circuit. This requires a transpulmonary gradient of 3–8 mmHg. This can be achieved if the CVP (PA) pressure is kept at 12–15 mmHg with an LVEDP of 5–10 mmHg. In addition, the cardiac rhythm must be kept in the sinus, and ventricular performance must often be supported pharmacologically.

# Chapter 5
# The Musculoskeletal System


**Thomas J. Mancuso**



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## Questions

1. Cerebral palsy, a movement and posture disorder:
   1. Is seen in 1–2/1000 children, making it the most common childhood movement disorder
   2. Is initially diagnosed when the child exhibits delayed motor development
   3. Does not have identifiable risk factors in most cases
   4. Has a changing clinical picture despite the static nature of the neurologic damage
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

2. Even though many cases of cerebral palsy (CP) do not have an identified etiology, there are known associations such as:
   1. Birth asphyxia
   2. Prematurity
   3. Intrauterine growth restriction (IUGR)
   4. Family history
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

3. Children with CP may also have which of the following:
   1. Seizures
   2. Normal intellect
   3. Mental retardation
   4. Communication disorders, hearing and visual dysfunction
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

## Answers

1. E. All of the above

This condition is the result of an anomaly or insult to the immature CNS, but in many, if not most cases, a specific antecedent event or cause cannot be identified. The term static encephalopathy is often used synonymously with cerebral palsy. The incidence of CP is 7:1,000 live births and prevalence is 5:1,000 of the population. Cognitive impairment is not a consistent feature of CP, although many affected children do have a lower than normal IQ. Between 30% and 70% of children with CP do have impaired intellect. Many children with CP also have seizures. CP is described by the clinical appearance:

Spastic diplegia
Spastic quadriplegia
Spastic hemiplegia
Extrapyramidal
Atonic
Mixed

2. A. 1, 2, 3

The association of CP with prematurity is changing as neonatal care improves. The incidence is decreasing in heavier preterm newborns, but VLBW (very low birth weight) infants have a higher incidence.
Clinical types of CP:

Spastic diplegia: Lower extremity involvement, seen in low birth weight infants, after intraventricular hemorrhage. Severe mental deficits less common than in other types
Spastic quadriplegia: All four extremities involved. More severe mental deficiencies, seizures likely. Scoliosis, feeding problems more common
Extrapyramidal: Decreased tone, choreoathetosis seen. Fewer seizures and more normal development seen in these patients
Atonic: Hypotonia, brisk reflexes seen only in this type, severe cognitive delays

3. E. All of the above

Overall, approximately 60% of CP patients have mental retardation (MR). Children with spastic forms have a higher incidence of MR, which increases with the number of limbs involved. Learning disorders, deafness, and sensory impairment are also seen in these children. Impaired oromotor function may lead to difficulties with speech or aspiration pneumonia. One-third of children with CP have seizures.

4. Treatments for CP include:
   1. Braces and/or splints
   2. Intramuscular injections of botulinum toxin and/or phenol
   3. Surgery
   4. Neuraxial administration of baclofen to decrease spasticity
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

5. Septic arthritis:
   A. Is slightly more common than hematogenous osteomyelitis in children
   B. Occurs more often in infants and young children
   C. May present in infancy with fever, poor feeding, and subtle asymmetry of soft tissue folds
   D. In infants most often, involves the hip
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

6. Talipes equinovarus congenita (clubfoot):
   A. Has an incidence of 1:1000
   B. May be bilateral or unilateral
   C. May be treated conservatively until the second birthday
   D. May be effectively treated with casting in up to 70% of cases
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

7. Which of the following are associated with or characteristic of osteogenesis imperfecta?
   A. Defects in collagen formation
   B. Bones with thin cortices
   C. Deafness
   D. B-cell immunodeficiencies
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

4. E. All of the above

Treatments are directed toward maximizing motor function. Physical therapy and positioning techniques may delay the development of contractures. Bracing is most often used for foot and ankle problems. Botulinum toxin or phenol injection may temporarily decrease spasticity.

5. E. All of the above

The role of arthrotomy vs. needle aspiration in the treatment of septic arthritis is controversial although many would opt for arthrotomy in cases involving the hip joint. IV antibiotics should be given for 4–6 weeks. The differential diagnosis of an infant with fever, joint pain, and elevated WBC count includes juvenile rheumatoid arthritis, cellulitis, and toxic synovitis.

6. A. 1, 2, 3

Clubfoot is more common in males. Casting, if done early (in the neonatal period), with the casts being changed every few days, may successfully treat mild forms of talipes equinovarus (from the Latin *talus* [ankle] + *pes* [foot]; *equino* indicates the heel is elevated like a horse's and *varus* indicates it is turned inward) in about one-third of cases.

7. A. 1, 2, 3

OI is a group of disorders characterized by brittle bones. Still's classification system has six types, with varying degrees of bone fragility, different associated findings, and inheritance patterns.

Associated findings in these patients include middle-ear deafness, blue sclera, short stature, and thin skin.

8. Regarding developmental dysplasia of the hip (DDH) , formerly called congenital dislocated hips (CDH):
   1. It can be diagnosed in the newborn with the Barlow and Ortolani tests.
   2. In the newborn, it is diagnosed with plain X-rays of the hips.
   3. It is more common in girls and newborns who were born in breech presentations.
   4. It is treated with surgery followed by bracing for 6 months.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

9. Slipped capital femoral epiphysis (SCFE):
   1. Is more common in males
   2. Often presents with limp
   3. Is often accompanied by obesity
   4. Is commonly bilateral
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

10. Scoliosis:
    1. Is defined as a lateral curvature of the spine
    2. May compromise pulmonary function
    3. Has both congenital and acquired etiologies
    4. Involves rounding of the back in the thoracolumbar area of the spine
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

8. B. 1, 3

DDH occurs more frequently in firstborns. More than 20% of children with DDH have a positive family history, and it occurs six times more frequently in girls than in boys. The degrees of hip dysplasia (in order of increasing severity) are dislocatable, subluxable, and dislocated hips. X-rays are of little value as a diagnostic aid before 6 months of age since bony changes are not apparent. Ultrasonography is used in some centers and in Europe, but interpretation is difficult.

9. A. 1, 2, 3

In SCFE, the femur is rotated externally from under the epiphysis. About one-fourth of children have bilateral involvement, but not simultaneously. Obesity is commonly seen in affected children. In Legg-Calve-Perthes (LCP) disease, which is seen in younger (4–8 years) children than SCFE, there is ischemic necrosis of the proximal femoral epiphysis and later resorption. With subsequent reossification, there may be collapse of the femoral head. As a group, affected children have shorter stature and delayed bone age compared to their peers.

10. A. 1, 2, 3

Types of scoliosis include idiopathic (80%), congenital (5%), neuromuscular (10%), and miscellaneous (5%). Miscellaneous causes include genetic disorders and connective tissue diseases.

Although idiopathic scoliosis requiring correction is much more common in girls than boys, mild curves are found equally in both genders. Scoliosis curves >25° are likely to increase if the child is still growing. Curves of 40°–50° will increase even if growth is complete, and curves >75° will affect pulmonary function.

Congenital scoliosis can be complete or partial and is often associated with other congenital anomalies. Associated anomalies include renal agenesis or obstructive uropathy, congenital heart disease, or spinal dysraphism. Congenital scoliosis is seen in children with VATER or Klippel-Feil syndrome and meningomyelocele.

11. Which of the following organisms are associated with sepsis occurring after orthopedic surgery?
    A. *Haemophilus influenzae* type B
    B. *Neisseria gonorrhoeae*
    C. Staphylococci
    D. Enteric gram-negative rods

12. Juvenile rheumatoid arthritis:
    1. Has a prevalence of 60–100/1000,000
    2. Is much more common in females
    3. Is divided into three subtypes: systemic-onset, polyarticular, and pauciarticular
    4. Generally first presents in young children, before the age of 6–7 years
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

11. C

Staphylococci. While perioperative antibiotics are important in the prevention of postoperative sepsis, overuse or extended administration of antibiotics has been implicated in the increased incidence of *Clostridium difficile* toxin-related diarrhea.

12. E. All of the above

JRA is one of the more common chronic illnesses of children. This disease affects approximately 200,000 children in the USA. It commonly presents at either 1–3 years of age or in adolescence.

Girls are affected twice as frequently as boys with both polyarticular and pauciarticular forms, while the sex incidence is equal in systemic-onset disease. Affected children often have growth retardation, anemia, and chronic uveitis. Severity is based on the degree of impairment in tasks of life. Treatment includes NSAIDs; disease-modifying antirheumatic drugs (DMARDs) such as methotrexate and sulfasalazine; tumor necrosis factor (TNF) blockers such as etanercept (Enbrel) and adalimumab (Humira); immune suppressants such as abatacept (Orencia), rituximab (Rituxan), anakinra (Kineret), and tocilizumab (Actemra); steroids; and gold to decrease inflammation.

Physical therapy, occupational therapy, and surgery are used to preserve function, and counseling and nutritional support round out the picture for interventions in this chronic disease. Differential diagnosis includes systemic lupus erythematosus, Lyme disease, or Kawasaki disease.

Clinical types:

Systemic: Ill appearance associated with high fevers, irritability, rash, splenomegaly.

Polyarticular: Involvement of >5 joints for 6 months or more. Subdivided into seronegative or seropositive. More common in girls.

Pauciarticular: Peak age at 2 years; large joints are generally involved.

# Chapter 6
# Hematology/Oncology


**Thomas J. Mancuso**



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## Questions

1. Clinical manifestations of sickle cell anemia include:
   1. Hand-foot syndrome, painful often symmetrical swelling of the hands and feet
   2. Painful, vaso-occlusive crises
   3. Acute chest syndrome
   4. More frequent bacterial infections
      A. 1, 2, 3
      B. 2, 4
      C. 4 only
      D. All of the above

2. Sickle trait:
   1. Is found in approximately 8% of the African-American population in America
   2. Is found in approximately 3% of the Hispanic population in America
   3. Is found in <1% of racial groups in America other than Hispanic and African-American
   4. Is not associated with hemolytic anemia
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

3. The acute chest syndrome:
   1. Is the leading cause of death in sickle cell patients after the age of 10 years
   2. Is only seen in infants with SS disease
   3. Is a syndrome of hypoxemia, CXR infiltrates, and pulmonary infection/infarction
   4. Is best treated with nebulized bronchodilators and vigorous hydration
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

## Answers

1. E. All of the above

Sickle hemoglobin differs from normal adult hemoglobin by one amino acid substitution, glutamic acid for valine, at position 6 on the beta chain. Individuals heterozygous for HbS are resistant to falciparum malaria. Affected homozygous SS individuals have severe hemolytic anemia since the sickled cells are poorly deformable and brittle. Clinical manifestations are rarely seen before 6 months of age with the hand-foot syndrome often seen in 1–2-year-old children.
SS disease is a chronic hemolytic anemia with associated crises such as splenic sequestration crises, aplastic crises, and vaso-occlusive crises. Pain crises are the most common type of vaso-occlusive crisis. Below is a list of common clinical manifestations seen in SS disease:

Cerebrovascular accidents, acute chest syndrome, priapism gallbladder disease, hematuria
Renal concentrating defect, cardiomyopathy, infections
A variety of psychological problems including school failure and depression

2. E. All of the above

Individuals heterozygous for HbS typically have no signs or symptoms of sickle cell disease. Rarely, these individuals have painless hematuria. The diagnosis of sickle trait is made with hemoglobin electrophoresis. The RBCs in people with sickle cell trait contain 30–40% HbS; thus sickling does not occur under normal circumstances. In unusual conditions such as shock, very high altitude, or extremely demanding exercise, a vaso-occlusive crisis may occur.

3. B. 1, 3

This clinical syndrome may occur as a complication of postoperative atelectasis. Initially, the child may not appear severely ill, but the condition can progress rapidly. Early detection of any pulmonary compromise in a child with sickle cell disease, followed by vigorous treatment (CPT, incentive spirometry, etc.), is essential given the high mortality of children who develop the syndrome.

4. Regarding infection in children with sickle cell disease:
   1. Osteomyelitis is relatively common, particularly with salmonella.
   2. Encapsulated organisms such as pneumococcus and *Haemophilus influenzae* type b are common etiologic agents.
   3. Serious infection is particularly common in the first 5–6 years of life.
   4. With the newer vaccines, infections are no longer a problem for these children.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

5. Therapy for vaso-occlusive crises includes:
   1. Adequate analgesia
   2. Antibiotics
   3. Adequate hydration
   4. Immobility
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

6. Acute lymphoblastic leukemia (ALL) :
   1. Is the most common leukemia in childhood
   2. Has its peak incidence in children at 10 years of age
   3. May relapse in the bone marrow or CNS
   4. Is treated with total body irradiation
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

7. Chemotherapeutic agents used in the treatment of low-risk ALL include:
   1. Prednisone
   2. Vincristine
   3. Intrathecal methotrexate (MTX)
   4. Bleomycin
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

4. A. 1, 2, 3

The polyvalent pneumococcal vaccines currently available are poorly immunogenic in children under the age of 5 years. Prophylactic penicillin is effective in preventing serious pneumococcal infections in these younger children. Full immunization status is especially important in children with sickle cell disease.

5. B. 1, 3

Children with frequent pain crises are difficult to assess. Children afflicted with this chronic condition often have a flat affect making observer assessment of their degree of discomfort unreliable. Analgesics should not be withheld if the child reports pain, however. Treatment is directed toward preventing complications such as the acute chest syndrome, uncovering etiologies such as infections (osteomyelitis, pneumonia), and providing adequate hydration, nutrition, and comfort.

6. B. 1, 3

ALL occurs with slightly greater frequency in boys than girls. It is subclassified on the basis of immunologic, cytogenetic, and molecular genetic markers. The median ages for the various types of ALL range from <1 year to 7 years. Presenting signs and symptoms are usually nonspecific and include anorexia, lethargy, and irritability. Pallor, bleeding and fever, and signs of bone marrow failure prompt medical attention. There are approximately 200 new cases of ALL/year. Based on survival, ALL is characterized into standard and high risk. Standard risk characteristics in addition to cytogenetic and immunologic factors include age 2–9, female gender, white race, absence of adenopathy, WBC count $<10 \times 10^9$, and absence of CNS disease.

7. A. 1, 2, 3

Without treatment of sanctuaries, relapses in the CNS and testicles were common. Induction generally consists of vincristine, prednisone, and asparaginase, accompanied by intrathecal methotrexate, hydrocortisone, and Ara-C. CNS irradiation is effective in minimizing CNS disease, but it also produces late neuropsychiatric effects.

8. Hodgkin's disease:
   1. Has a bimodal age distribution with peak incidences in the second and fifth decades of life
   2. Commonly presents with painless enlarged cervical lymph nodes
   3. Often causes enlarged mediastinal lymph nodes which may cause cough or other respiratory symptoms
   4. Is sensitive to both chemotherapy and radiation
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

9. Acute cervical adenitis, inflammation of one or more lymph nodes in the neck, is caused by:
   1. *Staphylococcus aureus*
   2. Atypical mycobacteria
   3. Group A streptococcus
   4. Adenoviruses
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

10. Relatively common noninfectious causes of cervical adenitis include:
    1. Hodgkin's disease
    2. Non-Hodgkin's lymphoma
    3. Neuroblastoma
    4. Hemangiomas
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

8. E. All of the above

The lymphadenopathy seen is usually in the cervical area, but axillary and inguinal nodes are sometimes part of the presentation. Hepatosplenomegaly is rare. Fever, weight loss, and night sweats are seen. Staging is important in prognosis and in determining treatment. Disease-free survival rates from 60% to 90% are achieved, based on the staging at diagnosis.

9. A. 1, 2, 3

With chronic infection, signs such as erythema, warmth, and fluctuance are absent. Nodes associated with malignancy are firm and may be fixed to underlying structures or overlying skin. Another infectious cause of cervical adenitis is Kawasaki's disease.

10. B. 1, 3

The most common presentation of Hodgkin's disease is painless, firm adenopathy. Cervical or supraclavicular nodes are commonly involved. Significant enlargement of the nodes in the anterior mediastinum leads to cough, respiratory distress, and cardiovascular embarrassment. This mediastinal involvement is seen the most in older children with Hodgkin's disease. Hodgkin's disease is staged using the Ann Arbor system:

Stage I disease involves a single LN area or a single extralymphatic site.
Stage II is more extensive but on one side of the diaphragm.
Stage III disease is seen on both sides of the diaphragm.
In stage IV, the malignancy is disseminated, involving greater than 1 extralymphatic site.

Neuroblastoma has a very varied presentation. Abdominal pain and mass are a common presentation, but in children with localized disease, adenopathy is also seen.

11. Neuroblastoma, the most common extracranial tumor of childhood and the most frequently diagnosed cancer in infants, presents in a variety of ways including:
    1. With a hard painless mass in the neck
    2. As an abdominal or thoracic mass
    3. With bone pain from skeletal and bone marrow metastases
    4. With seizures from CNS metastases
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

12. Wilms' tumor of the kidney:
    1. Commonly presents with an asymptomatic flank mass
    2. Is often diagnosed in children at 2–4 years of age
    3. Is associated with hypertension in up to 60% of cases
    4. Often metastasizes to the lungs
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

13. Manifestations of graft versus host disease (GVHD) in children include:
    1. Maculopapular rash
    2. Generalized erythroderma with bullae and desquamation
    3. Liver dysfunction manifested by elevated bilirubin
    4. GI disturbances manifested primarily by diarrhea
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

14. Brain tumors:
    1. Are the most common solid tumor in children
    2. In children between the ages of 2 and 12 years are most often located in the posterior fossa
    3. May present with signs of increased intracranial pressure
    4. May present with focal neurological signs
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

11. A. 1, 2, 3

This tumor has a variable presentation since it may develop at any site of the sympathetic nervous system. It is generally discovered as a mass or masses on exam or on a radiologic scan. Treatment varies depending on the stage at diagnosis, as does the chance for survival. Overall cure rate, with all stages included, is approximately 50%.

12. E. All of the above

This tumor accounts for most renal cancer in children. The asymptomatic mass is often discovered by a parent. Surgical removal is indicated even in cases where pulmonary metastases have occurred. With chemotherapy following surgery, survival ranges from 50% to <90% depending on histology and stage.

13. E. All of the above

GVHD occurs when there is a disparity of histocompatibility antigens between the recipient of a bone marrow transplant and the donor marrow. Donor T lymphocytes damage various tissues in the host especially the skin, GI tract (mucositis), and liver. There are acute and chronic forms of the disease with chronic having a worse prognosis.

14. E. All of the above

Brain tumors are the second most common reported malignancy in children and adolescents. Surgery and radiation are the mainstays of treatment. CNS tumors may present with headache, worse in the morning. These tumors are classified by location (infratentorial, supratentorial) and histology.

15. Regarding posterior fossa tumors:
    (a) They tend to present with symptoms of raised intracranial pressure.
    (b) The most common histology, cerebellar astrocytoma, also has the best prognosis.
    (c) Morning headache with associated vomiting may be part of the presentation.
    (d) Medulloblastoma is the second most common posterior fossa tumor.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

15. E. All of the above

Infratentorial (posterior fossa) tumors predominate in children 4–14 years of age. Nearly one-half of pediatric brain tumors arise in the cerebellum, frequently astrocytomas and medulloblastomas. In another classification system, medulloblastomas, which are poorly differentiated, very malignant tumors, are termed primitive neuroectodermal tumors (PNET).

# Chapter 7
# Surgery


**Thomas J. Mancuso**



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## Questions

1. Which of the following organisms are associated with sepsis occurring after abdominal surgery?
   A. *Haemophilus influenzae* type B
   B. *Neisseria gonorrhoeae*
   C. Staphylococci
   D. Enteric gram-negative rods

2. Gastroschisis differs from omphalocele in the following way(s):
   1. Gastroschisis is usually a 2–4 cm defect, often with a right paramedian location.
   2. In infants with gastroschisis, the bowel is not covered by membranes.
   3. Omphalocele is associated with a greater incidence of non-GI anomalies such as congenital heart disease, bladder exstrophy, and/or cloaca.
   4. A silo is often used in closure of larger defects.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

3. Which is the most common type of esophageal atresia?
   A. Esophageal atresia (EA) with a posterior tracheoesophageal fistula (TEF) near the carina
   B. "H"-type EA/TEF
   C. Esophageal connection to the trachea with a second, more distal TEF
   D. Esophageal connection to the trachea without a distal TEF

## Answers

1. D. Enteric gram-negative rods

The administration of prophylactic antibiotics in the OR and in the perioperative period is an important measure in reducing the incidence of infection. Overuse of antibiotics has been implicated as a cause for the development of *Clostridium difficile* toxin-associated diarrhea.

2. E. All of the above

Omphalocele is a midline defect of variable size involving the omphalos (Greek: "navel"). Omphalocele is associated with chromosomal abnormalities, other GI defects (35%), cardiac defects (20%), and also cloacal exstrophy. Only 10% of patients with omphalocele are born preterm. Omphalocele is part of the Beckwith-Wiedemann syndrome (macroglossia, hyperinsulinism, hypoglycemia, and gigantism). Omphalocele results when the intestines fail to return into the abdomen from the umbilical coelom. With omphalocele, the bowel is covered with membranes, decreasing fluid losses. In gastroschisis, a part of the small intestine herniates through the abdominal wall.

Sixty percent of patients with gastroschisis are born prematurely. A few affected patients have jejunal atresia, but other anomalies are not seen with this condition. Repair of either defect may involve a "silo" if the extruded intestines do not fit into the smaller abdominal cavity.

3. A. Esophageal atresia (EA) with a posterior tracheoesophageal fistula (TEF) near the carina

TEF is the failure of the linear division of the trachea and esophagus during embryogenesis. The most common type (85–90%) is a proximal blind pouch with a distal TEF. All patients present with aspiration at birth with respiratory distress and inability to handle oral secretions. Mortality is approximately 3% in term infants but can be much higher in preterm newborns and in those with other congenital anomalies. Forty percent of infants born with TEF have associated anomalies, with cardiovascular anomalies being seen most often. TEF is also seen as part of the VATER (vertebral anomalies, anal atresia or arterial anomalies, TEF, renal anomalies) or VACTERL (vertebral anomalies, anal atresia or arterial anomalies, TEF, renal anomalies, limb anomalies) associations.

4. The VACTERL complex includes a tracheoesophageal anomaly and:
   1. PDA, ASD, or VSD
   2. Renal defects
   3. Abnormalities of the bones of the forearm
   4. Spinal dysraphism
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

5. Regarding acute appendicitis in children:
   1. In many children younger than 2 years of age, the appendix is found perforated at operation.
   2. More males than females develop acute appendicitis.
   3. It is unusual for the child with acute appendicitis to have an appetite.
   4. Among school-aged children, the diagnosis is more often missed in girls than in boys.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

6. Midgut volvulus:
   1. Occurs when the bowel twists upon itself
   2. Occurs when *incomplete* intestinal rotation leads to a shortened mesentery
   3. Leads to vascular compromise of the bowel
   4. Is seen in over 60% of neonates with malrotation
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

7. Duodenal atresia:
   1. Often presents with vomiting which may or may not be bilious
   2. Is often accompanied by other intestinal obstructions in both the small and large intestine
   3. Is associated with trisomy 21
   4. Generally does not present until 1–2 months of life
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

4. E. All of the above

The VATER association includes vertebral anomalies, imperforate anus or arterial anomalies, TEF, and renal anomalies. A single umbilical artery is often seen. In VACTERL, the L stands for limb anomalies. An association does not have a single known etiology. The evaluation of an infant with TEF should include a search for the anomalies in the VACTERL association.

5. A. 1, 2, 3

The annual incidence of acute appendicitis is 4:1000. In adolescent girls, the diagnosis is more difficult because other causes of abdominal pain such as ovarian cysts, ovulatory pain, menstrual pain, and pelvic inflammatory disease mimic appendicitis. Other diagnoses in the differential include gastroenteritis, mesenteric adenitis, inflammatory bowel disease, RUL pneumonia, and urinary tract pathology.

6. E. All of the above

In 70% of patients with malrotation and volvulus, the presentation is within the neonatal period, and in half of these, the presentation is in the first 10 days of life. The balance of cases can present at any time, even into adulthood. Malrotation is twice as common in boys as girls. Presentation includes distention and bilious vomiting. X-ray studies (plain film or upper GI contrast study) confirm the diagnosis. In malrotation, the duodenum is seen in an abnormal position, with the duodenojejunal junction located to the right of the spine.

7. B. 1, 3

Duodenal atresia occurs in 1:20,000 live births. Common findings in affected infants include abdominal distention and jaundice. Maternal polyhydramnios is also often found. Associations in addition to trisomy 21 are congenital heart disease, TEF, and renal anomalies.

8. Intussusception:
   1. Has a peak incidence in infants less than 1 year of age
   2. Presents with colicky abdominal pain, bloody (currant jelly) stools, and vomiting
   3. May be reduced with a carefully performed enema
   4. Is nearly always caused by a "lead point" such as a polyp or duplication
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

9. Hirschsprung's disease, the absence of normal enteric ganglionic neurons:
   1. Has a much higher incidence in Caucasians
   2. Is limited to the rectum and sigmoid in over 50% of cases
   3. Is initially managed with stool softeners since some cases spontaneously resolve
   4. Rarely involves not only the rectum and sigmoid but the entire colon
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

10. Hirschsprung's disease can be diagnosed by:
    1. Rectal manometry
    2. Surgical rectal biopsy
    3. Suction biopsy
    4. Barium enema
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

11. Other than organomegaly, which of the following may cause abdominal distention in the neonate?
    1. Pneumoperitoneum
    2. Intestinal obstruction
    3. Ascites
    4. Pyloric stenosis
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

8. A. 1, 2, 3

Intussusception is seen in children from the age of 3–18 months. The incidence varies from 0.5 to 4/1000 live births. In almost all children under 1 year of age, no clear cause is found. The nonspecific signs of vomiting and colicky pain are nearly always part of the presentation. Later in the course of the illness, fever and lethargy are seen and finally the child may pass a currant jelly stool. A carefully done barium enema is used as a diagnostic and therapeutic tool. Excessive pressure in the performance of the enema will perforate the bowel. Up to 50% of patients with intussusception are successfully treated with hydrostatic reduction.

9. C. 2, 4

Hirschsprung's disease is a functional obstruction of the colon or rectum that results from failure of migration of ganglion cells in the developing colon. It is the cause for up to 25% of all cases of bowel obstruction in the newborn and is seen more often in males. The aganglionic segment does not permit normal colonic motility. More than 80% of cases involve only the rectum and a small part of the colon. Management is surgical since the aganglionic segment is permanently contracted. Generally, a colostomy is performed at the level of normal innervation (the so-called leveling colostomy) with a later colonic or ileal pull-thru.

10. E. All of the above

The diagnosis should be suspected in any newborn that does not pass meconium within the first day of life. A frozen section can be done with the biopsy material, which is stained for acetylcholine to identify abnormal nerve trunks. H&E stains confirm the absence of ganglion cells.

11. A. 1, 2, 3

Hepatomegaly and hepatosplenomegaly are possible causes of abdominal distention in the newborn. Pneumoperitoneum is usually a result of GI tract perforation, often in preterm newborns. Among the causes of neonatal ascites, urinary ascites is the most common, followed by cardiac and idiopathic. Pyloric stenosis is an incomplete obstruction at the gastric outlet not associated with ascites.

12. Abdominal masses in the newborn:
    1. Are generally not malignant
    2. Are retroperitoneal in approximately 66% of cases
    3. If retroperitoneal, are most often renal in origin
    4. If due to a tumor, are most likely abdominal teratomas
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

13. Which of the following have been implicated as having a role in the development of necrotizing enterocolitis (NEC)?
    1. Intestinal ischemia
    2. Bacterial colonization of the bowel
    3. Feeding
    4. Multiple births
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

14. Which of the following are associated with intestinal ischemia?
    1. Polycythemia
    2. Umbilical vessel catheterization
    3. Congestive heart failure
    4. An open patent ductus arteriosus
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

15. Patients with extrahepatic biliary atresia (incidence = 1:10–15,000 live births):
    1. Will be cured for life if a Kasai operation is done within the first 3 weeks of life
    2. Are generally well initially and then develop jaundice at 3–6 weeks of age
    3. Should be started on phenobarbital to induce liver enzymes in the remaining normal liver
    4. Will likely die within the first year of life without surgical intervention
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

12. E. All of the above

Approximately 10–15% of abdominal masses in the newborn are due to malignant tumors. Types of renal masses seen in newborns include hydronephrosis, polycystic disease, renal vein thrombosis, Wilms' tumor, and mesoblastic nephroma. Other retroperitoneal masses seen in this age group include neuroblastoma, ganglioneuroma, and sacrococcygeal teratoma.

13. A. 1, 2, 3

NEC is the most common gastrointestinal emergency in the infant. The incidence ranges from 1% to 4% of all NICU admissions, with a higher incidence in infants with birth weights <1000 g. The most commonly implicated etiologic factors are an ischemic insult to the gut and the presence of intraluminal bacteria or viruses and substrate (formula or milk). Approximately 93% of infants who develop NEC have been fed enterally. Because of the role of bacteria in NEC, antibiotics are often part of the therapy.

14. E. All of the above

Mesenteric blood flow in the newborn is affected by a variety of factors in addition to those mentioned. During hypoxia, the so-called diving reflex shunts blood from the mesenteric, renal, and peripheral vascular systems to the brain and heart. Polycythemia and also exchange transfusions have been implicated in intestinal ischemia. Other possible etiologies for NEC are RDS, hypotension, hypothermia, and birth asphyxia. The presentation of NEC may include abdominal distention, vomiting and gastric residual, lethargy, hypotension, apnea, and temperature instability.

Lab findings include pneumatosis intestinalis on X-ray, thrombocytopenia, blood and/or reducing substances in the stool, and metabolic acidosis.

15. C. 2, 4

This condition is defined as atresia or hypoplasia of any part of the extrahepatic biliary system.

The most common form includes atresia up to the porta hepatis and even intrahepatic ducts. Approximately 15% of affected children have other defects. Clinical presentation includes jaundice in the second to third week of life; acholic stools; enlarged, hardened liver; and splenomegaly. Conjugated bilirubin is elevated along with alkaline phosphatase, gamma-glutamyl transferase, and transaminases.

16. In pyloric stenosis:
    1. The child develops metabolic alkalosis due to continued vomiting of gastric contents.
    2. There is non-bilious vomiting which may contain “coffee ground” material.
    3. The child may develop hypochloremia.
    4. The diagnosis is best made with a barium swallow.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

17. Pyloric stenosis, the most frequently occurring cause of gastric obstruction in the newborn:
    1. Is seen in approximately 1:2500 live births
    2. Is equally common in all races and ethnic groups
    3. Is seen with the same frequency in males and females
    4. Is seen in infants as young as 2 weeks and as old as 12 weeks
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

18. Regarding children in MVAs who have a history or physical exam indications of lap belt injury:
    1. These injuries are most common in children under the age of 1 year.
    2. There may also be damage to the spinal cord in these children.
    3. Diagnostic peritoneal lavage is always indicated.
    4. Hollow viscus injury may not be apparent for 12–24 h.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

16. A. 1, 2, 3

Pyloric stenosis is much more common in males. It is seen in 1:150 males and 1:750 females. The presentation includes progressive, relentless vomiting generally starting between the second and fourth week of life with a range of 2–12 weeks. Typical electrolyte abnormalities seen in infants with pyloric stenosis include hypochloremia, hypokalemia, and hyponatremia along with azotemia. The severity of the child's condition is graded by the degree of hypochloremia. An infant with more severe dehydration will have, in addition to the metabolic acidosis, a paradoxical aciduria. The kidney, in an effort to maintain intravascular volume, absorbs as much sodium as possible, but the only anion available to absorb is bicarbonate. Thus, the urine is acidic despite the systemic alkalosis. Prior to surgical repair, the child should be adequately hydrated and the electrolyte abnormalities corrected. Given the nature of the pathology, gastric outlet obstruction leading to intractable vomiting, administration of contrast is a poor way to make the diagnosis. Ultrasound or air contrast plain X-rays are more current diagnostic tools.

17. D. 4

The incidence of pyloric stenosis is 1:2500 live births. It is seen predominately in whites and is most common in firstborn males. The average age of onset is 3–4 weeks with a range of 2–12 weeks. Interestingly, this anomaly is not seen at birth, and in cases where it is managed medically, the hypertrophic pylorus eventually (after 4–6 weeks) returns to normal and the child stops vomiting, all without surgical intervention. Pyloric stenosis is a common reason for surgery in the neonatal period.

18. C. 2, 4

Injuries to the pancreas can result from either lap belt injury or bicycle handlebars. Injury to the pancreas is difficult to diagnose. Elevations in amylase and lipase may not be seen until 1–2 days after the injury. Rapid deceleration while in a lap belt may also damage the intestines with perforation or even transection possible. If there is intestinal damage, in addition to a bruise over the abdomen in the area of the lap belt, back pain may be a symptom.

19. In the evaluation of the child who has suffered blunt abdominal trauma, organ damage may be indicated by:
    1. Left shoulder pain
    2. Hematuria
    3. Flank ecchymosis
    4. Bilious emesis
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

20. Gastroesophageal reflux (GER) in infants and young children can present with:
    1. Recurrent pneumonia
    2. Irritability
    3. Wheezing, stridor, or hoarseness
    4. Apnea
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

21. Treatment(s) for GER include(s):
    1. Thickened, small, and frequent feeds
    2. Magnesium sulfate to increase gastric pH
    3. H-2 receptor blockade
    4. Avoidance of high-carbohydrate meals
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

22. Esophageal foreign bodies in children:
    1. Are best diagnosed with a barium swallow
    2. Often lodge at or just below the cricopharyngeus muscle
    3. Always require removal if they pass into the stomach
    4. May cause stridor in infants
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

19. E. All of the above

The spleen is the abdominal organ most often damaged following blunt abdominal trauma in children. Kehr's sign, left shoulder pain resulting from pressure on the left upper quadrant, is suggestive of splenic injury. The severity of splenic injury is graded by CT from 1 to 5. Grade 1 is a tear in the capsule, while grade 5 indicates a completely ruptured spleen. Renal damage following blunt abdominal trauma is relatively common in children. Abdominal CT with renal contrast will make the diagnosis in most cases. Flank pain and bruising and urinalysis with blood and protein should raise the suspicion of renal damage.

20. E. All of the above

GER must not be confused with other causes of regurgitation in the newborn/infant such as pyloric stenosis, duodenal stenosis, annular pancreas, malrotation, or any of a host of metabolic diseases. Contrast studies and/or 12–24-h pH probe studies may confirm the diagnosis. Pneumonia and wheezing develop with aspiration of refluxed gastric contents, irritability is due to the pain of reflux esophagitis, and apnea is a possible reaction to the presence of aspirated gastric contents in the trachea or larynx.

21. B. 1, 3

Medical treatment of GER is directed to lowering the pH of the gastric contents and decreasing the amount of reflux. In the well, thriving child with a small amount of post-feeding reflux, observation and reassurance are all that is needed. In more severe cases, placing the child at an angle (30° head up) after feeding may limit the reflux as will the institution of frequent small feedings instead of larger ones.

22. C. 2, 4

A barium study of the esophagus may be useful in the evaluation of upper airway obstruction, often demonstrating posterior esophageal compression from a vascular ring. The cricopharyngeus muscle, located high in the esophagus, often stops a swallowed esophageal foreign body from progressing further. A foreign body which passes into the stomach will likely be passed through the entire GI tract, so retrieval is often not undertaken unless it is indicated by the specific nature of the foreign body (such as an open safety pin).

23. Extraintestinal manifestations of inflammatory bowel disease (IBD) include:
    1. Growth retardation
    2. Peripheral arthritis
    3. Anemia
    4. Reactive airway disease
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

24. Common findings in patients with IBD (ulcerative colitis or Crohn's disease) include:
    1. Iron deficiency anemia
    2. First-degree heart block
    3. Hypoalbuminemia
    4. Stool cultures positive for various enteric pathogens
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

25. Cleft lip:
    1. May be unilateral or bilateral
    2. Occurs with and without cleft palate
    3. Occurs in 1:600–1000 live births
    4. Varies from being a small notch in the vermilion border to a complete separation
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

26. Problems seen in children with cleft palate ± cleft lip include:
    1. Otitis media
    2. Feeding difficulties
    3. Malpositioned teeth and dental decay
    4. Problems with phonation
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

23. A. 1, 2, 3

Inflammatory bowel disease (IBD) is a term given to both ulcerative colitis and Crohn's disease. Ulcerative colitis is a chronic inflammatory illness limited to the mucosa and submucosa of the colon and rectum. The risk for cancer is estimated to be 10–20% per decade after the first 10 years of disease. Crohn's disease is a transmural inflammation involving any and all portions of the GI tract. Management of IBD centers on nutritional support, immunosuppression, and surgery. Extraintestinal manifestations of these illnesses differ, but growth retardation and anemia, due to poor nutrition and GI blood loss, are regularly seen. Arthritis, arthralgias, and various skin manifestations, such as erythema nodosum, are also seen in both diseases.

24. B. 1, 3

The hypochromic, microcytic anemia and hypoalbuminemia seen in IBD are due to both iron losses through subtle GI bleeding and to poor nutrition. Infection of the GI tract is generally not part of the problem in IBD. Except in cases of dehydration, electrolytes are generally within normal limits.

25. E. All of the above

Recurrence patterns of this problem do not suggest a simple pattern of inheritance. Isolated cleft palate appears to be a separate entity from cleft lip with or without cleft palate. Isolated cleft palate has an incidence of 1:2500 live births. Cleft lip with cleft palate is more common that either is seen in isolation. The frequency is higher than 1:1000 in Native Americans, Japanese, and Chinese people and lower in African-Americans. Other anomalies are seen in up to 25% of all patients with cleft lip, palate, or both and more often in children with bilateral cleft lip. The Robin malformation sequence includes cleft palate as well as micrognathia and glossoptosis. Up to 20% of patients with the Robin sequence have cardiac anomalies such as ASD, VSD, or PDA.

26. E. All of the above

Cleft lip and palate often occur as isolated anomalies. Various techniques and equipment are available for feeding these infants, and no single solution is suitable for all. The management team for these infants should include a maxillofacial surgeon, audiologist, speech pathologist, otolaryngologist, exodontist, and geneticist.

27. Chiari type II malformations, also called Arnold-Chiari malformations:
    1. Are seen in nearly all patients with meningomyelocele
    2. May be asymptomatic
    3. Can be a cause of headache, particularly with coughing or straining
    4. Can be associated with vocal cord paralysis
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

28. Tests used to confirm the diagnosis of Chiari II malformation include:
    1. Sleep studies
    2. PET scan
    3. EEG
    4. MRI
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

27. E. All of the above

A Chiari II malformation is a bony defect that includes caudal displacement of the cerebellar vermis, fourth ventricle, and lower brainstem below the plane of the foramen magnum. Chiari II malformations are often asymptomatic. Presentation is often a headache, in particular after cough or with flexion/extension of the neck, lower cranial nerve signs, and, if the Chiari malformation has led to development of a syrinx, long tract signs such as lower extremity weakness.

28. D. 4

Chiari malformations are graded by the distance the CNS structures (cerebellar tonsils) extend below the foramen magnum on MRI, i.e., >6 mm in children <10 years old and >5 mm in older children. In an MRI study of adults, the prevalence of Chiari II using these diagnostic criteria was 0.5–1.0% and >70% of these patients were asymptomatic. A syrinx was noted in 30% of these subjects.

# Part II
# Consultations in Pediatric Anesthesia

# Chapter 8
# Prematurity/Extreme Prematurity


**Thomas J. Mancuso**

A 2-week-old male, 900 g, born at 27 weeks' gestational age is scheduled emergently for exploratory laparotomy for free air in the abdomen.
VS: HR = 185/min; BP = 50/30 mmHg; right hand SpO2 = 92%; T = 36.2 °C. His hemoglobin is 13.0 g/dL. He is intubated with a 2.5 mm oral endotracheal tube.
Ventilator settings: FiO2 = 0.5; RR = 30/min; PEEP = 3 cm H2O; PIP = 22 cm H2O.



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## Preoperative Evaluation

### Questions

1. Is this patient premature? Why? The progress note by the neonatologist refers to this infant as ELGAN. What does this term mean? What organ systems are you concerned about as you plan your anesthetic for this child?

2. Does this patient have surfactant in his lungs? If not, when will it develop? Would the administration of artificial surfactant influence your anesthetic plan?

## Answers

1. Infants born before the 37th week of gestation are considered premature [1]. The term ELGAN means extremely low gestational age newborn and is replacing the term for birth weight used to classify preterm newborns (low birth weight (LBW), extremely low birth weight (ELBW), etc.). The Committee on Fetus and Newborn (COFN) of the AAP made this change since morbidity correlates with gestational age (GA) more closely than with birth weight [2]. This infant is ELGAN, with a GA of 27 weeks, a postnatal age of 2 weeks, and a postconceptual age of 29 weeks. The third trimester is the time when most organ systems mature. Of particular importance is the immaturity of the pulmonary, central nervous, renal, and hepatic systems. Gas exchange and management of mechanical ventilation require exquisite attention to detail in these tiny patients. CNS immaturity and the possible deleterious effects not only of hypoxia and metabolic derangements but also of the anesthetic, hypnotic, and analgesic agents themselves are an evolving issue. Preterm infants do not maintain fluid and electrolyte balance well, requiring care in the administration of IV fluids and electrolytes. Liver immaturity, both in synthetic and metabolic capacities, can result in much longer duration of action of IV agents.

2. An infant born at 27 weeks will almost certainly be deficient in pulmonary surfactant and develop respiratory distress syndrome (RDS) [3]. Surfactant is produced by type II pneumocytes. The amount and composition of the surfactant change throughout gestation. Lamellar bodies are first seen in type II pneumocytes 20–24 weeks' gestation. There is progressive accumulation of saturated phosphatidylcholine in the lung tissue until term. The surfactant present in the lungs of term newborns is composed of 50% saturated phosphatidylcholine, 20% unsaturated phosphatidylcholine, 14% other lipids, 8% phosphatidylglycerol, and 8 % surfactant proteins (SP-A through SP-D). The immature lung has decreased surfactant function, much less phosphatidylglycerol, and more phosphatidylinositol. Administration of artificial surfactant following delivery is very beneficial, decreasing surface tension in the alveoli as natural surfactant does in the term newborn. Practice varies among neonatologists with regard to administration of surfactant prophylactically in the delivery room or when needed later in the ICN. Generally no more than two doses are administered. If surfactant were administered, it should be expected that the compliance of the lungs would increase to closer to that seen in term newborns. When ventilating for a newborn treated with surfactant, it is important to consider this improved compliance. There are several manufacturers of surfactant. The primary difference is whether or not the product is derived from another species or the product is synthetic. A Cochrane review concluded that there is "some evidence that animal derived surfactant extract leads to better outcomes in babies with respiratory distress syndrome compared to synthetic surfactants that do not contains proteins" [4].

3. How can you evaluate the status of the central nervous system (CNS)? What signs and symptoms would you look for with regard to intraventricular hemorrhage? What factors contribute to the onset of intraventricular hemorrhage? How will it affect your anesthetic management?

4. How can you diagnose persistent patency of the ductus arteriosus? Is this significant for anesthetic management? Why/why not?

5. Does this patient need a workup for D.I.C.? Why? Will an assessment of fibrin split products help? Why/why not?

3. Periventricular-intraventricular hemorrhage (IVH) is a common occurrence in the preterm newborn and the most serious CNS lesion encountered in the newborn period [5]. It is a major cause of death in preterm newborns. In at least 90% of cases, the hemorrhage occurs in the first week of life. The incidence and severity of periventricular-intraventricular hemorrhage occurring vary inversely with gestational age. IVH grades of severity go from I to IV based upon the radiographic appearance of the extent of the hemorrhage, from an isolated germinal matrix hemorrhage to intraventricular and parenchymal hemorrhage:

   Grade I: Subependymal and/or germinal matrix hemorrhage
   Grade II: Subependymal hemorrhage also into lateral ventricles
   Grade III: Grade II plus ventricular enlargement
   Grade IV: Intraparenchymal hemorrhage

   Clinically, the occurrence of an IVH may be suggested by sudden cardiovascular instability or, if the IVH itself goes unnoticed, hydrocephalus, which may occur later. Numerous causes for IVH have been proposed, but it is often difficult to establish a definite cause and effect relationship. Loss of autoregulation of cerebral blood flow in these patients and rapid changes in cerebral blood flow and pressure are likely involved. Possible specific causes include neonatal asphyxia with low blood pressure, rapid volume expansion, and changes in serum osmolarity, abnormal coagulation, hypoxemia, hypercarbia, and large swings in systemic BP with excessive agitation in resisting mechanical ventilation.

4. PDA is nearly a normal finding in the preterm [6]. In the fetus, the ductus arteriosus is essential for adequate circulation and oxygen delivery. In postnatal life, as pulmonary vascular resistance decreases, a large PDA may lead to inadequate forward systemic flow and CHF from excessive pulmonary blood flow. Clinically, bounding pulses, tachypnea, cardiomegaly, and signs of pulmonary overcirculation are seen when the ductus is opened. The diagnosis can be confirmed with 2D echo with color Doppler. A functionally closed ductus in a patient such as the one in this case can be reopened by excessive fluid administration. This makes fluid administration in these critically ill infants challenging. Excessive fluid administration can lead to significantly worsened gas exchange and also decreased systemic flow, whereas inadequate preload will also lead to decreased LV output.

5. NEC is often associated with a coagulopathy. Thrombocytopenia is commonly seen as NEC worsens. In patients suspected of having NEC, serial platelet counts are followed as a measure of disease severity. The laboratory diagnosis of DIC can be difficult since, in the newborn, several tests of coagulation may be outside of the reference range [7, 8]. For example, D-dimers are often found in preterm newborns without DIC. In the clinical setting of NEC, thrombocytopenia, microangiopathic hemolytic anemia, and prolonged PT are indicative of DIC. Treatment of this infant's coagulopathy will involve transfusion of PRBCs, platelets, and also coagulation factors either in the form of FFP or cryoprecipitate. In the NICU, exchange transfusion may also be undertaken.

## Intraoperative Course

### Questions

1. Does this patient need an arterial line? Why/why not? A central line? Why? What site will you use to monitor temperature? Why? Where does temperature change first? Last? Will you monitor glucose levels? Why? How often?

2. Should you give atropine prior to induction? If this infant arrived in the OR with an endotracheal tube in place, on nasal cannula O2, would you give atropine prior to an "awake" intubation? Why/why not? Would pancuronium do just as well?

3. How would you ensure temperature homeostasis? What is your choice of fluid management? Why? What if hyperalimentation is running? Would you discontinue and change over to a D20 solution? D10 solution? Not change? What are the effects of narcotic anesthetic techniques in this age group? Is MAC different in this age group? In what way? Is ketamine a choice?

4. What muscle relaxant would you choose? Why? Is there any specific advantage to cisatracurium?

**Answers**

1. Direct arterial monitoring would be very helpful in this case, but technically difficult and associated with complications. Radial arterial cannulation is preferred. Femoral arterial cannulation may be technically less difficult, but circulation to the distal leg may be affected in a patient this small, even with a 22 g catheter. An alternative is a CVL through which blood products can be given and which also can be used for sampling. With a functioning oximeter, the need to measure arterial PaO2 is lessened. In many cases, pre- and postductal oximeters are used. Temperature monitoring is very important given the ease with which hypothermia may occur and the problems hypothermia will cause. Rectal measurement gives a good indication of central temperature, but care must be taken not to perforate the delicate rectal mucosa during insertion. An important part of intraoperative care of this infant will be frequent monitoring of serum glucose and electrolytes, platelet count, hemoglobin, pH, and blood gases. With the administration of either the existing hyperalimentation or D10, serum glucose will likely be maintained. A more complete discussion of glucose management is below in note 1 of "Additional Topics."

2. Atropine will help maintain a high infant heart rate during the induction and the case, but hypoxemia will still lead to bradycardia. Pancuronium also has a vagolytic effect on the heart rate but does not decrease oral secretions, as does atropine.

3. Temperature maintenance begins prior to the start of the case. The OR temperature should be turned up prior to the arrival of the infant. Inspired gases can be humidified and warmed. When this is done, water "rainout" must be drained out of the circuit, and not into the infant's lungs. Additionally, the temperature of the inspired gases must be monitored and kept below 39 °C to avoid burning the infant [9, 10]. Maintenance glucose, as D10, at 100–120 mL/kg/day, must be given throughout the case, preferably through a separate IV, while blood products and other crystalloids are given through another IV. Any hyperalimentation already running should be continued at the same rate, and once serum glucose is checked, the infusion rate can be carefully lowered. Opioid analgesics will decrease the stress response and help minimize postoperative catabolism, and in some cases high-dose opioid analgesia has decreased mortality. MAC varies with age, although there is no data on the preterm weight newborn, particularly the low birth weight (LBW), extremely low birth weight (ELBW), or ELGAN newborn.

4. Muscle relaxants are an important part of the anesthetic in cases such as these. Only very rarely are the infants able to tolerate a MAC of the inhaled agents, so muscle relaxants are needed to assure immobility of the patient.

5. As the case progresses, the airway pressure suddenly rises, there is a diminution of breath sounds bilaterally, and the SpO2 decreases steadily. What could be going on? What measures can you take to prevent this from occurring again?

## Postoperative Course

### Questions

1. When would you extubate? Why/why not? Is this baby at risk for post-extubation croup? What factors are important for subglottic stenosis in the premature infant? Might this infant develop tracheomalacia?

2. How can you assess postoperative pain in the neonate? How would you manage pain in this patient? Why? Are there differences in pharmacokinetics of opioids in this age group? Why?

5. During the case, the infant is moved away from the anesthesiologist down the OR table and is difficult to reach or even see. A kink in the endotracheal tube can easily occur. The circuit is stiff with low compliance and may easily kink the softened warmed endotracheal tube. Other common causes of increased airway pressure needed for ventilation through this 2.5 mm endotracheal tube include mucous plugging, the development of a pneumothorax, or resistance to mechanical ventilation by the patient. Humidification of the inspired gases will help keep tracheal secretions from occluding the endotracheal tube, but has its own problems such as "rainout" of water into the ventilator tubing and excessive heating of the inspired gases.

## Answers

1. Since the infant arrived intubated, required mechanical ventilation, was given opioids during the case, and now has an abdominal incision, extubation should not be done. The child would very likely hypoventilate or have apnea if extubated. If an infant is intubated with a tightly fitting endotracheal tube, post-extubation stridor or "croup" is a possibility, but not a certainty. Subglottic stenosis and tracheomalacia are complications of intubation to which newborns are subject. Although it is likely that the more trauma done to the trachea of a newborn, the greater the chance of complications such as subglottic stenosis or tracheomalacia, any preterm newborn who has been intubated is at risk for them.

2. Pain assessment in the newborn is even more challenging than assessment in patients with the ability to verbally communicate with the caregivers. This is part of the definition and explanation of the term "PAIN" found on the International Association for the Study of Pain (IASP):

- An unpleasant sensory and emotional experience associated with actual or potential tissue damage, or described in terms of such damage [11].
- *Note:* The inability to communicate verbally does not negate the possibility that an individual is experiencing pain and is in need of appropriate pain-relieving treatment. Pain is always subjective. Each individual learns the application of the word through experiences related to injury in early life.

  There are several scoring systems for the assessment of pain in the term and preterm newborn. The Neonatal Infant Pain Scale (NIPS) and the Crying, Requires oxygen, Increased vital signs, Expression, and Sleeplessness (CRIES) are composite measures of physiology and behavior and among those most commonly used. Postoperative pain in this patient could be managed by systemic opioid administration [12]. Morphine and fentanyl both have prolonged elimination half-lives in term and preterm newborns. There is evidence to support increased sensitivity to depression of respiratory drive in newborns, but opioids can certainly be safely administered to newborns provided there is appropriate

3. What are the long-term problems associated with necrotizing enterocolitis (NEC)? What is the history growth and development of survivors of NEC? Are there subsequent surgical problems? Nutritional status?

## Additional Topics

### Questions

1. Which infants are at risk for hypoglycemia? What threshold is used for the diagnosis of hypoglycemia in term infants? In preterm newborns? How would you treat hypoglycemia? What are the risks?

2. You need vasopressor support for a 2000 g infant undergoing a diaphragmatic hernia repair. Which vasopressor will you choose? Why? Does it matter what the pH is? Why? In which ways do catecholamine receptors differ in the newborn from those of the adult? Is the stressed newborn any different? Can you use ephedrine? Phenylephrine?

monitoring. Regional analgesia has been used in preterm newborns, but this patient still may have or develop bacteremia or coagulation abnormalities, two contraindications to regional analgesia.

3. Preterm newborns who have survived NEC may continue to have problems including malnutrition, intestinal obstruction, failure to thrive, and residual hepatic disease resulting from TPN. Their nutritional status depends greatly on the amount of intestine damaged by NEC itself and the amount removed at surgery. These infants and those whose NEC was treated medically (i.e., without surgery, NPO, antibiotics) will present with intestinal obstruction much later in childhood.

**Answers**

1. Hypoglycemia during general anesthesia can only be diagnosed by measurement of the serum glucose. Infants of diabetic mothers, SGA infants, and infants with syndromes such as Beckwith-Wiedemann and nesidioblastosis are at increased risk. Treatment is with a "mini" bolus of 200–300 mg/kg over 90 s followed by an infusion of 6–10 mg/kg/min. The specific number at which hypoglycemia is diagnosed is the subject of much controversy. The diagnosis traditionally has had three criteria and is not applicable to the OR setting. The criteria are the presence of clinical signs and symptoms (lethargy, pallor, cyanosis, jitteriness, apnea, seizures), a documented low serum glucose measurement, and a resolution of the clinical signs and symptoms promptly after correction of the low glucose measurement. In the OR, a safe lower threshold for treatment is 70 mg%. This number is somewhat above the threshold recommended for newborns who are not under general anesthesia, but, given the inability to suspect the diagnosis on clinical grounds, a higher threshold is needed. In contrast to earlier thinking, the absolute number used as the diagnostic criterion for hypoglycemia is not different for term or preterm newborns.

2. Dopamine use has been studied in term and preterm newborns. In the presence of normal pH, the dose range is similar to that used in older children and adults (2.5–20 mcg/kg/min). There is some indication that newborns respond to smaller doses than those commonly recommended. Acidosis significantly impairs the inotropic response to catecholamines. Epinephrine's potent inotropic and chronotropic effects are due to stimulation of alpha- and beta-receptors. Unless doses used are excessive, epinephrine can be safely used to support critically ill newborns. The usual range for infusion is 0.05–2 mcg/kg/min. Treatment of hypotension in critically ill newborns is best accomplished with direct-acting agents such as epinephrine.

3. How does continuous positive airway pressure (CPAP) work to improve pulmonary function in the premature infant? Which physics principle governs this? What is the role of surfactant?

4. You are performing a preanesthetic assessment of an 8-week-old term infant scheduled to undergo repair of a bilateral inguinal hernia. He has been to the ED for urgent reduction of the hernia 5 days ago, hence the scheduled repair. As you meet the family preoperatively, the parents are concerned about the possible deleterious effects of general anesthesia on the neurologic development of their child. Your response?

3. Preterm newborns are deficient in surfactant [2]. This deficiency leads to increased surface tension in alveoli that then tend to collapse at end-exhalation, decreasing FRC. Recall the Laplace equation which states that the pressure at the surface of a sphere is twice the surface tension divided by the radius of the bubble. Alveoli are not perfect spheres, but the relationship still has applicability:

$$P = 2T / r$$

On the basis of this relationship, pressure inside a small alveolus should be higher than that inside larger alveoli, leading to collapse of smaller alveoli and enlargement of larger alveoli. Surfactant promotes a decrease in surface tension even as the surface area of an alveolus is reduced, thus preventing collapse. The surface tension decreases to a greater extent than the radius, resulting in a diminishing transmural pressure gradient, stabilizing the smaller alveoli. Distending airway pressure helps to return the FRC to the volume it would be in a well newborn.

4. It is important to reassure the family of our vigilance and care during and after the anesthetic and surgery and also to present the differences between animal data and the experience and uncertainty of the relevance to the human newborn [13–18]. If the family is interested in further research on their own, a useful reference might be SmartTots, a partnership between the International Anesthesia Research Society and the FDA, and their recent statement intended for parents excerpted here:

   - In the United States alone, more than 1 million children, 4 years of age and under, undergo surgical procedures requiring anesthesia annually. While most children appear to recover well, findings from these animal studies call for further research to ensure the safety of every child undergoing anesthesia. Until this determination can be made, children requiring surgery essential to their health should proceed as directed by their physician. Young children usually do not undergo surgery unless the procedure is vital to their wellbeing. Therefore, postponing a necessary procedure may itself lead to significant health problems, and may not be an option for the majority of children
   - The entire statement is available on the SmartTots website: http://smarttots.org/faq-for-parents/.

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Papageorgiopu A, Pelausa E, Kovacs L. Chapter 22. The extremely low-birth weight infant. In: MacDonald MG, Mullett MM, Seshia MMK, editors. Avery's neonatology. 7th ed. Philadelphia: Lippincott Williams & Wilkins; 2016. p. 335–56. This chapter reviews the epidemiology of low birth weight newborns as well as NICU management of these patients. Respiratory, cardiovascular, and fluid management are reviewed. The authors also discuss management of many of the common clinical problems that affect low birth weight newborns. Morbidity seen in low birth weight newborns is also included.

Whitsett JA, Rice WR, Warner BB, et al. Chapter 27. Acute respiratory disorders. In: MacDonald MG, Mullett MM, Seshia MMK, editors. Avery's neonatology. 7th ed. Philadelphia: Lippincott Williams & Wilkins; 2016. p. 397–415. The authors review in detail the embryology of lung development, the development of surfactant, the pathophysiology of respiratory distress in the preterm newborn and management of RDS.

# Chapter 9
# Newborn Emergencies


**Thomas J. Mancuso**

A 14-hr-old male, 2400 g, born at 37 weeks' gestational age, is scheduled emergently for repair of an esophageal atresia with tracheoesophageal fistula. The newborn choked and gagged on the first glucose water feed. A contrast study confirmed the diagnosis. An NG tube is in place. The infant is receiving nasal cannula oxygen at 300 mL/min.

VS: HR = 158/min; BP = 88/52 mmHg; RR = 44/min; $T$ = 37.2 °C; $SpO_2$ = 95%; Hgb = 13.0 g/dl.



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## Preoperative Evaluation

### Questions

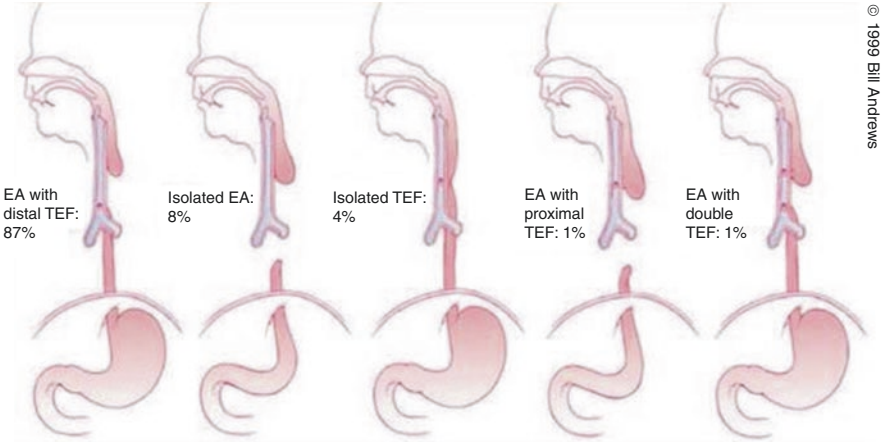
1. Is this an emergency? What historical information might have helped anticipate the diagnosis? Are there other, safer, means to make the diagnosis than a contrast study?

2. What other evaluations of the newborn are needed prior to undertaking the induction of anesthesia? What conditions are likely or associated with EA/TEF?

## Answers

1. While repair of the esophageal atresia (EA) with tracheoesophageal fistula (TEF) may not be a true emergency, it is, at the least, very urgent. The longer the newborn is unrepaired, the greater the risk for aspiration. Surgical correction should proceed very quickly, but proper preparation can be accomplished in short order. The diagnosis can be suspected in cases of maternal polyhydramnios. In the delivery room, inability to pass a suction catheter into the stomach should raise the suspicion of EA. A contrast study is not needed to make the diagnosis. Aspiration of oral contrast is a significant risk. Plain X-rays may show the dilated, air-filled esophageal pouch. A film with a radiopaque catheter coiled in that pouch will confirm the diagnosis. If there is no gas in the abdomen, it is possible that the child has EA without TEF.

2. It is important to ascertain which type of TEF is present. In cases of esophageal atresia, >90% have an associated tracheoesophageal fistula. The most common variant of a TEF, by far (90%), is esophageal atresia with a distal fistula between the posterior trachea near the carina and the stomach. The next most common, approximately 7–8%, is EA without TEF. Many other types and subtypes have been described. Up to 50% of patients with EA/TEF have other congenital anomalies. Cardiovascular anomalies make up one-third of the anomalies seen in these patients. The cardiac anomalies seen are, in order of occurrence, VSD, ASD, tetralogy of Fallot, and coarctation of the aorta. Other organ systems involved in these patients are musculoskeletal (30%), gastrointestinal (20%), and GU (10%). Patients with EA/TEF may have the VATER syndrome that consists of vertebral defects or VSD, anal/arterial defects, TEF/EA, and radial or renal anomalies [1, 2].



3. Is a preoperative gastrostomy with local anesthesia indicated? Would the situation be different if the patient were preterm with respiratory distress syndrome (RDS)?

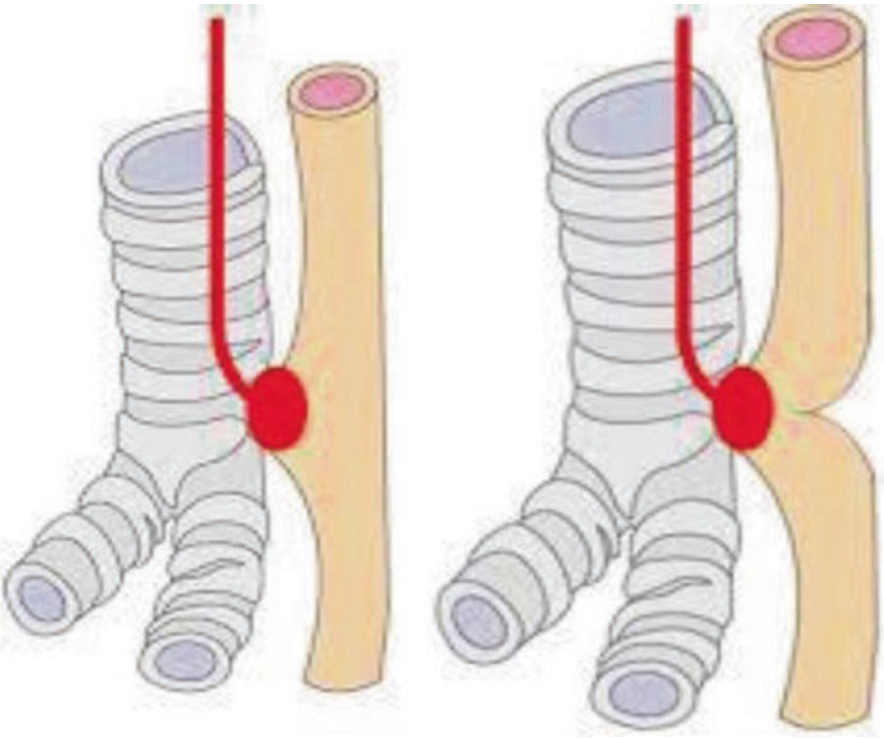
3. There are three methods used to decrease or eliminate insufflation of the stomach with the inspired gas from the endotracheal tube. The endotracheal tube tip can be placed beyond the fistula, just above the carina, but in some cases the fistula is actually at the carina making this procedure impossible. In cases where the newborn is having severe respiratory compromise and positive pressure ventilation has been instituted, a ventilator breath may follow a path from the trachea through the fistula and distend the stomach. The abdomen can become very distended, further compromising ventilation. In these dire situations, an emergent gastrostomy may allow the abdominal pressure to be relieved enough for ventilation to continue [3]. Approximately 25% of newborns with EA/TEF are born preterm, and in cases with respiratory distress, the situation is even more difficult since institution of positive pressure ventilation will require higher pressures. This will invariably also put gas into the stomach through the fistula. In cases when ventilation of the lungs is ineffective or incomplete, another option in addition to an emergency gastrostomy is placement of a balloon-tipped catheter through the fistula into the stomach, inflating the balloon and occluding the fistula. This can be accomplished by placing the balloon-tipped catheter through the fistula from the trachea with a rigid bronchoscope. Photo below shows the relative sizes of the right and left bronchi and the fistula with a positive pressure breath. Spontaneous breathing may not illustrate this as well, under direct vision.

## Intraoperative Course

### Questions

1. What monitors would you require for this case? Arterial line? Where? How will you assess intravascular volume? CVP catheter? How much information about preload would a Foley catheter give you for this case?

2. What might be done to minimize the effect of the fistula at induction? Is IV or inhalation induction preferable? How should the airway be secured? Does the presence of the fistula alter the techniques for intubation? What should be done with the NG tube once intubation is accomplished?



**Answers**

1. For otherwise well term newborns with EA/TEF, standard monitors with the addition of "pre-" (right hand) and "post-" (left hand or either foot) ductal pulse oximeters and a Foley catheter will often be sufficient. If there is pulmonary compromise, either from aspiration or because of prematurity, an arterial line is useful for frequent ABG determinations. A CVP catheter would not only give some information about intravascular volume but also be an excellent route for administration of resuscitation medications, should that be needed. If peripheral IV access is good in an otherwise well newborn with EA/TEF, the risk of placing a CVP line may not be justified. Urine output should mirror renal blood flow (GFR) but it is a secondary measure. In addition, the small volume produced may be difficult to accurately collect and measure. Nevertheless, this monitor can provide useful information for these cases.

2. In cases where the connection between the trachea and esophageal point of entry of the fistula is open, avoidance of positive pressure ventilation is important. Positive pressure ventilation will force gas through the fistula into the stomach. IV access should be secured prior to any attempts at induction of anesthesia or

3. Is controlled or spontaneous ventilation preferable for these cases? How would you determine whether or not a percutaneous gastrostomy is indicated prior to the definitive repair? Is a precordial stethoscope of particular importance for these cases?

4. After positioning and the start of the thoracotomy, breath sounds from the left axillary stethoscope markedly diminish and the $SpO_2$ decreases. What might be the cause? What would you do? Could the endotracheal tube have accidentally entered the fistula?

intubation. Awake intubation is often done, followed by spontaneous ventilation with the infant breathing oxygen plus incremental doses of a volatile anesthetic. Alternatively, an inhalation induction can be done, and when an adequate depth of anesthesia has been achieved and the airway anesthetized with the appropriate dose of topical anesthetic, laryngoscopy and intubation can be done. It has been commented that turning the bevel of the endotracheal tube anteriorly will decrease the chance of intubating the fistula, but this is unproven. It also has been suggested that since the fistula is often relatively low in the trachea, a deliberate right main stem intubation should be done and the endotracheal tube then withdrawn to a position just above the carina, hopefully distal to the fistula. Great care is required while advancing the endotracheal tube in the trachea, however. The fistula may be quite large, and the endotracheal tube may easily be placed into the fistula if it is advanced too far into the trachea [4]. As mentioned above, an alternative that will allow positive pressure ventilation is performance of a rigid bronchoscopy following induction of anesthesia and placement of an occluding balloon-tipped catheter through the fistula. The balloon is then inflated and the catheter pulled taut, thus closing the fistula [5]. This technique allows positive pressure ventilation to proceed without distending the stomach. The surgeon will likely ask that the NG tube be advanced during the procedure to facilitate identification of the esophageal pouch.

3. If the stomach distends after intubation, even with gentle assistance of respiratory efforts, and this distention is interfering with ventilation (leading to the use of higher ventilation pressures), percutaneous gastrostomy will allow some control of the situation. The usual position for surgery is left side down for a right thoracotomy. The surgeon retracts the right lung, leaving only the left lung for gas exchange. In this situation, a left axillary stethoscope will give the anesthesiologist immediate information about the adequacy of ventilation. The left bronchus is easily occluded by blood or secretions and may be kinked by the surgeon during the procedure; the anesthesiologist must be aware of these events as soon as they occur [6].

4. Secretions and/or blood may easily occlude the lumen of the trachea or right main bronchus. Additionally, the bronchus is often kinked by surgical retraction during the procedure. Even with occlusion of the fistula by a balloon-tipped catheter, given the relatively large size of some fistulae as seen in the photo above, there still may be room for the tip of the endotracheal tube to completely or partially enter the fistula, greatly decreasing or eliminating ventilation of the lungs [6].

## Postoperative Course

### Questions

1. Is this patient a candidate for extubation at the conclusion of the surgery? If not, how should the newborn be ventilated? Should the NG tube be removed?

2. What options are there for postoperative analgesia? Does the presence of VATER or VACTERL (vertebral anomalies, anal atresia, cardiac defects, tracheoesophageal fistula and/or esophageal atresia, renal and radial anomalies, and limb defects) syndrome affect your willingness to use regional analgesia? If the patient has no other anomalies, would regional analgesia be useful for this case? What drugs would you use?

3. Following extubation from minimal ventilator settings on postoperative day #1, the patient exhibits respiratory distress with inspiratory stridor. What might be the cause? What therapy would you begin? How would you decide whether or not to reintubate the child?

## Answers

1. For term infants who undergo a relatively uncomplicated repair, extubation is a possibility, but the intensive care nursery team who will care for the baby should be involved with the decision.

   The mode of ventilation, if extubation will be delayed, should be guided by the intraoperative course. It is often advisable to use a ventilator from the ICN for newborns in the OR since the anesthesia machine ventilators are not specifically designed for use in the newborn. The position of the NG tube is very important. It is generally left in a position such that the tip is just proximal to the esophageal anastomosis.

2. Postoperative analgesia can be provided by administration of local anesthetic into the epidural space [7] or via an ultrasound-guided right paravertebral catheter [8]. If the patient has the VATER association, epidural catheter placement may be problematic, but regional techniques should not be ruled out prior to review of an X-ray of the spine [9]. If regional analgesia is not undertaken, parenteral opioids can be used to provide analgesia. In either case, cardiorespiratory monitoring must be done [10].

3. Patients with EA/TEF may have significant tracheomalacia at the level of the dilated esophageal pouch. In utero, the dilated esophageal pouch may compress the developing trachea, leading to weakened cartilage. With vigorous inspiration, this area of the trachea may partially collapse, and inspiratory stridor will result. It is unlikely that treatment of this problem with inhaled racemic epinephrine will be effective as it usually is with infectious croup, but if subglottic edema is part of the problem, a trial of this treatment should be undertaken. The trachea should be reintubated if respiratory failure is imminent based on clinical and laboratory criteria. If reintubation is done, exquisite care must be taken with the NG tube, and esophageal intubation must absolutely be avoided.

## Additional Topics

### Questions

1. What are the major preoperative considerations in evaluating a patient for congenital diaphragmatic hernia (CDH)? What immediate interventions can be performed therapeutically? Sudden deterioration may indicate what? On which side?

2. What are the important differences between omphalocele and gastroschisis? What are the important anesthetic considerations during correction of these defects?

3. What are the considerations for anesthetic care during resection of a sacrococcygeal teratoma?

4. What are the anesthetic concerns for a child coming to the operating room for repair of pyloric stenosis?

**Answers**

1. In the preoperative evaluation of newborns with congenital diaphragmatic hernia, the size of the hernia is important in predicting the severity of cardiorespiratory compromise and ultimate prognosis [11, 12]. Eighty percent of the defects are posterolateral, most commonly on the left, through the foramen of Bochdalek. Twenty percent of newborns with CDH have associated cardiac defects, most often patent ductus arteriosus (PDA) [13]. Poor prognosis is associated with birth weight <1,000 g, gestational age <33 weeks, and an A-a gradient >500 mmHg [14]. Placement of a nasogastric tube may help ventilation by decompressing the stomach. Mechanical ventilation should be done with the lowest possible airway pressures [15]. Sudden deterioration may be due to the occurrence of a pneumothorax on the contralateral side from the hernia defect.

2. Omphalocele is herniation of the intestine into the umbilical cord, while gastroschisis is a defect in the abdominal wall. With omphalocele, a peritoneal sac covers the intestines (unless it is ruptured during delivery), but there is no covering in cases of gastroschisis. Infants with omphalocele are much more likely to have associated GI, cardiac, or craniofacial anomalies, but only approximately 25% are preterm or low birth weight. In Beckwith-Wiedemann syndrome, omphalocele occurs in association with macroglossia, hypoglycemia, organomegaly, and gigantism. A much higher percentage of newborns with gastroschisis are born preterm. Two important anesthetic considerations for these conditions are fluid management and possible compromise of ventilation and/or circulation during replacement of the abdominal contents and attempted closure of the abdominal wall [16].

3. Sacrococcygeal teratomas can be quite large with an extensive blood supply. Surgical excision can cause significant bleeding to the point that occlusion of the descending aorta may be needed as a temporary measure for hemostasis. The newborns' position may change from supine to prone more than once during the procedure [17].

4. Pyloric stenosis generally presents between 2 and 6 weeks of age with vomiting that is relentless and progressive but not bilious. The persistent vomiting may result in dehydration and hypochloremic metabolic alkalosis. Fluid replenishment and normalization of electrolytes should be accomplished prior to taking the child to the OR for a (often laparoscopic) Ramstedt pyloromyotomy. Suctioning of the stomach should precede induction of anesthesia. Several passes with an orogastric tube may be needed. Induction is by a rapid sequence technique. Rare occurrences of apnea in the postoperative period (possibly related to the still somewhat alkaline CSF) or hypoglycemia have been seen. These children generally do very well postoperatively, often taking POs within hours of the end of the procedure. Analgesia can often be provided with PO/PR acetaminophen [18–21].

5. An infant, several hours old, is brought to the OR by the surgeon for respiratory distress with a diagnosis of infantile lobar emphysema. Is infantile lobar emphysema a surgical emergency? Why/why not? Should the patient undergo bronchoscopy first? Why/why not? What muscle relaxant would you choose? Why? A colleague of yours suggests the avoidance of nitrous oxide; do you agree?

6. A newborn is scheduled for exploratory abdominal surgery for obstruction. The abdomen is distended, meconium has not yet passed, and the intestines are palpable through the anterior abdominal wall. What is the most likely surgical diagnosis? What respiratory management concerns do you have? How will you approach securing the airway? Is it likely that this baby's sweat chlorides will be normal?

5. There is a long differential diagnosis for a hyperlucent area in the lung of a newborn or infant that includes congenital pulmonary airway malformations (CPAM), localized pulmonary agenesis, bronchogenic cyst, airway foreign body, pneumothorax, or localized pulmonary interstitial emphysema (PIE). Congenital lobar emphysema is a relatively unusual cause of respiratory distress in the newborn and infant period. Presentation is usually within the first 6 months of life [22, 23] and includes tachypnea, tachycardia, and signs of respiratory distress [3]. The left upper lobe is the most commonly affected. Progressive air trapping leads to hyperinflation of the affected lobe. This lobe can then compress adjacent structures such as normal lung and vessels and even cause mediastinal shift. In infants who are rapidly worsening, this condition can certainly be a surgical emergency. Preoperative maneuvers such as chest tube placement or needle aspiration of the trapped air have not been successful in alleviating the respiratory distress in these children. Induction of anesthesia is a challenge. A slow inhalation induction with oxygen and sevoflurane, allowing the child to breathe spontaneously, will minimize the possibility of increasing the size of the emphysematous lobe and worsening the situation [22]. When the infant hypoventilates, gentle positive pressure ventilation must be performed. If positive pressure ventilation must be delivered, consideration should be given to the use of high-frequency ventilation [24]. Epidural catheters (caudal, lumbar, thoracic) have been used to provide analgesia for these procedures [25], and postthoracotomy analgesia also can be provided with ultrasound-guided paravertebral catheters.

6. Meconium ileus results from obstruction of the distal small intestine by abnormal meconium [26]. This problem occurs almost exclusively in infants with cystic fibrosis (CF); however, most infants with CF do not develop meconium ileus. This patient has CF until proven otherwise. The exocrine gland dysfunction in CF leads to pulmonary disease, pancreatic dysfunction, and abnormalities in sweat gland function that cause increased NaCl concentration in sweat. When a sweat chloride is measured at >60 mEq/L, the diagnosis of CF is confirmed. The pulmonary compromise is due to thickened secretions and abnormal mucociliary clearance of those secretions. Small airways become obstructed and portions of the lung become hyperinflated. There is an inconsistent response to bronchodilators. Induction of anesthesia in this newborn is complicated by full stomach considerations, the decreased FRC due to abdominal distention, and the possible pulmonary compromise due to CF. Once tracheal intubation is accomplished, the anesthesiologist should be prepared to suction the pulmonary secretions, possibly after lavage, to improve gas exchange and pulmonary mechanics.

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# Chapter 10
# Fetal Surgery


**Joseph P. Cravero and Thomas J. Mancuso**

You are asked to provide anesthesia for a 30-year-old female who is currently pregnant with a 22-week fetus who has a myelomeningocele. The surgery team would like to correct the defect at this point to avoid neurologic damage that will result from prolonged exposure of the fetal neural structures to amniotic fluid. The baby will be brought ex utero for the procedure and then returned for the duration of the gestation.



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## Preoperative Evaluation

### Questions

1. What are the maternal and fetal physiological considerations that should be taken into account prior to beginning this procedure?

2. How do you maintain fetal well-being during the course of the procedure? What are the primary considerations for maintaining adequate blood flow?

3. How would you prepare the patient(s) for surgery and anesthesia? What kind of monitors should be in place?

4. What is the normal fetal oxygen saturation? How would you monitor heart function?

## Answers

1. Pregnancy affects many aspects of maternal physiology. Oxygen demand is greater, so precautions must be taken to prevent periods of prolonged apnea or hypoventilation. Capillary permeability increases so the risk of pulmonary edema is elevated – particularly in the setting of magnesium used for tocolysis. The weight of the gravid uterus can decrease venous return, so left uterine displacement is an important consideration. In terms of the fetus, exposure to stress has been associated with increased cortisol and other stress hormones. Opiates have been shown to attenuate this response – so there is no doubt about the need for anesthesia in the fetus. The fetus requires less anesthesia than a child, but it should be recognized that although inhaled agents readily cross the placenta, they do not reach maternal levels for a prolonged period of time. The fetal heart has less contractile tissue and is sensitive to the cardiac depressant effects of anesthesia. When combined with fetal manipulations during surgery, hypotension, bradycardia, and cardiovascular collapse are a significant consideration. The circulating volume of a fetus is very small and blood loss is poorly tolerated. Fetal skin is not yet mature and subcutaneous tissue is lacking – thus leading to the tendency for hypothermia if exposed to ambient temperature environment for any period of time.

2. Fetal circulation is dependent on uteroplacental blood flow. Maternal volume status and blood pressure must be optimized. Uterine tone increases during contractions with a corresponding increase in vascular resistance. It is therefore critical to keep the uterus relaxed. Kinking of the umbilical cord must be avoided and corrected if it occurs. Increased pH and hypocarbia will decrease uterine blood flow and result in fetal hypoxia.

3. The operating room should be warmed to about 80°. O negative blood should be prepared for the fetus, and type-specific blood should be ready for the mother. The mother should receive metoclopramide and Bicitra® for prophylaxis because of her slow gastric emptying time and the risk of aspiration. A pulse oximeter should be prepared for both patients. An arterial catheter and transducer are indicated for the mother. Tocolysis should be aggressively pursued with an indomethacin suppository and IV magnesium as needed.

4. Normal fetal saturations are 60–70% – values above 40% are adequate during surgery. Echocardiography can be used to monitor heart rate and stroke volume. Fetal labs and blood gases can be obtained from the umbilical artery by the surgical team.

## Intraoperative Care

### Questions

1. Does the mother require rapid sequence induction and intubation? Why? How would you maintain anesthesia? TIVA? Inhaled agents? How do you anesthetize the fetus? What would you do to maintain uterine relaxation during the case?

2. What are your hemodynamic goals? At what point would you treat hypotension and what would you treat it with?

3. How would you provide fluid or blood to the fetus? How would you provide additional anesthesia once the baby is exteriorized and the procedure is being performed? What are your primary concerns once the fetus is returned to the uterus?

## Postoperative Care

### Question

1. Where should the mother be managed in the postoperative time frame? What are the most important aspects of her care? How should she be monitored and what should she be monitored for? What would be the best choice for postoperative analgesia? Regional analgesia? Systemic opiates? Why?

**Answers**

1. General anesthesia administered to the mother will provide adequate anesthesia to the mother and fetus. A rapid sequence induction is indicated. High levels of inhaled agent are needed to help maintain uterine relaxation in addition to tocolytics and IV nitroglycerin.

2. Hypotension can be treated with vasoactive medications such as ephedrine, phenylephrine, or dopamine to keep blood pressure approximately within 20% of baseline. When the fetus is exteriorized, intramuscular injection of muscle relaxant and fentanyl to the fetus augments the existing inhaled anesthesia, which will have crossed the placenta.

3. The surgeon can obtain umbilical arterial or venous gases and provide intravascular access if blood loss is a problem, thus allowing the administration of fluids and blood. After the fetus is returned to the uterus and the uterine incision is closed, the mother's cardiovascular status must be vigorously supported and aggressive tocolysis must be continued.

**Answer**

1. The mother should be monitored closely in an ICU environment. Tocolysis will be an important aspect of care, most prominently with magnesium sulfate. Mother needs to be monitored for any indication of labor with close tocodynamometry. She also needs to be monitored for signs of heart failure, and adrenergic drugs may be needed to manage this condition. An epidural catheter is often employed to maximize maternal comfort, minimize stress, and decrease the incidence of progressive uterine contractions.

## Additional Questions

### Questions

1. You are asked to provide anesthesia for a mother/baby pair to facilitate delivery and an (ex utero intrapartum treatment) EXIT procedure to secure the airway of the newborn who is known to be nearly completely obstructed by a cystic hygroma. What is an EXIT procedure? What is the best choice for anesthesia technique for this procedure? What is the overall goal of the EXIT procedure? Does this influence your choice of technique? Why is uterine atony so important for the success of the procedure? How can you design the anesthetic in order to avoid the adverse consequences of using very high end-tidal levels of volatile agents in order to achieve uterine atony? What specific medications would you use for tocolysis? What is the physiological importance of amnioinfusion following hysterotomy? What is your anesthetic management strategy following completion of the EXIT procedure?

2. A mother presents for laser ablation of placental vessels to prevent ongoing twin-twin transfusion. What is this procedure? Why is it performed? What are the risks? How would you provide anesthesia?

## Answers

1. Ex utero intrapartum treatment (EXIT) is an operation performed at the time of vaginal delivery or caesarean section. A portion of the fetus is delivered, and brief procedures such as endotracheal intubation or examination of a neck mass can be accomplished while the fetus is still connected to the placenta through the umbilical cord. Originally these procedures had to be very brief, but the duration has now lengthened to an hour or more in some cases.

   Uterine atony can be attained and maintained with high doses of inhaled anesthetic, which is the most important goal of the anesthetic strategy to preserve uterine perfusion. Sympathomimetics are needed to maintain maternal arterial pressure to within 10% of baseline to ensure fetal perfusion. Fetal monitoring can take place with pulse oximetry and echocardiography. Endotracheal intubation or LMA placement can be accomplished prior to uterine separation with tracheostomy then performed after delivery. Uterine atony must be reversed by the administration of intramuscular methergine (methylergonovine). Fluids and blood should be readily available since blood loss is generally greater during these procedures than for a normal vaginal delivery or c-section. Amnioinfusion is used to prevent uterine contractions and placental abruption. Partial rather than full exteriorization of the fetus further serves to preserve uterine volume and therefore decreases the chance of uterine contractions.

   Following completion of the EXIT procedure, the high dose of volatile anesthetic administered for uterine relaxation is decreased, and supplemental intravenous anesthesia is administered. This technique has become known as "SIVA." If an epidural was placed prior to the general anesthetic, it can be dosed in order to further decrease the level of inhaled agent needed.

2. Laser ablation of placental vessels involves fetoscopic laser photocoagulation of superficial unidirectional arteriovenous vessels on the placenta. The procedure reduces the amount of twin-twin transfusion. It has been found to prolong pregnancy and is associated with improved fetal outcomes and survival. Neurodevelopmental morbidity is also reduced. The procedure is performed by creating a "surgical window" sswith a combination of patient positioning and amnioinfusion. Risks of the procedure include rupture of the amniotic membrane, subchorionic bleeding, preterm delivery, and fetal death. The anesthetic will depend on the mother and her tolerance of emotional stress and discomfort associated with the procedure. Many centers accomplish this procedure with local anesthesia. Alternatively, spinal, epidural, or combined spinal epidural anesthesia has also been utilized. Fetal sedation (for movement control) can be achieved with sedatives such as fentanyl and midazolam administered to the mother.

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# Chapter 11
# Conjoined Twins


**Joseph P. Cravero**

The patients are 7-month-old conjoined twin females born at 32 weeks' gestation. They are joined at the sacrum. Planned surgery is separation.



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## Preoperative Evaluation

### Questions

1. What is the rate of conjoined twin birth? What is their chance of survival? How urgent is the surgery?

2. How are conjoined twins classified? How does their classification impact their prognosis?

3. Describe the nature of the preoperative evaluation for conjoined twins.

## Answers

1. Conjoined twins are a relatively rare occurrence with an incidence of approximately 1 in 50,000 to 1 in 200,000 births. They are always monozygotic and monochorionic twins. Forty percent of these pregnancies end in stillbirth. Of the 60% that are born, only about 20% live to be eligible for separation. The defect leading to conjoined twins is likely a fusing of overlapping or closely contiguous twin embryonic axis formative fields within a single embryonic disc. It is thought that these factors are responsible for the failure of twins to separate after the 13th day after fertilization. Conjoined twins have been created in amphibians by simply constricting the embryo so that two embryos form, one on each side of the constriction. Most often this surgery is not urgent, and time can be taken to optimize the planning for the environment as well as the underlying health of the twins. Many of these cases are performed when the twins are a year or more of age. On rare occasions, based on the shared anatomy, the health of one or both twins may be adversely impacted by their conjoined nature – in which case the need for surgery becomes more time-sensitive.

2. Conjoined twins are classified according to the region by which they are joined.
   Craniopagus twins are joined at the head. Thoracopagus twins are joined at the upper half of the trunk. (This is the most common form of conjoined twins making up 35–40%.) Omphalopagus twins are joined at the chest or abdomen – the second most common form of the anomaly at 30% of all cases. Most often these patients share a liver and prognosis for separation is generally good. Pygopagus are joined at the sacrum and constitute 19% of all cases. Separation is most straightforward in these cases since they do not generally share vital organs and survival is high.

3. Preoperative evaluation will vary with the site at which the twins are conjoined and the nature of shared organs. In this case, it would include routine blood and urine analysis, coagulation screen, plain X-rays, and ultrasound evaluations of the abdomen and pelvis. Computed tomography and digital subtraction angiography (DSA) delineate anatomic and bony detail including organ position, shared viscera, and vascular anatomy. In this case, a CT myelogram or MRI would be critical to determine the extent to which neural elements and spinal cord anatomy are shared. We preferred the use of the CT myelogram since that did not involve anesthetizing the patients where an MRI generally would require sedation/anesthesia. The determination of the blood supply to these elements would also be critical.
   Preoperative assessment and planning with interdisciplinary communication and cooperation is vital to the success of this type of procedure. A dedicated team of anesthesiologists for each twin is required. There must be two sets of all monitoring equipment and resuscitation equipment. All equipment and monitors need

## Intraoperative Course

### Questions

1. What monitors and vascular access are needed for this surgery?

2. How would you perform induction of anesthesia for this case?

to be labeled for each twin. It is often advised to color code the equipment that is to be used and separate between the two patients (one is green, the other yellow, etc.). It is helpful to color code the teams as well. It is advised to "trial" the operating room setup and simulate the operation and environment at least once prior to surgery. Plans for the surgical, anesthesia, OR technical, and nursing aspects of the case must be fully reviewed and trialed.

Prior to surgery, it may be desirable to test for the presence of cross-circulation. At times, this is very clear from the MRI or CT scans that are obtained for surgical planning. If the degree of shared circulation is in doubt, one simple way to determine this is by administering an anticholinergic such as atropine to one twin and monitoring if heart rate change is observed in the other twin. Alternatively, other agents such as Tc-99m, microcolloidal human serum albumin (HSA), and Tc-99m HIDA can be injected in one twin and detected in the other. Finally, indigo carmine can be injected into one twin with the examination of the other twin's urine for indigo carmine excretion. This is critical when considering how to safely induce anesthesia for these patients. If the patients share circulation, anesthesia must be induced simultaneously since any drug administered to one twin will necessarily affect the other. Conversely, if circulation is not shared, induction could be achieved separately.

## Answers

1. Surgery to separate conjoined twins is always a long process with the potential for significant blood loss and fluid shifts. A full set of ASA monitors is required for both patients. In addition, it would be appropriate to place a radial arterial line for both twins. The need for central access can depend on the exact nature of the conjoining of the twins. As a general rule, it is preferable to have central access for each patient in order to provide fluids, blood products, and vasoactive drugs and obtain some measure of central filling pressure. Urinary catheters should be placed.

2. The induction of anesthesia will depend on the nature of the airways involved and the presence of shared circulation of the twins. Conjoined twins may be positioned in such a way that their airways are difficult to access even though they may be anatomically normal. In addition, they may have craniofacial anomalies that could make their airway management challenging regardless of positioning. In this case, the twins were normal in appearance and facing away from each other. They underwent inhaled induction (with a single IV catheter in place on each twin) simultaneously, muscle relaxant was given after mask ventilation was established, and intubation was accomplished without difficulty. If the circulation were shared, it would be particularly important to induce anesthesia simultaneously since both twins would receive some effect from any drug admin-

3. Are there any other medications or precautions that are particularly important for this procedure?

## Postoperative Course

### Question

1. Would you extubate these babies postoperatively?

## Additional Question

### Question

1. A set of 9-month-old thoracopagus conjoined twins requires a CT myelogram to delineate an uncertain area of possible shared spinal anatomy. How would you accomplish sedation for the injection and CT scan?

istration. In any case, if twins are induced separately, it is critical to observe the nonanesthetized twin for any reaction or effect.

3. As these are very long procedures and blood loss is likely to be several blood volumes, it is important to plan for blood conservation and transfusion. The administration of tranexamic acid (TXA) or epsilon aminocaproic acid (Amicar®) would be reasonable to attempt to minimize fibrinolysis. If the children were large enough, blood salvage techniques should be in place. Fluids should include a maintenance fluid of D5 1/2NS and an isotonic solution such as lactated Ringers to account for third spacing and blood loss. Colloidal fluids such as albumin could be added when fluid replacement exceeds 50 mL/kg, but there is little indication that this changes outcome. The blood bank should be notified of this case, and appropriately typed, screened, and (if necessary) crossmatched blood should be available. The blood bank should be at least one to two units "ahead" at all times for each twin. The need for platelets and fresh frozen plasma should also be anticipated. If thromboelastography (TEG) is available, it can be very helpful in these cases to determine the nature of anticoagulation in the face of massive transfusion needs.

**Answer**

1. No. After any separation procedure, the duration of the procedure and the administration of large amounts of fluid and blood require a prolonged recovery. Airway edema and neurological status are not going to be appropriate for extubation. ICU beds should be available, and sedative/opiate infusions will be needed for (usually) a significant period while the twins recover.

**Answer**

1. We believe that in these cases the least amount of anesthesia that accomplishes the goals of the procedure would be most desirable. It is important to remember that if the circulation is shared, sedating one twin may suffice for both. Also, it is important to note that only one twin needs to undergo contrast injection. In this case, we chose the twin whose spine offered the easiest exposure and place EMLA cream at L3–L4. Sucrose pacifiers were given to both twins. While their nurse held them on her chest, a 22 G spinal needle was placed and clear CSF was accessed. The contrast was then administered. The twins were then positioned for the CT scan. One twin was administered 0.05 mg/kg of midazolam and 0.25 mg/kg of ketamine. Both became quiet. With standard ASA monitors in place, the scan was completed.

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# Chapter 12
# Neuroanesthesia

**Thomas J. Mancuso**

An active 2-year-old, 12 kg boy is scheduled for a frontal craniotomy for resection of a craniopharyngioma.

VS: HR = 100/min; BP = 110/60 mmHg; RR = 24/min; $T$ = 37 °C.

A heart murmur is detected on preoperative examination.



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## Preoperative Evaluation

### Questions

1. What is the possible significance of a heart murmur? How should it be evaluated? Are any lab tests helpful? Which ones? Should this patient have a preoperative echocardiogram? Why? Should a cardiology consult be obtained? Why? A small atrial septal defect is diagnosed. Is this of any significance?

2. What are the perioperative implications of this tumor? What is diabetes insipidus?

   How can diabetes insipidus be diagnosed preoperatively? If diabetes insipidus is present preoperatively, will this affect your preoperative fluid management? How do you assess the need for perioperative steroids? What are the possible implications of inadequate steroid replacement? What other hormones can be affected? What lab work would you require preoperatively? Explain.

## Answers

1. During routine random examinations, up to 30% of children will demonstrate an innocent murmur. There are several innocent murmurs of childhood, not associated with any cardiac pathology, with which a pediatric anesthesiologist should be familiar. The innocent murmur (Still's murmur) is characterized by a high-pitched, vibratory, short systolic murmur heard along the left midsternal border without radiation in children 2–7 years of age. An innocent venous hum resulting from turbulent flow in the jugular system may also be detected in the neck or upper chest. The hum can be changed or eliminated by position changes or light compression of the jugular veins in the neck. In some cases of increased cardiac output such as during febrile illnesses, murmurs of flow across normal semilunar valves are heard. The murmur of an ASD is similar to that appreciated in pulmonic stenosis. There is no murmur caused by the low-velocity left to right flow across the ASD itself. Because of the increased flow across the pulmonic valve in children with right to left flow through an ASD, a murmur can be heard. The murmur is characterized as a soft ejection-type (crescendo-decrescendo) murmur of relative pulmonic stenosis which is heard at the upper left sternal border. This murmur results from the excessive flow across a normal pulmonary valve. The second heart sound is louder and also widely and persistently split as a result of this excessive flow. The most common type of ASD is the secundum type with the abnormal connection between the atria, a result of incomplete formation of the second atrial septum. PVR remains normal throughout childhood and CHF is quite infrequent. Adults with uncorrected ASDs do develop CHF and/or atrial flutter or pulmonary hypertension, so correction of the ASD is generally undertaken in early childhood. The significance of an ASD is that of the possibility of a paradoxical embolus in which air or clots in the venous system cross the ASD and lead to complications in the systemic arterial circulation.

2. Craniopharyngioma, a tumor of Rathke's pouch, may descend into the sella turcica and destroy part or all of hypothalamic and pituitary tissues as it enlarges, leading to hypopituitarism [1]. Preoperatively, the child should be evaluated for adrenal or thyroid dysfunction [2]. If ACTH secretion is impaired by the tumor, the production of glucocorticoids and androgens by the adrenal cortex will be below normal. If not evaluated preoperatively, adrenal insufficiency should be assumed and the patient treated accordingly. Diabetes insipidus is unlikely to be seen preoperatively but certainly may occur during or after the procedure. It is diagnosed by the presence of a large volume of dilute urine (Osm <300 mOsm/mL) in the face of increasing serum osmolarity and increasing serum sodium. If replacement of urinary losses with dilute IV fluid such as D2.5W or D5 0.2NS is insufficient, an infusion of aqueous vasopressin should be started. The preoperative lab tests ordered depend upon the clinical presentation but may include electrolytes, fasting glucose, thyroid function tests, and a CBC. Of course, imaging studies ordered by the neurosurgeon should be reviewed as well.

## Intraoperative Course

### Questions

1. Would you administer a premedication on this child? Why? Would you require an intravenous catheter before starting the induction? Why? If not, or if the initial attempts are unsuccessful, what's next? A colleague suggests intramuscular ketamine. Agree? Why? Is an inhalation induction appropriate? Explain.

2. Would you insert an arterial catheter? Why? Is a central venous catheter needed? Why? If yes, where? Potential problems? Where do you want the tip to be? How do you confirm its position? What if multiple attempts are unsuccessful? Is a urinary catheter necessary? Why? Is a precordial Doppler necessary? Explain.

3. What agent would you use for induction? Why? Explain your choice of muscle relaxant, if you are using one. Explain your choice of agents for maintenance. Suppose the surgeon asked you to give mannitol. How much is appropriate? Would hypertonic NS be a better option? Why/why not? What is the expected effect of administering either of these agents? What are the potential problems with the administration of either of these agents?

## Answers

1. The possibility of raised ICP should be considered when planning whether or not to administer a premedication. In a child such as the one presented who does not have intracranial hypertension, an inhalation induction is appropriate, with or without a premedication, depending upon the patient's (and the family's) level of anxiety. Placement of an IV for induction is also appropriate and would allow a more rapid induction without the possibility of airway compromise that sometimes occurs during an inhalation induction and that would likely upset both the child and family. Ketamine is a potent cerebral vasodilator and also can cause sudden increases in ICP. Use of ketamine for this child is appropriate but may not be so as an induction agent in children with raised ICP. Intramuscular midazolam is another possibility for a particularly anxious, uncooperative child who refuses oral premedication. Barbiturates have some advantages in neurosurgical patients, since this class of drugs does lower both cerebral blood flow (CBF) and the cerebral metabolic rate for oxygen (CMRO2).

2. An arterial catheter is appropriate for cases such as this in which large fluid shifts or blood losses are possible and/or frequent monitoring of serum ABGs or electrolytes is planned. The radial artery is the most convenient and commonly used site, although the posterior tibial and dorsalis pedis arteries in the foot are also acceptable sites. Complications of arterial cannulation include arterial occlusion, flushing of emboli through indwelling catheters, ischemia distal to a catheter, and rarely, infection. A central venous catheter may be useful in this case as a measure of preload. For neurosurgical procedures, cannulation of the femoral vein is an attractive option. Not only is the insertion site accessible to the anesthesiologist who is at the patients side, but also venous drainage from the head is not impaired. A CVP catheter is not useful in treating venous air embolism (VAE) except as a route for administration of resuscitation medications, should that become necessary. Given the possibility of DI, a urinary catheter is an important monitor. VAE is a possible complication of pediatric neurosurgical procedures. A precordial Doppler is the most sensitive monitor of VAE, detecting even minute, clinically insignificant amounts of air. The precordial Doppler is of limited use during electrocautery. Supplementing the Doppler with another monitor of VAE such as the capnograph or end-tidal nitrogen monitoring is helpful since these monitors are not affected by electrocautery [3].

3. Induction of anesthesia can be safely accomplished with either an inhalation or IV technique in this active 2-year-old without evidence of raised ICP. Muscle relaxation should be part of the maintenance since any movement of the child once positioned would be dangerous. The goals of maintenance of anesthesia should include provision of a "slack brain" for the neurosurgeon and stable

4. During the craniotomy, the blood pressure decreases to 60/40 mmHg. What are the possible causes? The end-tidal $CO_2$ decreases as well. Cause? How can hypovolemia be differentiated from venous air embolism? Are there different treatments for each of these? PEEP? Does the presence of the atrial septal defect influence your management? After your first intervention, the hypotension persists. What is your next move? When are vasopressors indicated? Which would you choose? Why? Would you continue/restart the nitrous oxide? Explain.

5. The urine output is 4 mL/kg/h. What is your differential diagnosis? Which lab tests might be helpful? The serum sodium is 155 mEq/L. Diagnosis? Treatment? Which intravenous fluids would you use? Why? Should they contain glucose? Why? How do you determine the rate of administration?

hemodynamics. The technique should allow for a rapid emergence at the conclusion of the procedure. Administration of opioid prior to pin placement and local anesthetic infiltration along the proposed incision will help minimize hemodynamic derangements. Mannitol administration may help reduce ICP and decrease the size of the brain, allowing better surgical exposure. Starting doses, in the range of 0.25–0.5 mg/kg IV, raise serum osmolality by approximately 10 mOsm. If given too rapidly, mannitol may cause transient hypotension. Repeated and large doses may increase serum osmolality to >320 mOsm, a dangerous level. Hypertonic (3%) saline has been used more recently in the treatment of raised ICP in patients with traumatic and nontraumatic cerebral edema and is an option to consider in the operating room. Interest in this treatment has undergone a resurgence. Penetration of sodium across the blood-brain barrier is low. Sodium has a reflection coefficient higher than that of mannitol and shares with mannitol both the favorable rheologic effects on CBV and osmolar gradient effects. Hypertonic saline exhibits other theoretical benefits, such as restoration of cell resting membrane potential, stimulation of atrial natriuretic peptide release, inhibition of inflammation, and enhancement of cardiac performance.

4. Venous air embolism (VAE) is a distinct possibility in pediatric neurosurgical procedures. The incidence varies with the sensitivity of the detection device used, but up to 30–40% of children undergoing intracranial procedures have VAE. Maintenance of a generous circulating blood volume and the use of positive pressure ventilation help decrease the likelihood of a VAE. Once detected or suspected (unexplained hypotension), the anesthesiologist must alert the neurosurgeon who will flood the field, while the anesthesiologist ventilates with 100 % oxygen and treats any hemodynamic instability. Vasoactive, inotropic agents may be needed to maintain the blood pressure at normal levels. Enhancing cardiac contractility may help to move any air from the right ventricle into the pulmonary circulation. The presence of an ASD in this patient is particularly troubling since air in the right atrium may cross to the left atrium and then travel to the cerebral or coronary circulation [4]. If hemodynamic instability persists, turning the patient to a left side down and head down position (Durant's maneuver) may help move the air out of the right ventricular outflow tract and improve the hemodynamics. Hypovolemia may present similarly to VAE, and if vigorous fluid administration is ongoing, the Doppler sounds may be difficult to interpret. In this situation, monitoring end-tidal nitrogen may help differentiate VAE from hypovolemia.

5. DI is a common complication of surgery for a craniopharyngioma [5–7]. It is caused by disruption of the ADH-secreting cells. Diagnosis is made when the patient produces a large volume of dilute urine in the face of hypernatremia. The diagnosis is confirmed when the serum sodium is >145 mEq/L, the serum osmolality is >300 mOsm/L, the urine output is >4 mL/kg/h, and the urine osmolality is <300 mOsm/L. Treatment, outlined above, is directed at replenishing urine output and maintaining normal serum osmolality. Since the administration of water is not an option in the anesthetized patient, dilute IV fluids can be given to

6. How much blood loss is acceptable prior to transfusion? Why? Are there alternatives? What are the risks?

7. The operation takes 10 h. Is the child a candidate for extubation in the OR? Pros/cons? How would you minimize straining at extubation? Do you anticipate hypertension at the end of the case? Is this a problem? Why? Prevention/treatment?

## Postoperative Care

### Questions

1. The urine output remains high postoperatively. How long do you anticipate this polyuria will persist? What treatment is indicated? What is vasopressin? Can you use it? How? Would DDAVP be an option in the immediate post-op period? Dangers?

2. Eight hours postoperatively, the child has a seizure. What is your differential diagnosis? What treatment is indicated?

replenish the excessive water losses in the urine. If D2.5 is used, hyperglycemia may result. If the serum osmolality remains high, an infusion of vasopressin offers the greatest flexibility in the maintenance of fluid balance. An infusion of vasopressin, starting at 1 mcg/kg/h, has begun and slowly increased until the urine output decreases to <2 mL/kg/h.

6. The lowest permissible hemoglobin depends upon the patient and the situation during the procedure. Measurement of an ABG or central venous blood gas ($SvO_2$) may give some information about the adequacy of oxygen delivery to the patient. Elevated serum lactate or lower than normal $SvO_2$ could indicate an imbalance between global oxygen delivery and oxygen consumption. The potential for continued bleeding is an important factor in deciding whether or not to administer blood/blood products. The risk of transmitting an infectious agent to a person via a transfusion varies from 1:100,000 for hepatitis A to 1:1–2,000,000 for HIV. Hemolytic transfusion reactions occur as often as 1:15–20,000 transfusions. Other results of transfusion include nonhemolytic transfusion reactions, urticaria or other allergic-type reactions, and possibly immunomodulation.

7. The usual criteria apply in considering whether or not to extubate this child. However, following neurosurgical procedures, it is important to assess neurological function and much easier to do so in an extubated, nonsedated patient. If opioids were a part of maintenance and the inhaled agents decreased as closure of the wound took place, straining and coughing prior to extubation should be minimal. Deep extubation is an option for this patient, but experience with this technique is essential prior to undertaking it. Also, the anesthesiologist must be certain that the child has a very good mask airway while anesthetized prior to performing a deep extubation.

**Answers**

1. DI may persist for several days following surgery for craniopharyngioma and may even be permanent [5]. Management using an IV infusion of vasopressin offers greater flexibility, but once longer-term therapy is indicated, the route of administration should be switched to intermittent IV and then intranasal. Oral desmopressin is available in addition to the intranasal form. The usual starting dose is ten times the intranasal dose.

2. Other postoperative complications seen after this procedure include hyperthermia and seizures. Retraction of the frontal lobes during this lengthy procedure may be responsible for this postoperative problem. On occasion anticonvulsants are begun intraoperatively and continued postoperatively. Hyperthermia may result from damage to the hypothalamic thermoregulatory mechanisms.

## Additional Topics

### Questions

1. What respiratory complications can you anticipate in the life of a child born with myelomeningocele? How does this happen with each problem? At what ages would you expect these various problems to present clinically?

2. A 3-year-old showed up in the pre-op clinic scheduled for the removal of an orbital dermoid. He has four cafe au lait spots on his trunk and buttocks. He has had headaches recently, and his pediatrician noticed his blood pressure was elevated (135/105 mmHg). Any more information needed? His mom has noticed some "flushing" spells recently. What do you think may be going on? There is a family history of neurofibromatosis. Are there any associated endocrinopathies?

3. What is the Glasgow Coma Scale? What is the maximum total score you can get? How many points do you get for being dead by neurologic criteria?

## Answers

1. Myelodysplasia is an abnormality of fusion of the neural groove during the first month of gestation. The resulting saclike herniation of the meninges is called meningocele, and if neural elements are contained within the sac, then it is called myelomeningocele. There are often accompanying abnormalities such as hydrocephalus, tethered cord, and Arnold-Chiari type II malformations present in these children. At birth, fluid losses through the defect may lead to dehydration. Intraoperatively, during the initial repair, high third-space fluid losses are an important consideration. Since the majority of myelomeningoceles are in the lumbar region, as the child grows older, the resulting urinary insufficiency leads to electrolyte abnormalities [8]. In addition, as they age, various bladder augmentations and other procedures are often done on these children leading to additional difficulties with electrolytes. The paralysis at and below the level of the lesion leads to the development of thoracolumbar scoliosis. As the scoliosis worsens, pulmonary function is impaired [9–11].

2. Neurofibromatosis is differentiated into two forms, NF-1 (90%) and NF-2 (10%). This patient has NF-1. This disease can affect nearly every organ system. The tumor characteristics of the condition are overgrowths of Schwann cells and endoneurium. Clinically, café au lait spots, axillary or inguinal freckling, neurofibromas, bone lesions, and optic gliomas are seen. Precocious sexual development is seen as a result of invasion of the glioma into the hypothalamus. CNS tumors account for significant morbidity in these children. In addition, the incidence of pheochromocytoma, rhabdomyosarcoma, Wilms' tumor, and leukemia is higher than in the general population.

3. The Glasgow Coma Scale is used to assess cortical and brainstem function. Patients dead by neurologic criteria get a GCS of 3. The GCS has been changed from what is used to evaluate adults for use in infants and children [12].
   *Infants*
   - Eye opening
     - Spontaneous 4
     - Opens to verbal stimuli 3
     - Opens to painful stimuli 2
     - No response 1
   - Verbal response
     - Coos and babbles 5
     - Irritable cry 4
     - Cries to pain 3
     - Moans to pain 2

- Motor response
  - Spontaneous purposeful movements 6
  - Localizes to pain 5
  - Withdraws to pain 4
  - Flexion to pain (decorticate) 3
  - Extension to pain (decerebrate) 2
  - No response 1

*Children*

- Eye opening
  - Spontaneous 4
  - Opens to verbal stimuli 3
  - Opens to painful stimuli 2
  - No response 1
- Verbal response
  - Oriented 5
  - Confused 4
  - Inappropriate words 3
  - Incomprehensible words 2
  - No response 1
- Motor response
  - Obeys commands 6
  - Localizes to pain 5
  - Withdraws to pain 4
  - Flexion to pain (decorticate) 3
  - Extension to pain (decerebrate) 2
  - No response 1

*Adults*

- Eye opening
  - Spontaneous 4
  - Opens to verbal stimuli 3
  - Opens to painful stimuli 2
  - No response 1
- Verbal response
  - Oriented to person, place, time 5
  - Confused 4
  - Inappropriate words 3
  - Incomprehensible words 2
  - No response 1

4. A patient with cerebral palsy and spasticity needs to have his heel cords (Achilles tendons) lengthened. Is he likely to have swallowing problems? How will you evaluate him for the potential to reflux and aspirate? Should he receive a rapid sequence induction? Which IV agents are best? Your choice of muscle relaxant? What if the child has “no veins”?

- Motor response
  - Obeys commands 6
  - Localizes to pain 5
  - Withdraws to pain 4
  - Flexion to pain (decorticate) 3
  - Extension to pain (decerebrate) 2
  - No response 1

4. Cerebral palsy is a static encephalopathy that has a changing clinical presentation over time. It is a disorder of posture and movement often associated with seizures, resulting from a lesion in the developing brain. Children with CP often have surgical procedures as treatment for contractures, scoliosis, gastroesophageal reflux, and other problems [13]. If a rapid sequence induction is planned, succinylcholine may be used. Its use in children with CP has been studied, and serum potassium increases as it does in patients without CP given succinylcholine [14]. In CP patients who are bedridden, the fact of very limited mobility may make them unsuitable for succinylcholine, as with all such patients. If there is no IV access, IM administration of ketamine, glycopyrrolate, and succinylcholine is an option.

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# Chapter 13
# Orthopedics I: Spine Surgery


**Robert S. Holzman**

A 12-year-old, 36 kg girl with cerebral palsy, spastic quadriparesis, and moderate-severe thoracic dextroscoliosis is scheduled for a T3-L5 posterior spinal fusion. She has a well-controlled seizure disorder (none in the last 8 years) and is fed by gastrostomy. She does have reflux. She is 3 weeks post placement of halo traction. The etiology of her cerebral palsy is a severe meconium aspiration at birth with cardiopulmonary arrest. She has had many anesthetics in the past for upper endoscopies, soft tissue tendon releases, Botox/phenol injections, and bilateral femoral and Pemberton periacetabular osteotomies. She takes baclofen 1 mg, twice a day. She is hypotonic and wears a neck brace for support. She also has sialorrhea. Her hemoglobin/hematocrit is 13.3 gm/dL and 41%.



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## Preoperative Evaluation

### Questions

1. What is the clinical significance of the different causes of scoliosis? What is different specifically about neuromuscular scoliosis? What longer-term comorbidities should you expect? How will you evaluate the respiratory as well as the cardiovascular system? Will you need any specific laboratory tests? Why? What other systems are you concerned about in this patient? Why?

## Answers

1. There are three main types of scoliosis – idiopathic, neuromuscular, and dystrophic. Idiopathic is the most common form and can occur in healthy children and adolescents, without a specific known cause. It affects girls eight times more than boys and often runs in families. There are three types that typically occur at different ages: infantile idiopathic scoliosis (children between birth and 3 years of age), juvenile idiopathic scoliosis (between 3 and 9 years of age), and adolescent idiopathic scoliosis (between 10 and 18 years old). Approximately 80 percent of all pediatric cases of idiopathic scoliosis fall into this category [1]. Neuromuscular scoliosis is the second most common form of scoliosis and is associated with disorders such as cerebral palsy, spina bifida, and spinal cord injury. Dystrophic scoliosis is associated with neurofibromatosis type I (NF-1) (there is also a nondystrophic form, which is treated similarly to idiopathic scoliosis).

   Dystrophic scoliosis is caused by bony dysplasia or intraspinal pathology and presents early and progresses rapidly. Pseudarthrosis is common. In this circumstance, a thorough workup for intraspinal lesions must be undertaken with a view toward early surgical intervention, as bracing is ineffective. The surgical approach is often anterior and posterior, with a generous fusion area.

   Scoliosis surgery most commonly involves prone positioning for posterior instrumentation or lateral positioning for anterior approaches. For combined approaches, it obviously involves both. There is extensive tissue trauma and considerable blood loss, which is usually greater in the more debilitated and complex patients who are often less mobile. Extensive bone dissection may result in fat or air embolism. In addition, there is the potential for spinal cord ischemia.

   The significance of a high degree of curvature is that as the curvature increases above 65 degrees, it produces rotational spinal deformity, narrowing of the chest cavity, and maldistribution of ventilation and perfusion. Progression of the rotational deformity may predispose to spinal cord ischemia during the surgical correction. The restriction of lung volumes from the primary disease process has its greatest effect on vital capacity. The vital capacity is further diminished by 60% on the first postoperative day and gradually recovers over the following 7 days. Further ventilatory compromise can occur from splinting of muscles due to inadequate pain control after surgery and diminished central respiratory drive from opioid analgesics. Therefore, many patients, particularly those with significant comorbid conditions, require postoperative ventilatory support.

   Respiratory function can be assessed by pulmonary function tests (PFTs) and a room air arterial blood gas (ABG) although these are not necessary for moderate forms of idiopathic scoliosis. I would use an ECG to evaluate rhythm abnormalities and conduction system abnormalities, particularly in patients with significant clinical impairment of the cardiorespiratory system.

2. Of what significance is the baclofen treatment for planning the anesthetic?

3. How would you evaluate the significance of the gastroesophageal reflux disease (GERD)?

## Intraoperative Course

### Questions

1. How will you induce anesthesia for this patient? What kind of access will you need? Any difference for a child with neuromuscular scoliosis than idiopathic scoliosis? Why? Is a central venous line necessary/a good idea? Why? Will end-tidal $CO_2$ sampling be accurate in this patient? Why/why not? Is it likely that evoked potentials will be used to monitor this patient? Why/why not? Are somatosensory (SSEP) as well as motor-evoked potentials (MEP) indicated? Is a wake-up test indicated? Despite being the "gold standard," what are its drawbacks?

Long-standing severe scoliosis can increase pulmonary vascular resistance by several mechanisms: reduced pulmonary vasculature and increased alveolar capillary pressure by compression from deformed ribs, hypoxemia, and hypercarbia. More advanced testing, when indicated by clinical findings, might include echocardiography for structural abnormalities as well as assessment of myocardial performance. A cardiac MRI would be useful if the chest is severely deformed and the echo imaging window is restrictive.

2. Oral baclofen, a muscle relaxant commonly used in conditions resulting in spasticity, has drowsiness as its most prominent side effect. In addition, it may lower seizure thresholds. It also has the typical but rare common side effects such as nausea, vomiting, GI upset, etc. Respiratory depression has been reported but has not been known to pose a problem for anesthetics. In patients with poorly controlled seizures, it may lower the seizure threshold and therefore increase the risk of perioperative seizures.

3. As usual, the risk of GERD should be evaluated in the context of frequency, severity, and precipitating conditions such as the eating of meals late at night vs. GERD occurring during an NPO interval. Depending on the frequency and seriousness of the problem, it may be prudent to attempt an intravenous placement and rapid sequence induction of anesthesia following preoxygenation.

**Answers**

1. A mask induction is fine provided that venous access can be readily established and the patient does not have significant active gastroesophageal reflux. These patients often have difficult IV access and a mask induction will enhance your ability to place an IV. In addition, they often have alterations in the geometry of their stomach and its relation to the lower esophageal sphincter because of prior abdominal surgery as well as an abnormal body habitus with a large upper body in relation to a smaller lower body. On the other hand, if reflux is severe, the potential for aspiration may overrule the patient's fear of needles. That said, the fear of needles is not trivial if it results in crying and aerophagia in the preoperative holding area. The fear of needles can be minimized, however, by providing oral anxiolytic premedication and the application of a topical anesthetic. In addition, difficult IV placement can be facilitated by ultrasound. In any case, large-bore IVs will be needed because the blood loss is likely to be significant because of the neuromuscular scoliosis. That makes it virtually mandatory to have an arterial line for the case. Strong consideration should be given to a central line or a PICC line for patients with difficult venous access who are undergoing major operations because they will need vascular access and blood draws for many days after surgery.

2. Having had numerous prior procedures, the patient is terrified of needles and requests a premed. Your response? Why? How do you justify it to the patient? Is administering a g-tube premed safe in a patient with reflux? What if she had a jejunostomy tube?

3. What anesthetic agents would you use for maintenance? Why? Would the use of somatosensory-evoked potentials affect your answer? Why/why not?

$ETCO_2$ may not be accurate in patients with severe scoliosis. With asymmetrical ventilation, one lung will be more compressed, will have a higher airway resistance, and will be more poorly ventilated and therefore hypercapnic (reservoir of $CO_2$), while the other lung will have a lower airway resistance and will tend to be hyperventilated and therefore relatively hypocapnic. Because of the differential lung emptying as well as the different reservoirs of $CO_2$, there will be a "double hump" on the $ETCO_2$ trace [2].

It is very likely that evoked potential monitoring will be used, and depending upon the patient's neuromuscular competence, it is likely that somatosensory-evoked potentials, measuring the integrity of the dorsal (sensory) pathways, as well as motor-evoked potential monitoring will be used. While almost all known anesthetic agents cause a decrease in both amplitude and latency in both motor-evoked potentials (MEPs) and somatosensory-evoked potentials (SSEPs), it is still possible to obtain signal reproducibility with low doses of volatile agents. The alternative is total intravenous anesthesia (TIVA) with propofol as the hypnotic agent. Motor-evoked potentials (MEPs) use magnetic stimulation of the motor cortex and detection of action potentials in the corresponding muscle groups. The wake-up test monitors voluntary motor function of the lower limbs once the vertebrae have been distracted. It involves a gradual decrease in anesthesia depth to the point when patients are able to respond to a verbal command. The end point for the wake-up test is voluntary movement of the lower extremities in response to a verbal command, so patients must be prepared for and capable of executing this request.

Notwithstanding the fact that it is the "gold standard," because it is not a continuous monitor and is only a point in time, spinal and ischemic events occurring before and after the wake-up test may not be detected.

2. A prepyloric gastrostomy tube will not necessarily prevent aspiration, which depends more on the geometry of the stomach, the integrity of the lower esophageal sphincter, and esophageal motility. A post pyloric jejunostomy has a better chance of minimizing aspiration risk, but aspiration may still, although rarely, occur [3]. A premed is probably a good idea for this veteran of the operating room.

3. My choice would be a balanced anesthetic consisting of a potent opioid such as fentanyl or sufentanil supplemented with a low dose of volatile agent. Beginning with a mask induction, at least one IV could be placed and, if chosen, a short- to medium-acting nondepolarizing muscle relaxant could be given to facilitate intubation. This single dose will be metabolized while additional lines and a Foley catheter are placed and before motor-evoked potentials will begin. Deep intubation with a volatile agent without a muscle relaxant can also be considered, if appropriate. It is important to check with the neurophysiology monitoring technician regarding the effect of any anesthetic technique chosen on the reliability of the signals compared to baseline [4].

4. During surgery the patient is bleeding more than usual; you are about halfway through and she has lost 750 mL. What concerns do you have? Is this typical for patients with neuromuscular scoliosis? What measures can you take to decrease the bleeding? How does tranexamic acid work?

5. The neurophysiology monitoring technician informs you that there has been a decrease in the amplitude and latency of the SSEPs. How do you interpret these findings? What are the possible causes? How can you systematically address them? Should you proceed to a wake-up test? Why/why not?

## Postoperative Course

### Questions

1. As the patient begins pulling at the endotracheal tube at the end of the case, you loosen the tape, and she vomits. With retching, you can hear airflow in the pharynx, and the patient phonates. Your considerations? What therapeutic interventions will you make? Why?

4. The loss of 750 mL of blood represents about 30% of the circulating blood volume, and this will decrease her Hct from 41% to 28%. Depending on the volume of blood lost per unit time and the rate of fluid replacement, she has a large blood volume loss and as the case goes on will lose oxygen-carrying capacity. If autologous blood were available, it would be ideal, although not completely free of risk because of the chance of administrative error. Hetastarch may also be utilized as a volume expander. However, infusion of more than 1L may cause platelet dysfunction and increase surgical bleeding. Intraoperative blood conservation can be achieved by salvaging red blood cells with Cell Saver technology, acute hemodilution, and deliberate hypotension, although only a mild amount should be utilized during anemic states. Autologous blood donation is probably the most effective and safest strategy.

   Antifibrinolytics such as tranexamic acid (TXA) have proven efficacy with regard to blood conservation during surgery [2]. Tranexamic acid is a synthetic derivative of the amino acid lysine that exerts its antifibrinolytic effect through the reversible blockade of lysine-binding sites on plasminogen molecules. Intravenously administered tranexamic acid has decreased blood loss and transfusion requirements in patients undergoing cardiac surgery with cardiopulmonary bypass, in patients with upper gastrointestinal bleeding, after oral surgery in patients with hemophilia, and in patients undergoing orthotopic liver transplantation or transurethral prostatic surgery.

5. Volatile anesthetics tend to affect the amplitude of the SSEP more than the latency [3]. When significant changes occur, the cause may be technical, physiologic, anesthetic, or patient-related (positioning, related to surgery). All of these possibilities should be investigated. The end-tidal volatile anesthetic concentration should be reduced. The mean arterial pressure should be checked and, if needed, raised. In addition, the SSEP in other limbs should be checked.

## Answers

1. The endotracheal tube has been dislodged from the glottis. Preparation has to be made for suctioning of secretions and assisting ventilation with 100% oxygen via a facemask. The patient's mental status may permit coughing on command or reflexive coughing if she is close enough to emergence anyway. If she remains unresponsive and is unable to cough reflexively or voluntarily, she may need to be reintubated with a rapid sequence induction with cricoid pressure.

2. After the patient is in the PACU for an hour, she appears more difficult to arouse than before, although her heart rate is slow (60) and she appears comfortable. Her oxygen saturation is 94% with a simple mask with 5 L/minute flow. Your considerations? Why? How would you evaluate the patient? How would you intervene in this patient, specifically? How do perioperative respiratory considerations differ for patients with neuromuscular scoliosis? Should she go to the ICU overnight? What would you monitor specifically? Would pulmonary function tests help?

## Additional Topics

### Questions

1. A 14-year-old female has a history of neurofibromatosis (NF1) and progressive dystrophic scoliosis. What differences are there between dystrophic and idiopathic scoliosis? The spine surgery will require an anterior release as well as a posterior spinal fusion with multiple sublaminar wires. Any specific implications for SEP monitoring? For anesthetic technique? Any specific comorbidities that you would be concerned about with NF1?

2. A 12-year-old with Duchenne muscular dystrophy, wheelchair bound, and progressive scoliosis requires spine fusion. Why? What is the significance of being wheelchair bound at 12 yo? Any likely comorbidities of his DMD? His resting heart rate is 110 – is that reassuring? What is the significance of tachycardia in DMD? His echocardiogram shows an EF of 48%, a reduced velocity of circumferential fiber shortening corrected for rate (VCFc) of 1.2, and a shortening fraction (SF) of 23%. Is this helpful? Do you expect any airway difficulties?

2. An oxygen saturation of 94% with supplemental oxygen being delivered suggests that she has significant shunting and perhaps some alveolar collapse. After evaluating her chest exam, it would be reasonable to obtain a chest X-ray and look for areas of consolidation or collapse and proceed with a standard evaluation for postoperative respiratory care. It might involve incentive spirometry, repositioning, CPAP or BiPAP application, noninvasive mask-based ventilation, or even BiPAP or noninvasive ventilation during the perioperative period. Neuromuscular scoliosis patients, because of their primary disease, should be anticipated to require more postoperative support over a longer period of time. She would likely need a more closely monitored environment like the ICU or a respiratory care step-down unit. Pulmonary function testing would be a quantitative adjunct if there were baseline studies to compare, but in the end, it is a clinical decision with regard to respiratory adequacy during recovery.

**Answers**

1. Dystrophic as well as nondystrophic scoliosis can occur with NF-1 [4–6]. Dystrophic scoliosis occurs earlier and is typically more severe, as well as associated with bony dysplasia or intraspinal tumors. Nondystrophic scoliosis is more similar to idiopathic scoliosis, although this may vary. Dystrophic curves are typically more sharply angulated and the spine is usually more rigid. Ectasia, or thinning of the dura, is a common finding. Dystrophic scoliosis will often require an anterior release and a posterior spinal fusion, which may be done as two sequential operations or in one step. It is crucial to intervene early in the dystrophic form because of its rapid progression and secondary organ system effects. The role of the sublaminar wires is to distribute correctional force more equitably and therefore stabilize and distribute stress more equitably across the instrumentation and fusion/repair. The importance of utilizing an anesthetic technique that preserves the highest-quality signal for evoked potential monitoring during sublaminar wire tightening is that at least 50% of true positive changes in EPs occur during tightening. Careful attention at this time to dose of volatile anesthetic, neuromuscular competency, and blood pressure is crucial.

2. Neuromuscular scoliosis due to muscular dystrophy is a result of progressive muscle incompetence and resulting weakness. Axial imbalance leads to unbalanced mechanical forces on the spine [5, 6]. Being wheelchair bound at 12 years old, a relatively common finding in DMD, suggests that his disease process has progressed steadily and significantly and that he may be at substantial risk for comorbidities such as cardiac and respiratory involvement as well as progressive scoliosis. Not all forms of muscular dystrophy are accompanied by parallel progression in cardiac disease; good cardiac function may stay relatively well preserved, or patients with milder forms of muscular dystrophy may, in fact, have substantial myocardial impairment. A resting heart rate of 110 in a 12-year-

3. What are the risks for an adolescent with symptomatic spondylolisthesis? What signs and symptoms would concern you as the anesthesiologist? Any special monitoring or anesthetic technique considerations?

4. A 9-year-old girl with juvenile ankylosing spondylitis of the cervical spine has just undergone a cervical spinal fusion and instrumentation from C2 to C7. She was a grade 3 laryngeal view following induction, and she was electively intubated with a fiber-optic bronchoscope without any difficulty. The case proceeded uneventfully over the course of several hours, with a blood loss of 150 mL during an otherwise remarkable repair. She was in the prone position the entire time. Do you anticipate any difficulty with extubation? Would you consider this a "difficult extubation"? Is there anything you can do to assess any anticipated airway difficulty? Any devices you could have available to lessen the risk? What could you do clinically to evaluate the airway with regard to extubation safety? What is the data on extubation risk in this population?

old may or may not be reassuring; a resting heart rate of 60–100 in "normal" children over 12 years of age is normal. On the other hand, patients with progressively compromising DMD tend to have a high incidence of sinus tachycardia. It is best to obtain indices of myocardial contractility echocardiographically in patients with symptomatic muscular dystrophy. It is crucial to remember that dilated cardiomyopathy can develop even in mild cases of muscular dystrophy. Systolic performance is often abnormal; about half of patients have a reduced (<28%) shortening fraction and reduced VCFc [7]. Evaluation of load-independent measures of contractility (the end-systolic wall stress-VCFc relation) tends to be lower. Cardiac conduction defects are also associated with Emery-Dreifuss muscular dystrophy and may require pacing [8]. DMD patients can have a difficult airway based on progressive infiltration of the tongue, especially the base of the tongue, with fibrotic muscle, as well as limitations in movement of the head and neck from prolonged immobility and fibrous infiltrate.

3. Spondylolysis is a stress fracture of the pars articularis of the fourth or fifth lumbar vertebrae. Risk factors include genetic predisposition and overuse activity such as gymnastics, with repeated hyperextension of the lumbar vertebrae. Subluxation of one vertebra from another due to spondylolysis results in spondylolisthesis. This condition can be associated with compression of the spinal nerves and a narrowing of the spinal canal. The treatment of spondylolysis is always conservative but surgical treatment is sometimes necessary. The treatment for spondylolisthesis is posterior or anterior fusion with instrumentation over two to three vertebrae. Most adolescents can be treated conservatively, but those who demonstrate neurologic signs or a high-grade slip (>50%, progression of the slip and lack of response to conservative measures) or postural deformity or gait deformity are candidates for surgery. Patients are generally healthy, similar to those with idiopathic scoliosis. Neuromonitoring can be used during the case but a wake-up test is not usually necessary. An arterial line is optional, and surveillance lab work intraoperatively is not usually necessary because the blood loss is typically less than 500 mL. Similar considerations for anesthetic technique apply because of the use of SSEP and MEP monitoring.

4. Ten to 20% of patients with ankylosing spondylitis become symptomatic before 20 years of age, with a female/male ratio of 2–3:1. It is characterized by inflammation of skeletal ligamentous bony insertion sites especially in vertebral and sacroiliac articular structures. Intervertebral disks calcify and syndesmophytes form and attach to ligaments, bridging across calcified intervertebral disks. The inflammation results in an ingrowth of synovial material into joints with destruction and joint fusion. There may be autoimmune comorbidities such as polyarthritis, aortic valve thickening, aortic regurgitation, aortitis, restrictive lung disease, renal amyloidosis, and uveitis or iritis. Notwithstanding the relative ease of tracheal intubation, the results of prone positioning, cervical spine instrumentation, and local edema will narrow the anteroposterior diameter of the

oropharynx.The postoperative edema ± the potential for hematoma along dissection planes may make extubation difficult and reintubation impossible.

Available clinical options for the "difficult extubation" consist of waking the patient up completely so that you have a reliable mental status with regard to following commands and self-reporting adequacy of breathing. It is helpful to test for a "leak" by deflating the cuff of the endotracheal tube, although a direct laryngoscopy will provide more information about swelling around the laryngeal inlet. Most patients are successfully extubated following cervical spinal fusion; extubation failures (in adults) are more frequently associated with long surgical times and significant blood loss. An excessive cervical fixation angle may also contribute to difficulty in postextubation breathing as well as direct laryngoscopy for reintubation. Finally, an airway exchange catheter can be utilized for several minutes postextubation to ensure the patient's capability of breathing with a natural airway. The airway exchange catheters are typically very well tolerated in patients in the immediate postoperative period.

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# Chapter 14
# Orthopedics II


**Robert S. Holzman**

A 6-year-old girl is scheduled for a Pemberton osteotomy for developmental dislocation of the hip (DDH). She is otherwise well, in first grade, and, although a little nervous, very conversational.



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## Preoperative Evaluation

### Questions

1. How common is this form of hip dysplasia? How does it occur? Is it an isolated finding, or should you expect syndromic associations? Risk factors? What is the goal of the surgery?

2. How is this disorder treated at various ages? Why?

3. Do you require any preoperative lab work? What are the risks/benefits?

## Answers

1. Developmental dysplasia of the hip (DDH) refers to abnormal development of the acetabulum, femur, labrum, and capsule. At late gestation, femoral head grows more rapidly than the acetabular cartilage so that at birth the femoral head is less than 50% covered. The acetabulum is at its most shallow and most lax in order to maximize hip ROM which facilitates the delivery process. After several weeks, acetabular cartilage develops faster than the femoral head, which allows progressively more coverage. The femoral head becomes displaced from the acetabulum in the perinatal period, which results in abnormal development of the hip joint. It is more common in females, firstborn children, and breech deliveries. Despite the routine use of Barlow's test (posteriorly dislocatable hip) in the newborn exam, less than 2% of infants with DDH will have a positive Barlow's test. The vast majority of unstable hip examinations improve by age 2 months (60% at 1 month; 88% at 2 months). Several neuromuscular syndromes such as meningomyelocele, arthrogryposis, and cerebral palsy can be associated with hip dislocation. Children who are prepubertal or pubertal are at risk for slipped capital femoral epiphysis (SCFE). The goals of the surgery are to stabilize the hip joint to minimize early osteoarthritis.

2. Treatment of infants younger than 6 months is usually done with bracing in a Pavlik harness. Closed reduction is considered after 6 months of age in children with hip dislocation, and open reduction is usually done after 2 years of age because of the risk of avascular necrosis and the increased failure of closed reduction. At this point, open reduction with a pelvic osteotomy, usually through an anterolateral approach, is used.

3. Because there are no associated syndromes or comorbidities, it is reasonable to proceed without preoperative lab work. A baseline hematocrit can be obtained at the time the IV is placed, and if needed, a type and screen or crossmatch can be set up at that time.

## Intraoperative Course

### Questions

1. Your anesthetic plan for induction? Does the patient need premedication?

2. What kind of access do you plan? Any special considerations with regard to patient positioning? Is an arterial line necessary? A central venous line?

3. What is your strategy for blood conservation? Controlled hypotension? Hemodilution? Cell Saver?

4. What is your approach for a regional anesthetic for this patient? Is it necessary to avoid neuromuscular blockers for the block?

**Answers**

1. The preoperative conversation with the family will determine the need for premedication. This is usually an easy age group to work with, and if the child is conversational with strangers, having been in school for a few years, it is often easy to find some common ground to talk about. A parent-present induction without premedication would probably work out well, for this patient.

2. While it is extremely unlikely to require a transfusion, there is nevertheless a steady amount of bleeding during the osteotomies which may be underappreciated unless the surgical field is carefully watched. I would place two relatively large IVs after induction. I would not anticipate any sudden blood loss nor massive blood loss, and in a healthy child, the need for an arterial line or a central venous line to monitor gas exchange or metabolic status would probably not be necessary. It is important to consider this question because of the restricted extremity access for this procedure, so you need to be relatively sure from the beginning. Another factor is the age/size of the patient – the smaller the patient, the more likely that an arterial line would be an added safety benefit.

3. Blood conservation strategies should include the use of a Cell Saver and tranexamic acid. Intraoperative cell salvage can reduce allogenic blood transfusion by 50% in adults; however, the smaller the patient, the less likely this will make an impact on this statistic because the red cell mass that is salvaged is simply very small – unless there is massive blood loss [1, 2]. Nevertheless, blood transfusion was significantly reduced in volume and frequency when used in craniofacial, major orthopedic surgery (acetabuloplasty), scoliosis correction, and complex cardiac surgery [3]. Continuous systems are faster and require only small volumes of salvaged blood for processing and produce blood with high hematocrit, making them more suitable for pediatric cases. Tranexamic acid is a very useful strategy for decreasing blood loss and should be routinely used [4]. Controlled hypotension and hemodilution are techniques that have largely been replaced.

4. A regional anesthetic as a component of perioperative care is very useful. Lumbar plexus blocks as an adjunct to light general anesthesia for the procedure as well as postoperative analgesia are rapidly replacing epidural blocks in some centers. Some advantages of the more specific nerve block include a shorter duration of urinary catheterization and earlier ambulation. The majority of these blocks are now done with ultrasound guidance, but rarely, they need to be confirmed with the use of a nerve stimulator; therefore, it is good to avoid neuromuscular blockade until after the block is in. We typically use a continuous catheter technique as a way to provide perioperative continuous infusion.

5. How do you plan to keep this patient warm?

## Postoperative Course

### Questions

1. The patient is crying and in obvious pain in the PACU following the procedure. Should the lumbar plexus block be providing total anesthesia for this procedure? What areas does it not cover? The surgeon comes by and wants to give the patient diazepam – is this a rational suggestion?

2. Despite your best intraoperative efforts, the patient arrived in the PACU with a temperature of 34.8 °C and is shivering so badly that his teeth are chattering and there is no tracing on the ECG. Is this temperature dangerous? Would meperidine help? Why? Is it likely that an equianalgesic dose of fentanyl or morphine would work just as well?

5. Heat loss is a major consideration – there is much surgical exposure, the rooms are typically cold in order to keep personnel comfortable because they are covered in many layers including lead aprons, and because of patient size, there is not much surface area to heat. The wide prep involves the legs up to the nipple line. The forced hot air warmer is typically utilized as well as the use of low fresh gas flows or a closed-circuit anesthetic technique, reducing fresh gas flows to the minimum amount to meet metabolic needs. The most important aspect of keeping the patient warm is to not lose the heat in the first place, so these strategies should begin after induction, and not wait until the patient is prepped and draped and the surgeon is ready to begin – that can often be an hour after induction!

## Answers

1. A lumbar plexus block provides analgesia to the three major nerves of the lumbar plexus, blocking sensation to the upper anterior leg and the lateral femoral cutaneous nerve. This block also anesthetizes the distal branches of the lumbar plexus, including the iliohypogastric, ilioinguinal, and genitofemoral nerves that innervate the groin area. The gluteal area and the posterior thigh are not blocked by a lumbar plexus block but rather a sciatic block, which is not routinely done in these patients, so there may very well be some spasm in the muscles of the posterior thigh. Diazepam is a very rational approach to treat spasm in this area (which may remain undetectable by physical diagnosis) and can be given in incremental intravenous amounts in the PACU until an effect is obtained.

2. Meperidine would likely help decrease the shivering. Although the mechanism is not totally understood, it is thought that the special antishivering efficacy of meperidine results at least in part from an uncharacteristically large reduction in the shivering threshold rather than from an exaggerated generalized thermoregulatory inhibition. This pattern differs from that produced by alfentanil, clonidine, propofol, and the volatile anesthetics, all of which reduce the vasoconstriction and shivering thresholds comparably [5, 6].

## Additional Topics

### Questions

1. A healthy 17-year-old high school football player wants to wake up with as little pain as possible following his ACL reconstruction and is particularly interested in regional anesthesia as an adjunct. He makes it clear he wants a general anesthetic as his primary anesthetic. What can you offer him as a plan?

2. What is a club foot? At what age do you typically encounter these patients? Any special considerations for their anesthetic? What are the advantages and disadvantages of offering a regional anesthetic as an adjunct?

**Answers**

1. There are several effective approaches to local anesthetic/regional techniques as adjuncts to perioperative analgesia. First, it is important to acknowledge that a regional block is not required for effective postoperative analgesia; much success has been achieved with multimodal pain approaches, particularly as they have been used to reduce opioid requirements. That said, the earliest consistent nerve blocking technique was the femoral nerve block, introduced about 20 years ago. A few years later, an adductor canal approach was introduced, in an effort to improve ambulation postoperatively. Recently, comparative studies have examined these two techniques, which appear to be equally effective in analgesia, with simple instillation of local anesthetic in the joint space. All three appear to be, at least statistically, without significant difference. It is likely that the optimal combination would include multimodal approaches along with nerve block or instillation [7–9].

2. Club foot (talipes equinovarus) occurs in 10:10,000 births with equal prevalence in males and females. There can be neurogenic causes (arthrogryposis, spina bifida, and tethered cord), connective tissue disorders (Larsen syndrome), and mechanical causes (oligohydramnios, intrauterine band disorders), or it can be an isolated idiopathic deformity. These associations should be carefully considered in the preoperative evaluation. These children are seen in infancy. Talipes equinovarus surgery involves lengthening some tendons and releasing tight ligaments in order to place bones and joints in normal position. Surgery is generally done during infancy, and the foot is placed in a cast to maintain the correct position while the tendons and ligaments heal. There is minimal blood loss during surgery because surgical tourniquets are used. Regional anesthesia can be considered, but only after weighing the benefits and risks. What has to be kept in mind is that pain control can be accomplished with narcotic analgesics and NSAIDs. Children who have talipes equinavara surgery are usually placed in bilateral short leg casts postoperatively and are at risk for postoperative swelling which can cause nerve, muscle, and skin damage. These children need to be monitored closely for this complication. A regional block can interfere with evolving signs of such complications in an infant. As long as this is continuously evaluated, then a caudal or lumbar plexus block would be reasonable choices. There are occasions and associated comorbidities when a regional anesthetic is preferred as the primary anesthetic, with supplemental sedation [10].

3. A 5-year-old with a right Sprengel deformity requires correction. What else would you like to know, as the anesthesiologist? How does this anomaly develop? Should you expect any associated disorders?

3. The Sprengel deformity is the most common congenital malformation of the shoulder girdle with a male to female ratio of 3:1. The condition is usually sporadic although it can be familial with an autosomal dominant pattern of inheritance. Embryologically, the scapula begins as a cervical appendage that differentiates opposite the fourth, fifth, and sixth cervical vertebral bodies at approximately 5 weeks of gestation. Normally it descends into the appropriate position approximately 90 days later. Any obstruction to this migration will result in a hypoplastic, elevated scapula. There may be some limitation of motion due to an omovertebral connection between the medial border of the scapula and the lower cervical spinous process. This rhomboid- or trapezoid-shaped cartilage or bone can be found in approximately one-third of these patients. Additionally, the prescapular muscles may be contracted and fibrotic. These deformities can vary in severity and have been classified by Cavendish:

   - Grade 1 Very mild. Shoulders almost level. Clothing masks deformity.
   - Grade 2 Mild. Shoulders almost level, but superomedial portion of high scapula is visible.
   - Grade 3 Moderate. Shoulder is elevated 2–5 cm higher on the affected side.
   - Grade 4 Severe. Scapula is very high with superomedial angle at occiput with neck webbing and brevicollis.

   The Sprengel deformity is usually associated with other malformations such as fused ribs, chest wall asymmetry, cervical ribs, congenital scoliosis, and cervical spina bifida. It can also be associated with a number of syndromes: Klippel-Feil sequence, Moebius sequence, Poland sequence, VACTERL, velocardiofacial syndrome (22q11 deletion), and Goldenhar syndrome [11].

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# Chapter 15
# Congenital Bone and Connective Tissue Disorders


**Robert S. Holzman and Joseph P. Cravero**

A 1.5-year-old, 6 kg, with osteogenesis imperfecta is on the add-on list for open reduction internal fixation of a left femur fracture using an elastic nail. His preoperative temperature is 39.4 °C. He has no signs of a URI and, in fact, doesn't look sick. He has a long arm cast on his right arm for a humerus fracture from last week. Despite attempts in the ER, no one succeeded in placing an IV before he arrived in the pre-op holding area.



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## Preoperative Evaluation

### Questions

1. He has no signs of a URI; how will you evaluate this? Could this be a urinary tract infection? Is this temperature normal? Why?

2. What else will you evaluate preoperatively? What if his platelet count is normal, but he has a history of easy bruising as well as easy breaking? How can platelet aggregation abnormalities be evaluated? How will it affect your anesthetic planning and management? Hematologic support? Does he need platelets?

3. Any anticipated airway concerns about this patient?

## Intraoperative Course

### Questions

1. What is your induction plan? Does it include a pre-induction IV? Assume no pre-induction IV; what else are you planning? What are your considerations for an inhalation induction? How will you judge adequate anesthetic depth to start an IV? What are the limitations in your access?

## Answers

1. The preoperative evaluation should rule out pulmonary and other infections. This temperature can, however, be normal for children with osteogenesis imperfecta. Hyperthermia occurs in OI patients due to a baseline hypermetabolic state from the high turnover of bone degeneration and reformation.

2. The tendency toward bruising is due to defect in platelet adhesion; the platelet count is expected to be normal. Bleeding time is a good clinical indicator of this platelet aggregation defect, which can cause excessive bleeding during surgery. Transfusion of platelets will improve coagulation function and minimize surgical bleeding. Platelet transfusion is indicated if there is unexpected excessive intraoperative bleeding.

3. Patients with the more severe forms (and this patient probably has OI type 3, the most severe) typically have triangular facies with frontal bossing. This can lead to a difficult mask fit as well as a difficult intubation. Further intubation difficulties may be as a result of a small mouth and a large occiput. Patients with basilar impression, a craniovertebral junction anomaly with invagination of the margins of the foramen magnum upward into the skull, are at risk for neurological compromise, especially with neck flexion during intubation.

## Answers

1. A smooth induction is the best induction, particularly in osteogenesis imperfecta patients.

   Inasmuch as IVs have already been attempted, the patient will not likely welcome any further attempts before anesthetic induction, so I would begin with a mask induction. If it is helpful to have a parent there (and in this age group it is likely to be), then I would proceed with a parent-present induction. This is particularly important because the parent can handle the physical aspects of any restraint, if necessary, rather than well-meaning strangers who have not handled this particular patient. It is reason to try to induce in a semi-upright position because of the emergency nature of the case, a chance to minimize the risk of regurgitation, and improved diaphragmatic excursion and therefore a little bit of preservation of the functional residual capacity. The usual criteria for signs of depth apply, from the loss of consciousness and the eyelash reflex to softening of the abdominal muscles, to a decrease in blood pressure and moderate slowing of the heart rate. Once this is done, then vascular access should be established, preferably in the left upper extremity, but if needed and after discussion with the surgeon, the right lower extremity may be used.

2. He is 6 kg and hyperthermic; how will you manage his fluid therapy? What kind of IV fluid do you think he needs?

3. Examining this baby was difficult when he was awake; now you see that his neck mobility is extremely limited in flexion, extension, and rotation. His tongue is large. How would you secure his airway? Would neuromuscular blockade help?

4. One of your partners suggests that this patient needs an arterial line instead of a blood pressure cuff. Your thoughts?

## Postoperative Course

### Questions

1. The patient has features of a hypermetabolic state, likely ventilatory restriction due to rib malformations and fractures as well as impaired muscle strength, and perhaps inadequate nutrition. How will you know if it is safe to extubate?

2. Fluid replacement requires careful management because some patients with OI may experience hyperhidrosis and may need additional fluid replacement of insensible losses. In addition, there will be a greater requirement for free water in hypermetabolic states, approximately a 7% increase for every degree C. This 6 kg 18-month-old is very small for his age and it is likely that muscle and liver glycogen store are not very large, so hypoglycemia is a concern. A maintenance glucose-containing solution should be provided and the blood glucose should be checked intraoperatively.

3. Since this baby has had other anesthetics, much can be gleaned from his prior airway management such as whether he was easy to mask, required a nasal or oral airway, needed positive pressure to maintain his airway, etc. I would expect that during the inhaled induction he might preserve his ventilatory drive a bit better because with an elevation in his oxygen consumption because of his temperature, he would also have an increase in his $CO_2$ production and therefore his minute ventilation. Of course, this will be opposed by the progressively rising level of end-tidal anesthetic agent. It would be worthwhile, if his respiratory drive is well defended and you are able for him to tolerate a deep inhaled anesthetic, to take a look with a videolaryngoscope. This will give a high-fidelity view of the airway and at the same time enable control of his large tongue. This should all be done after venous access is established. At this point, when a reassuring view of the larynx is obtained, I would go ahead with neuromuscular blockade in order to facilitate intubation and minimize laryngeal reflexes.

4. An arterial line is a safe alternative to a blood pressure cuff if the OI is severe. Alternatively, the use of an ultrasonic Doppler or aneroid strain gauge blood pressure device allows the use of the lowest inflation pressures. Repeated inflation of the blood pressure cuff to high pressures such as with the use of oscillometric device may cause fracture of the fragile bones.

## Answers

1. Based on the procedure, the goal is to extubate at the end of the case. Most of these patients have adapted remarkably well to a disease they have had their whole life, even though that life (1.5 years) has been short. Although anatomic dead space may not be “normal,” ventilation-perfusion matching has had a lifetime to compensate, so these patients, on the whole, do remarkably well postoperatively. Nevertheless, it is important to recognize that they may need special nursing care in the postoperative period, and an ICU “step-down” unit may be appropriate overnight. Most importantly, it is important to educate everyone taking care of the postoperatively about their tendency for bone fractures and the need for careful positioning and padding. The involvement of the parents in post-op care is extremely important [1].

## Additional Topics

### Questions

1. Describe the findings associated with the following radial dysplasia and associated syndromes:
   - Radial aplasia and Hallermann-Streiff syndrome
   - Radial aplasia and Cornelia-de Lange syndrome
   - Radial aplasia and Holt-Oram syndrome
   - Radial aplasia and Fanconi anemia
   - TAR syndrome
   - VATER syndrome

2. A 6-month-old comes in for a hernia repair; he has rickets. What are the anesthetic implications? Any lab tests needed for this particular patient? Which ones? Is there any particular anesthetic technique that would be safer than any other? Why/why not? Risk of postoperative mechanical ventilation? Why is the serum calcium level typically normal? Then how is the metabolic component of the diagnosis made?

## Answers

1. Radial dysplasias and associated syndromes:
   - Radial aplasia and Hallermann-Streiff syndrome: narrow upper airways, obstructed nares, micrognathia, tracheomalacia, obstructive sleep apnea and cor pulmonale, and structural heart disease. Bird-like facies and ocular defects such as microphthalmia, cataracts, coloboma, glaucoma, and retinal degeneration.
   - Radial aplasia and Cornelia-de Lange syndrome: short neck (66%), anticipate difficult airways, and difficult access. Severe developmental delay. High-arched palate.
   - Radial aplasia and Holt-Oram syndrome: an autosomal dominant heart disorder (ASDs, VSDs, and conduction system defects) associated with skeletal malformations including hypoplastic thumb and short forearm. Patients may have radioulnar synostosis, accessory carpal bones, or carpal coalition or tarsal coalition. Hyperphalangism and preaxial polydactyly are additional anomalies seen in some patients.
   - Radial aplasia and Fanconi anemia: skeletal defects in association with bone marrow failure. Microcephaly with ptosis, strabismus, and microphthalmia may occur, along with hydrocephalus. Patients have short stature, with small or aplastic thumbs and radial aplasia, clinodactyly, syndactyly, or other radial abnormalities. There can be additional axial defects in the ribs or vertebral bodies, as well as abnormalities of the kidneys and genitals.
   - TAR syndrome (thrombocytopenia-absent radius syndrome): similar to Fanconi anemia, it may be associated with micrognathia and congenital heart disease (tetralogy of Fallot, coarctation, ASD). Bilateral radial aplasia ± ulnae. The thumb is always present. Thrombocytopenia with diminished or absent megakaryocytes in the bone marrow.
   - VATER syndrome: V (vertebral anomalies), A (anal atresia), T (tracheoesophageal fistula), E (esophageal atresia), R (radial and renal dysplasia) in any combination. The VACTERL association is an acronym for V (vertebral anomalies), A (anal atresia), C (cardiac malformations), T (tracheoesophageal fistula), E (esophageal atresia), R (renal anomalies), and L (limb anomalies).
2. Patients with rickets usually have hypocalcemia that may cause arrhythmias and potentiate nondepolarizing muscle relaxants. Patients with untreated rickets may have soft bones, which predispose to fracture with mild pressure. Poor muscle tone is associated with kyphosis. Lab tests should include serum ionized calcium, phosphorus and alkaline phosphate concentrations, and parathyroid hormone. Avoidance of muscle relaxants may reduce the potential of enhancing the muscle weakness. Hypocalcemia and hypophosphatemia potentiate nondepolar-

3. An 11-year-old boy with achondroplasia is scheduled for a leg-lengthening procedure. He has no history of prior surgery and is otherwise healthy and active. What anesthetic concerns do you have about this form of dwarfism? The patient can extend his neck through an arc of 15–20°. Does this influence your choice of anesthetic induction techniques? Does this patient need soft tissue films of his neck? Why/why not? An airway MRI? C spine films? Why/why not? How will it affect your anesthetic management? Should this patient be extubated (assume odontoid hypoplasia and decreased neck range of motion). Why/why not? Is it safer if he pulls his tube out in the middle of the night in the ICU? What criteria will you use for extubation?

izing muscle relaxant effects. The need for postoperative ventilatory support is high due to baseline poor muscle tone, restrictive chest wall disease (pectus, scoliosis), residual inhalation anesthetic effect, and postoperative opioid-induced suppression of central respiratory drive. The serum calcium is normal because of the compensatory increase of parathormone secretion in response to the initial low serum calcium. Increased parathormone secretion mobilizes calcium and phosphorus from the bone (producing osteomalacia), normalizes serum calcium, raises the serum alkaline phosphatase, and lowers serum phosphorus due to inhibition of reabsorption of phosphorus from the renal tubules. The metabolic component of rickets is completed by analysis of plasma calcium, phosphorus, alkaline phosphatase, and vitamin D hepatic and renal components (vitamin D3, D2, 25(OH)D3, 1,25(OH)2D3, 24,25(OH)2D3).

3. Failure of bone growth in this syndrome may result in stenosis of the foramen magnum and spinal canal and vertebral malalignments, most likely in the lumbar spine. Embryologically, the problem is primarily a defect of mesoderm, i.e., the failure of the cartilage to form bone. This patient does not need soft tissue films of the neck. Achondroplasia is a disease of underdevelopment of bone. Mid-face hypoplasia is expected with this disorder. MRI of the airways can be helpful to determine the extent of airways involvement. As a disorder of bone underdevelopment, achondroplasia can be associated with dysgenesis of the odontoid process, resulting in atlantoaxial instability. The presence of atlantoaxial instability requires protection of the spinal cord from compression by stabilization of the spine in extension during tracheal intubation maneuvers and throughout the perioperative period. Rigid videolaryngoscopy will likely result in a better view of the larynx and glottis without significant manipulation of the head and neck. It is reasonable to anticipate a difficult intubation due to mid-face hypoplasia, micrognathia, or macrognathia, and prepare the patient and the equipment accordingly. Determining whether the patient with a difficult airway should get extubated will depend on the intraoperative course as well as the potential for postoperative complications. For concerns about the ease of reintubation, the endotracheal tube could be removed over an airway exchange catheter for a period of time, facilitating replacement if necessary. I would ensure that the patient is alert and responds appropriately; there is a leak around the endotracheal tube, and patient does not have residual effect of neuromuscular blockade. This patient may not fulfill standard spirometric or inspiratory force criteria due to restrictive chest wall disease because of scoliosis and hypotonia. Many patients with achondroplasia are cognitively impaired and will not cooperate with spirometric assessment. For those patients with a normal mental status, it often suffices to ask them if they are getting enough air to breathe as an aid to extubation.

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# Chapter 16
# Otolaryngology


**Robert S. Holzman**

A 10 kg, 2 1/2-year-old boy is scheduled for adenotonsillectomy for sleep-disordered breathing. He had a sleep study with an apnea-hypopnea index of 12. His parents say they can hear him snoring two doors away at home. They proudly brought in his report card from preschool where his teacher indicated that he is the best behaved boy in the class because he always takes his nap at naptime and never has a problem falling asleep. Occasionally he has to go to the school nurse because of headaches. Admission vital signs: BP 128/85, HR 130, RR 45/min.



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## Preoperative Evaluation

### Questions

1. Why is this patient underweight for his age? Why is this of importance to you as an anesthesiologist? What else would you like to know specific to his growth that would influence your anesthetic plan? Why does this happen? Is it more or less common for children with OSA to be underweight?

2. How do the other symptoms and signs influence your plan? What is the significance of his good behavior at naptime? Are headaches common in children with OSA? Why? Why is he hypertensive and tachycardiac? What is the significance of his respiratory rate? Do you think he has "white coat hypertension" or could this be something else? Does this patient need a preoperative echocardiogram? What specifically on the echo would be worrisome or reassuring to you as an anesthesiologist? Would a preoperative ECG suffice?

## Answers

1. In contrast to the popular association of hypoventilation and airway obstruction with the Pickwickian (obesity-hypoventilation) syndrome, most pediatric sleep-disordered breathing is associated with either a normal body habitus or weight below normal, even to the point of failure to thrive. The reason for this is that often children with chronic airway obstruction are slow eaters because they are forced to chew and swallow between episodes of mouth breathing. Many will choose to avoid foods that require a lot of chewing, such as meats, and therefore they will limit their own diet. They may also have impaired taste and smell if they have substantial nasal obstruction.

   Interestingly, following adenotonsillectomy, approximately 75% exhibit an increase in growth hormone, insulin-like growth factor 1, and significant weight gain. Comorbidities that may be associated with prolonged upper airway obstruction involve effects on the cardiovascular system such as pulmonary hypertension, systemic hypertension, morphometric facial changes ("adenoid facies"), and sleep fragmentation.

2. Behavioral abnormalities typically exist on a spectrum of irritability to somnolence and are difficult to separate out from normal toddler development. The constellation of symptoms is more important, and chronic airway obstruction is often associated with parental nighttime complaints of fears that their child will stop breathing, loud snoring, gasping, choking, coughing, periods of apnea, restless sleep, and the child's head extension in an effort to unconsciously resolve the airway obstruction. Morning or daytime headaches are a frequent complaint and may be a result of systemic hypertension, nighttime fragmented sleep, head and neck muscular pain, or various combinations. Hypertension in children may be associated with a visit to the doctor's office, so-called white coat hypertension, but in this patient population, elevations in blood pressure, especially systolic pressure, are also associated with sleep-disordered breathing in direct relationship to the apnea-hypopnea index. Moreover, these patients may have biventricular dysfunction on echo features of left ventricular hypertrophy, pulmonary hypertension, and impaired right ventricular performance. It would not be unreasonable to seek this kind of evaluation in patients with long-standing sleep-disordered breathing because of these possibilities. An ECG may not reveal an increase in right ventricular forces but an echocardiogram might reveal findings consistent with pulmonary hypertension such as right ventricular or pulmonary hypertension, interventricular septal shifting, or stiffening.

3. Would you premedicate this patient? What are your concerns? What are the advantages? What agent(s) would you consider? Would you modify your anesthetic technique as a result?

## Intraoperative Course

### Questions

1. Does this patient need any monitors beyond standard noninvasive monitoring? What monitors do you have available among your standard monitors that help you evaluate the pulmonary circulation and the right heart?

2. Mask vs. intravenous induction—are both acceptable? Is an intravenous induction safer? Mom insists on being present for induction—is this a good idea? As the patient enters Stage II, he develops "see-saw" breathing with phonation and profuse salivation, is moving all extremities in a flexion pattern, and seems to be struggling. What's happening and why? His $SpO_2$ is 94%, but in 5 seconds it goes to 88%—your next move? Choices of drugs? How and where would you deliver them? The child was given an adequate dose of intramuscular succinylcholine and atropine (40 mg and 0.4 mg), but his heart rate has slowed to 62 although you are now moving the chest with adequate ventilation. What is your next move? Should you begin CPR?

3. Premedication would depend on assessment in the preoperative period; the strategy has to take into account the concerns of the parent about the child's behavior, the judgment of the anesthesiologist with regard to the effects of immediate preoperative stress on the cardiopulmonary system, and concerns about any abnormalities of ventilatory control being aggravated by the premedication. Benzodiazepines alone would not be likely to shift the $CO_2$ response curve much if at all, but residual effects may very well emerge in the immediate postoperative period following inhalation anesthetics and/or opioids, so this must be kept in mind.

## Answers

1. No further monitoring is needed beyond standard noninvasive monitors. That said, the available standard monitors should be regarded with an additional level of interpretation to reflect possible aggravation of pulmonary hypertension and biventricular performance. Oxygen saturation will depend to some extent on pulmonary artery pressures as well as right-heart performance; blood pressure will be a reflection of this as well. Left ventricular dysfunction may also be reflected in abnormal blood pressure responses, but it will depend on the balance of chronically elevated catecholamines along with any myocardial contractility impairment, especially with propofol or higher doses of inhalation anesthetics. Because of the solubility of carbon dioxide, it is not likely that patients will exhibit significant alterations of ETCO2 as a reflection of elevated pulmonary artery pressures. ECG findings in severe circumstances might include bradycardias, right-heart strain patterns, or ectopy.

2. While both are acceptable, there is no doubt that a mask induction would be easier, unless there was a pre-existing IV. That is not very likely, as this child is probably coming in from home. However, an IV induction should not be completely ruled out, because it affords a more rapid induction, bypassing the likelihood of passing through an excitement stage during an inhalation induction and allowing the rapid administration of neuromuscular blocking agents to minimize the chance of laryngospasm, which is higher in this patient population. There are also recent reports on increased safety with intravenous inductions of anesthesia. Mom can certainly be present for the induction, with careful discussion beforehand that the primary job of the anesthesiologist remains the care of the patient, and not the parent, and the use of the euphemism "falling asleep" is just that—it is, after all, a euphemism, and not really "sleep."

3. He now has an IV. How will you plan your anesthetic at this point? Do these events influence your plan? The surgeon suggests a deep extubation because he thinks the wakeups are less problematic. Do you agree? Would you choose that strategy? Why?

4. Does this patient need to go to the ICU?

The excitement stage during an inhalation induction is associated with the increased elaboration of endogenous catecholamines, which may be associated with all of these signs of reactivity and in addition will produce an increase in oxygen consumption and therefore carbon dioxide production. Some positive pressure in the circuit by closing the APL valve is a reasonable strategy as is the insertion of an oral airway to improve upper airway patency. Occasionally, laryngospasm can only be treated with neuromuscular blockade; a depolarizing agent such as succinylcholine can be co-administered with an anticholinergic intravenously or intramuscularly (if prior to IV insertion), or a nondepolarizing drug such as rocuronium can be used. If the heart rate continued to decrease for a few seconds following administration of neuromuscular blockade, and the chest was rising and adequate gas exchange was the result, I think I would wait a few more seconds to see if the saturation began to improve. There is a little bit of a delay in the display of the $SpO_2$ in relation to improvement in ventilation. If the heart rate continued to decrease and there was evidence of impaired gas exchange, worsening hypoxia, and bradycardia, then CPR should be quickly initiated to augment the circulation of further resuscitation drugs and augment cardiac output and cerebral blood flow.

3. Once an IV is in place, then the anesthetic planned for this patient can be continued. This may or may not include a "deep" extubation, which has the advantage of a well-anesthetized airway at the end of the case so coughing and "bucking" are avoided, but also has the disadvantage of an anesthetized unprotected airway at the end of the case. Those experienced in the technique typically have very few difficulties with either strategy; those less experienced should perfect their technique in healthy tonsillectomy patients first before using this technique in patients with significant comorbidities and a tendency to more irritable airways.

4. The ICU is the appropriate perioperative destination for this patient, in accordance with typical concerns about their perioperative risk of airway obstruction and ongoing disordered breathing, especially following a general anesthetic. It is now part of the guidelines of the American Academy of Otolaryngology-Head and Neck Surgery for tonsillectomy in children as well as the American Academy of Pediatrics. Children less than 3 years of age should be kept overnight in the intensive care unit.

## Postoperative Course

### Questions

1. The patient is brought to the ICU, extubated. His $SpO_2$ on arrival, with blow-by oxygen, is 94% and he is sleepy. What are the possibilities? On auscultation, he has diffuse, moist and coarse breath sounds without wheezing. You think you can hear rales at the lung bases. Is this kind of patient at risk for post-tonsillectomy pulmonary edema? How does this happen? What is the appropriate course of action at this point? At what point will it be safe to transfer him from the ICU?

2. What will you counsel the parents about with regard to his recovery course for the next few days? Over what period of time will he actually "normalize" his cardiopulmonary system alterations to chronic upper airway obstruction?

## Additional Topics

### Questions

1. What is the CHARGE syndrome? Of what importance is choanal atresia in the first week of life? What special considerations are there for the surgical correction? Are there specific anesthetic implications?

## Answers

1. Impaired oxygenation in the immediate post-op period can have a variety of reasons—aspiration, somnolence, atelectasis, or a phenomenon well recognized with tonsillar hypertrophy: post-extubation pulmonary edema. While an abnormal chest X-ray aids the diagnosis, rales will often reveal themselves prior to radiological confirmation, as will impairment in oxygenation. Likewise, improvement in oxygenation and auscultation will be more rapid than radiological resolution of abnormalities. Appropriate intervention may simply include elevating the inspired fraction of oxygen using a non-rebreathing mask, but diuretics, CPAP, or reintubation and positive pressure ventilation may be required in severe cases. Discharge directly from the ICU after an overnight stay is possible with complete normalization in room air; otherwise, continued inpatient observation may be warranted if resolution is slower.

2. There will be no "immediate" effect on snoring or respiratory control in the first few postoperative days, although there will be noticeable improvement in snoring over the first few weeks. If there is a central control component, that will improve in the first few weeks to months after surgery. Pulmonary hypertension and right-heart dysfunction will improve in most patients over weeks to months following surgery.

## Answers

1. The CHARGE syndrome is an acronym for *c*olobomas of the eye, *h*eart disease, *a*tresia of the choanae, *r*etarded growth, *g*enital anomalies, and *e*ar anomalies. It is a more extreme form of choanal atresia, which may occur unilaterally or bilaterally. In addition to the acronym findings, CHARGE patients often have CNS abnormalities such as olfactory bulbs that are abnormal, cranial nerve abnormalities, and pharyngeal dyscoordination leading to aspiration. Because these anomalies occur early in embryological development, there can be varying stages of severity for each, along with impairments of development of other contiguous structures such as the branchial arches and occipital somites. It is therefore not uncommon to have a short neck, short mandible, small mouth, clefting of the lip or palate, a range of cardiac anomalies of varying severity, and failure to thrive. In the newborn period, severe respiratory distress may occur which cannot be relieved by a nasal airway. Likewise, nasogastric intubation for decompression or feeding may not be possible.

2. A 12-year-old boy presents with a growth in the nasopharynx and is scheduled for biopsy. What concerns do you have? Of what significance is the diagnosis of juvenile nasal angiofibroma? What implications does it have for anesthetic management? Is it important to know about the extent of this tumor prior to anesthesia? Why? What implications does it have for management? The patient will first be scheduled for coil embolization under anesthesia in the interventional radiology suite. How will your anesthetic management be influenced for this procedure? How should the $CO_2$ be controlled? Is there an optimal choice of anesthetic agents? Should the patient be managed with controlled hypotension? Why/why not?

3. A 15-year-old is scheduled for incision and drainage of a peritonsillar abscess; she has trismus and is frightened. What are your considerations for anesthetic induction? Should this patient undergo an awake intubation? Rapid sequence induction? Should she receive a muscle relaxant? What about an awake transnasal fiberoptic intubation? How is this situation different than a Ludwig's angina patient?

Surgical correction at this point has progressed to transnasal endoscopically guided membranous and bony resection in the majority of cases, although transpalatal approaches are still utilized.

The surgical goal is to create choanal patency, preserve mucosal integrity, and minimize the chance of re-stenosis. If stenting is utilized, when the patient returns for stent removal after about 3 weeks, dilation or dilation plus injection of mitomycin-C is often used to promote epithelial growth.

Anesthetic considerations include the possibility of midfacial dysmorphism, congenital heart disease, developmental delay, and pharyngeal dysfunction with aspiration. Midfacial dysmorphism with a foreshortened nasomaxillary complex may make the mask fit as well as direct laryngoscopy difficult, and the medical consequences of prolonged upper airway obstruction and its cardiopulmonary consequences challenging.

2. Most childhood tumors in the nasopharynx are benign, but can have significant consequences nevertheless. Encephaloceles, dermoids, and benign teratomas can occur as congenital remnants, in which case they present at an early age with airway obstruction or more insidiously in older patients. The juvenile nasal angiofibroma is the most aggressive of these benign tumors presenting in early adolescence, usually in boys. They extend locally into the surrounding nasopharyngeal tissue and cranially through the skull base. They are typically evaluated radiologically by CT scan, MRA, and/or angiogram and at the same time embolized in order to reduce the vascularity for subsequent surgical resection. Because transit time in vascular areas is related to volume, pressure, and pH, all three can be positively influenced by the anesthetic technique. For placement of embolization devices, increased volume, normal to slightly higher than normal blood pressure, and moderate controlled hypercarbia may facilitate coil placement. For the surgical procedure, this physiology should be reversed, so that bleeding may be decreased through the use of carefully controlled hypotension, volume reduction, and positive pressure-controlled hyperventilation.

3. This is a very typical presentation in an adolescent with a sore throat, difficulty swallowing, often sick for a few days, and occasionally, and experiencing a change in voice with difficulty talking. She may even have some mild respiratory distress. Although the majority will have been treated successfully with antibiotics, those coming to surgery have usually failed such therapy. Most patients will have had CT scans of their upper airway preoperatively, and therefore the extent of the peritonsillar abscess is easy to evaluate. Trismus is difficult to evaluate with regard to predicting the ease of direct laryngoscopy and endotracheal intubation. It is typically relieved following induction with a hypnotic agent and the use of a muscle relaxant unless the inflammation and edema have been progressive over several days. Assuming that the clinical exam and radiological evaluation do not suggest anatomic difficulties with a rapid sequence induction of anesthesia, direct laryngoscopy, and intubation of the trachea, this would be the

4. A 5-year-old Haitian girl who speaks only Creole is scheduled for partial reduction glossectomy for cystic hygroma. How will you evaluate her? Her tongue protrudes 7 inches beyond her mouth, and the distal 3 inches are desiccated and macerated. How will you begin your induction sequence? Is it likely that you will be successful at orally intubating her? Nasally intubating her? How would you pre-oxygenate? What are your postoperative considerations?

5. Differentiate epiglottitis, croup, and bacterial tracheitis. How does it make to your anesthetic management to consider these differences?

optimal choice. Secondary choices include topical anesthesia, sedation, and an "awake" look or placement of an endotracheal tube; however, the risks of patient discomfort, coughing, and potential disruption of the abscess may outweigh the benefit. An alternative would be the maintenance of spontaneous ventilation either following intravenous induction with a hypnotic agent or mask induction with a volatile agent. A transnasal fiberoptic intubation would be very hazardous because the peritonsillar abscess often extends into the upper pole of the tonsillar bed, right at the junction of the soft palate, and instrumentation of the soft tissue in the area could be a significant risk for abscess disruption.

4. There are several important features here; first of all, the inability to provide positive pressure ventilation or even supplemental oxygenation by mask is most influential on the anesthetic plan. Secondly, the patient does not speak English and therefore will have a more difficult time cooperating with an anesthetic plan that involves sedation, topicalization, and an "awake" intubation.

   Her ability to cooperate must be carefully assessed with the aid of a translator in the presence of the parents who can help to explain what the anesthesiologist will be doing. An IV should be established first in order to provide sedation to the point of arousable somnolence. A variety of medications can be used for this purpose, but a combination of midazolam and fentanyl would probably be my choice. Topical anesthesia can be provided by lidocaine (a 2% concentration in this age group should be enough) to the level of the laryngeal inlet. Depending on the choice made, standard or video direct laryngoscopy or fiberoptic laryngoscopy (transnasal or transoral) can be accomplished. Supplemental oxygen can be delivered by an assistant while asking the patient to take deep breaths. Postoperatively, depending on the duration and extent of the surgery, she may have swelling that would make nasal reintubation more comfortable for the patient and more secure for her ICU stay.

5. Epiglottitis refers to the acute bacterial infection of the supraglottic larynx that had historically been caused by Haemophilus influenzae type B. The typical clinical picture is the sudden onset of fever and airway distress in the absence of a URI in a toxic-appearing young child. They are often sitting, rather than lying down because they can breathe more easily. Radiographically, they typically have a thumb sign of the epiglottis. Croup, or laryngotracheobronchitis, is usually more gradual in its onset, preceded by several days of URI-like symptoms, and caused by URI-related organisms such as parainfluenza. Many patients have a typical "barking" cough with or without stridor, while others can have significant upper airway obstruction. Biphasic stridor supports the diagnosis of laryngotracheobronchitis. The age group is somewhat younger, usually 6 months to 3 years of age. Radiographically, a "steeple" sign is present in the subglottis. Bacterial tracheitis may present with fever, stridor, voice change with a brassy quality, and a toxic appearance. The trachea usually has purulent debris, crusting, ulceration, and membranes that may require removal. A range of gram-positive and gram-negative organisms are often the culprits. These patients often

6. How does a laryngeal cleft occur embryologically? What are the implications for airway management? Should these patients receive a tracheostomy in the newborn period? Why/why not?

7. What are your considerations for using jet ventilation as part of your management of the airway during laser surgery for juvenile laryngeal papillomatosis?

need to be supported with endotracheal intubation and perioperative intensive care. It is important to differentiate the disorders because perioperative airway support is often necessary for epiglottitis and bacterial tracheitis, while airway instrumentation and endotracheal intubation should optimally be avoided for croup, so as not to have a foreign body in an injured and subsequentially healing area.

6. Failure of complete separation of the primitive foregut into the trachea and esophagus can result in varying degrees of residual communication between the two. It can be as subtle as a small communication between the arytenoid cartilages indicating incomplete formation of the interarytenoid muscle or complete communication at the cranial portion of the larynx and upper third of the trachea, making them functionally one tube. These infants have an abundance of pharyngeal secretions, recurrent aspiration pneumonias, choking episodes, and respiratory distress, typically associated with attempts at feeding. A significant portion also have tracheoesophageal fistulas. A tracheostomy may be ineffective in establishing an airway because of the tendency for the tracheostomy tube to pass through the posterior wall of the trachea into the esophagus. It is usually better to attempt a primary closure of the mucosa separating the trachea and esophagus through suspension laryngoscopy while maintaining spontaneous respiration. The alternative, open repair and separation of the trachea and esophagus, is often fraught with hazards of perioperative tissue breakdown and formation of fistulous communications.

7. Jet ventilation accomplishes several things that intubation with a metal tube or foil-wrapped tube cannot. First of all, it provides unimpaired access to the airway and complete visualization for the surgeon. Secondly, it decreases the risk of fire by not having any combustible material within the airway at all. The hazards are several: there is a risk of barotrauma and dissection of tracheal, pretracheal, or pharyngeal tissue if the driving pressure of the jet is too high, and for that reason, a pressure/compliance curve is an optimal strategy, where the amount of driving pressure is just enough to ventilate the patient, assessed by using chest wall movement and/or breath sounds as an endpoint for adequate ventilation. Transcutaneous carbon dioxide (TcCO2) can provide a quantitative measure of gas exchange. The airway is unsecured, and therefore, debris and smoke can be "jetted" into the unprotected airway, so efforts at ventilation should be made in concert with the surgeon's laser resection. The more severe forms require extracorporeal membrane oxygenation (ECMO) or cardiopulmonary bypass (CPB).

8. You are called into a colleague's room because the laser aperture was accidentally left open and the laser fired on a polyvinylchloride endotracheal tube that remained in place just prior to extubation and after the suspension laryngoscope was placed; the patient has flames and smoke in the tube and singed lips. Your colleague's sleeve has caught fire, and he is preoccupied with that. What do you do next?

8. The colleague will probably be adequately cared for by the OR personnel, who likely know how to handle this straightforward situation. The patient's airway situation is more complicated. Working in conjunction with the surgeon, ventilation of the lungs has to be discontinued and all anesthetic gases including oxygen have to be discontinued as well. The flames should be extinguished with saline, and the endotracheal tube removed. All of these steps should take place virtually simultaneously. At that point, the patient's lungs should be ventilated by mask, and the surgeon should prepare to evaluate the trachea endoscopically for damage and burns. Depending on the degree of burn and damage, endotracheal intubation and perioperative mechanical ventilation may be required.

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# Chapter 17
# Craniofacial and Maxillofacial Surgery


**Robert S. Holzman**

An 18-year-old male with a Class II malocclusion is scheduled for a LeFort I osteotomy, bilateral mandibular sagittal split osteotomy, and genioplasty. He has a prior history of cleft lip and palate repair in infancy and earlier childhood, and a pharyngeal flap for velopharyngeal insufficiency at around 10 years old. In addition, he has dental crowding and doesn't like the way he looks.

He is otherwise healthy, active, and entering college next year.



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## Preoperative Evaluation

### Questions

1. What is a malocclusion? What are the different classes and what implication do they have for the anesthetic? What problems is this patient likely to have in his history? What is the role of orthodontics in preparation for successful orthognathic surgery?

2. He has a prior history of several procedures; do any of those have specific implications currently?

## Answers

1. A malocclusion is a misalignment between the teeth of the two dental arches when they approach each other as the jaws close. It is a common finding that does not require treatment most of the time. More severe malocclusions require orthodontic ± orthognathic surgery in order to decrease the risk of tooth decay, relieve pressure on the temporomandibular joint, and improve aesthetics. More than a century old, Angle's method of classification is typically used: Class I (the molar relationships are normal but spacing, crowding, and over- or under-eruption of teeth occur), Class II (retrognathism or overbite; there are two divisions to this class as well), and Class III (prognathism, anterior crossbite, or underbite, where the upper molars are posterior to the lower molars). The advantages of this most commonly accepted categorization are simplicity and reproducibility, despite there exist many other, more complete classifications. Crowding is usually treated with orthodontics or extraction before orthognathic surgery. Other comorbidities would include a tendency for obstructive sleep apnea. The need for a LeFort I osteotomy is not surprising; it may be required in up to 25% of cleft lip and palate patients.

2. With a history of a pharyngeal flap for velopharyngeal insufficiency, it is crucial to plan for the possibility of increased bleeding, tissue disruption, and possible posterior pharyngeal wall dissection with passage of a nasotracheal tube. Topical vasoconstriction with oxymetazoline will help to decrease bleeding. Care must be taken in administering the nasal spray, however, because it is possible, especially if administered with the patient supine, that a significant overdose can be delivered, with hypertensive consequences. Pharyngeal flaps may be either superiorly or inferiorly based, although most are superiorly based as they have fewer complications. This leaves two lateral apertures at the choanal level, which may make passage of a nasotracheal tube more difficult. A sphincter pharyngoplasty results in a vascularized and innervated myomucosal flap from the palatopharyngeus muscle. This flap leaves a central passage, through which it is typically easier to pass a tube.

## Intraoperative Course

### Questions

1. How will you plan for nasotracheal intubation in this patient?

2. In dissecting the medial maxillary buttress, you note that the peak inspiratory pressure has dropped, the end tidal $CO_2$ trace has changed from a plateau appearance to a parabola, and the surgeon says he can smell "anesthesia in the field." He is complaining about the bubbling as well and asks you to fix the tube leak. What do you think is going on? How will you determine the cause? How can you fix the problem? Other than replacing the tube, what else can you do?

3. The surgeon is now ready for the "downfracture" and mobilization. What is that? What are the concerns about the downfracture?

4. During the downfracture, the heart rate suddenly decreases to 32 bpm from 82. Why? What can you do acutely? Is there a way to prevent it from happening again?

## Answers

1. For the above reasons, in this patient I would choose direct visualization with a fiberoptic scope to tube passage. If a difficult airway is not anticipated based on physical exam, the intubation can be accomplished after the induction. While the best alternative is probably direct visualization with a fiberoptic scope, it is not the only method. A small, softened nasogastric tube can be passed as a guide, and a softened nasotracheal tube can be passed over it, similar to a Seldinger technique, in order to enter the oropharynx. At that point, conventional laryngoscopy or videolaryngoscopy can be employed for tracheal intubation.

2. There are several possibilities to account for these findings. A partial circuit disconnect could have occurred, which should be relatively quick to diagnose as the circuit is examined at the 15–22 mm connector. The cuff of the tube may have herniated into the glottis, which would result in an incomplete airway seal. That possibility should prompt a direct laryngoscopy for rule out. Because of the timing, however, it is important to consider whether the pilot tube or the endotracheal tube itself was cut during the osteotomy. There are two choices at that point—using a throat pack to obtain a better seal at the laryngeal inlet (often not a perfect choice) or replacement of the endotracheal tube. It would be safest to use a tube exchanger like a Cook catheter for this purpose, preserving the already established pathway to the trachea as well as the ability to oxygenate the patient. As a last result, for deteriorating conditions, a tracheostomy may be necessary.

3. The downfracture refers to the physical maneuver of mobilizing the maxilla once it is freed from the pterygoid plates, creating a pterygo-maxillary disjunction. This is the time of greatest bleeding, which may be arterial in nature from the ascending palatine branch of the facial artery and the anterior branch of the ascending pharyngeal artery. Mobilization is further accomplished, once the maxilla is free, by stretching the soft tissue and improving the range of motion [1].

4. Bradycardia and asystole have been noted during the most stimulating portions of the LeFort procedure such as during the dissection of the posterior buttress of the maxilla or during the downfracture, when traction can activate the trigeminovagal reflex. It may also occur during a mandibular sagittal split osteotomy during subperiosteal retraction on the medial aspect of the ascending ramus [2–4].

## Postoperative Course

### Questions

1. The patient will be in rubber band fixation postoperatively. How will you prevent postoperative nausea/vomiting? Should he have a nasogastric tube?

2. You are called to the bedside in the PACU. The patient has nasal bleeding. Does it matter if it is unilateral or bilateral? From the side that had the nasal tube?

## Additional Topics

### Questions

1. A 1-year-old, 9 kg boy is scheduled for repair of orbital hypertelorism for Apert's syndrome. He appears congested but afebrile, with a blood pressure of 92/55 mmHg, pulse 120 bpm, respiration 32/min, and temperature 37 °C. Hematocrit is 33%. He is on no medications, and his parents report that he is terribly afraid of doctors. Will this patient have a difficult airway? What monitors will you select for this case? What are your considerations for induction of anesthesia? Any anticipated difficulties with vascular access? Should you consider tranexamic acid for blood conservation?

## Answers

1. Postoperative nausea and vomiting, in this context, should not be considered together but as two separable entities. Nausea as an unpleasant sensation does not pose significant risks with regard to head and neck surgery, but vomiting can potentially result in hematoma, wound dehiscence, dehydration, electrolyte imbalances, and, in extreme cases, esophageal damage or aspiration. Dexamethasone has antiemetic effects and is administered in this context to decrease tissue swelling but has a synergistic effect with other anti-nausea medications. In addition to dexamethasone, I would plan on giving ondansetron (5-HT3 receptor antagonist) and haloperidol (dopamine D2 receptor antagonist). Avoiding nitrous oxide and minimizing opioids may be helpful as well. Nasogastric tubes, whether passed at the end of the case and removed or remaining in for several hours or even overnight postoperatively, do not seem to decrease the incidence of nausea or vomiting [5].

2. Bleeding after a LeFort I is usually epistaxis, which can be anterior or posterior or both. Unilateral anterior epistaxis may be due to a traumatic intubation or to stripping the nasal mucosa off the underlying nasal floor and septal areas. Hemorrhage from both nares is suggestive of an injury to an artery posteriorly, such as the internal maxillary artery or its branches (sphenopalatine or ascending palatine). Epistaxis is typically treated with packing and expectantly. If arterial injury is suspected, angiography should be expeditiously arranged for.

## Answers

1. That depends on what you mean by a difficult airway. The midface is hypoplastic, and the eyes appear to be proptotic, although that is more a reflection of the hypoplastic midface and orbits but normal size eyes. The hypoplastic skull base contributes to abnormal development of the sphenoid, frontal, and maxillary sinuses, which in turn often lead to an appearance of chronic congestion, simply because sinus and nasal drainage is impaired. The branchial arches, however, are typically not affected, so that mandibular development proceeds normally. The combination often results in chronic upper airway congestion, moderately difficult fit for a mask, but relatively easy laryngoscopy and intubation. The surgical approach is a bifrontal craniotomy with multiple osteotomies and (hopefully)

2. What is Goldenhar syndrome? How does it develop? What are the anesthetic considerations?

   Are there particularly significant associated anomalies? Do these patients tend to get easier to take care of over time?

3. What is the developmental history of a cleft lip and palate? Are there associated anomalies you should expect? How can you assess the potential for intubation difficulty? Would you use a muscle relaxant as part of your anesthetic technique? Why does a cleft palate develop with Pierre-Robin Sequence?

preservation of an intact dura. There can, however, be neurosurgical consequences if there is a dural puncture, or even with the prolonged reconstructive surgery, dural exposure, and large blood loss and fluid shifting that occurs. Polysyndactyly of the hands and feet is common, and therefore, intravenous access may be difficult. Tranexamic acid, an antifibrinolytic, has been shown to clearly be of benefit in decreasing blood loss during craniofacial surgery in infants [6].

2. Goldenhar syndrome is the eponym for hemifacial microsomia, an anomaly characterized by variable hypoplasia of the mandibular division of the first branchial arch, including the mandibular ramus, body and temporomandibular joint, hypoplasia of soft tissue components of the face and jaw, and hypoplasia of the facial nerve. The more severe forms are typically very difficult intubations because of the inability to open the jaw on the affected side. Furthermore, breathing through a natural airway (and therefore support through a mask airway) may be difficult because of a small pharynx. Because this anomaly occurs early in embryological life, anomalies of other contiguous areas are not uncommon, including fusion of occipital somites that can give rise to the Klippel-Feil anomaly of cervical vertebral fusion. Cardiac defects such as atrial and ventricular septal defects may also occur. A paramedian cleft palate may also occur. Anomalies of the first branchial cleft such as low set, misshapen ears, and sensorineural hearing loss are common as well. Managing the airway of these patients tends to get more difficult with time.

3. Typical cleft lip formation occurs along a line joining the primary and secondary palates through the middle of the ipsilateral ala. Clefting of the primary and/or secondary palate may also occur in continuity with a cleft lip. Clefting of the soft palate may also occur, because palatal fusion occurs progressively from anterior to posterior portions of the palate. There may be associated anomalies of structures or organ systems that are developing at the same time as the midface and palate, such as the heart and occipital somites. A cleft palate may also occur in association with glossoptosis (Pierre-Robin Sequence), in which case it is more properly considered a deformation by interference with the progressive midline fusion of the maxillary shelves. A paramedian palatal cleft can occur with branchial arch abnormalities such as hemifacial microsomia (Goldenhar syndrome) [7]. It is important to carefully evaluate any association with branchial arch abnormalities because of the potential for difficult laryngoscopy and intubation. The use of any specific anesthetic technique should be directed to the desired end point of the surgery as well as any anticipated difficulty with extubation. These patients will occasionally have significant obstructive or mixed sleep apnea; attention to perioperative monitoring for apnea and hypoxia is probably more important than any particular anesthetic technique.

4. What are the advantages and disadvantages of using oxymetazoline (Afrin®) as a vasoconstrictor for nasal intubations?

4. Oxymetazoline (Afrin®) is now the topical vasoconstrictor of choice for use on the nasal mucosa prior to nasotracheal intubation. Although many vasoactive agents have been used in the past such as cocaine, epinephrine, and phenylephrine, none have been shown to be superior to oxymetazoline with regard to vasoconstriction and have cardiovascular and potentially central nervous system side effects of considerably greater frequency than oxymetazoline [8]. As an imidazole derivative, oxymetazoline is rapidly absorbed across mucosal membranes in children. Hence, toxicity generally develops within minutes. While fractionated doses during pharmacokinetic studies demonstrate relatively little clinical toxicity, this is hardly the way oxymetazoline is delivered clinically by anesthesia personnel in the operating room, especially when delivered after induction. Given the supine position of patients on the operating room table, it is common practice to hold the bottle inverted and squeeze it, delivering a much larger and potentially toxic amount, particularly with small children.

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A broad general review of the embryology and clinical considerations for congenital anomalies of the head and neck

# Chapter 18
# Ophthalmology


**Robert S. Holzman**

A 6-week-old otherwise healthy infant is scheduled for an eye exam under anesthesia to evaluate unilateral leukocoria noted by his parents; it is suspected that he has retinoblastoma. He has an uneventful birth history and currently weighs 5 kg.



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## Preoperative Evaluation

### Questions

1. At first glance, this seems to be a relatively straightforward anesthetic. What should you anticipate in the short- and long-term issues confronting this infant and his family?

## Answers

1. Histologically, retinoblastoma is a primitive neuroectodermal malignancy that occurs intraocularly. The term "leukocoria" indicates that the (typical) presentation is with a white pupil. Lens, vitreous, or retinal abnormalities can produce the same finding, as can many medical disorders. Nonetheless, about 50% of the time, retinoblastoma is the etiology. There are heritable and nonheritable forms, and the significance for the anesthesiologist is that one may anesthetize additional family members during a comprehensive evaluation. A thorough staging will be required (assessments by EUA as well as imaging – therefore, additional anesthetics) in order to stratify intraocular from extraocular extensions. As treatment has evolved, there has been a shift from external beam radiation to chemotherapy because of the increased risk of patients developing secondary tumors following radiation. Because leukocoria is the most common presentation of retinoblastoma, all infants and children with an abnormal red reflex during routine pediatric exams (including prior to discharge from the newborn nursery) require immediate referral to an ophthalmologist [1].

   The examination in the operating room is thorough and not at all quick, so due consideration must be given to the patient's age as well as positioning requirements. The head will likely be turned from side to side and the OR table rotated 90° (field avoidance positioning). Furthermore, the room will be dark. A low threshold should be set for intubating the trachea. After dilation and bilateral fundoscopic exams, B-scan ultrasonography will be done. MRI may be part of the same exam or done separately. For patients with extraocular extension, bone marrow examination, lumbar puncture, and placement of central lines for chemotherapy may be done as well. Fluorescein angiography may be utilized as well.

   Enucleation remains the definitive treatment of intraocular retinoblastoma, particularly in the majority of patients who present with advanced unilateral disease with poor visual prognosis. Globe salvage strategies include systemic chemotherapy with focal consolidation and intra-arterial chemotherapy. For small tumors, focal consolidation therapy alone can be effective. This includes cryotherapy, laser photocoagulation, hyperthermia, and plaque irradiation. External beam radiation therapy (EBRT) is avoided if possible given the associated side effects and should be avoided in children <12 months of age but still has a role in the treatment of recurrent tumors and seeding. The risk of second malignancy is increased more than threefold by EBRT, especially if the patient is less than 1 year of age. Intravitreal chemotherapy is a relatively recent addition and has supplanted EBRT for the treatment of vitreous seeding. Systemic chemotherapy in the setting of retinoblastoma is often termed chemoreduction because it is administered to shrink the tumor and allow subsequent treatment with focal

2. Following his initial and lengthy visit to the operating room during which he underwent bilateral EUA, ultrasound, and fluorescein angiography as well as numerous photographs, he is scheduled for four sessions over 4 months of intra-arterial chemotherapy in the interventional radiology suite. What are your specific considerations?

consolidative therapy. 38.2% of patients with hereditary retinoblastoma will develop a secondary malignancy with an associated long-term mortality rate of 26% [2].

Significant recent attention has been focused on intra-arterial delivery of chemotherapeutic agents. Based on early attempts in Japan to cannulate the carotid artery directly to deliver antineoplastic drugs to the eye, Abramson and colleagues modified the Japanese protocol to cannulate the ophthalmic artery. This technique has been called supraselective intra-arterial chemotherapy. The total dose of whole-body radiation given with multiple fluoroscopies also remains undefined. Intravitreal chemotherapy for control of vitreous seeding has also been reported.

When an eye is enucleated for retinoblastoma, the goal is to remove at least 1 cm of optic nerve to ensure that the cut end of the nerve is free from tumor to reduce both the risk of orbital relapse and the need for adjuvant therapy. Some centers harvest fresh tumor tissue in the operating room for genetic studies and tumor bank storage. Tissue harvesting should be done cautiously so as not to destroy the globe's anatomy or create artifacts that might jeopardize pathologic evaluation. At the time of enucleation, the largest possible orbital implant is placed to encourage normal development of the pediatric orbit. These children will also need fitting for an ocular prosthesis.

Children who undergo any form of eye salvaging treatment (external beam radiation, chemoreduction) need frequent follow-up examinations under anesthesia to monitor for recurrence. A typical schedule would require examinations under general anesthesia every 4–8 weeks until age 3, followed by less frequent examinations if the disease becomes quiescent.

Recurrence of retinoblastoma from treated lesions is very common and can occur years after treatment.

2. Dosing of chemotherapy drugs when administered by intra-arterial injection is lower than dosing for systemic chemotherapy; therefore, the anticipated side effects should be lower, as well as the geographical distribution of those side effects. Much lower doses of chemo are used in this approach, so the side effects tend to be limited to the eye area. Possible side effects include:

- Swelling around the eye
- Detachment of the retina from the back of the eye
- Bleeding inside the eye
- Weakening of the muscles that move the eye
- Drooped eyelid
- Loss of eyelashes

Possible long-term side effects are not yet clear. This approach also exposes the child to some radiation, because real-time X-rays are used to help guide the catheter into place. It's not yet clear if (or how much) this might raise cancer risk later in life. Nonetheless, a complete regression rate of up to 88% has been reported [3, 4].

## Intraoperative Course

### Questions

1. You are asked to administer fluorescein 10% for the angiography portion of the case. The dose was double-checked and verified at 7 mg/kg, an appropriate pediatric dose. What is the appropriate rate of injection for fluorescein? How quickly after injection does luminescence typically appear in the retinal and choroidal vessels? Within 3 minutes you notice an increase in the peak airway pressure, listen to the lungs, and there is bilateral wheezing. What do you think is going on? Would you do anything else to verify the diagnosis? What about turning the room lights on (the ophthalmology team doesn't want you to do this).

It is also unclear what effects there may be from repeated anesthetics during infancy and early childhood, particularly when they occur within a short time range of each other. While there are no specific anesthetic techniques indicated, the overarching consideration in the interventional radiology suite remains motionlessness, particularly with the use of image intensifiers and magnification. Patients should receive liberal IV fluids because of the osmolar effects of contrast dyes employed during the angiography. It is routine to administer nasal oxymetazoline in appropriate doses into the ipsilateral nostril in order to enhance ocular blood flow and two puffs of albuterol through the endotracheal tube because of the known association of bronchospasm.

This approach is not free of complications. Femoral artery occlusion for 1 week and leukopenia in fewer than 10% of cases have been noted and avascular retinopathies resulting in blindness.

## Answers

1. Fluorescein should be administered rapidly intravenously, on the order of 1 mL/second; the anticipated lag time until visualization is 7–15 seconds. Bronchospasm may well be the diagnosis – there are wheezing and an increase in airway pressure. This may also be caused by the tip of the endotracheal tube at the carina or entering a mainstem bronchus, and it would not be unusual for that physical stimulus to elicit some airway reactivity. The capnograph trace may reflect a slow rate of rise on exhalation if the cause is bronchospasm. If there is a camel hump configuration on the exhalation curve, it may be more compatible with air trapping and differential emptying of the lungs. Anaphylaxis may be the cause as well, so the lights should be turned on and the patient's skin carefully examined for urticaria (which are not always seen during general anesthesia). Hypotension and cardiovascular collapse may accompany anaphylaxis under anesthesia, and appropriate measures, including the administration of epinephrine in dilute doses (0.1 mcg/kg), should be undertaken. A continuous infusion may be necessary because of ongoing degranulation and the inability to decrease the patient's exposure to the inciting agent at this time.

## Postoperative Course

### Questions

1. You are called to the bedside in the PACU because the patient has become acutely jaundiced. What do you think is going on? How would you evaluate? What time course of resolution do you expect?

## Additional Topics

### Questions

1. A 2-year-old, 8.5 kg girl is scheduled for bilateral lateral rectus recession with adjustable sutures. She was born at 32 weeks, required some supplemental oxygen for a few days but not intubation or mechanical ventilation, and went home with an apnea monitor for a month that "never alarmed" according to the parents. Her vital signs are blood pressure of 92/55, pulse 120, respiration 32, and temperature 37 °C. Hematocrit is 32. She has never had any previous surgery.

   Is the concentration of inspired oxygen a concern for her surgery? Is it safe to avoid premedicating her with an anticholinergic such as atropine or glycopyrrolate? Would you premedicate her with atropine if she were scheduled for a medical rectus resection? Is she at risk for malignant hyperthermia? As part of the plan, the surgeon would like to adjust the correction in the PACU via the adjustable sutures.

   - What will the surgeon need from you in order to obtain the best result?
   - What medication will you use?
   - Are there any premedication administration criteria that you use to determine the patient's fitness for this portion of the procedure?
   - What monitoring will you choose?

## Answers

1. The most common reaction following fluorescein injection is temporary yellowish discoloration of the skin and urine. The urine may have a bright fluorescent yellow color, like Day-Glo. Discoloration of the skin usually fades in 6–12 hours and usually fades in urine in 24–36 hours.

## Answers

1. She may have retinopathy of prematurity, which has been associated with a history of supplemental oxygen therapy in prematures and many other neonatal factors as well; however, the index of suspicion for severe retinopathy is probably low, because the treatment was of short duration. Nevertheless, it would be reasonable for the ophthalmologist to do a quick exam just to answer the question about the appearance of the fundus so that it can be documented, and it is likely that with this history an exam would have been done anyway. It would not dissuade me from delivering a routine anesthetic consisting of an $FiO_2$ of 0.3–0.4 following 100% oxygen prior to intubation. I would not premedicate her with an anticholinergic in order to prevent an oculocardiac reflex but would rather be mindful of it during traction of the lateral rectus muscles. While I would be more concerned about a greater incidence with traction on the medial rectus muscles (greater incidence of bradycardia) [5], I still would monitor and interact with the surgeon before placing reliance on treating preemptively with atropine.

   Malignant hyperthermia has been associated in the past with strabismus, principally through case reports in the ophthalmology literature, although a higher than background rate (fourfold higher) association with masseter muscle spasm has been described in strabismus patients who underwent anesthetic induction with halothane and succinylcholine. The association between masseter muscle spasm and the subsequent development of malignant hyperthermia remains unclear. I would proceed, therefore, with a routine inhalation induction but nevertheless try to avoid succinylcholine for the more typical reason of its higher incidence of masseter muscle spasm and other possible adverse effects such as rhabdomyolysis or hyperkalemia in children. In addition, many ophthalmologists prefer not to have the sustained extraocular muscle contracture, produced as a side effect of succinylcholine, influence their measurements for the procedure.

   Adjustable suture strabismus surgery is used to "fine-tune" the position of the extraocular muscles in the postoperative period. An adjustable suture is placed on the extraocular muscle(s) that was repaired via standard strabismus surgery, and then the eye alignment is checked in the immediate postoperative period [6].

2. A 5-year-old just fell from the Jungle Gym at McDonald's with a Big Mac in his mouth and landed on a rock causing an open eye injury. The ophthalmologist wants to go to the operating room right away because the eye is salvageable and the surgery should be accomplished as quickly as possible. The child is crying in the emergency room when you see him. He is otherwise healthy, with hematocrit 36%. What anesthetic plan and counseling will you give to the parents?

3. What are the systemic effects of the following antiglaucoma drugs that are of significance to the anesthesiologist?

   - Timolol
   - Brinzolamide
   - Pilocarpine

The adjustment suture can be adjusted by sliding the knot and then securing it into a permanent position. While adults and some adolescents can undergo this adjustment with topical anesthesia, most children will require an anesthetic in order for the surgeon to work in a motionless field and maximize patient safety and comfort. Continuous infusion propofol in the PACU is typically sufficient to provide these conditions without requiring a trip back to the operating room. Standard noninvasive monitoring is utilized, including end-tidal $CO_2$ via nasal cannula, plastic face mask, or a Jackson-Rees breathing circuit.

2. This is one of the "classical" controversies in pediatric anesthesia, balancing the risk of the full stomach and aspiration of gastric contents with the risk of extrusion of a portion of the vitreous. It is also important not to forget that, in dealing with the whole patient, there may be other associated injuries, in this case the possibility of loss of consciousness or orbital fracture. Assuming these other possibilities are ruled out and it is decided to proceed to the operating room as quickly as possible in order to obtain the best possible result, my priority would be a calm, anxiolytic induction of anesthesia. If an IV is in place, this would probably entail sedation with an intravenous anxiolytic such as midazolam. If an IV is not in place, then I would choose an oral premed with midazolam. It is tempting to consider a rapid sequence induction of anesthesia with cricoid pressure and succinylcholine; however, succinylcholine has been associated with an elevation of intraocular pressure, even with the administration of intravenous induction agents and/or "precurarization." A good alternative would be to use the priming method with a nondepolarizing neuromuscular blocking agent, with the administration of one-tenth of the intubating dose, waiting for 1–2 minutes for a partial depletion of acetylcholine quanta at the neuromuscular junction and then intravenous induction (with propofol) followed by an intubating dose of the nondepolarizing neuromuscular blocking agent. The onset of adequate relaxation for tracheal intubation usually follows within 90 seconds, during which time gentle ventilation with positive pressure up to 10–12 cm. $H_2O$ and cricoid pressure may be delivered. Although part of the counseling for the parents should include the seriousness of the situation with regard to balancing the aspiration considerations against eye salvage, practically speaking, if the patient has already been crying, he may suffer no more risk with anesthetic induction than he already has prior to surgery [7].

3. Antiglaucoma medications include cholinergic agonists (miotics), sympathomimetics, beta-adrenergic antagonists, carbonic anhydrase inhibitors, alpha-2-selective agonists, and prostaglandin analogs.

   *Beta-blockers* block beta-adrenergic receptor sites decreasing aqueous production in the ciliary body. Beta-blockers have a wide range of side effects including bradycardia, hypotension, depression, arrhythmias, bronchospasm, apnea, and dizziness. One drop of 0.5% *timolol* can reach cardiac beta-blockade levels in infants under 2 years of age, and timolol is commercially available as a 0.25% or 0.5% solution or in a gel form. Timolol is more effective in children

over 10 years of age, in cases where glaucoma is mild, and where it is the sole agent. *Betaxolol* is cardioselective with less effect on the pulmonary system. Neonates have developed Cheyne-Stokes breathing and apneic spells lasting up to 30 seconds that resolved after timolol 0.25% was discontinued, and there has been a report of multiple severe asthma exacerbations in a toddler.

It is thus contraindicated in children with cardiac arrhythmias and bronchospasm and should be used in the lowest possible dose for healthy children. *Timoptic*, a gel-forming, sustained-release preparation of timolol administered once daily, was shown in adult patients to be as efficacious in reducing intraocular pressure as timolol ophthalmic solution (0.25% and 0.5%) twice daily, with measurably less systemic absorption. The 0.25% dosage of either sustained-release preparation is often sufficient and is the preferred agent in pediatric glaucoma, due to the lower concentration, decreased systemic absorption, and once-daily dosing.

*Carbonic anhydrase inhibitors* are sulfonamide derivatives used topically or systemically to inhibit production and secretion of aqueous humor. The systemic forms are used as adjuncts to topical glaucoma treatment. The topical preparations are used for short-term treatment before laser surgeries to prevent postoperative pressure elevations. *Dorzolamide* 2% and *brinzolamide* 1% are eye drops, and *acetazolamide* is administered orally as a liquid suspension at a pediatric dose of 8–30 (generally 10–15) mg/kg/day. *Cosopt*® is a fixed combination of timolol 0.5% and dorzolamide 2%. Side effects of acetazolamide include gastrointestinal upset. Anorexia is frequently seen when used in infants and hyperpnea is also common. Urinary frequency may develop but usually normalizes after several weeks. Less frequently, renal calculi are seen. This class is contraindicated in patients with severe kidney or liver disease, as well as reduced sodium or potassium serum levels, or adrenal failure. It is also contraindicated in those with true sulfa allergies. Metabolic acidosis severe enough to require administration of bicarbonate, blood dyscrasias, and Stevens-Johnson syndrome have also been reported.

*Prostaglandin analogs* are a newer class of medications with impressive potency in adults, once-daily dosing, flat diurnal curve effects, and few side effects. This class includes *latanoprost* 0.005%, *bimatoprost* 0.03%, *travoprost* 0.004%, and *unoprostone* 0.15%. These agents are analogs of endogenous F2-alpha prostamides, and they activate matrix metalloproteinases to remodel the extracellular matrix of the uveoscleral pathway, facilitating flow of aqueous humor. This effect is enhanced by their ability to relax nocturnal ciliary muscle tone.

*Miotics and sympathomimetics* are historically first-line agents that are rarely used today. *Pilocarpine* is a cholinomimetic-miotic used for glaucoma. It causes miosis and therefore increases the size of the canals of Schlemm, promoting outflow of the aqueous humor. Miotics may be used pre- or postoperatively in glaucoma surgery to constrict the pupil and pull the iris away from the anterior chamber angle. Because the muscle-tendon attachments are not well formed in infants, miotics have only a minor influence on aqueous outflow in this age

group. Sympathomimetics work by decreasing aqueous production via vasoconstriction of blood vessels in the ciliary body. With prolonged use, they improve aqueous outflow, likely in part by desensitization of beta-adrenergic responses. To a lesser extent, they improve uveoscleral output as well.

For the most part, timolol gel once daily and brinzolamide are recommended first-line glaucoma agents often used together, with timolol gel providing the advantage of low cost and once-daily dosing and brinzolamide preferred by patients for comfort. Brinzolamide is contraindicated in the presence of sulfa allergies. Additional medications are added as required for optimal control. Miotics, such as pilocarpine, while well tolerated, are infrequently used because of the required frequency of dosing. Brimonidine is contraindicated in young children and the sympathomimetics usually add little to treatment.

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A developmentally-based perspective on pediatric eye disease and anesthetic consider ations.

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Excellent overview of typical pediatric ocular medications.

# Chapter 19
# Respiratory System


**Robert S. Holzman**

A 15-year-old boy without a significant past medical history experienced nausea, vomiting, diarrhea, productive cough, runny nose, fever, and shortness of breath, worsening over the last 6 days. He vapes regularly (nicotine + marijuana) "near constantly" for the past several months and orders a mint-flavored juice from the Internet. He has a history of depression and takes sertraline and clonidine. He has had a history of a 10 lb weight loss over the last few weeks. He is scheduled for a diagnostic flexible bronchoscopy with bronchoalveolar lavage and fungal washings. VS: heart rate 120 bpm; blood pressure 126/84; and temp 102.6 °F. His room air saturation is 86%. His breathing is nonlabored, but tachypneic. His breath sounds are clear. He has no crackles or wheezing and no lymphadenopathy. Currently he is on high flow nasal cannula oxygen, FiO2 = 0.4.



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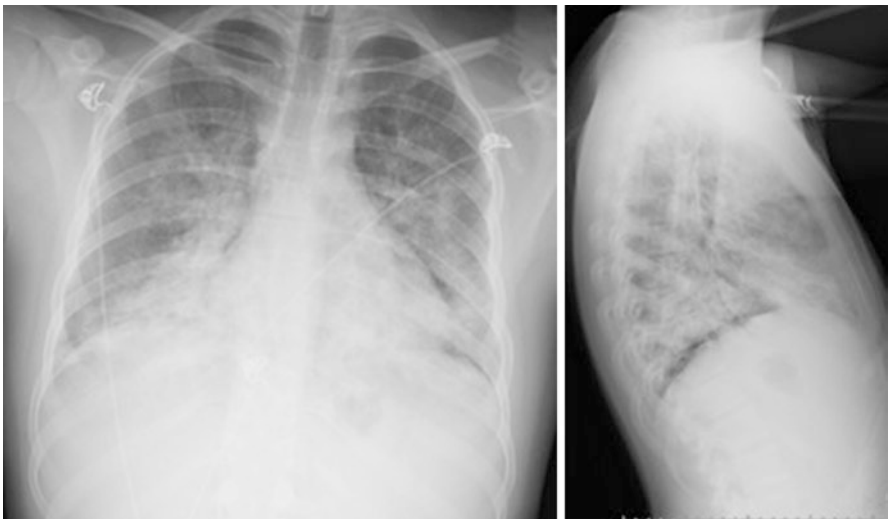
## Preoperative Evaluation

### Questions

1. How would you evaluate his chest X-ray? Of what significance are these findings to you? Would you order further tests? Which ones? How will you incorporate the findings into your anesthetic plan?



## Answers

1. The chest X ray shows bilateral, multifocal areas of airspace opacities consistent with pneumonia. The cardiac silhouette is clear; no pleural effusions are seen. There may be some cephalization of flow in the pulmonary vasculature. This correlates with his clinical picture. One should expect, with the increased work of breathing and elevated temperature, an increase in oxygen consumption, impaired ventilation/perfusion matching and decreased efficiency of oxygenation, and a smaller margin of oxygen reserve, particularly following anesthetic induction

2. The pulmonary team obtained pulmonary function tests, the results of which were:

| Spirometry (BTPS) | | Predicted | Pre Bronchodilator | |
|---|---|---|---|---|
| Parameter | | Value | Actual | % Pred |
| FVC | L | 4.49 | **4.57** | 102 |
| $FEV_1$ | L | 3.89 | **3.75** | 96 |
| $FEV_1$ / FVC | % | 87 | **82** | 94 |
| PEFR | L/s | 7.79 | **8.63** | 111 |
| $FEF_{50}$ | L/s | ---- | **4.25** | ---- |
| $FEF_{75}$ | L/s | 2.20 | **1.79** | 81 |
| $FEF_{25-75}$ | L/s | 4.40 | **3.88** | 88 |
| FIVC | L | 4.49 | **3.51** | 78 |
| FIF50 | L/s | ---- | **3.01** | ---- |
| SVC | L | 4.49 | **4.54** | 101 |
| MVV | L/m | 136.2 | **----** | ---- |

| Lung Volumes (Box) | | Predicted | Pre Bronchodilator | |
|---|---|---|---|---|
| Parameter | | Value | Actual | % Pred |
| TLC | L | 5.36 | **6.14** | 115 |
| VC | L | 4.49 | **4.57** | 102 |
| FRC | L | 2.85 | **3.26** | 114 |
| RV | L | 1.26 | **1.57** | 125 |
| RV/TLC | % | 24 | **26** | ---- |

| Diffusion (Uncorrected for Hb) | | Predicted | Pre Bronchodilator | |
|---|---|---|---|---|
| Parameter | | Value | Actual | % Pred |
| DLCO [Unc] | mL/min/mmHg | 29.57 | **23.41** | 79 |

What is your interpretation of the spirometry data? The findings on the lung volumes? What do you think about the diffusion capacity for carbon monoxide? What does this suggest? How will any of this information help guide you with regard to his care during anesthesia? Plan for postoperative care?

3. The pulmonary physicians also noted that he is sexually active, with one partner. Of what significance is this with regard to his illness?

2. The dynamic lung values of forced vital capacity (FVC), forced expiratory volume in 1 second ($FEV_1$), and peak expiratory flow rate (PEFR) are all severely reduced, which strongly suggests (assuming the patient's cooperation was adequate) bronchiolar collapse, obstruction, or gas trapping. The $FEV_1$ as a fraction of the FVC which forcefully expired in 1 second indicates a severe reduction in flow; this is supported as well by the compromise in the PEFR. Additional information gleaned from the relationship between $FEV_1$/FVC is that there is a significant restrictive component as well, because that relationship is normal. The high residual volume may reflect the patient's difficulty with exhalation. Among the most worrisome of the findings is the carbon monoxide diffusing capacity (DLCO) of half of predicted. The DLCO is an overall measurement of the efficacy of diffusion across the alveolar-capillary membrane, and this finding supports severe impairment of oxygenation at the alveolar-capillary membrane level. All of these findings would worsen the margin of safety with regard to oxygenation. Since this will not improve and might conceivably worsen, with the procedure and the bronchoalveolar lavage, the patient and family should be counseled for postoperative mechanical ventilatory support, and the resolution of impaired oxygenation may be longer.

3. Sexual activity in an adolescent with respiratory distress introduces the possibility of human immunodeficiency virus (HIV) and/or *Pneumocystis jiroveci* pneumonia (PJP), formerly known as *Pneumocystis carinii* pneumonia (PCP), the most common opportunistic infection in persons with HIV infection. It is officially classified as a fungal pneumonia (although unresponsive to antifungal treatment).

4. The pulmonary team would like "as little anesthesia as possible" because they are worried about his overall status; they would also like to do the bronchoscopy through a laryngeal mask airway and have you avoid using an endotracheal tube. What do you think?

## Intraoperative Course

### Questions

1. How would you premedicate this patient? What medication(s) would you choose? Aside from anxiolytics, what additional medications would you consider prior to induction? If his PFTs failed to demonstrate post-bronchodilator improvement in his airflow, would additional medications be of any benefit? Does the patient require a rapid sequence induction? How would you accomplish that (i.e., which choice of medications)? Are there strategies that would minimize airway irritability? The bronchoscopy is supposed to take less than 15 minutes – does this influence your choices?

2. The bronchoscopy does indeed take 15 minutes and your medical student asks you for the calculation for a neostigmine and glycopyrrolate reversal. What is your response? What are your criteria for reversing and extubating this patient? Is there more than one reason you would wish to avoid neostigmine? Any concerns about using sugammadex? What if the patient was a 15-year-old girl?

4. This is a management conundrum – and a frequently asked question – because it is influenced by the conditions you would like to have available if the patient gets into trouble with ventilation or oxygenation. "As little anesthesia as possible" reflects the concerns our colleagues often have about respiratory depression, hypoxia, mechanical airway misadventures like esophageal or mainstem intubation, and oxygen desaturation from numerous causes. However, I would prefer to control the airway with an endotracheal tube, thereby ensuring predictable ventilation in the setting of severe lung damage and already established impaired oxygenation and also in anticipation of perioperative respiratory support. A variety of ventilation management strategies are then available – no PEEP, higher PEEP, higher frequency/low tidal volumes, periodic sigh breaths for alveolar expansion, etc. Notwithstanding this strategy, there is little question that a potential price to pay would be an indwelling foreign body (the endotracheal tube) with the potential for further airway irritability.

## Answers

1. Premedication with an anxiolytic depends entirely on the patient's maturity and state of mind.
   Adolescents can be a difficult age group to deal with, commonly mature and straightforward, but also, not uncommonly, riddled with fears that affect them emotionally, but they cannot often articulate [1]. With an IV present, fractionated doses of midazolam can be given to decrease anxiety (e.g., crying, aerophagia with an increase in gastric size, endogenous catecholamines) without having a significant effect on respiratory drive. In addition, I would administer an anticholinergic such as glycopyrrolate in order to decrease secretion and also act as a bronchodilator, primarily in the conducting airways because of the anticipated irritation of the endotracheal tube as well as the cold, anhydrous gases used. Even without the suggestion on PFTs of a bronchodilator response to the impaired flow/volume loops, the multiple sources of airway irritability during an airway exam under anesthesia justify the use of an anticholinergic for prophylaxis.

2. The adequacy of response of the neuromuscular junction for reversal should be assessed, especially in view of the fact that the time course has only been 15 minutes. The "twitch" monitor may indicate that the patient lacks responsiveness, or perhaps has 1 twitch out of 4. Even in the presence of some post-tetanic facilitation, it is likely too soon to expect reversibility, even with a "full" (i.e., 70 mcg/kg) dose of neostigmine. In addition, the use of an anticholinesterase for reversal

## Postoperative Course

### Questions

1. Following extubation and transfer to the ICU, the patient is now trying to sit up in bed and is breathing 60 times per minute, with an oxygen saturation of 86%. How would you intervene? Does he need more sedation? He has biphasic wheezing – what does this mean? How will you assess the contribution of the biphasic wheezing to his degree of hypoxia and respiratory distress? Is this likely to be improved if you reintubate him, or is there another solution?

2. Two hours after the patient's arrival in the ICU, the nurses call you for your advice about the patients severe, unremitting cough. He's very uncomfortable and feels like he can't breathe well because of all the coughing, and the nurses are afraid he is going to fracture a rib. What can you do to help him?

of neuromuscular blockade can cause bronchospasm or exacerbate the underlying bronchospasm due to its anticholinesterase effects. Sugammadex should strongly be considered inasmuch as its mechanism of action does not result in acetylcholinesterase inactivation nor does it have the adverse muscarinic side effects that neostigmine possesses. For (female) patients on hormonal contraceptives, the package insert warns that another method of nonhormonal birth control should be utilized for 7 days following sugammadex administration.

## Answers

1. In this circumstance, the severity of his parenchymal lung disease is contributing both to his mechanical respiratory distress and his hypoxia. He may very well need the help of tracheal intubation and mechanical ventilation, but there is a downside as well – it will be yet another airway irritant and will not necessarily optimize ventilation-perfusion matching, which is always better with spontaneous breathing through a native airway. First, he needs optimal delivery of supplemental oxygen, which may be by a nonrebreathing mask at high flows. Second, he probably could benefit from some sedation (if that is felt to be a component) because the increased work of breathing (and anxiety) also results in a large increase in his oxygen consumption and carbon dioxide production. Sitting him up, if possible, will promote better excursion of the diaphragm and a better defense of the functional residual capacity, which will add to his margin of oxygenation safety. Finally, a decrease in his respiratory rate will likely promote improved distal alveolar airflow (such as it is with his significantly diseased parenchyma).

2. The coughing is likely due to the ongoing accumulation of secretions in the inflamed airways, and the nurses have a legitimate concern about the ribs (although probably not so much in this age group) but in addition should be concerned about a pneumothorax and the integrity of other intrathoracic structures such as the esophagus. Unfortunately, neither the cause of coughing nor its treatment is all that is well understood. In broad categories, treatments are divided into nonnarcotic and narcotic medications. The nonnarcotics, such as dextromethorphan (ironically derived from opioids) and benzonatate (Tessalon®), are moderately effective. Dextromethorphan may have significant serotoninergic effects. The narcotics (typified by codeine) act primarily at mu-2 receptors.

## Additional Topics

### Questions

1. A 3-year-old male, 15 kg, is scheduled for functional endoscopic sinus surgery, bilateral myringotomy and tubes, and nasal ciliary biopsy. His PMI is in the right precordium. He has a productive cough and mild expiratory wheezing, does not have a runny nose, and is completing a 14-day course of ampicillin. He uses a beclomethasone (Qvar) inhaler twice daily. Why is this patient wheezing? What are the implications of dextrocardia for this patient? Is this patient adequately treated with his current medication regimen? What would you add? Why? The surgeon would like to apply 4% cocaine to the nose and mucosa of the maxillary antrum for topical analgesia and vasoconstriction – is this reasonable? How much will you tell him he can use?

2. A 14-year-old girl is admitted for evaluation of dysphagia, chest pain, and a weight loss of approximately 7 kg over the course of 3–4 weeks. At 5 years of age, she received an allogenic bone marrow transplant for aplastic anemia that was complicated by the chronic sclerodermoid form of severe graft vs. host disease with generalized skin involvement, muscle wasting, and restrictive lung disease requiring supplemental oxygen (0.5 L/min) and nocturnal biphasic intermittent positive airway pressure (BIPAP) ventilation. She is afebrile, with a heart rate of 110–140 beats per minute and a respiratory rate of 18–24 breaths per minute. Her room air oxygen saturation is 91%, increasing to 98% with 0.5 L/min of oxygen. The rigid chest wall shows almost no expansion during inspiration. Pulmonary function tests are:

   - FVC = 17% of predicted
   - FEV1 = 18% of predicted
   - FEV1/FVC = 96% of predicted

## Answers

1. Wheezing, in the setting of asthma or other diseases with bronchoreactivity, is a result of airway narrowing due to inflammation and the accumulation of airway secretions. In chronic conditions, the inflammatory response, particularly in children, may result in tracheo- or bronchomalacia, worsening the wheezing because of the loss of integrity of the cartilaginous matrix of the airways and airway collapse with increased work of breathing. The increased work of breathing exacerbates wheezing by producing more turbulent flow and worsens the underlying metabolic homeostasis of the patient because the harder they work to breathe, the less efficient their breathing becomes. The dextrocardia may be an isolated finding but is more likely related to Kartagener's syndrome, which is characterized by ciliary dysmotility and abnormal polymorphonuclear (PMN) leukocyte motility. Patients develop chronic otitis media, sinusitis, recurrent respiratory infections, and bronchiectasis. Sterility in males is due to abnormal spermatozoa motility. Situs inversus or dextrocardia is associated with the syndrome.

   Single-drug medical management is probably inadequate; beclomethasone is acceptable as one component of long-term management but does not completely address all of the mechanisms of chronic airway reactivity or acute exacerbations. Sympathomimetic and anticholinergic therapy may be required for the unstable preoperative patient. Cocaine has been used historically to provide topical local anesthesia and vasoconstriction to mucosal surfaces, but the margin of safety at calculated maximum doses of 3 mg/kg is much narrower for children. Oxymetazoline (Afrin®) nasal spray is as effective with a greater margin of safety, although care should be taken in its administration as well.

2. Because of the severe restrictive ventilatory defect and the esophageal obstruction, I would begin the induction of anesthesia in the sitting position while continuing spontaneous ventilation. The patient has severe $CO_2$ retention, and I would expect a severe decrease in the minute ventilation response to hypercarbia, so it might be worthwhile to consider a respiratory analeptic to shift the $CO_2$ response curve, such as doxapram [2]. It would probably be necessary to use a small amount of continuous positive airway pressure to maintain the FRC. As the patient lost consciousness, she could be placed in a more supine position, with cricoid pressure applied, and the trachea could be intubated at that point. She is indeed a "full stomach," but, in my opinion, the risks of rapid sequence induction and commitment to positive pressure ventilation are outweighed by the advantages of spontaneous breathing with a careful and slow induction of anesthesia with the preservation of spontaneous breathing. Positive pressure may cause interventricular septal shifting that will significantly decrease her stroke volume and cardiac output, so I would be concerned about delivering

- TLC = 64%
- RV = 194%
- RV/TLC = 78%
- ABG: (0.5 L/min O2) pH = 7.27, pCO2 = 95 mm Hg, p02 = 183 mm Hg

Her ECG is unremarkable apart from a sinus tachycardia. Her echocardiogram is as follows: ruled out right ventricular dysfunction but is suspicious for elevated right ventricular pressure. Her barium swallow demonstrated a web at the cervical esophagus with mild distal narrowing. She is coming to the OR for an endoscopic esophageal dilatation that the gastroenterologists feel should take about one-half hour to 45 minutes.

What are your plans for intraoperative anesthetic technique and postanesthetic care? What are the likely complications and pitfalls?

3. A frightened 8-year-old with a large anterior mediastinal mass presents for supraclavicular lymph node biopsy, and his parents want to know whether he can be asleep for the procedure? What does the answer depend on? Why? Under what circumstances would you want a perfusion team present?

positive pressure ventilation and committing her to controlled ventilation if it could be avoided. Even a small amount of controlled ventilation may be enough, particularly in this patient, to drive her $CO_2$ below apneic threshold that $CO_2$ required for spontaneous breathing. She has a severe restrictive ventilatory defect with incipient changes in the pulmonary circulation and right heart. She has a "cuirass" type of pulmonary physiology which prevents deep breaths either during spontaneous ventilation and most importantly during controlled ventilation [3]. Because of the progressive vascular sclerosis that is characteristic of graft versus host disease, placement of peripheral arterial catheters may carry significant risk of ongoing vascular obstruction, even after the arterial catheter is removed. Notwithstanding her history (and anticipated complication) of elevated pulmonary vascular resistance and right heart problems, these are well understood and changes intraoperatively can be interpreted in light of these findings and supporting evidence from noninvasive monitors such as $ETCO_2$ and pulse oximetry [4].

3. An anterior mediastinal mass may or may not cause symptoms depending on its encroachment on the tracheobronchial tree and the right heart and pulmonary circulation. Physical diagnosis is not always helpful with regard to the severity of the chest disease, because even patients with more than 50% airway narrowing may only be symptomatic with orthopnea; the superior vena caval syndrome is relatively rare in pediatric patients [5–7]. Pulmonary function tests may be helpful in demonstrating inspiratory and expiratory compromise. At 8 years of age, a MAC plus good local anesthesia by the surgeon is a very acceptable plan. The patient should also be placed in semi-Fowler's position for optimal comfort for ventilation and gas exchange. Premedication with oral benzodiazepines may be a very rational plan and will lessen the patient's anxiety and improve the chances for a successful operating room course. For patients whose risks increase because of greater than 50% airway encroachment or significant tumor impingement on the pulmonary circulation or the right ventricle, it is not a bad idea to have a rigid bronchoscope available. In the highest-risk situations, when the patient has had episodes of syncopy which may be related to a drop in pulmonary blood flow as a result of right heart obstruction, it may be worthwhile to have both groins prepped and draped and have a perfusion team standing by to cannulate and go on to cardiopulmonary bypass immediately.

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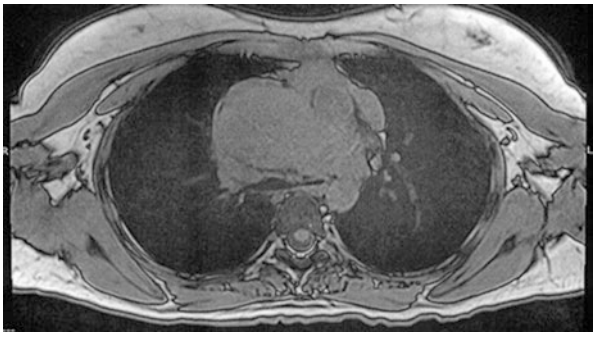
# Chapter 20
# Thoracic Surgery

**Robert S. Holzman**

A 13-year-old girl, 41 kg, with a middle mediastinal mass is scheduled for a thoracoscopic biopsy. A prior biopsy was consistent with a lymphoproliferative disorder. She does not have a cough, has no difficulty lying flat, and is not short of breath. Her initial presentation several months earlier was with shortness of breath and a pericardial effusion requiring drainage (1200 mL).

On MRI, the mass is 8.8 (transverse) × 4.9 (anteroposterior) × 8.3 (longitudinal) cm. The mass is just behind the SVC, extending along the right paratracheal region, and is insinuated in the middle mediastinum, anterior to the carina. There is displacement of the main pulmonary artery leftward as well as downward displacement and compression of the right main pulmonary artery branch. The thoracic aorta is normal in caliber. There is compression of the right upper lobe airway and also compression of the left mainstem bronchus by the inferior component of the patient's mediastinal mass (see image). The central airway is patent.



To facilitate the biopsy, the surgeon requests one-lung ventilation.



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## Preoperative Evaluation

### Questions

1. Given the findings, would you anticipate any comorbidities? Where is the middle mediastinum, and how does morbidity differ from patients with an anterior mediastinal mass?

2. Are there any further tests you would want in preparation for the surgery? Would further imaging be useful? An echocardiogram? What about pulmonary function tests? What specifically would you look for – dynamic airway exam, static airway exam, or evidence of a diffusion problem?

3. Do you have any specific considerations in preparing for one-lung ventilation for this case?

## Answers

1. The middle mediastinum is the central portion of the inferior mediastinum (the portion of the mediastinum caudal to the thoracic plane, drawn from the sternal angle to the intervertebral disc of T4–T5.) The middle mediastinum is bounded by the pericardial sac. While the clinical significance of the middle mediastinum with regard to tumors typically involves lymphadenopathy or metastatic lung disease, pediatric masses of the middle mediastinum typically include lymphoma or tuberculous nodes. These tumors can often affect contiguous structures by their mass effect. This is evidenced at the level of the carina and mainstem bifurcation in the CT scan slice as compression of both mainstem bronchi, with a greater effect on the left than the right.

   Anterior mediastinal masses, with regard to pathology, may also be lymphomas but can also be teratomas, thymomas or enlarged thymus glands, pericardial cysts, diaphragmatic hernias, or cystic hygromas. These may also have a mass effect on the trachea and heart and may, in addition, present with superior vena caval syndrome [1].

2. The location of the tumor and the CT scan evidence of bronchial compression should prompt a more complete investigation of the airway and cardiac implications. Pulmonary function testing would examine any component of dynamic airway obstruction as well as air-trapping leading to higher static lung volumes. Both of these findings, when abnormal, can be helpful in guiding mechanical ventilation, such as the use or avoidance of PEEP, alterations in the I:E ratio, or selection of tidal volumes based on compliance [2]. Because of the location in the middle mediastinum, the heart should be evaluated for the tumor's effect on the pulmonary circulation and impedance to the right ventricular outflow tract and the atria. The fact that the patient is basically asymptomatic is reassuring but not necessarily completely predictive when ventilation is controlled and the patient is anesthetized.

3. One-lung ventilation is indicated to facilitate the thoracoscopic approach, and in a patient this size can be accomplished with a smaller double-lumen endobronchial tube. A double-lumen TT is labeled by the OD of the entire tube, with the measurement reported in French (Fr). Even though the OD is standardized, the ID of the double-lumen TT can vary between manufacturers. There is no need to memorize sizing tables, as one can obtain the ID of a lumen of any TT by simply looking on the package or in the package insert. In order for a well-lubricated bronchoscope to physically fit inside the lumen of the TT (and not seize up from friction), the OD of the bronchoscope (ODB) needs to be <90% of the ID of the

## Intraoperative Course

### Questions

1. What are your considerations for anesthetic induction? Is it safe to go ahead with a routine intravenous induction of anesthesia? Why? Would you use neuromuscular blockade to facilitate tube placement? Would it be safer for the patient to breathe spontaneously? Under what circumstances is this a good idea?

2. Would you place a single-lumen tube first before the double-lumen tube? How will you select the proper size for this patient? What method will you use to confirm proper placement of the tube? Does the surgeon's choice of surgical approach influence the tube you will select? Why? What are the methods for confirming correct placement? Is ultrasound useful?

TT (IDTT). This can be written as ODB/IDTT <0.9. Bronchoscopes can become deformed from repeated use and cleaning, and the tip may have a larger diameter than documented. It is always a good idea to test the fit of the bronchoscope inside a TT before use in the patient. During bronchoscopy, even if the CSA ratio is <50%, there will be an increase in airway resistance and a decrease in ventilation. So, the proper-sized bronchoscope and the proper-sized double-lumen tube need to be preselected. I would plan on a 32 French (French = outer diameter); tracheal tubes are usually cited as inner diameter, with the conversion of 3:1 French/mm. I would plan for a 32 French double-lumen tube. The lumen size of each tube varies with the manufacturer but is typically around 4.5 mm ID, so I would plan on using a 2.8 mm bronchoscope to guide the tube into the left mainstem bronchus.

## Answers

1. It is reassuring that the patient is asymptomatic while awake with regard to respiratory distress. This won't necessarily be true with apnea, positive pressure ventilation, and a general anesthetic. The CT scan suggests that air-trapping may occur because of extrinsic collapse of the airways at the level of the mainstem bronchi. Preservation of spontaneous breathing, which can be accomplished with incremental doses of propofol and a volatile anesthetic until the patient is completely anesthetized, and then cautious conversion to controlled ventilation in the absence of neuromuscular blockade would be a worthwhile exercise. If tolerated without air-trapping, then neuromuscular blockade can be introduced, cautiously. Relying on spontaneous breathing will not work after the introduction of a pneumothorax for the thoracoscopy.

2. This is certainly an acceptable technique if a difficult airway is anticipated or impaired gas exchange is encountered, but in most circumstances, mask ventilation can be followed by placing the double-lumen tube directly. The proper size in this age group would be about a 32 French; as an outside diameter, this would be 10.7 mm (normal tracheal diameter about 13 mm) [3]. While there are different ways to confirm tube placement, it is most reasonable to intubate over an appropriately sized fiber-optic bronchoscope selectively placed into the proper bronchus. Most clinicians tend to choose only left-sided endobronchial tubes because of the higher likelihood of occluding the right upper lobe bronchus when a right-sided tube is chosen. For this case, it would depend on whether a left-sided tube can be advanced in the left mainstem bronchus, given the extrinsic compression. A carefully placed right-sided tube may need to be chosen because of the extrinsic compression to the left mainstem bronchus. The pre-

3. Does this patient need an arterial line? Does it matter if the surgeon intends to do a formal lateral position thoracoscopic approach vs. a "bump up" at 300? How is this likely to affect gas exchange, oxygenation, and ventilation?

4. What is your plan for postoperative analgesia? Would a regional anesthetic be indicated? The surgeon tells you he doesn't want the patient to have a Foley catheter postop. Does this influence your choice of regional technique?

5. With lateral positioning and insufflation, the patient's SpO2 decreases to 85% on 100% oxygen. What do you think is going on? What is likely to make it better? Will it improve if you just "wait it out"?

formed endobronchial tube will not necessarily enter the left mainstem bronchus when placed blindly, but if blind placement is chosen, it can be confirmed by auscultation or chest X-ray. As clinicians use ultrasound with greater frequency, one can look for the loss and resumption of pleural sliding when the endobronchial lumen is occluded and then ventilated, respectively.

3. Although not strictly required for this case, an arterial line is important for close monitoring of the hemodynamic consequences of the pneumothorax and insufflation on gas exchange and myocardial performance, especially when superimposed on any comorbidities resulting from the mass effect in the middle mediastinum. In addition, because of the vital structures in the area and the chance for any complications, closer monitoring can be accomplished with an arterial line. Numerous factors influence the efficiency of gas exchange during a VATS procedure, especially in a lateral decubitus position – the acute change in hydrostatic pressure gradient, the influence of abdominal viscera on diaphragmatic length and excursion, the influence of the anesthetic technique on hypoxic pulmonary vasoconstriction, compression of the dependent lung parenchyma, the shift in the mediastinum and its influence on blood return and the cardiac output, etc. Older children and adolescents behave much like adults with regard to V/Q matching during one-lung ventilation; a period of V/Q mismatch is typically followed by progressive adjustment particularly when lower levels of volatile agents are utilized because of less depression of hypoxic pulmonary vasoconstriction. Infants and small children do not typically fare as well, and OLV frequently cannot be accomplished in this age group, resulting in the surgeon's need to manually compress the lung for exposure or to tolerate periodic reinflation maneuvers during the procedure.

4. With a VATS approach, analgesia may be accomplished by opioids, primarily. Adjunctive regional techniques would include intercostal nerve blocks performed by the surgeon under direct vision through the thoracoscope, the anesthesia team percutaneously, a paravertebral block, or a thoracic epidural. While a thoracic epidural can be limited to segmental blockade and have a higher likelihood of preserving bladder detrusor function, it is reasonable, when concerned about avoiding a Foley catheter, to use a paravertebral approach instead.

5. This is not surprising when getting started and with the initiation of OLV. The strategy for avoiding hypoxemia is to maximize pulmonary vascular resistance on the operative side and minimize PVR in the dependent lung. Defense of the FRC in the ventilated lung will enhance normal pulmonary vascular resistance and will counter the paralysis, lateral positioning, the open or insufflated contralateral hemithorax, and the weight of the mediastinal structures. The paO2 and SpO2 may continue to fall/readjust for 45 minutes after OLV is started [4]. CPAP for the dependent or operative lung can also be tried when intraoperative hypoxemia is encountered, although the surgeon's view may be more difficult. Multiple strategies in varying combinations may need to be tried in order to achieve an optimal shunt fraction.

## Postoperative Course

### Questions

1. The patient is complaining of pain in the PACU, rating it 8/10. How is this possible if the paravertebral block was sited correctly? Is there anything you can offer to provide additional pain relief, other than a systemic opioid?

2. You note bubbling within one of the chambers of the Pleur-Evac three-chambered chest tube drainage system. Why are there three chambers? What do they represent? What is the significance of the bubbling? What does it represent anatomically? What can you do to fix it?

## Additional Topics

### Questions

1. A 2-day-old girl with tachypnea and chest X-ray evidence of a right upper lobe congenital cystic adenomatoid malformation (CCAM) needs to come urgently to the operating room for a procedure. Her delivery was straightforward; she is at term, with a birth weight of 4 kg. Is this an unusual problem? Common? Relatively common for neonatal surgical emergencies? For neonatal thoracic surgical emergencies? Would your considerations be any different if this was a congenital lobar emphysema or a pulmonary sequestration with regard to planning the anesthetic?

   What are your considerations for the bronchoscopy exam which has to precede the thoracotomy? Is there any way you can offer one-lung ventilation in order to facilitate the surgeon's exposure? Does it matter if the CCAM is on the left or on the right? How can you place a selective endobronchial tube in this age patient? What are the pulmonary consequences of doing so in a 2-day-old? What are the cardiovascular consequences?

## Answers

1. A paravertebral block will not necessarily provide complete analgesia, particularly in the immediate perioperative period although the overall success rate is comparable to thoracic epidural analgesia. The incidence of hypotension, urinary retention, itching, nausea, and vomiting (7–17%) is lower than with intravenous analgesia or thoracic epidural analgesia. Other adverse events which are different from those for epidural analgesia include pneumothorax, pleural puncture, and vascular puncture, most of which are moot for thoracic surgery. The use of ultrasound guidance has substantially reduced these risks.

2. The three chambers have three different purposes – a collection chamber, a water-seal chamber, and a suction control chamber. The collection chamber drains fluid, blood, or air. The water seal chamber holds a column of water (e.g., 2 cm) which prevents air from being sucked into the pleural space with inspiration, and the suction chamber may use a water column or controlled suction to generate up to −25 cm H2O for the water column or −40 cm H2O for suction. The presence of bubbling indicates an air leak, which may be accentuated by the patient coughing. In addition to a pleural tear, bubbling may indicate a migrated tube with drainage holes outside the skin or an inadequate closure of the chest tube insertion site [5].

## Answers

1. CCAMs comprise approximately 25% of all congenital lung malformations, with an estimated incidence of 1:25,000 to 1:35,000. Eighty percent of affected neonates present with some degree of respiratory distress due to pulmonary compression or hypoplasia. As a developmental outgrowth of the tracheobronchial tree, these anomalies may become overdistended due to air-trapping.

   Secondary causes of respiratory insufficiency are mediastinal shift and spontaneous pneumothoraces. If there is time for a portable chest X-ray, it might be worthwhile to look for evidence of worsening intrathoracic volume of the CCAM and mediastinal shifting. Blood work, such as arterial blood gases, will not help at this point. If there is no time for a CXR, then air-trapping and mediastinal shifting with a possible pneumothorax should be anticipated and preparations made for chest tube drainage and respiratory support. The bronchoscopy exam should be accomplished with spontaneous breathing so as not to deliver positive pressure, or if positive pressure is required, it should be the lowest possible to

2. Under what circumstances would you insist on a preoperative cardiology evaluation and echocardiogram prior to a TE fistula repair?

3. A19-year-old male, 6 feet tall and 60 kg, is scheduled for a left VATS pleurodesis for a recurrent pneumothorax. His initial pneumothorax was 2 weeks ago, treated with oxygen overnight, and then he was discharged home. What is your anesthetic plan? Would you administer a muscle relaxant as part of the anesthetic induction? Why or why not? Are there any special considerations for mechanical ventilation during the surgery? How would you set the I:E ratio on the ventilator? Why does this matter? Several bullae were excised – how does this influence your anesthetic management? What postoperative problems can you anticipate in relation to this?

avoid insufflation of the CCAM. Each of the three congenital lung lesions has several important features. Congenital lobar emphysema (CLE) involves abnormal emphysematous lung that communicates with a bronchus; overexpansion is a major concern. CCAM tissue does not participate in gas exchange but may communicate with the tracheobronchial tree, and therefore the same concern remains with regard to air-trapping. Pulmonary sequestration lesions involve nonfunctional lung tissue without a bronchial connection, and the blood supply is from anomalous systemic arteries. Selective endobronchial intubation can be accomplished with a bronchial blocker in this age group, under the guidance of a flexible fiber-optic bronchoscope or a rigid bronchoscope/telescope. Children in this age group, however, may not tolerate one-lung ventilation well and do not have the same capability as older children and adults to utilize hypoxic pulmonary vasoconstriction as a compensatory mechanism for oxygen desaturation [6].

2. TE fistula may occur as an isolated finding or be a component of the VACTERL syndrome (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities). In particular, cardiac defects occur in a range of 40–80%; therefore, in association with the presence of more than one anomaly, a thorough cardiac evaluation should be accomplished. Normal physical findings and the absence of radiological abnormalities are favorable for lack of associated congenital heart disease [7].

3. It is highly desirable to have lung isolation for this procedure, which will facilitate the surgical approach to bullae excision as well as a pleurodesis. It is not surprising that the patient is a male; the ratio is about 3:1 male/female, usually tall and thin body habitus. The anesthetic plan may include a regional block such as a thoracic epidural or an ipsilateral paravertebral block. Often the surgeons will do intercostal blocks under direct vision with the thoracoscope. It is ideal to intubate these patients with an appropriately sized left-sided double-lumen tube while breathing spontaneously, in order to avoid positive pressure. If positive pressure is required, then the lowest practicable peak inflation pressure should be used, together with an I:E ratio that allows for optimal lung emptying. Small tidal volumes are justified, although the minute ventilation is not and, in this age group, is often elevated because of the increase in oxygen consumption. Once the lung is isolated, neuromuscular blockade can be administered to facilitate a motionless surgical field. With the excision of the bullae, one has to be sure that the staple line is intact without any air leak. This is why the lung is re-expanded under direct vision once the bullae have been excised [8].

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# Chapter 21
# Cardiac I


**James A. DiNardo**

A 3-year-old male with a diagnosis of tetralogy of Fallot and a classic right Blalock-Taussig shunt (right subclavian artery to right pulmonary artery) created at 1 month of age presents with a history of gradually decreasing exercise tolerance and increasing frequency of hypercyanotic episodes. He had a hematocrit of 69% 2 months previously. He is scheduled for complete repair of his lesion.



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## Preoperative Evaluation

### Questions

1. What is the tetralogy of Fallot? Is it really four lesions? What is the principal anatomic lesion?

2. What are the reasons that this child would have received a palliative shunt for his first surgical procedure rather than a definitive repair? Are there any long-term consequences of a Blalock- Taussig shunt (BTS) of importance to you as an anesthesiologist?

3. The patient is extremely apprehensive and very scared of needles. How will you gain his confidence so that you can perform a physical exam? How will you assess him for volume status and dehydration? Would you expect his hematocrit to be any different today than it was 2 months ago? What do you expect his oxygen saturation to be? Any additional lab studies you would like to see? Any additional diagnostic cardiac information you would like to have? What would you expect to find on echocardiogram?

## Answers

1. The primary lesion in tetralogy of Fallot is a conoventricular malalignment ventricular septal defect (VSD). As a result of this lesion, there is anterior and superior displacement of the aorta and "crowding" of the pulmonary outflow tract. This lesion produces aortic override (50% or more of the aorta over the VSD), hypoplasia of the pulmonary outflow tract (pulmonic stenosis), and dynamic right ventricular outflow tract (RVOT) obstruction due to anterior deviation of the conal septum and muscle bundles in the RVOT. Right ventricular hypertrophy (RVH) occurs secondary to fixed and dynamic RVOT, not as a primary manifestation of the lesion.

2. A palliative shunt is typically done in institutions not versed in infant cardiac surgery utilizing cardiopulmonary bypass (CPB) or in resource-limited environments. The long-term adverse consequences of such a shunt would be (1) poor growth of the native pulmonary arteries due to preferential blood flow to one lung, stenosis at the anastomosis site, or chronic low pulmonary blood flow. In the worst case scenario, the branch pulmonary arteries might become discontinuous. (2) Progression of RVH as placement of a shunt does not address RVOT obstruction and thereby does not remove the stimulus for continued hypertrophy.

   Following a classic BTS, the right arm will not be a reliable source of either noninvasive or invasive BP monitoring as perfusion to the arm is provided via collateral vessels around the shoulder and scapula. Following a modified BTS (graft from innominate artery to right pulmonary artery), accurate blood pressures might be obtainable, but this will have to be determined by comparison with noninvasive pressures from the contralateral arm.

3. Volume status would be assessed in this child as in any child of similar age. The erythrocytosis associated with cyanosis is progressive; however, hematocrits above 70% are rare. A baseline oxygen saturation of 60–70% with desaturation episodes into the 40–50% range would be expected. A platelet count would be useful as erythrocytosis is associated with thrombocytopenia. A PT and PTT would be useful as chronic cyanosis is associated with poorly defined coagulation abnormalities. It would be necessary to know the status of the pulmonary vasculature. Specifically, it is necessary to know whether the pulmonary arteries are continuous and of normal size. The echocardiogram will demonstrate severe RVH, and there will be bidirectional flow across the VSD. It is possible that the flow across the VSD could be entirely right to left with all pulmonary blood flow supplied by the Blalock-Taussig (BT) shunt.

4. He has a “tet spell” in the examining room – how do you manage it? What is a “tet spell,” physiologically and anatomically? Why do you choose the maneuvers you choose? Would it be any different in the preoperative holding area? How about in the operating room during induction intraoperative course?

4. A "tet spell" is simply an exacerbation of the dynamic component of RVOT obstruction that results in increased right to left shunting across the VSD. It will be precipitated by physiologic perturbations that reduce the caliber of the RVOT: (1) reduced venous return and (2) increases in shortening and thickening of the free wall and septum of the RV. Treatment is directed at these causes. Alternatively treatment can be directed toward increasing systemic vascular resistance (SVR) which will reduce right to left shunting at the VSD but does nothing to treat the underlying cause of the "tet spell." The only difference between the holding area and the operating room is the extent of monitoring, qualified personnel, and drug choices available.

## Treatment Modalities

**Improved venous return**

- Volume infusion – at least 10–15 mL/kg
- Sedation – reduces tachypnea (morphine is used most frequently)
- Administration of an alpha agonist (phenylephrine) – reduces venous unstressed volume and improves venous return

**Relaxation of the RVOT**

- Sedation
- Negative inotropes – beta-blockers (esmolol). Heart rate reduction may also increase the caliber of the RVOT by increasing RV end-diastolic and end-systolic volumes.

**Increasing SVR**

- Administration of an alpha agonist (phenylephrine)
- Aortic or femoral artery compression – manual compression of the abdominal aorta, hip flexion

## Intraoperative Course

### Questions

1. Are there any specific drugs you would like to have ready in the OR, besides the "usuals"?

2. What kind of monitoring will you select? Place prior to or post induction? Any specific considerations for placement of the monitors? Invasive lines? ECG?

3. How will you induce anesthesia? Maintenance? Fluid management strategy? Any particular considerations for the Blalock-Taussig shunt?

4. What are your considerations as regards the hemodilution associated with cardiopulmonary bypass?

5. Do you expect any coagulation problems? Would you use any particular pharmacological management strategy in this regard?

6. What are your considerations as regards weaning from cardiopulmonary bypass?

## Answers

1. Phenylephrine, infusions of dopamine, epinephrine, milrinone, and tranexamic acid

2. ECG (5 lead including V), pulse oximeter X 2 (upper and lower), BP X 2 (upper and lower, arterial line and BP cuff), CVP. ECG, pulse oximeter, and BP cuff prior to induction. Right arm may not be a reliable source of central BP measurement given the presence of a right Blalock-Taussig shunt (BTS).

3. Induction and maintenance of anesthesia could be accomplished with any agent or combination of agents that preserve myocardial contractility and pulmonary blood flow. Maintenance of pulmonary blood flow via the BTS is dependent on maintenance of systemic BP and a normal or low pulmonary vascular resistance (PVR). Appropriate PVR can be obtained with a high $FiO_2$ and pH of 7.45 or above. pH can best be maintained in this range with a mild respiratory alkalosis ($PaCO_2$ of 30–35 mm Hg in the absence of a metabolic acidosis). The ventilatory settings to accomplish this should be achieved with the lowest mean airway pressures possible to avoid mechanical impairment of pulmonary blood flow. This usually is accomplished with a tidal volume of 10–12 mL/kg, an I:E ratio of 1:3, and a rate of 10–15 breaths per minute.

4. Hemodilution is an expected consequence of CPB. In this instance, the high hematocrit will likely lead the perfusionist to prime with a little or no blood in order to attain a target hematocrit of 25–35%. The prime will most likely be a crystalloid or colloid solution. The danger to this patient will be a dilutional thrombocytopenia and factor deficiency secondary to the reduced plasma volume associated with erythrocytosis. This could be countered by adding FFP rather than a crystalloid or colloid solution.

5. The clotting abnormalities have been previously delineated. Use of a lysine analog antifibrinolytic agent such as tranexamic acid (TXA) or epsilon-aminocaproic acid (EACA) is warranted. The serine protease inhibitor aprotinin is currently unavailable for use.

6. The major CPB weaning issues are:

   - Reduced RV systolic function as a consequence of less than optimal preservation of the hypertrophied RV and the ventriculotomy that may be necessary to close the VSD/augment the RVOT.

7. What are your transport considerations from the OR to the intensive care unit?

## Postoperative Management

### Questions

1. What is the systemic inflammatory response syndrome (SIRS) and how does it affect the postoperative course?

2. What arrhythmias will you anticipate occurring in the ICU? Why?

- Reduced RV diastolic function for the reasons outlined above.
- Residual volume load on the RV from either pulmonary insufficiency (from a transannular patch) or a residual VSD. A residual VSD will be very poorly tolerated as it presents an acute volume load to the LV and to a chronically pressure-overloaded RV.
- Right to left shunting at the atrial level if the foramen ovale has been left open as a pop-off valve.
- Atrial arrhythmias, particularly junctional ectopic tachycardia (JET), if the VSD was closed across the tricuspid valve.

7. Transport to the ICU with continuous BP, pulse oximetry, and ECG. Blood or colloid for volume infusion. Hemodynamic stability while the patient is being ventilated with the transport bag should be obtained prior to leaving the OR.

## Answers

1. SIRS is characterized by tissue and endothelial injury leading to enhanced capillary permeability (capillary leak syndrome) and transmigration of leukocytes into interstitial fluid with subsequent activation of sequestered leukocytes, elaboration of chemoattractants, and amplification of the inflammatory process. The spectrum of SIRS-induced responses ranges from tissue edema to end-organ dysfunction and failure. It is not uncommon to see hyperglycemia in children with SIRS or to see an elevation of hematocrit due to extravasation of intravascular fluid to tissue spaces. The magnitude of this response is enhanced in neonates/infants and small children due in part to the high circuit surface area to blood volume ratio as compared to adults.

2. JET (junctional ectopic tachycardia) is the most likely. The substrate for this arrhythmia is injury to myocardium in and around the AV node occurring as consequence of surgical retraction of the tricuspid annulus. JET typically manifests with a junctional rate only slightly faster than the sinus node rate and is the only narrow complex tachycardia in which the atrial rate is less than the ventricular rate (A:V ratio <1:1). Much less commonly (10%), there may be retrograde activation of the atrium with inverted p waves noted and an A:V ratio of 1:1. In either case, there is loss of AV synchrony (loss of atrial kick). At a HR <160–170 bpm, this arrhythmia may be well tolerated. It is unlikely to be tolerated in the presence of restrictive diastolic function at any rate. JET with HR >170 bpm is associated with hemodynamic instability and increased postoperative mortality.

## Additional Topics

### Questions

1. What differences are there in the architecture of the immature and adult myocardium, and what physiological implications does this have? How would it affect your anesthetic management? What would the length-tension curve look like compared to an adult myocardium?

2. Why should systemic to pulmonary artery shunts be clamped or ligated as soon as bypass begins? Of what importance is this to your anesthetic management?

3. A 12-month-old child with trisomy 21 presents for reduction of macroglossia. On examination you hear a murmur. Would you be prepared to continue with this case? Are there any cardiac abnormalities you would expect? Why? How does this happen? What implications are there for anesthetic management?

Neither cardioversion nor adenosine is effective. Treatment of JET is atrial pacing at a rate slightly faster than the junctional rate reinitiating A-V synchrony. This therapy is effective unless the junctional rate is very fast (>160–170 bpm) at which point atrial pacing at a faster rate is unlikely to improve hemodynamics because the reinitiation of A-V synchrony is offset by the reduction in diastolic filling time present at these rates.

## Answers

1. The architecture of the immature myocardium is characterized by reduced contractile elements and smaller myocytes than adults. In addition, the fetal myocardium has a decreased sarcoplasmic reticulum and a poorly developed or absent T-tubule system as compared to adults. This results in a greater dependence on trans-sarcolemmal calcium influx for generation of contractile function and a greater susceptibility to the negative inotropic properties of anesthetic agents. The fetal tension-length curve is characterized by greater resting tension (impaired diastolic relaxation) and lower developed active tension (impaired systolic function and a propensity to develop afterload mismatch).

2. All systemic to pulmonary artery communications whether anatomical or surgical need to be controlled and clamped or transected prior to initiation of CPB to prevent recirculation of CPB pump flow into the pulmonary circulation. In the presence of uncontrolled communications, some portion of CPB flow will go to the lungs and the left atrium. This will produce systemic hypoperfusion and distention of the nonbeating, unvented heart. If the shunt cannot be controlled immediately and a vent is placed in the left atrium to prevent distention of the left heart, CPB pump flow must be increased to compensate for this "steal" from systemic blood flow.

3. The most likely defect in this patient would be an endocardial cushion defect comprised of a primum atrial septal defect (ASD), an inlet ventricular septal defect (VSD), and a common AV valve. This is associated with a large left to right physiologic shunt (high Qp:Qs) and high pulmonary artery pressures. Eventually this child would be at risk for development of elevated PVR due to pulmonary hypertensive vascular disease (PHVD) with a gradual decrease in left to right shunting.

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# Chapter 22
# Cardiac II


**James A. DiNardo**

A 2-year-old boy presents for an exploratory laparotomy for treatment of a presumed small bowel obstruction. He has a Holmes heart type of double-inlet left ventricle (single ventricle, great vessel concordance, hypoplastic pulmonary outlet chamber). He was palliated with a bidirectional Glenn shunt at age 10 months for progressive cyanosis. His native pulmonary artery has not been oversewn. There is a 60 mm Hg echocardiographically derived gradient across the pulmonary outflow tract and some antegrade pulmonary blood flow. He tolerates exercise well and has a Hct of 45%. His baseline $SpO_2$ is 75–80%.



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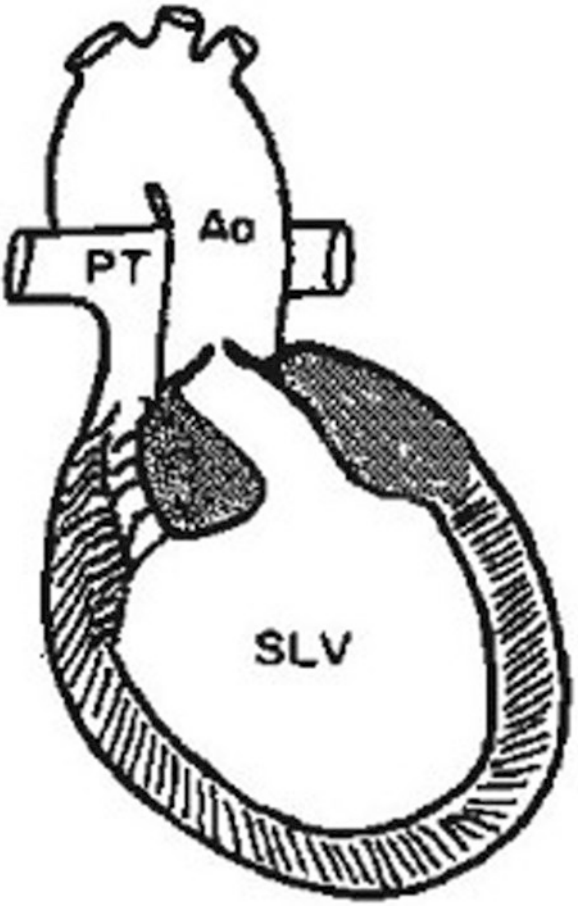



## Preoperative Evaluation

### Questions

1. What is the difference between a single inlet and a double-inlet ventricle? Why would it matter? What are the morphologic components of any ventricle? Is it important to your anesthetic management to understand the outlet component? Is there a difference whether or not the outlet component is concordant or discordant with the ventricular morphology? Why?

## Answers

1. A double-inlet ventricle is one in which both AV valves drain into one ventricle resulting in complete mixing of systemic and pulmonary venous blood. Subsequent delivery of this mixed blood to the systemic and pulmonary circulations then occurs in parallel (not series) from one ventricle. These two features are the basis of single ventricle physiology. The morphological components of a ventricle are the inlet, the body, and the outlet. A "ventricle" which is composed of only an outlet portion is called a bulboventricular foramen (BVF). Transposition of the great vessels (TGV) refers specifically to the anatomic circumstance

2. What is a bidirectional Glenn shunt? How does it differ from a Fontan procedure? In what way are the differences relevant for anesthesia? Are there any specific differences with regard to anesthesia for venous to pulmonary artery shunts compared to systemic arterial to pulmonary artery shunts such as the modified Blalock-Taussig shunt (mBTS) or Waterston shunt? When would it matter?

3. Of what significance are the exercise history and the hematocrit? What if he was simply short of breath with exertion but had a hematocrit of 45%? What if the hematocrit was 57% and he was short of breath at rest? Does this patient need to come into the hospital the night before surgery for intravenous hydration? Why/why not? What would your choice of fluid be? Full maintenance rate, one-half maintenance? Why?

wherein there is concordance of the atrioventricular connections associated with discordance of the ventriculoarterial connections. Corrected transposition (C-TGV) refers specifically to the anatomic circumstance wherein there is discordance of the atrioventricular connections associated with discordance of the ventriculoarterial connections. In the case of single ventricle physiology, concordance or discordance of the ventriculoarterial connections becomes academic as systemic and arterial flow are derived from one ventricle. The relevant issue is whether the outflow to either circulation from the single ventricle is obstructed. In the case described here, there is potential for subpulmonary obstruction at the level of the VSD communication to the BVF and pulmonary outflow tract.

2. A classic Glenn shunt is an end-to-end anastomosis of the superior vena cava (SVC) to the right pulmonary artery (RPA) in which the RPA is separated from the main PA. Thus, all pulmonary blood flow is derived from the SVC to RPA anastomosis. A bidirectional Glenn (superior cavopulmonary anastomosis) is currently the procedure of choice. It involves an end-to-side anastomosis of the SVC to the right pulmonary artery in which the RPA is in continuity with the main PA and LPA. Thus, all pulmonary blood flow is delivered from the SVC to the RPA and LPA which are in continuity. All IVC blood is delivered to a common atrium (mixing with pulmonary venous blood) following a Glenn shunt. These shunts are dependent on systemic venous pressure (SVC pressure) to provide pulmonary blood flow and not on systemic arterial pressure as is the case with systemic to pulmonary artery shunts such as the mBTS (usually graft between innominate artery and RPA) or Waterston shunt (ascending aorta to RPA anastomosis).

   The Fontan procedure creates continuity between the IVC and the SVC, and the pulmonary artery creates a total cavopulmonary connection. Following a bidirectional Glenn shunt, Fontan physiology would be created by surgically creating continuity between the IVC and the underside of the pulmonary artery. Following a Fontan, all systemic venous blood is delivered to the pulmonary circulation. Cardiac output is dependent on delivery of systemic venous blood across the pulmonary vascular bed to the single ventricle. The driving pressure for delivery of this blood is the difference between the systemic venous pressure and the pressure in the common atrium into which the pulmonary veins drain: the transpulmonary gradient (TPG).

3. The hematocrit is consistent with adequate pulmonary blood flow and systemic oxygen delivery as there is no compensatory erythrocytosis. The good exercise tolerance is consistent with preserved ventricular function and chronotropic reserve such that cardiac output and systemic oxygen delivery can be increased. In patients with a palliated congenital heart lesion, loss of chronotropic reserve may be a component of impaired exercise tolerance. A hematocrit of 57% and shortness of breath would warrant further evaluation, specifically an echocardiogram to evaluate ventricular function, AV valve regurgitation, and patency of the

4. Should this patient receive a premedication prior to coming to the operating room? What if he was extremely apprehensive? What would be the potential advantage? If the patient became profoundly sedated and difficult to arouse to anything except vigorous shaking, would that be a problem? Why?

## Intraoperative Course

### Questions

1. Other than routine noninvasive monitors, does this patient need any invasive monitoring? Why/ why not? Would a central line be helpful? Why? How about a pulmonary artery catheter? Would you go to the cardiac catheterization lab before surgery to place one? Does ECG lead placement need to be configured in any particular way for a single, morphological left ventricle? What would you expect to see with standard ECG lead placement?

2. How would you induce anesthesia for this patient? What would be important in your decision? Is an inhalation anesthetic better than high-dose fentanyl? Is ket-

Glenn pathway. Cardiac catheterization would be necessary to rule out decompressing venous collaterals from the Glenn pathway to the IVC or directly to the pulmonary venous system. These collaterals would diminish pulmonary blood flow and increase right to left shunting (systemic venous blood to the common atrium). In addition, cardiac catheterization would delineate the extent of pulmonary arteriovenous malformations (AVMs); these are a source of intrapulmonary right to left shunting (systemic venous blood delivered directly to the pulmonary veins). These AVMs develop in patients with Glenn shunts due to a lack of hepatic venous blood being delivered to the lungs.

Delivery directly to the lungs (without passing through another capillary bed first) of an unidentified hormonal factor produced by the liver is necessary to prevent development of AVMs. The liver must receive and process gut blood flow to create this factor. A similar phenomenon occurs in cirrhotic patients, not as a result of deprivation of the pulmonary bed of hepatic venous blood but as a result of severely impaired hepatic synthetic function. This patient should be admitted the night before surgery and given full maintenance fluids. This is necessary to prevent dehydration in the setting of preexisting erythrocytosis.

4. The decision to premedicate should be individualized to the patient's preoperative physiological status. Oversedation of this patient would likely cause pulmonary venous desaturation as a result of hypoventilation and development of pulmonary V/Q mismatch and shunt. This patient is no more at risk for development of this than a normal patient; however, the consequence in terms of systemic arterial saturation is more severe given that this patient's arterial saturation is a weighted average of the volume and saturation of pulmonary venous blood and the volume and saturation of systemic venous (in this case IVC) blood.

**Answers**

1. A central line via the right internal jugular vein would end up in the RPA if a classic Glenn was present and in the MPA if a bidirectional Glenn was present. In either case the transduced pressure would be the mean PA pressure. The difference between the MPA pressure and the mean common atrial pressure would be the transpulmonary gradient (TPG). Placement of a pulmonary artery catheter would allow measurement of the MPA and the common atrial pressure indirectly via a pulmonary artery occlusion pressure (PAOP). However, placement of such a catheter would be highly impractical as it would require fluoroscopy in the cardiac catheterization laboratory and would provide little additional information. An arterial line would be a necessity. Standard ECG lead placement would reveal qRS forces directed leftward and anterior and would be consistent with left ventricular hypertrophy (LVH).

2. The major anesthetic goal here would be avoidance of elevated PVR. In the case of a classic Glenn shunt, dynamic obstruction to pulmonary blood flow across the VSD

amine an acceptable choice if the patient is screaming and crying uncontrollably in the pre-op area? Why/why not? Would you be worried about the patient under this circumstance?

3. The patient is given an intramuscular injection of ketamine/midazolam and glycopyrrolate in the midst of crying in the preoperative holding area and over the course of 30 seconds remains as cyanotic as he was when he was crying; but now he is not crying and is still cyanotic. Your management? Is oxygen likely to help? What else would you do? What effect would manipulating resistance in the pulmonary and systemic circulation have? How can you do this in the holding area?

4. How will you manage his intraoperative mechanical ventilation? What effect will management of $PaCO_2$ have on pulmonary blood flow?

## Postoperative Management

### Questions

1. What criteria will you use to extubate? Is this patient likely to have an increased dead space to tidal volume ($V_d/V_t$) ratio? Why? Is he likely to have impaired efficiency of oxygenation (significant shunt)?

2. One of your colleagues suggests that postoperative analgesia would best be provided by a local anesthetic infusion into an epidural catheter. Do you agree? Why/why not? Is this approach associated with any particular advantages or disadvantages in a patient with Glenn physiology in terms of ventilation? How will local anesthetic agents in the epidural space affect venous return? Particular concerns in this patient?

and BVF would result in a clinically relevant reduction in the quantity of antegrade pulmonary blood and total pulmonary blood flow. In the case of a bidirectional Glenn, dynamic pulmonary outflow obstruction would be largely irrelevant in terms of reduced pulmonary blood flow. An inhalation induction in this patient would be prolonged given the reduced Qp:Qs. The only disadvantage to this is that a longer period of airway management (along with the risk of aspiration, laryngospasm, etc.) as compared to an intravenous induction would be necessary. Ketamine administration is acceptable as long as it is not accompanied by hypercarbia and/or hypoxia.

3. Oxygen by mask followed by initiation of positive pressure ventilation with oxygen if necessary. There is no danger of overcirculation in this patient (i.e., PVR reduced to the point where Qp:Qs is so high that systemic perfusion is impaired) given that Glenn physiology is present.

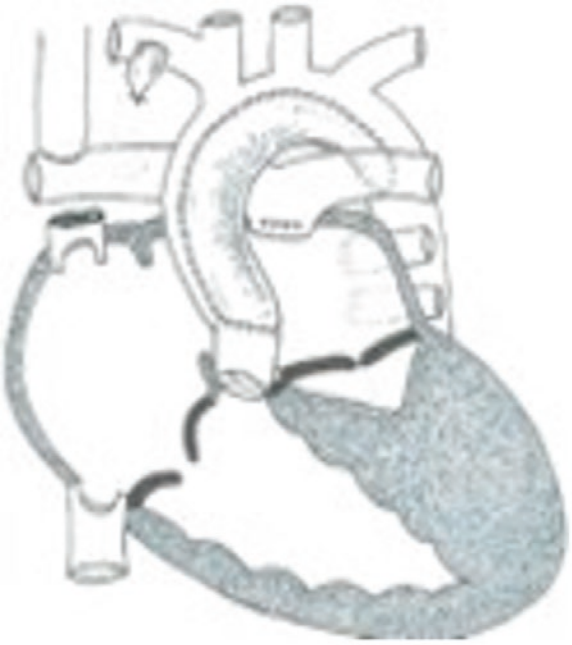
4. Mechanical ventilation should be managed to provide appropriate alveolar ventilation while minimizing mean airway pressure. This can be accomplished with tidal volumes of 8–10 mL/kg, respiratory rate of 10–15, I:E of 1:2 to 1:3, and judicious use of PEEP (3–5 cm $H_2O$) to improve pulmonary compliance. High mean airway pressure will mechanically obstruct pulmonary blood flow and will make ventilation less efficient by increasing the dead space to tidal volume ratio (V/V). Alveolar ventilation should be adjusted to obtain mild hypercarbia and a respiratory acidosis. In theory, mild hypocarbia can reduce PVR and increase pulmonary blood flow. However, the majority of SVC blood in this child comes from the brain, and mild hypercarbia ($PaCO_2$ 45–55 mm Hg) has been demonstrated to increase pulmonary blood flow by increasing cerebral blood flow.

## Answers

1. $V_d/V_t$ will be increased in a patient with a Glenn shunt due to an increase in Zone 1 ($P_{airway} > P_{alveolar} > P_{pulmoary\ venous}$) lung. This is the direct result of low pulmonary artery pressure (i.e., SVC pressure). In this patient, a physiologic right to left shunt is present due to IVC blood (systemic venous blood) that is directed to the aorta. Increasing $FiO_2$ will have no direct effect on the oxygen-carrying capacity of this portion of the cardiac output. Increasing $FiO_2$ will improve the oxygenation saturation of pulmonary venous blood if there is intrapulmonary V/Q mismatch.

2. Lumbar or thoracic epidural analgesia with a local anesthetic will have no direct effect on pulmonary blood flow in a patient with a Glenn shunt. However, good pain relief is more likely to lead to early extubation and resumption of spontaneous ventilation with appropriate alveolar ventilation. Spontaneous ventilation generates negative intrathoracic pressure and is likely to result in better pulmonary blood flow

than that seen with controlled, positive pressure ventilation. Pooling of venous blood (conversion of stressed volume to unstressed volume) due to an increase in venous capacitance can occur as a result of epidural sympatholysis. The subsequent reduction in mean venous pressure will reduce venous return to the heart. This effect can be counteracted by infusion of large volumes of intravenous fluids or by administration of drugs such as phenylephrine and dopamine that reduce venous capacitance. In this patient, the hemodynamic effects of epidurally administered local anesthetics will not be substantially different that that seen in a patient with a normal heart. In a patient with a dynamic source of obstruction to pulmonary blood flow and without an alternate source of pulmonary blood flow, an acute reduction in ventricular volume would lead to a drastic reduction in pulmonary blood flow.

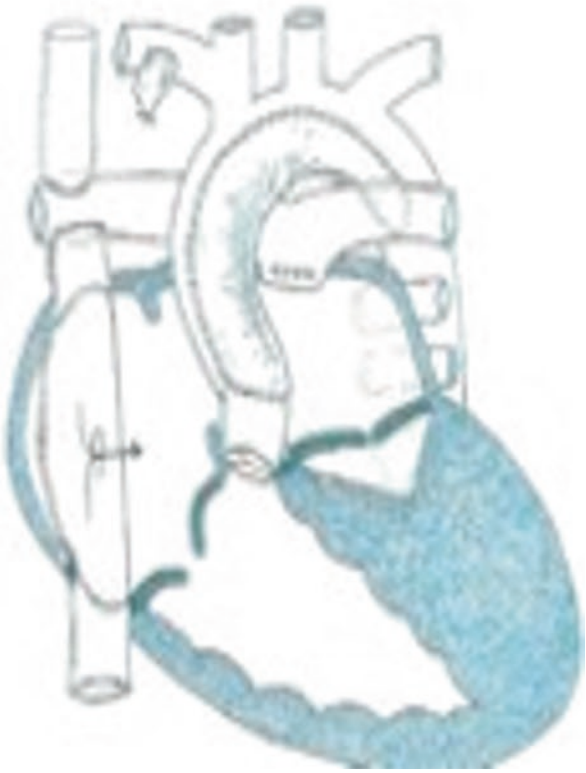


Bidirectional Glenn

## Additional Topics

### Questions

1. A 14-year-old male presents for posterior spinal fusion. He has a history of a univentricular heart and has been palliated with a modified Fontan procedure, s/p fenestration. Describe briefly the modified Fontan operation. What is the function of the fenestration? Does it require a separate procedure to close? What are the anesthetic goals in a patient of this sort undergoing a spinal fusion?



Fenestrated Fontan

## Answers

1. The Fontan procedure creates continuity between the IVC, the SVC, and the pulmonary artery. Following a Glenn shunt, Fontan physiology would be the result of surgically creating continuity between the IVC and the pulmonary artery. Following a Fontan, all systemic venous blood is delivered to the pulmonary circulation. A fenestrated Fontan involves placing a 4 mm hole in the baffle of the tunnel connecting the SVC and IVC to the pulmonary artery. When the transpulmonary gradient (TPG) is high, transpulmonary blood flow and delivery of blood to the systemic atrium are low, resulting in a low cardiac output syndrome. In this circumstance, the fenestration permits systemic cardiac output to be maintained at the expense of systemic oxygen saturation by allowing a small right to left shunt at the atrial level with delivery of venous blood to the systemic atrium. These fenestrations are closed in the cardiac catheterization laboratory following hemodynamic evaluation with temporary balloon occlusion.

2. A 3-year-old female presents for bilateral ureteral reimplantation. She has tetralogy of Fallot, a functioning right Blalock-Taussig shunt, and a resting oxygen saturation of 87% on room air and a hematocrit of 69%. What are your thoughts? What are the risks of erythropheresis? Is erythropheresis effective? How would you perform it?

3. A child with Wolff-Parkinson-White (WPW) syndrome is anesthetized for ablation of the reentrant tract. Prior to commencing the procedure, the child is anesthetized for ablation of the reentrant tract. Prior to commencing the procedure, the patient's heart rate increases to 285. ST depression is evident. The cardiologist says not to worry – if the patient stays in the rhythm, it will speed up the EPS mapping. What do you think? What would you do? What would you tell the cardiologist?

2. This child has outgrown her shunt and has erythrocytosis in response to chronic hypoxemia. Erythropheresis is effective in reversing the coagulation abnormalities associated with erythrocytosis and potentially in reducing the risk of cerebrovascular accident. It is generally performed using either isotonic crystalloid solution or FFP. Systemic air embolism is a risk of this procedure. Chronic erythropheresis will lead to development of a microcytic (iron deficiency) anemia. The symptoms of this type of anemia are similar to those of hyperviscosity syndrome from erythrocytosis.

3. The patient needs to be either chemically or electrically cardioverted. This is an unstable rhythm that will likely deteriorate to ventricular fibrillation (VF).

## Further Reading

1. Nasr VG, DiNardo JA. Pediatric cardiac anesthesia handbook. New Jersey: Wiley; 2017.
2. Kussman BD, DiNardo JA. The cardiovascular system. In: Holzman RS, Mancuso TJ, Polaner DM, editors. A practical approach to pediatric anesthesia. 2nd ed. Philadelphia: Lippincott Williams & Wilkins; 2015. p. 306–74.

# Chapter 23
# Cardiac III

**James A. DiNardo**

A 2-month-old with hypoplastic left heart syndrome (HLHS), s/p stage I palliation (Norwood with Sano shunt), presents with gastroesophageal reflux and failure to thrive. She is scheduled for placement of a gastrostomy tube and a Nissen fundoplication. Baseline $SpO_2$ is 75–80%, weight 3.8 kg, and Hct 42%.



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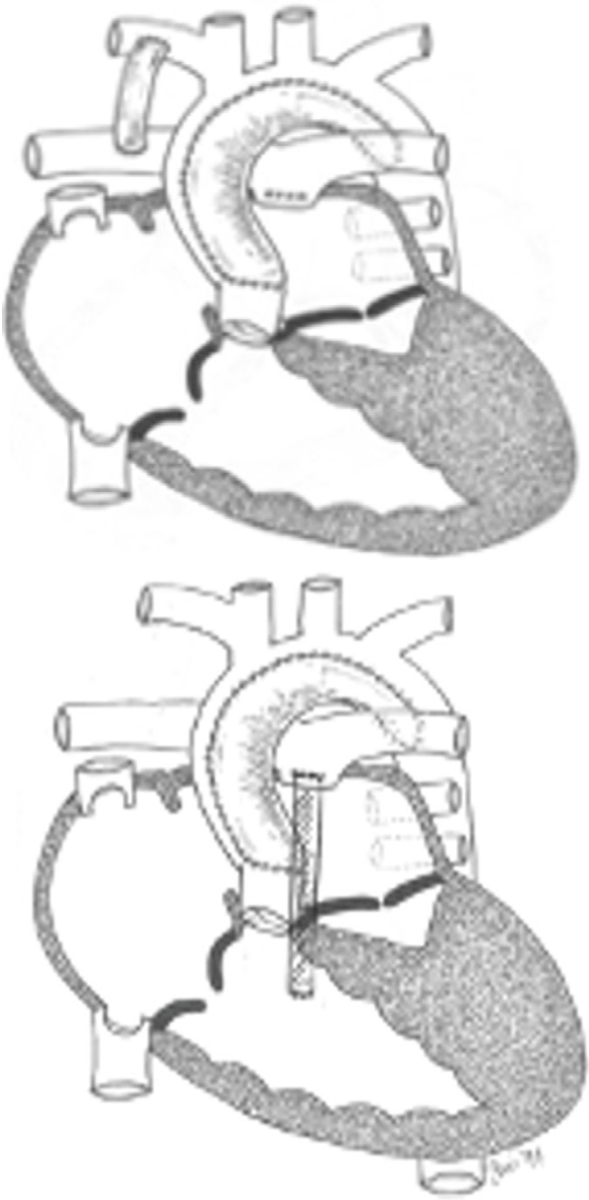
## Preoperative Evaluation

### Questions

1. What is a Norwood procedure?

**Answers**

1. Norwood with BT shunt and Norwood with RV-PA conduit (Sano)



2. What does the term “parallel circulation” mean?

3. What anatomic problems may arise after the Norwood procedure

## Intraoperative Course

### Questions

1. How would you induce anesthesia for this patient? Maintenance technique?

The Norwood procedure (stage I palliation) as it applies to HLHS involves resection of the intra-atrial septum to allow unrestricted delivery of pulmonary venous blood to the systemic ventricle (RV) via the right atrium, a Damus-Kaye-Stansel procedure wherein the proximal main pulmonary artery and pulmonary valve are used to provide continuity between the RV and proximal aorta, an aortic arch reconstruction, and coarctectomy; an aortic arch reconstruction and coarctectomy is performed using autologous pericardium or graft material and either a modified Blalock-Taussig shunt (mBTS) or RV to main pulmonary conduit (Sano shunt) is constructed to provide pulmonary blood flow.

2. Single ventricle physiology (SVP) is used to describe the situation wherein complete mixing of pulmonary venous and systemic venous blood occurs at the atrial or ventricular level and then this mixed blood is distributed to both the systemic and pulmonary beds in parallel rather than in series. As a result of this physiology, the ventricular output is the sum of pulmonary blood flow (Qp ) and systemic blood flow (Qs); distribution of systemic and pulmonary blood flow is dependent on the relative resistances to flow (both intra- and extra-cardiac) into the two parallel circuits; oxygen saturations are the same in the aorta and the pulmonary artery. This physiology can exist in patients with one well-developed ventricle and one hypoplastic ventricle as well as in patients with two well-formed ventricles where there is complete obstruction of outflow from one of the ventricles such as with truncus arteriosus or tetralogy of Fallot with pulmonary atresia.

3. Common problems following the Norwood procedure include RV dysfunction with tricuspid regurgitation, residual aortic arch obstruction, and inadequate pulmonary blood flow secondary to shunt stenosis or patient growth. Differentiating poor systemic cardiac output and heart failure symptoms due to RV dysfunction, TR, and arch obstruction from low pulmonary blood flow symptoms due to shunt obstruction or inadequate shunt size can be difficult. Transthoracic echocardiography is the diagnostic procedure of choice. Cardiac catheterization may be indicated to further delineate hemodynamic and to intervene on lesions such as residual coarctation or shunt stenosis.

## Answers

1. Induction techniques that do not depress ventricular function and that prevent precipitous increases in SVR and PVR should be employed. Ketamine (1–2 mg/kg) in combination with a low dose of fentanyl (2–5 μg/kg) or remifentanil (0.5–1 μg/kg) and rocuronium are reasonable if an intravenous catheter is in place. In the absence of an intravenous catheter, oral or intramuscular premedication with ketamine and midazolam or inhalation of sevoflurane not to exceed 4% while maintaining spontaneous ventilation can be considered to facilitate IV placement. Anesthesia can be maintained with additional opioids delivered as a bolus or infusion in conjunction with isoflurane or sevoflurane.

2. What monitoring would you use? Is an arterial catheter necessary?

3. The surgeon would like to perform this procedure via a laparoscopic approach. Agree or disagree? Wsshy?

4. How would you treat hypotension (systolic BP <60 mm Hg) and a reduction in $SpO_2$ to 70%? occurring 30 minutes after insufflation of the abdomen with $CO_2$?

Precipitous increases in SVR without an increase in single ventricle output result in diversion of output to the pulmonary circulation at the expense of the systemic circulation leading to reduced systemic oxygen delivery despite what initially appears to be "normal" BP and $SpO_2$. Failure to recognize or prevent this scenario will ultimately result in cardiovascular collapse. In patients with depressed ventricular function and/or tricuspid regurgitation, judicious use of inotropic support (dopamine 3–5 μg/kg/min) may be necessary to maintain systemic oxygen delivery.

2. There should be a low threshold to employ invasive blood pressure monitoring as meticulous attention to both systemic oxygen delivery (ABG analysis) and systemic perfusion pressure (beat to beat BP) is necessary.

3. Laparoscopic procedures are increasingly being utilized, and it is important to recognize the effects of abdominal insufflation of $CO_2$ on both hemodynamics and the reliability of end-tidal $CO_2$ as a surrogate measure of $PaCO_2$. There is currently no strong evidence to support one approach over the other in terms of safety and efficacy.

4. The first step is to rule out excessive abdominal insufflation pressures leading to compromise of venous return, elevation of systemic and pulmonary afterload, and compromise of alveolar ventilation. In the absence of hemodynamic or respiratory compromise from abdominal insufflation, a more careful analysis is necessary. In patients with SVP, hypotension associated with a fall in SpO2 should be assumed secondary to a fall in cardiac output (systemic oxygen delivery) due to the dependence of $SaO_2$ on $SvO_2$. A fall in SVR will cause a fall in BP, but a simultaneous fall in $SaO_2$ is unlikely unless there is a significant pressure gradient across the shunt. There are two ways to increase cardiac output:

   - Increase HR.
   - Increase stroke volume.
     - Augment preload and increase end-diastolic volume.
     - Increase contractility and decrease end-systolic volume.

   It should be kept in mind that preload reserve or the ability to augment stroke volume via the Frank-Starling mechanism is limited in SVP patients due to the volume-loaded nature of the single ventricle. Small doses of ephedrine (0.05–0.1 mg/kg) may provide sufficient increase in HR and contractility to improve cardiac output in this situation if use of a judicious fluid bolus (10–20 mL/kg) is insufficient. There is no role for administration of a pure alpha-adrenergic agent such as phenylephrine in this setting.

## Postoperative Management

### Questions

1. Should this patient be brought to the ICU intubated for a period of postoperative ventilation or should they be extubated in the OR?

2. Following extubation in the operating room, the baby is awake and alert but the $SpO_2$ is 65–70%. What are the major determinants of $SaO_2$ in this patient?

## Answers

1. Extubation in the OR is possible but a postoperative ICU bed should always be available in the event postoperative ventilatory or hemodynamic support is necessary. If there is any concern about the ability of the patient to maintain a normal level of consciousness and normal gas exchange, the patient should remain intubated until the sources of these deficiencies are resolved.

2. With single ventricle physiology, the arterial saturation (SaO2) will be determined by the relative volumes and saturations of pulmonary venous and systemic venous blood flows that have mixed and reach the aorta. This is summarized in the following equation:
   The determinants of SaO2 are:

   Aortic saturation = ([(systemic venous saturation)(total systemic venous blood flow) + (pulmonary venous saturation)(total pulmonary venous blood flow)]) / ([total systemic venous blood flow + total pulmonary venous blood flow])

   - The ratio of total pulmonary to total systemic blood flow ($Q_p$:$Q_s$). A greater proportion of the mixed blood will consist of saturated blood (pulmonary venous blood) than of desaturated blood (systemic venous blood) when $Q_p$:$Q_s$ is high.
   - Systemic venous saturation. For a given $Q_p$:$Q_s$ and pulmonary venous saturation, a decrease in systemic venous saturation will result in a decreased arterial saturation. Decreases in systemic venous saturation occur as the result of decreases in systemic oxygen delivery or increases in systemic oxygen consumption. Systemic oxygen delivery is the product of systemic blood flow and arterial oxygen content. Arterial oxygen content is dependent on the hemoglobin concentration and the arterial saturation.
   - Pulmonary venous saturation. In the absence of large intrapulmonary shunts and/or V/Q mismatch, pulmonary venous saturation should be close to 100% breathing room air. In the presence of pulmonary parenchymal disease, pulmonary venous saturation may be reduced. The V/Q mismatch component of pulmonary venous desaturation will be largely eliminated with a $FiO_2$ of 1.0, while the intrapulmonary shunt contribution will not be eliminated. For any given systemic venous saturation and $Q_p$:$Q_s$, a reduction in pulmonary venous saturation will result in a decreased arterial saturation.

## Additional Topics

### Questions

1. A 13-day-old child with transposition of the great arteries/intact ventricular septum arrives in the hospital on Friday afternoon and is booked for repair on Saturday morning. The child is receiving prostaglandin E1, is not intubated, and appears comfortable with a saturation of 89% on room air. When you question the cardiologist regarding the cost-effectiveness of operating on a weekend, how do you defend your position? What is the likely concern of the cardiologist? Discuss involution of the ventricular mass of the physiological right ventricle postnatally. What is this due to? How rapidly does it occur, i.e., when does it become physiologically significant?

2. A 5-year-old male, S/P truncus arteriosus repair as a newborn, regularly followed by his cardiologist, but full details not available because he recently moved to the area, presents to the ER with an acute abdomen. The general surgeon suspects acute appendicitis and wants to proceed with surgery as soon as possible. What is a truncus arteriosus? What problems may develop as a result of the repair? How can you assess these clinically? What preoperative investigations are indicated? How would you perform a rapid sequence induction? What monitoring would you use?

## Answers

1. The exact time frame of involution of LV mass is unknown but clinical experience suggests that the LV remains prepared to accommodate systemic afterload for 2–4 weeks following ductal closure in transposition of the great arteries with intact ventricular septum (TGA/IVS). The gradual postnatal decrease in pulmonary artery pressure following ductal closure and the subsequent reduction in LV afterload are responsible for this involution. The continued administration of PGE places the child at risk for apnea and an ongoing volume requirement with accumulation of interstitial edema. Independent of considerations regarding involution of ventricular mass, there are concerns that delaying surgery beyond 2 weeks of age is associated with impaired brain growth and slower language development. Impaired cerebral oxygen delivery and poor nutritional status have been implicated as potential causes associated with this finding in neonates undergoing delayed repair.

2. Truncus arteriosus

3. A 3-month-old with Shone's syndrome, S/P coarctation repair, has progressive inspiratory and expiratory stridor. He is in the emergency room with significant retraction and respiratory distress. He nevertheless has improved after a single dose of racemic epinephrine and now is monitored in the ICU. Saturation is 98% in humidified head box oxygen with soft biphasic stridor, but he is still tachypneic and is scheduled for direct laryngoscopy and bronchoscopy. What is Shone's syndrome? What is the etiology of the pulmonary hypertension? Describe your clinical evaluation. What might you find on the CXR? How would you induce this patient? Is an inhalational induction safe? Is it indicated for this procedure? What is the effect of airway obstruction on pulmonary artery pressure and cardiac output? How would you maintain anesthesia in this patient particularly as the ORL service wants to perform a functional examination to rule out tracheomalacia?

4. A 10-year-old with coarctation of the aorta is scheduled for repair by thoracotomy. Describe your clinical assessment of this patient. What preoperative investigations are important? How will they affect surgical management? Would you perform one lung anesthesia? What is the smallest double-lumen endotracheal tube available? What is the smallest patient you would consider a candidate for a double-lumen endotracheal tube? Are there any alternatives? What are the possible complications during coarctation repair? What methods are you aware of for spinal cord protection? Which are proven effective? How do you manage hypertension during aortic cross clamping? How will you control systemic hypertension during emergence form anesthesia?

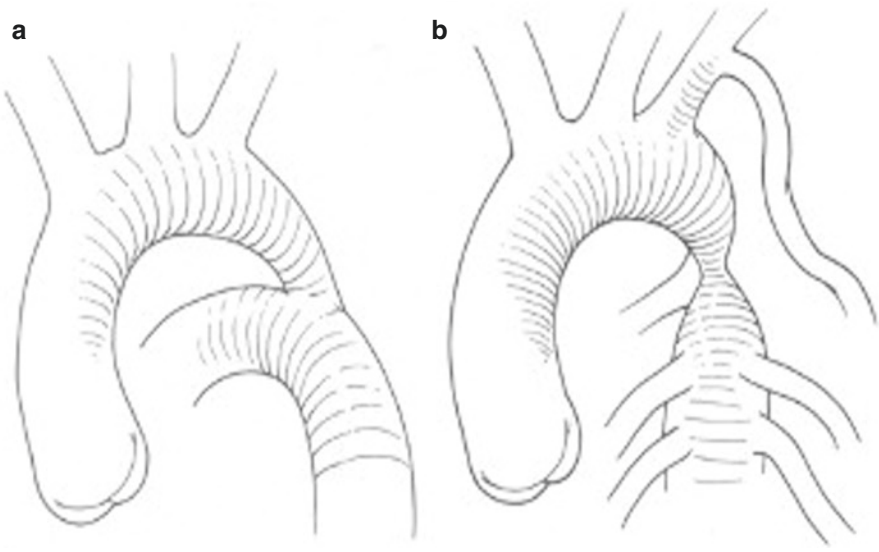
The truncal valve (common aortic and pulmonary valve) may have more than three leaflets and may be dysplastic, with both stenosis and regurgitation. Definitive repair for truncus arteriosus involves (1) VSD closure such that the truncal valve becomes the neo-aortic valve and (2) creation of RV to PA continuity with a valve conduit. The most likely sequelae following repair of truncus arteriosus are conduit stenosis/insufficiency and truncal valve (neo-aortic valve) insufficiency. Conduit dysfunction with significant RV dysfunction will be obvious on physical exam (enlarged liver, systemic venous hypertension, pleural effusions). Significant truncal valve dysfunction will have similar physical findings to aortic insufficiency or stenosis. A preoperative echocardiogram is warranted.

3. Shone's complex, strictly defined, is the combination of parachute mitral valve (mitral stenosis), subaortic stenosis, aortic stenosis, and coarctation of the aorta. Pulmonary hypertension is the direct result of left atrial (LA) hypertension that in turn is the consequence of obstruction to both LV inflow (mitral stenosis and LVH) and outflow.

   Clinical evaluation would include assessment for both left sided heart failure (pulmonary edema, pulmonary vascular congestion on CXR) and right sided heart failure (systemic venous congestion). Induction of anesthesia should be directed toward avoidance of tachycardia (increased MV gradient and increased LA pressure and increased PA pressure), hypotension (inadequate subendocardial perfusion of hypertrophied LV), and hypercarbia/hypoxemia from airway obstruction (increased PVR, RV dysfunction, and reduced preload delivery to LA). Following premedication, an inhalational induction could be performed utilizing sevoflurane not to exceed 4% inspired concentration and spontaneous ventilation. Small (0.5 mg/kg) intermittent doses of ketamine could be used to augment the depth of sevoflurane anesthesia while maintaining spontaneous ventilation during the procedure.

4. In patients presenting beyond infancy, upper extremity hypertension or a murmur is the most common presentation for coarctation of the aorta. The systolic murmur of a narrowed descending thoracic aorta may be best heard along the left paravertebral area between the spine and scapula. Continuous murmurs may be heard along the chest wall due to collateral vessels supplying tissues beyond the coarctation. These collaterals may originate from the internal thoracic, intercostal, subclavian, and/or scapular arteries. Other murmurs can be due to coexistent aortic valve stenosis and/or VSDs. In the setting of extensive collateral development, it is not uncommon for even a severe coarctation to be completely asymptomatic. Symptoms when present include exercise intolerance, headache, chest pain, nosebleeds, and lower extremity claudication. There is an increased incidence (as high as 10%) of cerebral artery aneurysms. Similar abnormalities may occur in the spinal arteries. Intracranial hemorrhage or subarachnoid bleeds are a risk for these patients. The smallest double-lumen tube (DLT) is a 26 French

with an external diameter similar to a 6.0 mm cuffed ETT. This DLT is appropriate for use in child 8–10 years old. Alternatives for this child would be a 5F Arndt bronchial blocker, a Fogarty catheter as a bronchial blocker, or a 3.5 or 4.5 Univent tube. There are no proven methods of spinal cord protection although mannitol administration and mild systemic hypothermia (34–35 °C) are popular.




Typical appearances of coarctation of the aorta. A. Coarctation associated with hypoplasia of the aortic isthmus and ductal-dependent perfusion of the descending aorta present at birth. Closure of the ductus arteriosus will likely result in LV afterload mismatch, compromise of somatic perfusion and oxygen delivery, and presentation in infancy. B. Coarctation of the aorta in a patient who has tolerated ductal closure and developed extensive collateralization. Extensive fibrosis in the juxtaductal region in association with growth of the native aorta has produced a discrete hourglass constriction. Presentation will be as a child or young adult.

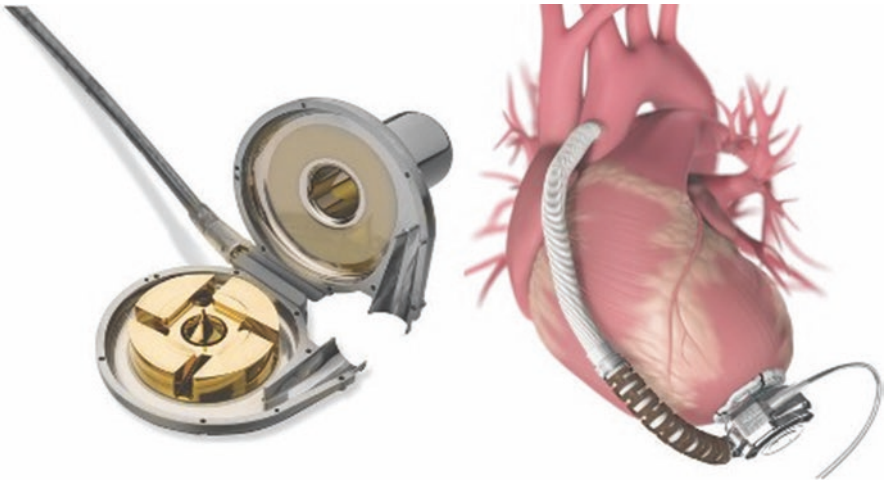
## Further Reading

1. Nasr VG, DiNardo JA. Pediatric cardiac anesthesia handbook. New Jersey: Wiley; 2017.
2. Kussman BD, DiNardo JA. The cardiovascular system. In: Holzman RS, Mancuso TJ, Polaner DM, editors. A practical approach to pediatric anesthesia. Philadelphia: Lippincott Williams & Wilkins; 2015. p. 306–74.

# Chapter 24
# Cardiac IV: Mechanical Support

**James A. DiNardo**

A 13-year-old 30 kg female with a dilated cardiomyopathy (DCM) and progressive heart failure is being considered for orthotopic heart transplantation (OHT). Due to a precipitous decline in the patient's cardiovascular status culminating in intensive care unit admission and initiation of maximal medical therapy, the decision has been made to semi-electively provide mechanical circulatory support (MCS). The chosen device is a HeartWare centrifugal pump as a left ventricular assist device (LVAD).



HeartWare centrifugal LVAD. Inflow to the device is via the LV apex with outflow directed to the ascending aorta.



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## Preoperative Evaluation

### Questions

1. What are the indications for MCS in children with heart failure? What would be the ultimate goal of device implantation in this patient?

2. How are MCS devices classified?

## Answers

1. The indications for initiation of MCS in patients with heart failure vary by institution, but generally the following parameters are considered:

   - A progressive increase in inotropic support such as epinephrine >0.3 μg/kg/min or the requirement of a second inotrope
   - Cardiac index <2.0 L/min/$m^2$
   - Mixed venous oxygen saturation <40%
   - Lactic acidosis
   - Poor end-organ perfusion evidenced by oliguria (<1 mL/kg/h), rising serum creatinine, rising liver enzymes, inability to tolerate enteral feeding, immobility or extreme fatigue, or altered mental status
   - Invasive or noninvasive mechanical ventilatory support

   The goal in this patient would be to use a LVAD to serve as a bridge to OHT. In the current era, approximately 20–30% of pediatric patients awaiting heart transplant are bridged with VAD resulting in a 50% reduction in pediatric waitlist mortality. Other indications for MCS are as a bridge to myocardial recovery, destination therapy (DT), and bridge to decision. When used for bridge to recovery, the expectation is that sufficient myocardial recovery will occur following a treatable disease exacerbation or following a postcardiotomy insult such that the device can ultimately be explanted. A bridge to DT is more commonly employed in adults with other comorbidities that contraindicate OHT. In children, DT may be considered for patients such as those with advanced dystrophinopathies, where posttransplant outcomes remain poor. Bridge to decision involves use of a short-term support device such as ECMO to determine if sufficient end-organ recovery will occur to place a long-term device for bridge to recovery, OHT, DT. In the absence of end-organ recovery in this setting, redirection of care is considered.

2. There are numerous classification schemes for MCS; the following is a practical scheme:

   - Short-term or intermediate to long-term. Extracorporeal membrane oxygenation (ECMO) is the most commonly employed MCS device in children and is short-term support device. A child must generally remain intubated and sedated throughout the period of ECMO support. ECMO support can only be maintained for 1–3 weeks before complications which severely compromise end-organ function and recovery occurs. VADs are used for an intermediate- to long-term support. These devices allow tracheal extubation such that

3. How would you comprehensively evaluate this patient's preoperative physiologic status?

4. What other information should be solicited in this particular patient during the preoperative assessment?

patients can ambulate, take enteral nutrition, and assume activities of daily living. Unlike ECMO, these devices do not require anticoagulation with unfractionated heparin so that less aggressive anticoagulation with enteral agents (aspirin, dipyridamole, clopidogrel, and coumadin) and/or subcutaneous low-molecular-weight heparin can be undertaken.

- Paracorporeal or intracorporeal VAD. Paracorporeal devices have inflow and outflow cannula within the thoracic cavity; the cannula exits the thoracic cavity to attach to a device which is located outside the body. The Berlin Heart Excor is such a device. Intracorporeal devices are implanted entirely within the body with the only externalized component being the drive line which is connected to an external control device. The HeartWare centrifugal pump is such a device.
- Pulsatile or nonpulsatile VAD. The Berlin Heart Excor is a pulsatile VAD. The prosthetic ventricle is a rigid case divided into a blood and a pneumatic chamber by a flexible, noncompliant diaphragm. Unidirectional inflow and outflow valves are located in the blood chamber. The pneumatic chamber communicates with the pneumatic drive system via the pneumatic driveline. Vacuum can be applied to the pneumatic chamber during diastole. Nonpulsatile VADs use either an axial or a centrifugal pump to create continuous nonpulsatile flow. They have the advantage of being compact and valveless. The HeartWare HVAD is a nonpulsatile centrifugal pump. It has been used successfully in children with a BSA as low as 0.6 $m^2$ and a weight of 13–15 kg.

3. Children requiring placement of a MCA device are likely to have overt or occult end-organ dysfunction in addition to cardiac dysfunction. In particular, renal and hepatic dysfunction should be evaluated. Calculation of glomerular filtration rate (GFR) using a standard formula (Schwartz or MDRD) should be done rather than relying on blood urea nitrogen (BUN) and creatinine (Cre) values. Children with a GFR <60 ml/min/1.72 $m^2$ should be assumed to have occult renal dysfunction despite near-normal BUN and Cre values and are at risk for acute renal injury or failure. A full panel of liver function tests and a full coagulation profile should be performed to rule out hepatic failure as distinct from passive congestion.

4. Particular attention during the physical exam should be paid to assessment of arterial and venous access sites. A review of recent echocardiographic and cardiac catheterization data will give valuable information regarding vascular access. Pacemaker and/or implantable cardioverter defibrillators should be reprogrammed prior to surgery. Relying on magnet placement is not acceptable.

## Intraoperative Course

### Questions

1. What monitoring is required for this procedure? If a patent foramen ovale (PFO) has not been ruled out preoperatively, why must it be ruled out in the operating room?

2. What are your primary considerations during induction of anesthesia in this patient? What is unique about DCM patients as regards catecholamine support?

3. What factors determine whether a LVAD will function optimally? What effect will high systemic vascular resistance (SVR) have on the output of the centrifugal HeartWare pump? How would you expect an arterial waveform to appear following LVAD implantation?

## Answers

1. In addition to standard American Society of Anesthesiologists monitoring, all patients require arterial and central venous pressure (CVP) monitoring catheters. Unless contraindicated, a transesophageal echocardiography (TEE) probe is placed after induction of anesthesia.

   The existence of a PFO or any site for intracardiac shunting must be excluded by TEE before placement of any MCA that is capable of reducing LA pressure below RA pressure. Decompression of the LV and reduction in LA pressure below RA pressure will produce right-to-left intracardiac shunting and hypoxemia. This is particularly likely in the setting in which RA pressure is elevated, such as when there is concomitant RV dysfunction or elevated PA pressure.

2. It is important to recognize that DCM patients have limited contractile reserve, limited preload recruitable stroke work, and limited ability to respond to acute increases or decreases in afterload. In addition, the circulation time will be slow, given the low cardiac output state. These patients require an induction that will blunt the hemodynamic response to laryngoscopy and tracheal intubation without undue myocardial depression, vasodilation, and subsequent hypotension. In DCM patients, elevated catecholamine levels, downregulated beta-receptors, and reduced myocardial norepinephrine stores are present. Consequently, the myocardial depressant effects of induction agents will predominate. Preoperative inotropes should be continued.

3. LVAD generally functions well as long as there is sufficient intravascular volume and RV function to fill the pump. Proper functioning of the LVAD requires an RV that functions well enough to match the combined output of the LV and the LVAD. An RV that is capable of delivering only 3 L/min to the pulmonary circulation and LA will severely compromise optimal functioning of a LVAD that can deliver 6 L/min to the systemic circulation.

   Centrifugal pump output is determined by the pump revolutions/minute (RPM) and the pressure gradient (dP) between the inflow cannula (LV) and the outflow cannula (aorta). Pump flow is inversely related to dP, such that a high dP as is seen with a high SVR and a low LV pressure will reduce pump flow (systemic cardiac output) for a given pump RPM.

   The arterial waveform will be largely nonpulsatile much like an extremely dampened normal arterial waveform. This is because flow from the LVAD is primarily continuous and nonpulsatile. At a given RPM, during ventricular systole when LV pressure increases and dP falls, LVAD flow will increase, and there will be some pulsatility to the arterial wave even in the absence of aortic valve opening.

4. How does LVAD implantation affect RV function?

## Postoperative Management

### Questions

1. In the immediate postoperative period, the patient manifests low LVAD output in conjunction with an elevated CVP. What is the likely cause of these findings and how will you further delineate the etiology?

2. An echocardiographic examination demonstrates tricuspid regurgitation, RV distention, and RV free wall hypokinesis. How would you intervene?

3. Is use of milrinone valuable in this setting?

4. LVAD insertion results in global impairment of RV systolic function due to loss of the LV contribution to RV function from leftward septal shift. In addition, LVAD implantation generally acutely decreases mitral regurgitation while worsening tricuspid regurgitation (TR) both acutely and chronically. Poor apposition of the tricuspid valve leaflets after LV decompression with a LVAD occurs as the result of a leftward shift of the intraventricular septum and the increased RV volume load. There is generally dilation of the tricuspid annulus and leftward deviation of the septal components of the tricuspid subvalvular (chordae and papillary muscles) apparatus. Normally, this is offset by increased RV preload and reduced RV afterload (due to a reduction in LA pressure and PVR) such that RV output is maintained or increased. However, some patients have a component of PVR that is not immediately reduced by decompression of the LV in addition to underlying RV contractile dysfunction, particularly of the intraventricular septum.

**Answers**

1. These findings are consistent with RV dysfunction and failure to deliver adequate preload to the LVAD to maintain output. An echocardiographic examination should be performed.

2. Treatment of RV dysfunction following LVAD implantation requires treatment of reduced RV contractility with inotropes, optimization of RV preload, reduction of PVR with inhaled NO or prostacyclin, and attention to the extent of septal shift induced by LV decompression.

3. Milrinone is commonly mentioned as an intervention in this setting because it is an inodilator with the ability to reduce PVR. Enthusiasm for this drug should be tempered with the fact that it is a more potent vasodilator than an inotrope and while this property is useful in reducing PVR, it also reduces SVR. This can be detrimental in the setting of systemic hypotension because it reduces RV coronary perfusion and potentially exacerbates RV ischemia. It may be necessary to administer a vasoconstrictive agent such as norepinephrine or arginine vasopressin simultaneously if milrinone is administered.

## Additional Topics

### Questions

1. What is the definition of pulmonary hypertension (PH) in children? How is this different than the definition of pulmonary arterial hypertension (PAH)?

2. What is bivalirudin and how is it utilized in children?

3. What is Williams syndrome? Why are these patients at risk for cardiovascular adverse events in the perioperative period?

**Answers**

1. PH is defined with a specific threshold of PAP. Currently guidelines define PH as a mPAP >20 mm Hg in children >3 months of age. This definition holds regardless of the underlying cause of the elevated PAP. PAH, or precapillary PH, is a term used to define a specific cause of PH using strict hemodynamic criteria as follows:

   - mPAP >20 mm Hg
   - PAWP or LVEDP ≤15 mm Hg
   - PVR index ≥3 WU · $m^2$

2. Bivalirudin is a bivalent direct thrombin inhibitor (DTI) that binds to both the active and fibrin-binding sites of thrombin. As opposed to unfractionated heparin (UFH) that has a limited ability to inhibit clot-bound thrombin, bivalirudin is able to bind to both the circulating and clot-bound forms of thrombin with similar affinity. Bivalirudin is metabolized via proteolytic cleavage (80%) as well as renal mechanisms (20%). The estimated average half-life of bivalirudin in the pediatric population is 15–18 minutes and appears to be age dependent with increased weight-based clearance as compared to adults. Bivalirudin's safety and efficacy as a procedural anticoagulant for both catheter-based and surgical interventions in the adult population is well supported. Although bivalirudin utilization has increased in the pediatric population primarily as an alternative to UFH in cases of suspected or confirmed heparin-induced thrombocytopenia and thrombosis (HITT), there are at present no FDA-approved indications or established dosing guidelines in this age group.

3. Williams syndrome, also known as Williams-Beuren syndrome, is a rare congenital disorder involving the cardiovascular and central nervous systems. The phenotypic features of Williams syndrome include typical facial features, an outgoing "cocktail personality," and hypercalcemia. Williams syndrome is caused by a de novo deletion on chromosome 7q11.23, which encodes for the protein elastin. These patients are at risk for cardiovascular adverse events to the coexistence of ventricular hypertrophy and compromised coronary blood flow. These patients may have both systemic (supravalvar aortic stenosis) and pulmonary (pulmonary artery stenosis) arterial disease leading to the development of concentric ventricular hypertrophy. The affected aorta has a typical hourglass narrowing at the sinotubular junction. Coronary artery abnormalities (stenosis or dilation) have been identified in up to 45% of patients. Mechanical impairment of coronary blood flow is a frequent and often unappreciated feature of Williams syndrome. Adhesion of the right or left aortic leaflet edge to the narrowed sinotubular junction can restrict coronary blood flow into the sinus of Valsalva. Pulmonary arterial stenoses are present in approximately 40% of patients and occur mostly in the branch and peripheral pulmonary arteries.

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# Chapter 25
# Genitourinary Disorders

**Thomas J. Mancuso and Joseph P. Cravero**

A baby in the first day of life presents for closure of an exstrophy of the bladder. He is the product of 36-week gestation; he is 2.3 kg, with a preoperative hematocrit of 56%, BP 65/35 mmHg, HR 130 bpm, and RR 24/min RA SpO2 = 96%.



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## Preoperative Evaluation

### Questions

1. Is this baby preterm? How can you differentiate premature from small for gestational age (SGA) babies? What difference would it make in your anesthetic technique? What problems would you expect related to prematurity? Should a regional anesthetic be utilized? Narcotics? Why/why not? Would you hope to extubate this baby at the end of surgery?

## Answers

1. This baby is preterm because the infant was born before 37 weeks' gestation. Infants born between 36 and 37 weeks' gestational age are categorized as borderline preterm. Those born between 31 and 36 weeks' GA are considered moderately preterm and those born between 24 and 30 weeks' gestation are considered severely preterm. Small gestational age (SGA) babies weigh less than the tenth percentile for their gestational age. Babies born at 40 weeks who weigh less than 2.5 kg are SGA. The more premature the infants are, the greater the risk for perioperative complications. Preterm babies are born with structurally and physiologically underdeveloped vital organs. They are unable to maintain body temperature due to immature thermal regulation. Hyperthermia metabolic rate is linearly between 36 and 28 °C, therefore increasing oxygen consumption, which can lead to hypoxemia, acidosis, apnea, and respiratory depression. Preterm infants tend to lose body heat at a faster rate than term or older infants because of a higher body surface/volume ratio and lack of brown fat. Heat stress is equally detrimental because preterm infants are unable to sweat (dissipate heat by evaporative heat loss), and body heating causes dilation of peripheral vessels. During anesthesia the infant's body and head should be covered with plastic or cotton wrap to decrease heat and water loss [1, 2].

   Infants are unable to sustain ventilation due to poorly developed ventilatory centers in the brainstem and inefficient respiratory mechanics. Preterm infants are also at risk for respiratory distress syndrome due to impaired amounts or lack of surfactant. They may also develop intraventricular hemorrhage from rapid changes in blood pressure or cerebral ischemia from hypoperfusion due to impaired cerebral autoregulation. In addition, this population is at risk for left to right shunting via the ductus arteriosus soon after birth (within 3–5 days after birth). Premature infants born before 34 weeks' gestational age have a decreased glomerular filtration rate (GFR). Even term neonates have only 40% of an adult's GFR at birth. In addition, there is decreased tubular reabsorption capacity and a relative inability to absorb water, salts, glucose, protein, phosphate, and bicarbonate. Hyperglycemia and glycosuria can act as an osmotic diuretic and cause obligatory sodium as well as free water loss. Hepatic catalyzing enzymes are less active in premature infants. Oxidizing, reducing, and hydrolyzing enzymes are relatively inactive. Conjugation enzymes (conjugation with acetate, glycine, sulfate, and glucuronic acid) are also less active except for sulfonation. Therefore, the metabolism of various drugs, particularly opioids, can be impaired. These enzymes mature between 6 and 12 months of age, to adult capacity. A regional anesthetic should be used whenever feasible. Opiates could be used with caution, in reduced doses, and the infant's respiratory status should be closely monitored. I would hope to extubate if the vital signs (HR, RR, BP, temperature) are acceptable, successful epidural analgesia is provided, and minimal opiates are

2. Is it common for bladder exstrophy to occur in males? What do we call bladder exstrophy in a female? Why does this happen? Are there any future problems the patient can expect? Is early closure better than later closure? Why/why not? Is it likely that there is more surgery in the future for this baby? What type? Why?

## Intraoperative Course

### Questions

1. Does this baby need an arterial line? Why/why not? Should a central line be placed? Where would you place the IVs? Why? Can you only get an IV in the foot? What's next? If the case will take 8 hours, do you need to obtain surveillance blood gases? Why/why not? Would you treat if the pH were 7.34? 7.22? 7.14? Why?

administered intraoperatively. Prior to placement of a caudal or epidural, radiological images of the spine (which almost certainly would already have been part of this child's workup) should be evaluated to ensure the anatomy is normal.

2. Bladder exstrophy is more common in males. The male/female ratio is 2: 1. In a female, bladder exstrophy is known as a cloaca. At 5–6 weeks of gestation, the cloacal membrane prevents the normal migration of mesoderm (originator of anterior abdominal muscles and pelvic bones) of the infraumbilical area, resulting in failure of fusion of the rectus muscles and the pubic symphysis; the urethra fails to close dorsally (epispadias), and the anterior wall of the bladder wall may remain open. The urinary tract is everted exteriorly. Future problems include incontinence and sexual dysfunction. Early closure (within 24–48 hours) of the bladder and abdomen may allow an optimal anatomical and functional outcome. This child will likely require many further reconstructive surgeries to correct epispadias at age 2–3 years and urinary continence (the bladder neck) by age 4–5 years. Other possible procedures include bladder augmentation if the bladder is of small capacity, ureteral reimplantation for ureteral reflux, and creation of a continent urinary (e.g., Mitrofanoff) stoma [3].

**Answers**

1. An arterial line would be very helpful as it would enable surveillance blood gases and monitoring of accurate and continuous blood pressure (which are critical in cases with significant blood loss and large fluid shifts at this age). A central line is not necessary unless a prolonged postoperative course is expected along with difficulty in intravenous access. In that case, it may be wise to place a percutaneously inserted central catheter (PICC) line as part of the procedure. IVs should be placed in the upper extremities if possible because the lower extremities are usually prepared and draped within the surgical field and because of potential for loss of and lower extremity infusate from iliac veins within the surgical field. Surveillance blood gases are a good idea for a patient this age undergoing prolonged surgery to monitor accurate and continuous blood pressure, blood loss, and large fluid shifts. I would be cautious with a pH of around 7.25 and treat a pH less than 7.22 because it is associated with impaired cardiovascular performance.

2. Can you do this case with an epidural or caudal catheter and sedation? Would you choose to do so? Why/why not? A general anesthetic with an endotracheal tube is chosen. Would you place an oral or a nasal tube? Why? Should narcotics be avoided? Which would you choose? Why/why not? What about muscle relaxants? Surgeon wants you to avoid them. (Why do you think?) Is nitrous oxide contraindicated? Relatively contraindicated? What problems might you expect? When would you expect them?

3. The surgeon has turned the baby prone and is in the process of doing bilateral iliac osteotomies. The saturation drops to 94% and the end-tidal C02 has disappeared from the screen. What do you think is going on? The blood pressure cuff is not reading but recycling. The electrocardiogram (ECG) heart rate is 110? Then it becomes 80? What is Durant's maneuver? Should the baby be placed in the left or right lateral decubitus position?

## Postoperative Course

### Questions

1. Should this patient be extubated? What criteria would you use? The patient is vigorous, and you extubate, but saturation is 92% on supplemental oxygen $_2$ by face shield? Your next move? Face mask fails to improve saturation; would you be happy with 93%? What if it was 91%? Should patient be reintubated? For a respiratory rate of 24/min? 34/min? 54/min? What could be going on to account for the findings? How would you manage V/Q mismatch at this time?

2. This case cannot be done with a regional technique alone. This is a prolonged procedure that would require a dense block sustained for a long period of time. The use of an adequate surgical concentration of local anesthetics in premature infants could result in systemic toxicity due to impaired hepatic elimination of amide and ester local anesthetics, resulting in prolonged elimination half-lives. Moreover, sedation would be required in addition to regional anesthesia and could result in hypoventilation, periodic breathing, and apnea due to immaturity of the respiratory centers. The amount of sedation required, even with an optimal block, would result in alveolar collapse, shunting, and respiratory embarrassment with a small infant positioned very far away from the anesthesiologist. A nasal or oral endotracheal tube would be acceptable. I would use a nasal tube because it would be more secure intraoperatively and comfortable for the infant if he requires postoperative ventilatory support. Opiates should not be avoided. I would choose fentanyl, because the mixed enzyme oxidase enzymes are adequately mature to metabolize fentanyl effectively. The mixed oxidase enzymes that metabolize fentanyl are more active in early infancy. Muscle relaxants should be avoided if the surgeon plans to stimulate and assess sphincter function or identify major nerves. While nitrous oxide is not absolutely contraindicated, it is relatively contraindicated due to the fact that it can accumulate in the bowel causing distension or aggravate air embolism, should it occur.

3. Possible air embolism, occlusion or dislodgment of the endotracheal tube, or severe hypotension; most likely this is an air embolism. Durant's maneuver involves positioning the patient in steep head down and left lateral decubitus position. Left lateral decubitus allows the buoyant foam (blood/air mixture) to remain in the right ventricle and prevent it from occluding the pulmonary arteries.

## Answers

1. It is perfectly reasonable to plan for extubation, assuming some specific criteria are met: if the infant is awake, exhibits adequate strength (flexing at the hips for 5 s, tight fists, strong bite, furrowing his eyebrows as a sign of attention), is breathing regularly, and has an empty stomach and the muscle relaxant (if used during surgery) is reversed, with a return of train of four and no fade at 50 Hz (for 5 s tetanus). If oxygenation were poor after extubation, I would provide 100% oxygen with a face mask. I would not be happy with a saturation of 93% or 91% and would reintubate if this persisted. Hypoxemia and hypercarbia are likely due to ventilation/perfusion mismatch which should be improved with positive end-expiratory pressure (PEEP) and appropriate peak ventilator pressures. I would plan controlled ventilation after reintubation of the trachea.

2. How would you manage a continuous morphine infusion for pain relief? What about an epidural? How would you constitute the epidural solution? With fentanyl or Dilaudid? Why? A colleague stops by as you are putting in the epidural needle and says that he always gets a spine film in babies with bladder exstrophy because he is worried about *spinal anomalies that would complicate placement and impair appropriate spread of the local anesthetic*. Is he right? What difference would it make in your anesthetic management? Could you place a continuous caudal catheter instead? Or would you just stay away from the back?

## Additional Questions

### Questions

1. What is the difference between a neuroblastoma and nephroblastoma (Wilms' tumor)? Any other syndromes associated with neuroblastoma? Can both be endocrinologically active? What is the difference between a ganglioneuroma and a neuroblastoma? What makes a neonatal Wilms' tumor (congenital mesoblastic nephroma) different from a regular Wilms' tumor?

2. What is the natural history of "infantile" polycystic kidney disease? How does it affect anesthetic management? What are the nonrenal considerations for perinatal form?

2. I would start the morphine infusion at 15 mcg/kg/h after intubation or 10 mcg/kg in a non-intubated spontaneously breathing infant. Placement of a caudal to thoracic or lumbar catheter and infusion of local anesthetics is a safe alternative to IV opioids. Appropriate placement could be confirmed by ultrasound, epidurogram, or electrical stimulation. A solution of chloroprocaine 1.5% with or without fentanyl (1–2 mcg/cc) is safe in this age group and provides appropriate analgesia. If the trachea is extubated, it may be prudent to avoid neuraxial opioids to avoid the possibility of opioid-induced periodic breathing and/or apnea. My colleague is probably worried about possible associated vertebral anomalies and is correct about his concern. Identification of normal anatomy is a prerequisite for insertion of neuraxial needles and catheters. A continuous caudal catheter could be placed if there is no associated sacrococcygeal agenesis or anomaly. Placement of an epidural indwelling catheter is possible if the spine X-ray reveals normal anatomy at the site of the insertion, and ultrasound or MRI of the spine reveals no cord tethering [4].

**Answers**

1. Neuroblastoma is a neural crest malignancy that arises from primitive blast cells of the postganglionic sympathetic chain and adrenal glands. Nephroblastoma (Wilms' tumor) is a malignancy that arises from abnormal metanephric differentiation of the renal blastema (undifferentiated renal cells). Neuroblastoma can be associated with pheochromocytoma and neurofibromatosis type 1 (NF- I, von Recklinghausen disease). Neuroblastoma and nephroblastoma can both be endocrinologically active; 75% of neuroblastomas may secrete catecholamines. Ganglioneuroma is a benign tumor arising from well-differentiated and mature sympathetic ganglia. Neonatal Wilms' tumor is a benign nephroma arising from metanephric blastema or secondary mesenchyme, whereas regular Wilms' tumor (nephroblastoma) is a malignant tumor of the undifferentiated metanephric blastema.

2. Infantile polycystic kidney disease varies in severity. When oligohydramnios presents early in pregnancy, the outlook is extremely poor due to fetal pulmonary hypoplasia in addition to renal insufficiency. The condition sometimes presents later in infancy with reduced renal function. It may not become symptomatic until adolescence, when it represents a milder expression of the disease. The extent of pulmonary hypoplasia determines the difficulty of ventilation. The degree of renal impairment determines the clearance of anesthetic agents. The obstetrical team may consider cesarean section, particularly in bilateral polycystic kidney disease, because of the large body size and the risk of renal rupture during vaginal delivery. The newborn may require control of ventilation and treatment of high blood pressure.

3. You wish to perform a peripheral nerve block for hypospadias repair. Which nerves do you wish to block and what anatomical structures will your needle pass through on the way to those nerves?

3. Penile tissues are innervated by the dorsal penile nerve (S 2, 3, 4) and the perineal cutaneous nerve (branch of the pudendal nerve) at the root of the penis. The dorsal penile nerve is best reached at the base of penile shaft from the dorsal surface. The needle has to traverse the skin, subcutaneous tissue, and Buck's fascia. The fascial planes can be visualized with ultrasound, which has been shown to improve the success of this block [5]. A subcutaneous ring block of the penis will include a portion of the perineal nerve, but this will depend on the proximal vs. distal location of the hypospadias.

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# Chapter 26
# Transplantation


**Thomas J. Mancuso**

A 14-year-old female with a diagnosis of Budd-Chiari syndrome presents for liver transplantation. She has developed decreasing mental status, hyponatremia, and a reduction in urine output.



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## Preoperative Evaluation

### Questions

1. How would liver failure affect pulmonary function? What evaluation of pulmonary function would be helpful for this case? Would bronchodilator therapy be indicated preoperatively?

2. What cardiovascular abnormalities would you expect in this patient?

3. Is this patient exhibiting hepatic encephalopathy? Does she have raised ICP? What treatments should be given for each of these problems?

## Answers

1. Children with liver disease severe enough to be candidates for transplantation nearly always have abnormal pulmonary function [1–3]. Restrictive lung disease is caused by ascites in these children. In addition to the peritoneal space, there may be abnormal transudation of fluid in the pleural space. Pleural effusions will also compromise pulmonary function. Abdominal distention also decreases the functional residual capacity (FRC). These children are often malnourished, and the muscles of respiration, the diaphragm, and intercostal muscles are weakened, leading to a further decrease in the FRC. In addition to restrictive pulmonary pathophysiology, these children have other reasons for hypoxemia. The hepatopulmonary syndrome of hypoxemia and intrapulmonary shunts in these patients contribute to pulmonary morbidity. Intrapulmonary shunting of blood and impaired hypoxic pulmonary vasoconstriction lead to lower hemoglobin saturation. Pulmonary hypertension with increased pulmonary vascular resistance (PVR) can affect right ventricular performance. A small subset of patients with severe liver disease will manifest pulmonary hypertension.

2. Children with end-stage liver disease (ESLD) presenting for transplantation have significant derangements of cardiovascular function [4, 5]. These children have an increased cardiac output, increased ejection fraction, and lowered systemic vascular resistance (SVR). Peripheral vasodilatation and arteriovenous shunts account for the lower SVR. The circulating plasma volume is increased. The etiology of the hyperdynamic state of the circulatory system is unclear. Although children with liver failure generally have preserved cardiac function, those with severely advanced disease can exhibit impaired left ventricular performance. SvO2 is often elevated, probably due to the A–V shunts and to decreased oxygen delivery to the tissues. The RBCs in these patients are depleted of 2, 3-DPG and deliver less oxygen to the periphery. These children nearly always have a low albumin as part of ESLD. The Child-Pugh classification system includes serum albumin (along with bilirubin, prothrombin time (PT), and degree of encephalopathy) as one of the factors in determining the severity of liver insufficiency.

3. Patients with severe liver disease often have CNS changes. The cause for hepatic encephalopathy is not known, but the severity of the CNS dysfunction does parallel the severity of the liver disease. Possible causes for hepatic encephalopathy are the elevated levels of ammonia and other products of metabolism that accumulate as the liver fails, or the appearance of false neurotransmitters derived from amino acids that had not undergone degradation. The encephalopathy usually improves when appropriate therapy for the liver failure is started. Acute worsening of hepatic encephalopathy is usually an indication that the underlying liver disease has also worsened. In situations where more is demanded of the

4. What is the relationship between ESLD and impaired renal function? How can the presence of the hepatorenal syndrome be diagnosed in this patient?

5. Is this patient at risk for gastrointestinal hemorrhage? Would the presence of a portosystemic shunt affect the pre-op evaluation?

6. What disorders of the hematologic system would be expected in their patient?

liver such as GI hemorrhage or increased protein intake, hepatic encephalopathy will worsen. Infections or dehydration will also worsen hepatic encephalopathy. Treatment of hepatic encephalopathy includes restriction of protein intake, enteral lactulose, and neomycin and maintenance of as normal a metabolic state as possible [6–9]. If the patient is in fulminant hepatic failure, raised ICP is likely to be present. The exact etiology of the cerebral edema is not known, but vasogenic and cytotoxic mechanisms are thought to contribute. As the cerebral edema worsens, the ICP increases and the patient becomes more and more encephalopathic. Treatment is supportive and includes the usual measures used in the treatment of raised ICP [10]. These include intubation, sedation, and ventilation to modest hypocarbia, mild hypothermia, and treatment of blood pressure to maintain adequate cerebral perfusion pressure (CPP = MAP − ICP or CVP). Placement of an intracranial pressure monitor is necessary to have an accurate measurement of ICP.

4. Patients with ESLD often also have impaired renal function, secondary to lowered GFR resulting either from dehydration or from having developed the hepatorenal syndrome. Urine sodium concentration is generally low (<10 mEq/L) in both conditions, but in patients with prerenal azotemia, urine output increases, and serum BUN and Cr levels decrease following expansion of the intravascular volume. Patients with hepatorenal syndrome have oliguria and increased BUN levels that are generally not responsive to volume administration. Affected individuals also have ascites, and overly aggressive treatment of the ascites with diuretics may play a role in the development of the syndrome. Often dialysis is needed to reverse the pathophysiologic alterations of the hepatorenal syndrome until liver transplantation, which reverses the syndrome, can be accomplished [11, 12].

5. Portal hypertension is often part of liver failure [13]. Bleeding from esophageal and gastric varices are the major consequences of portal hypertension. A moderately severe episode of GI hemorrhage may tip a patient in tenuous condition into fulminant hepatic failure. Even if the bleeding is controlled, as the blood in the GI tract is metabolized and absorbed, the encephalopathy will worsen, and the episode of hypotension associated with the bleeding episode will worsen the renal ischemia, with the possible development of hepatorenal syndrome. Breakdown of liver glycogen is an important mechanism in the maintenance of normoglycemia. In liver failure, there is diminished breakdown of liver glycogen, making these patients susceptible to episodes of hypoglycemia.

6. Coagulation abnormalities are quite likely in patients with severe liver insufficiency or failure. In addition, these patients usually are anemic and thrombocytopenic. Fibrinogen, prothrombin, plasminogen, and many other coagulation factors synthesized by the liver are greatly diminished in patients with liver dysfunction/failure. Many patients with liver failure produce an abnormal fibrinogen molecule. In addition, bile salts are needed for absorption of fat-soluble vitamins

7. How should she be evaluated for metabolic abnormalities?

## Intraoperative Course

### Questions

1. What are the effects of liver disease on pharmacokinetics and dynamics of medications?

2. What preparations should be made with the blood bank and laboratory support for this case? How will rapid transfusion be accomplished?

that include vitamin K, a cofactor in the production of many coagulation factors. Many interventions by anesthesiologists, such as NG tube placement, intubation, and cannulation of vessels, have the potential to cause bleeding so that correction of coagulation abnormalities often is undertaken prior to the induction of anesthesia. Treatment of the coagulopathy seen in patients with liver failure may require replacement of factors and vitamin K. If platelet dysfunction is evident, DDAVP may be needed.

7. Patients with liver failure have derangements of many serum electrolytes. Common abnormalities are hypoglycemia and hyponatremia. Elevated BUN and Cr as a result of renal dysfunction are present, and elevated levels of ammonia are thought to be responsible for the encephalopathy [12].

**Answers**

1. There is a complex set of effects on the action and distribution of medications in patients with liver failure. These patients have a decreased serum albumin, which would lead to an enhanced effect of IV medications given at the usual dose on a mg/kg basis. These patients also have impaired hepatic metabolic function as well as impaired renal function. As a result of these abnormalities, the serum levels of medications will remain high for longer periods of time and will be less bound to protein. In addition, these patients may have depressed cardiovascular and pulmonary function prior to the induction of anesthesia.

2. Preparation of the OR should be for a long case in which massive blood loss is expected, temperature maintenance will be problematic, invasive hemodynamic monitoring will be needed, and many metabolic derangements will occur [14]. The OR table should be particularly well padded since these cases may last for many hours. Devices for rapid transfusion should be, at the very least, available or fully prepared. In the past, in larger patients, venovenous bypass was used, with the expectation that bowel edema and bleeding would be decreased compared with cases in which the vena cava was simply clamped. This practice is generally no longer used, however, simplifying the intraoperative management of liver transplant patients. The blood bank should be given as much notice as possible in order that the proper amounts and types of blood products are available. As a general guideline, ten units of PRBCs, ten units of FFP, and six to ten units of pooled platelets should be immediately available, with the expectation that more may be needed. Of course, these amounts should be adjusted upward or downward based upon the size and condition of the patient. Throughout the case, many ABGs, sets of electrolytes, coagulation profiles, CBCs, etc. will be sent. It may be necessary to have additional laboratory personnel to run these frequent and multiple tests.

3. Describe special consideration for monitoring and vascular access for this patient and procedure.

4. How will induction be performed? Does this patient have a full stomach? How does ascites affect the induction plan?

5. Describe the conduct of maintenance during the procedure.

3. In addition to routine monitors, temperature should be measured in more than one location. Rectal or bladder probes can be used in addition to esophageal probes. Several large IVs are needed, preferably in the upper extremities. During the anhepatic phase of the procedure, when the inferior vena cava (IVC) is clamped, lower extremity venous return will be limited to collateral veins or the venovenous bypass if it is used. Similar considerations apply to the arterial catheter. In some cases, the aorta will be clamped during the arterial anastomosis. Some centers use two arterial catheters. The direct arterial pressure tracing is unavailable during the frequent sampling, and if, because of frequent use, one arterial line fails, the second line will be available. A large, sheath-type central line is used in these cases for monitoring of central venous pressure, administration of vasoactive medications, and also administration of fluids and/or blood products. In general, pediatric patients need not be monitored with a pulmonary artery catheter. On occasions when peripheral IV access is difficult, two central lines may be used.

4. The patient should be comfortably positioned on the padded OR table prior to induction. In the induction of general anesthesia in unintubated patients, full-stomach precautions should be observed. Since these patients often have pulmonary dysfunction including a diminished FRC caused by ascites and abdominal distention and hepatopulmonary syndrome, thorough preoxygenation is essential prior to induction. Regardless of the specific IV hypnotic chosen, the dose should be adjusted based on the altered pharmacodynamics previously discussed. There is no specific contraindication to the use of succinylcholine. Often a combination of a hypnotic in a lowered dose and small doses of a benzodiazepine and opioid is used with the goal of rendering the patient unconscious without significant hypotension or heart rate alterations. As in most patients, the use of succinylcholine is associated with an increase in serum potassium of 0.5–0.7 mEq/L. If the patients have significant hyperkalemia prior to induction, the increase in potassium concentration may lead to cardiac arrhythmias. On the other hand, if the patient is compromised with a very small FRC, it is likely that significant hypoxemia will occur in the time required to achieve good intubating conditions using a nondepolarizing relaxant.

5. Although no particular technique has been proven to be advantageous or deleterious to children undergoing liver transplantation, it does seem prudent to avoid high doses of inhaled agents. High doses of inhaled agents have been shown to decrease splanchnic blood flow, possibly placing the graft at risk. A combination of an infusion of relaxant and an opioid with low-dose isoflurane or sevoflurane with additional benzodiazepines will likely achieve the goals of maintenance of an anesthetized state in the patient with minimal decrease in cardiac function. Since the procedure will last at least several hours and the child will generally remain intubated for the first postoperative night, concerns about the prolonged effect of IV medication affecting emergence are not important considerations.

6. What problems are expected during the preanhepatic phase?

7. What is important for the anesthesiologist during the anhepatic phase?

8. What problems are likely to occur during reperfusion?

6. The preanhepatic phase is the time of greatest blood loss. The surgeons are working to dissect free the failed liver. There may well be adhesions from previous procedures. Of course, during this time, the patient may be hemodynamically unstable and almost certainly has a coagulopathy. With significant bleeding and the massive transfusion required to maintain hemodynamic and metabolic stability, hyperkalemia, hypocalcemia, hypothermia, and hemolysis may all occur [15]. The use of washed PRBCs or newer PRBCs will decrease the amount of potassium in each unit. Ionized calcium and serum magnesium must be checked frequently during times of rapid transfusion since the citrate in the PRBC units chelates both divalent ions. Even with the administration of warmed blood products and fluids, the child's temperature may decrease during the preanhepatic phase. The abdomen is open and evaporative losses of fluid are significant. During this part of the procedure, ABGs, coagulation profiles, and electrolyte determinations should be done as often as every 30 min depending upon the amount of bleeding, transfusion requirements, and the degree of stability or instability of vital signs. In addition to blood loss, hypotension during the dissection phase may be due to either hypocalcemia or torsion of the liver during dissection with sudden decreased venous return.

7. The anhepatic stage of the procedure begins when the old liver is removed from the circulation, not with physical removal of the liver. When the infra- and suprahepatic cavae, portal vein, and hepatic artery are clamped, the child is anhepatic. Vigorous bleeding may still continue at the beginning of the anhepatic phase. In most pediatric liver transplants, femoral-axillary bypass is not used. Children tolerate clamping the vena cava during placement of the graft. While the old liver is out and the new liver not yet in the circulation, the child may demonstrate significant hemodynamic changes. There may be decreases in systemic blood pressure, central venous pressure, and cardiac output. As the child cools, oxygen consumption decreases with a concomitant decrease in carbon dioxide production. Also, during the anhepatic phase, any contribution the failing liver was making to glucose homeostasis is eliminated. The anesthesiologist should follow serum glucose frequently during the anhepatic phase and be prepared to treat hypoglycemia.

8. Once the vascular connections are complete, circulation is allowed into the new liver. The postreperfusion syndrome will occur in a significant number of patients once this happens. This syndrome includes hypotension and bradycardia, possibly even cardiac arrest. One preventable cause is inadequate flushing of the preservative solution from the graft. This solution is hyperkalemic, acidotic, and quite cold. If the graft is not thoroughly flushed, the patient will have profound hemodynamic instability once the preservative enters the circulation. The postperfusion syndrome can occur even if the graft is completely flushed of the preservative solution, however. Treatment is resuscitation with IV fluids and vasoactive agents. In some cases, only one or two doses of epinephrine are needed to maintain hemodynamic stability, but in others infusion of inotropes is

9. What problems are expected during the neo-hepatic/biliary reconstruction phase?

## Additional Questions

### Questions

1. A 6-year-old s/p cardiac transplant requires inotropic support due to acute rejection.

   What would your choices be to enhance cardiac output? Are anticholinergics effective? What are the relative effects of denervation on the adrenergic and cholinergic competency of the transplanted heart? Would milrinone be effective in enhancing contractility?

needed for several hours after the graft has been open to the circulation. In addition to the postperfusion syndrome, all patients have a rapid increase in end-tidal and arterial carbon dioxide once the IVC is unclamped.

9. As the new liver is connected to the recipient's hepatic veins and artery, coagulation problems begin to diminish. Hepatic artery thrombosis is more of a problem in pediatric liver transplantation, largely due to the smaller size of the vessel. Although no specific management of coagulation in the post-transplantation period has been shown to decrease the incidence of hepatic artery thrombosis, many anesthesiologists do not aggressively pursue complete normalization of PT/PTT as the liver is connected to the circulation unless significant, diffuse bleeding is ongoing. Correction of coagulation abnormalities generally begins as the new liver is connected to the circulation. However, if the patient has hypothermia or hypocalcemia, coagulation will be affected. These patients are sent to the ICU postoperatively with plans for mechanical ventilation [16]. Even in cases where the blood loss was not great, for example, less than half a blood volume, it is prudent to delay extubation, to later in the post-op period in the PICU or POD #1. The large incision will limit the child's ability to breathe. In addition, after such an extensive procedure, it may take some time to achieve hemodynamic stability.

**Answers**

1. Cardiac transplant recipients present several challenges to the anesthesiologist [17, 18]. Denervated hearts do not respond normally to input mediated via the autonomic nervous system. Drugs which act through stimulation of the autonomic nervous system may have little or no effect on a denervated heart. The usual bradycardic response to hypertension, mediated through the vagus nerve, occurs rarely if at all in these patients. In general, these patients do not tolerate decreased preload well.

   Pharmacologic enhancement of cardiac output is best accomplished with direct-acting agents such as epinephrine, isoproterenol, or dopamine. A drug such as ephedrine which has both direct and indirect effects in patients with innervated hearts will have only the direct effects on denervated hearts. Catecholamines such as dobutamine, dopamine, epinephrine, and norepinephrine will, via a direct effect on the myocardium, increase cardiac output. Atropine or pancuronium, two agents pediatric anesthesiologists rely upon to increase heart rate, will not be effective in cardiac transplant recipients. The alpha agonist phenylephrine, on the other hand, will increase vascular tone, but the baroreceptor response of lowered heart rate will not occur in the denervated heart. The phosphodiesterase inhibitors such as amrinone have direct effects on myocardial

2. An adolescent s/p heart transplant for viral cardiomyopathy 6 years ago presents to the emergency room with hypotension, J-point depression on ECG, but without angina. Please explain.

3. An adult living-related kidney is being transplanted into a 4-year-old recipient. What problems should you anticipate with unclamping? What is the most common cause of renal failure leading to kidney transplantation in children? What age is the greatest risk period for difficulty with perioperative medical management? Does anemia resolve after renal transplantation? What long-term effects should you anticipate with immunosuppression after renal transplantation, and how will it affect your subsequent anesthetic management of patients?

cells, increasing cAMP levels with resulting increased contractility. The systemic effects on preload will also occur, but baroreceptor responses to decreased preload will be absent or partially and inconsistently present in patients with denervated hearts.

2. Unexplained hypotension in a cardiac transplant recipient may very well be due to myocardial ischemia. These patients often have coronary artery disease after transplant. Rejection remains a major problem limiting survival in heart transplant recipients. The coronary arteries are affected with atherosclerosis when rejection occurs. Coronary artery vasculopathy accounts for approximately 30% of deaths after 1 year in transplant recipients. Angina may not occur in children with coronary artery disease since their hearts are denervated. Evaluation of cardiac transplant recipients for coronary atherosclerosis (vasculopathy) has been done in the cardiac catheterization lab using angiography. Dobutamine stress echocardiography has been used safely in children as a screen for coronary vasculopathy. Anesthetic management of children with coronary vasculopathy undergoing surgical procedures should be similar to techniques used for adults with coronary artery disease, with particular attention paid to the balance between oxygen supply and demand. The ECG may show ST segment changes with ischemia. Depending upon the procedure, consideration should be given to placement of a CVP or TEE. Children who have had cardiac transplantation who then return to the OR for procedures present several other problems to the anesthesiologist in addition to those outlined above relating to coronary artery disease. The denervated heart will not respond to autonomic input. Medications that affect cardiac rate or contractility via indirect mechanisms will not have those effects on the denervated heart. Direct-acting drugs such as epinephrine, norepinephrine, dopamine, and isoproterenol will affect cardiac performance. Baroreceptor responses to blood pressure changes are absent. Heart rate changes in response to decreased intravascular volume occur inconsistently. In addition, slowing of the heart rate, mediated by the vagus nerve, will not occur in these patients.

3. In cases where the kidney to be transplanted is from a person substantially larger than the recipient, significant hemodynamic consequences are likely, particularly when the graft is perfused. Hypotension may result not only from the release of graft preservative solution but also from depletion of intravascular volume as the new, large graft is perfused. Prior to opening the vascular clamps, the anesthesiologist should have given a generous amount of IV fluids, enough to elevate the CVP. Graft survival is dependent on adequate perfusion. The anesthesiologist must administer additional fluid as needed and/or use inotropes such as dopamine to maintain systemic blood pressure [19]. Although there are many causes of renal failure, obstructive uropathy, renal dysplasia/hypoplasia, and primary glomerular disease are the most common causes of ESRD in pediatrics. Younger transplant recipients present greater challenges with regard to perioperative anesthetic management as well as surgical technique [20]. Although renal trans-

4. Is flumazenil effective for hepatic encephalopathy? What is lactulose, and why is it used? What is the importance of a low-protein diet in liver failure?

plantation offers the best chance for normal growth and development in children with ESRD, nearly all such patients are maintained with either peritoneal or hemodialysis for varying lengths of time prior to renal transplantation.

The anemia seen in patients with ESRD has many causes such as decreased erythropoietin production, inadequate intake of iron and folate, low-grade hemolysis, and episodes of bleeding. In many patients, the hemoglobin will remain at approximately 6–9 mg/dL. With the administration of erythropoietin, the hemoglobin can be maintained at 10–11 g/dL. Following renal transplantation, most recipients are as anemic as they were pre-op. Many of the medications used to prevent rejection have deleterious effects on bone marrow production of red cells. For example, calcineurin inhibitors such as cyclosporine or tacrolimus (FK506) and antimetabolites such as azathioprine have bone marrow toxicity as a side effect. As the medications are adjusted to decrease all side effects, patients' hemoglobin increases, but it is not unusual for renal transplant recipients to be treated with erythropoietin (Epo) to increase the red cell mass.

Immunosuppression treatment for recipients of renal allografts includes steroids, cyclosporine, and tacrolimus. Cyclosporine is an 11-amino-acid peptide that inhibits T cell function by a variety of mechanisms, one of which is inhibition of interleukin (IL)-2 formation and action. Without IL-2, T cell activation is significantly diminished. Cyclosporine is metabolized by the cytochrome P450 system, and its metabolism is affected by co-administration of a variety of other medications. This drug has significant side effects. Neurotoxicity, manifested as tremors, paresthesias, headache, confusion, etc., hepatotoxicity, and hypertension and renal toxicity may limit the use of cyclosporine. Tacrolimus (FK506), a macrolide antibiotic similar to streptomycin, has similar immunologic effects as cyclosporine, inhibiting Il-2 and IL-2 receptor expression. Although prednisone has many deleterious side effects, it remains a part of the immunosuppression strategy used after renal transplantation. Side effects of importance include hypertension, growth failure, GI bleeding, pancreatitis, and osteoporosis. Post-transplantation lymphoproliferative disease (PTLD) is a very serious complication of immunosuppression [21]. This complication occurs in 1–3% of renal transplant recipients and can be seen at almost any time after the transplant. PTLD may result from B cell activation after a viral illness. The proliferation is seen in the GI tract and lymph nodes. The tonsils may be significantly enlarged as part of the presentation. Affected children may present for tonsillectomy/biopsy to confirm the diagnosis.

4. There are several possible explanations for the development of hepatic encephalopathy in the setting of liver failure. Ammonia, false neurotransmitters, and GABA (gamma aminobutyric acid) are all often elevated significantly in patients with liver failure and hepatic encephalopathy. Although ammonia levels are often elevated in patients with liver failure accompanied by encephalopathy, it is not unusual for an individual patient to exhibit encephalopathy prior to having elevated serum ammonia. GABA is an inhibitory neurotransmitter thought to play a role in hepatic encephalopathy. It is produced by intestinal bacteria as is

ammonia. Both are elevated in patients with liver failure. This molecule binds to CNS benzodiazepine receptors. Evidence in favor of this hypothesis is the fact that administration of flumazenil, a benzodiazepine antagonist, has partially reversed hepatic encephalopathy [7]. False neurotransmitters are also considered a possible cause of hepatic encephalopathy. Specifically, in liver failure the concentration of aromatic amino acids increases. These aromatic amino acids cross the blood-brain barrier and participate in the production of neurotransmitters [9]. Management of liver failure includes limiting protein intake as well as therapies to decrease serum ammonia levels [8]. Lactulose converts ammonia in the intestinal lumen into nonabsorbable ammonium. Minimizing the intake of protein also helps limit the production of ammonia. In addition, if there is less protein breakdown, fewer aromatic amino acids will be produced.

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# Chapter 27
# Minimally Invasive Surgery

**Robert S. Holzman**

A 14-year-old boy is scheduled for right-sided VATS resection of a posterior mediastinal/paraspinal mass (5 × 3 × 8 cm) found incidentally during a workup for persistent cough. He has a past medical history of exercise- and allergy-induced asthma and growth hormone deficiency, for which he takes growth hormone. He has a significant family history of hypertrophic cardiomyopathy. Labs unremarkable; VS unremarkable; 50 kg.



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## Preoperative Evaluation

### Questions

1. What respiratory effects would you expect with a posterior mediastinal mass? Are they always without respiratory effects? Would it matter if it was a left-sided or a right-sided mass? Other considerations for anesthesia and surgery?

2. What are your recommendations for postoperative analgesia? Any special considerations for implementing this plan?

3. Does this patient need an ICU bed to be booked postoperatively?

## Answers

1. Masses of the posterior mediastinum are usually of neurogenic origin (neurofibromas, schwannomas, and benign ganglioneuromas) from the sympathetic chain. Tumors derived from the neural crest, such as neuroblastoma and pheochromocytoma, may also, rarely, occur [1]. These tumors can grow quite large without significant encroachment on adjacent structures or exerting a mass effect on the middle or superior mediastinum, so it is rare for patients with posterior mediastinal masses to have respiratory distress, unlike patients with an anterior mediastinal mass. A CT scan or MRI will reveal the relevant thoracic relationships. For patients with signs and symptoms suggesting respiratory obstruction such as stridor or even persistent cough, it will be worthwhile to obtain pulmonary function tests in order to anticipate treatment with bronchodilators or postoperative mechanical ventilation. For masses that are large enough, venous drainage from the head and neck may be impaired, more likely with a right-sided rather than left-sided mass, resulting in the superior vena cava syndrome. Such patients may require vascular access below the chest, rather than in the upper extremities.

2. Adequate postoperative analgesia can be accomplished by a variety of means, including patient-controlled analgesia as well as a few different nerve block strategies. The surgeons could perform intercostal blocks under direct vision through the thoracoscope, although this would be a single-injection technique which would probably last for 6–8 hours. Continuous catheter techniques would provide longer duration analgesia and could be furnished via a unilateral paravertebral approach, an epidural approach, or an erector spinae approach. Each has specific advantages and disadvantages. An important consideration for this specific case is that it would be worth waiting until the end of surgery to make sure that intervertebral foramina were free of tumor before placement of any catheters, regardless of approach. If there is any doubt, local infiltration and PCA remain the reasonable strategies.

3. In all likelihood the patient will not need an ICU bed unless intraoperative events dictate otherwise.

## Intraoperative Course

### Questions

1. In addition to routine noninvasive monitors, do you need an arterial line for this case? Central access? Why/why not? What other non-routine monitors might be required?

2. How can you accomplish one-lung ventilation in this in a 50 kg adolescent? What are the available choices? What effective lumen size will you be dealing with, based on your choices? Why can't you find a right-sided double-lumen tube in the anesthesia workroom? What ventilatory adjustments will you need to make as you change over from two-lung to one-lung ventilation? Does this changeover affect minute ventilation?

3. Assume that the patient did have abnormal vital signs preoperatively with a heart rate of 110 and blood pressure of 150/104; what are your considerations for diagnosis of this posterior mediastinal mass? What is your basis for suspecting this diagnosis? Does the case need to be canceled for these vital signs? What if your first blood pressure was 185/122?

**Answers**

1. The need for an arterial line is influenced by the extent of the tumor and its local and potential systemic effects as well as how much anticipated dissection there will be according to the surgeons. In addition, if there is a component of airway compromise because of the size of the tumor and its mass effect on the middle or superior mediastinum, an arterial line should be considered along with ICU monitoring postoperatively. Adequate vascular access is crucial, and this patient should have (at least) 2 large-bore IVs; if the tumor results in a superior vena cava syndrome, then IV access should probably be in the lower extremities. A CVP might assist with assessing intravascular volume but the absolute number would not be diagnostic nor could it be relied upon intraoperatively as long as intrathoracic pressures were being manipulated and scopes were surrounding it. As the vena cava is dissected, the assessment may become unreliable as a result of mechanical displacement or intrathoracic pressure from insufflation [2].

2. There are several devices that are useful in small adults. Double-lumen tubes for a 50 kg adolescent boy can be utilized. Depending on the patient's appearance, a size up to 35 French can be utilized. A 32 French would not be unreasonable if the 35 was felt to be inappropriate.

   A 28 French would have a very small intraluminal diameter for one-lung ventilation during the case and is not really clinically acceptable. A bronchial blocker technique or Univent or Arndt tube can be utilized and would allow a larger lumen endotracheal tube. Right-sided DLTs are relatively hard to find. Roughly speaking, the total available lung parenchyma will be halved, but the minute ventilation requirements will be the same because the CO2 production will remain the same. Again, roughly speaking, I would decrease the tidal volume by 50% and look at the tidal volume/compliance to determine the volume I could deliver at an acceptable peak airway pressure. At that point, the minute ventilation required (should be the same as with two-lung ventilation) needs to be divided by the (tolerable) tidal volume in order to calculate the respiratory rate.

3. Posterior mediastinal masses are often neurogenic in origin, commonly arising from derivatives of the neural crest and its caudal migration in fetal life [3]. This germ line gives rise to pheochromocytomas, neuroblastomas, and more rarely, ganglioneuromas, ganglioneuroblastomas, and paragangliomas. With an elevation in the resting heart rate and blood pressure, and especially with the significantly increased blood pressure that may occur preoperatively or following anesthetic induction, one has to carefully consider pheochromocytoma or catecholamine-secreting neuroblastoma in the differential. Inasmuch as this is an urgent but not emergent case, it would be very reasonable to not proceed with

4. In the left lateral position with one-lung ventilation and the surgeons dissecting in the mediastinum, the oxygen saturations have decreased to 89% with an FiO2 of 1.0. Why is this happening? Are there equipment-related causes? Mechanical causes? Pulmonary causes? Cardiac causes? What will likely correct the situation? How would each of these interventions work? No matter what you do, you are not able to get the saturation above 93%; what maneuvers are left?

5. The SpO2 is now 88% on 100% O2 and the blood pressure 60/40 mmHg; how does this influence your analysis? What would you do? What do you think is going on? Can altering your ventilation affect this situation? Would you ask the surgeons to do anything differently?

## Postoperative Course

### Questions

1. Because the surgeon had concerns about placement of a regional block before he could fully evaluate the extent of the mass, he requested that any regional blockade be placed at the end of the case. What options are available to you? Which would be most effective for perioperative analgesia? What are the risks and benefits to each of these alternatives?

surgical resection, but retain the lines that were placed, go to the ICU, and spend the next 24 hours restoring blood volume and blocking the patient in order to attenuate the endogenous catecholamine response.

4. There are several potential causes for hypoxemia with one-lung ventilation, either by themselves or in combination. First of all, the change in position to left lateral decubitus changes ventilation perfusion matching in the lungs, and therefore physiological shunting will contribute to a change in the efficiency of oxygenation. There can be mechanical problems with incomplete location of the left-sided endobronchial tube into the left mainstem bronchus. An increase in airway reactivity may also be associated with lighter anesthesia, underlying reactive airway disease under the influence of a foreign body at the level of the carina. Finally, there may be suboptimal alveolar expansion of the "down" lung, therefore influencing closing capacity as well as adequacy of pulmonary circuit blood flow. All of these may be improved by carefully examining and confirming tube position and adequacy of the left mainstem bronchus seal; optimal ventilatory strategies including "best tidal volume" in relation to lung expansion, oxygen saturation, and capnograph morphology; passive oxygenation of the "up lung" with low flow oxygen (low enough not to provide continuous positive airway pressure of its own to the alveoli in the "up" lung); and clearing of secretions with attentive pulmonary toilet. Airway reactivity may be treated with bronchodilators (inhalation or systemic) and mechanical suctioning of the airway. If all of these maneuvers do not result in improvement, then the tube may have to be pulled back to a mid-tracheal position and used to ventilate both lungs.

5. There is cardiovascular compromise likely as a result of impaired venous return, mediastinal shifting, and shifting of the interventricular septum (or some combination thereof) because of the influence of pressure in the right chest. I would ask the surgeons to immediately decompress the chest after making sure that there was no immediately obvious surgical injury/vascular injury as a result of the dissection. A lower insufflation pressure will likely help this situation.

**Answers**

1. Regional anesthetic techniques include thoracic epidural anesthesia, thoracic paravertebral blockade, multiple intercostal blocks, interpleural analgesia, erector spinae plane block, and serratus anterior blocks. The current literature is characterized by enthusiastic advocates and often enthusiastic detractors of the various techniques. In addition, the routine use of ultrasound guidance has

## Additional Topics

### Questions

1. A 15-year-old girl is scheduled for a laparoscopic cholecystectomy. She has sickle cell disease.

   What are the relative advantages of the laparoscopic approach with regard to her perioperative care in the setting of this particular disease? Any disadvantages or special risks intraoperatively? What? Why? Would she benefit from a neuraxial anesthetic for postoperative pain control? Are there any risks for her? Her hematologist, pulmonologist, nephrologist, and cardiologist are all waiting to talk to you on the phone. What do you think they want to know, and what are you going to tell them?

2. An 8-year-old, 27 kg boy, with a right-sided empyema and a respiratory rate of 80 breaths/minute with a SpO2 of 89%, is in the ICU, getting ready to come to

allowed this proliferation of choices. Thoracic epidural analgesia was long revered as the gold standard for chest wall pain, but myofascial plane blocks (serratus anterior, ES) and PVBs have become alternative options, and all reduce perioperative opioid use. Not all are equally effective with regard to complete analgesia of the chest wall and the specific risks and benefits should be considered when making recommendations to patients.

## Answers

1. Cholecystectomy in a teenager with sickle cell disease is a relatively common procedure. A laparoscopic approach will allow her to have much less pain and a better ability to breathe postoperatively. Compared to a right upper quadrant incision with an open cholecystectomy, the laparoscopic approach will restrict her ventilation far less, improve her ability to maintain her FRC and take deep breaths, and lessen the chance that atelectasis will contribute to postoperative VQ mismatch and hypoxemia. Intraoperatively, one has to be careful to ventilate her with optimal tidal volumes in order to allow for adequate chest wall expansion and prevention of alveolar collapse. In addition, an adequate minute ventilation should defend her against respiratory acidosis and hypercarbia. It would be good to discuss an epidural with her for postoperative pain relief, but the majority of her pain may be as a result of diaphragmatic irritation with referred shoulder pain, and therefore, a high lumbar or thoracic epidural may not be of more help than a PCA.

   The various specialists are probably all concerned about the myriad end-organ effects of sickle cell disease and the kind of anesthetic technique that would minimize such effects. In order to decrease the chance of further sickling, the patient should have an adequate (or slightly above adequate) volume status, be kept warm and well oxygenated, and have a Hb SC level of less than 30–40% [4]. The prevention of chest crises can best be accomplished by adequate gas exchange intraoperatively and an optimal plan for postoperative pulmonary toilet, which can probably be accomplished with (1) laparoscopic surgery, (2) adequate analgesia via the epidural or PCA route, and (3) supplemental oxygen to maintain oxygen saturation at reasonable levels. The nephrologist will be concerned about her renal function, particularly in the renal medulla, which is susceptible to sickling in a hypoxic environment. Again, adequate volume repletion, warmth, and pain control will help renal blood flow and the maintenance of red cell integrity.

2. This patient is in severe respiratory distress, in all likelihood febrile with a significantly increased oxygen consumption, and will need a general anesthetic

interventional radiology for an evacuation and chest tube placement. What are your considerations for anesthetic technique? Can this case be accomplished with sedation and local anesthesia? What are the benefits and risks of each?

3. A 2-½-year-old, 13 kg girl is scheduled for a retroperitoneal laparoscopic nephrectomy for a unilateral polycystic kidney. She had a cold 3 weeks ago but is now afebrile and appears well. Are there anesthetic considerations in differentiating candidates for laparoscopic compared with open approaches for the same surgical procedure? What are the anesthetic implications with the retroperitoneal approach that are different than a transperitoneal approach? They plan to do the procedure with the patient in the prone position—how does this affect your anesthetic management? Is their recovery likely to be different? What are your considerations for management of intraoperative mechanical ventilation?

4. A 2-month-old for endoscopic suture repair for sagittal synostosis—what is the effectiveness of this procedure, and what implications does it have for the anesthesiologist?

with controlled ventilation, especially because he is also located remotely from you in the interventional radiology suite. The effect on adjacent lung parenchyma from an empyema of significant size may require him to remain mechanically ventilated postoperatively, so I would probably plan to return to the ICU with him intubated. The anesthetic technique may not matter very much; a primarily volatile agent-based or narcotic-based technique would probably be fine. Nitrous oxide should not be used because the pleural space will be invaded and a loculated air-filled pocket may expand even in the presence of a chest tube.

3. A laparoscopic approach to nephrectomy will allow for less postoperative pain, a shorter hospital stay, better cosmetic results, and a quicker return to normal activities [5]. Transperitoneal approach is commonly used for performing laparoscopy because of its familiarity, a larger working space, and a natural orientation to the natural landmarks. However, it requires mobilization and reflection of the colon. Surgically retroperitoneal laparoscopy (RPL) might be advantageous over TP laparoscopy (TPL) due to safe port placement, visceral handling with a lesser risk of injury, more rapid access to the renal pedicle, and easier renal artery control. Conversely, RPL may be technically more challenging because of the smaller working space and port proximity with resulting problematic ergonomics. Guillonneau et al. [6] in a retrospective study found both RP and TP approaches to have similar rates of complications and length of hospital stay but that the RP approach had longer operative times. There are mixed reviews of whether the retroperitoneal approach results in a greater burden of insufflated CO2 that results in hypercarbia. This finding may also be due to retained CO2 in retroperitoneal tissues that acts as a depot store within subcutaneous emphysema. Nevertheless, these findings do not seem to result in a clinically significant difference for patients. Mechanical ventilation requirements include a need for greater minute ventilation because of the additional exogenous CO2 insufflation, similar to transperitoneal insufflation, and the restriction to chest wall expansion imposed by prone positioning. There may also be lower perioperative analgesic requirements with the retroperitoneal approach because the abdominal viscera are not manipulated [7].

4. Endoscopic suture repair for craniosynostosis consists of a combination of endoscopic suture strip craniectomy and postoperative orthotic treatment. Key to understanding the success of the technique is that they are designed to restore normal anatomy, as opposed to reconstructing the skull at the time of surgery. They rely on subsequent brain and skull growth to obtain the correction [8]. Sagittal synostosis is the most common craniosynostosis. As experience has been gained with this technique, concerns about safety have given way to an appreciation for the speed of operation, extremely low rate of transfusion compared to open craniosynostosis repairs, and cost savings. There appears to be a much lower rate of venous air embolism when compared to standard open craniosynostosis repair [9]. Most patients are discharged the next day, and the rate of transfusion is about 2%. Cases are typically accomplished with two IVs, no arterial line, and patients are typically extubated at the end of the procedure and recover on the regular ward.

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# Chapter 28
# Ambulatory Surgery Procedures


**Thomas J. Mancuso and Joseph P. Cravero**

A 6-month-old female is scheduled for inguinal hernia repair. The mother reports that the child has had some nasal congestion for the last several days but has not had fever or other signs of systemic illness. The child is otherwise well except for a heart murmur, which has been followed by the child's general pediatrician.

VS: Temp 37.5, HR 122/minute, RR 32/minute, BP 90/60 mmHg RASpO2 97%.



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## Preoperative Evaluation

### Questions

1. Which pediatric patients are candidates for outpatient surgery? Which patients would *not* be candidates for outpatient surgery?

2. How old does a child need to be in order to qualify for outpatient surgery? What are the risk factors that would increase outpatient surgery risk?

3. What is the significance of an upper respiratory tract infection (URI) for outpatient surgery in this patient? If surgery is postponed, how long should you wait to perform anesthesia?

## Answers

1. Many pediatric patients are appropriate for outpatient surgery. There are certain categories of patients and procedures that are not appropriate however. In terms of patient groups, newborns are not appropriate for same-day surgery. Patients with significant systemic disease or malignant hyperthermia risk are likewise *not* appropriate for day surgery. In addition, patients undergoing procedures that are accompanied by large amounts of blood loss, respiratory compromise, or severe pain are not appropriate for outpatient management. Patients should be in a stable social environment where caregivers will be able to administer appropriate postoperative medications and/or interventions to manage discomfort, observe dressings, and monitor behavior and activity.

2. The age of patients appropriate for outpatient procedures depends on the underlying health and history of the patient. The primary concern is the incidence of apnea and bradycardia—which may occur in very young patients after general anesthesia. Apnea is strongly and inversely related to both gestational age and postconceptual age. A patient may be discharged to home after brief general anesthesia after approximately 5–6 weeks of age *if* he/she was born at full term and has had no other health issues—particularly no apnea or bradycardia. For patients who were born premature, the risk of apnea and bradycardia is significantly greater. Any former premature infant (born at less than 37 weeks' gestation) should be admitted after anesthesia for approximately 12–24 h of observation if they are less than 54 weeks postconceptual age. In addition, if a child was born at term and has had any issues with apnea and bradycardia, or if they have a sibling who experienced sudden infant death syndrome (SIDS), that child should likewise be admitted for observation until 58–60 weeks postconceptual age. In the past, it was thought that patients who received only regional anesthesia (such as a spinal) were not at risk for apnea; however, more recent data indicates that apnea may occur perioperatively in this population as well (although at a reduced incidence), and these patients should likely be admitted for observation [1–3].

3. URI illnesses are extremely common in children particularly in infancy and toddler age groups where the point prevalence in the middle of winter is approximately 30%. URI is defined as an illness limited to the head and neck, which may be associated with increased nasal secretions, but is *not* associated with systemic signs of illness such as fever or chills. A URI is also *not* associated with any lower respiratory symptoms such as wheezing, rhonchi, or rales. When anesthesia is administered to children who have a URI, there is an increased incidence of adverse respiratory events such as bronchospasm, laryngospasm, and coughing. On the other hand, anesthesia in children with these illnesses has not

4. What is the significance of the child's heart murmur? Should there be a cardiology consultation prior to the anesthesia? What specific questions would you have for a cardiologist?

## Intraoperative Course

### Questions

1. The parents request an inhaled induction—is that appropriate? How would you advise inducing anesthesia? The parents would like to be present for induction—would you agree to this?

been found to be associated with an increased incidence of serious morbidity such as respiratory or cardiac failure requiring ICU admission—or death. The period of increased airway reactivity after a significant URI lasts between 2 and 4 weeks. If elective surgery is postponed because of illness, it should not be rescheduled for at least this period of time. In this case, I would proceed with the surgery and anesthesia but would inform the mother of increased risk of minor respiratory events prior to beginning the case [4, 5].

4. Innocent heart murmurs in infants and young children are common. The two most common murmurs that fall into this category would be Still's murmur or the murmur associated with peripheral pulmonic stenosis. Still's murmur is due to resonance of blood as it flows through the left ventricular outflow tract during systole. It is "vibratory" or "musical" in quality and is heard most prominently at the left upper sternal border during systole. The murmur of peripheral pulmonic stenosis is heard best at the superior aspect of the left lower sternal border and is limited to systole. These "functional" murmurs are characteristic in that they are "soft"—less than 3/6 intensity. They may be positional—that is, heard in the supine position but not when sitting or standing. The child is otherwise healthy with no concerns about growth and no symptoms of heart failure, and the child tolerates periods of exertion (feeding and vigorous crying) without developing cyanosis or symptoms of heart failure such as dyspnea. Innocent murmurs are not be associated with a palpable thrill and are generally limited to systole. In this case, if the child is appearing well and the pediatrician believes this murmur is characteristic of an innocent murmur of infancy, I would accept the diagnosis after examining and confirming the history with the parents. If the child was sent to a cardiologist, I would want to know (1) if the cardiac anatomy was normal, (2) if the ECG was normal, (3) if the ventricular function was normal, and (4) if there was any evidence of shunting [1].

**Answers**

1. For a "well" 6-month-old, both an inhaled induction and IV induction are acceptable alternatives. The choice would be very much up to the preference of the anesthesia team and the OR system in which the anesthesiologist works. If the child was not appropriately NPO or had severe reflux requiring a rapid sequence induction, or if there were significant airway concerns, I would start an IV before inducing anesthesia. Since this is not the case with this patient, I would initiate anesthesia with inhaled sevoflurane and start an IV after induction. The presence of parents during anesthesia induction is a complex topic. As a general rule, parental presence does not decrease anxiety for the parents (unless accompanied

2. Can this case be done strictly with regional anesthesia? If general anesthesia is chosen, is regional anesthesia appropriate as an adjunct? What type of regional block would you perform in this case? The surgeon wants to perform a blind "field" block—is that appropriate?

3. What would you use for maintenance of anesthesia? Is total intravenous anesthesia (TIVA) appropriate?

by an extensive preoperative preparation program), and it does not change the behaviors of children around induction—particularly in a child 6 months of age. Parents are generally more satisfied with their perioperative experience if they have been allowed to be present for induction, but there are few observable improvements in behavioral or psychological outcomes that would argue for this practice. In this case, with a child who is not yet at the age associated with "separation anxiety," I would discourage parental presence for induction. In older children, I would accept parental presence for those who desire this, but I would administer a preoperative sedative such as midazolam (0.5 mg/kg) for children who are extremely anxious since parental presence alone is unlikely to improve this situation [1, 2].

2. Procedures that involve lower lumbar and sacral innervation can be performed under a spinal anesthetic. (In this case, if laparoscopic assistance were planned, a general anesthetic would be required.) Spinal anesthesia in infants has been variably adopted—with some centers very enthusiastic and others that rarely use regional anesthesia in this age group. Spinal anesthesia for infants and newborns is most commonly utilized in young infants (3 months old and younger). It is particularly popular for premature newborns/infants and those with complex respiratory diagnoses that could be adversely impacted by induction of general anesthesia and intubation. Studies in very young and complex infants have shown that spinal anesthesia can decrease the frequency of postoperative apnea and bradycardia although recent data suggests that the incidence is not completely eliminated. In this particular case, a 6-month-old is likely to be quite vigorous if disturbed (even with an effective spinal in place). Unless the surgeon and parents were very motivated to have a spinal anesthetic, I would not choose this option. Regional analgesia is an excellent idea for hernia surgery in a young infant. It would provide some component of the operative anesthetic (thus decreasing the MAC equivalents required for the surgery), and it would decrease the pain on emergence from anesthesia, thus decreasing the immediate requirement for opiates postoperatively. Regional analgesia could be provided by an ultrasound-guided ilioinguinal nerve block or a caudal block (with or without ultrasound guidance). The best block will depend on the training and preference of the anesthesiologist, but there is really no definitive evidence to suggest one block technique over another in this age group. A "field block" by the surgeon may provide analgesia, but the reliability and extent of coverage is unlikely to be as good as that provided by the other options [3, 4].

3. Both an inhaled maintenance of anesthesia and a TIVA anesthetic would be acceptable in this case. The primary advantages of a propofol-based TIVA in children include decreases in the incidence of nausea and vomiting and a decrease in the incidence of agitation on emergence. In the case of a 6-month-old infant, neither of these issues is particularly problematic; therefore, I would choose a general inhaled anesthetic to go along with regional analgesia.

## Postoperative Course

### Question

1. How would you control pain postoperatively? What non-opioid medications would be appropriate? Would you use opioid medications? Which ones would be appropriate? What are the issues associated with the use of opioids in this age group? How is a child at this age different than a newborn with respect to opioid administration?

## Answer

1. I would use regional analgesia (caudal block) to provide a component of pain control immediately on emergence. In addition, I would provide analgesia with acetaminophen and nonsteroidal anti-inflammatory agents. Acetaminophen works by decreasing the formation of prostaglandins and is safe in this age group. It has been shown to decrease the need for opiate medications by 20–30% in infants. Acetaminophen could be given by mouth (10–15 mg/kg) after emergence from anesthesia. It could be also be given by the rectal route in doses of approximately 30 mg/kg initially. The resulting acetaminophen levels are somewhat more variable than oral dosing. Subsequent doses can be given in 6 h intervals for 24 h, but the dose should be decreased to 20 mg/kg after the initial dose. Acetaminophen is also available in an IV preparation (12.5 mg/kg) and has been shown to have good effectiveness although it is much more expensive than the oral or rectal route. Ketorolac is a nonsteroidal anti-inflammatory agent that is effective in preventing the formation of prostaglandin 2 in children and adults. It has been shown to decrease opiate requirement by 30% after painful surgery. It can be given in a dose of 0.5 mg/kg during surgeries that are not at risk for significant blood loss—such as inguinal hernia [6]. Opioids are likely *not* needed in this case if regional anesthesia is present and non-opioid medications have been administered. Respiratory depression is always a risk with the use of opioids along with pruritus and nausea/vomiting. After 6 months of age, infants are no more sensitive to respiratory depressant effects of opioids (on a mg/kg basis) than toddlers and children; however, the small size of these patients makes drug calculation errors more of a risk than with larger patients. Newborns are more sensitive to the respiratory depressive effects of opioids. This is due to several issues including lower liver blood flow, a decreased activity of the cytochrome p450 enzymes, a less protective blood-brain barrier, and a greater intrinsic sensitivity to the respiratory depressant effects of opioids [1].

## Additional Question

### Question

1. A 17-year-old female is scheduled for laparoscopic ablation of endometriosis as an outpatient. After a previous surgery 1 year ago, she had to be admitted for nausea and vomiting. How would you attempt to minimize PONV in this case?

**Answer**

1. This patient is in a high-risk group for postoperative nausea and vomiting (PONV). There are several scales that grade risk of PONV—all of which include young age, female sex, gynecological surgery, history of previous nausea and vomiting, and nonsmoking status. The presence of these risk factors warrants efforts to decrease the baseline level of PONV as well as the use of prophylactic medications. I would attempt to mitigate her risk by providing the lowest risk anesthetic possible. Since general anesthesia with inhaled agents is associated with a higher incidence of PONV than total intravenous anesthesia, I would choose a TIVA anesthetic with propofol and low-dose remifentanil or a propofol infusion in addition to a reduced amount of inhaled agent. I would also administer multimodal pain medications with acetaminophen and NSAIDs in the perioperative time frame to minimize exposure to opiates. During the surgical intervention, I would use beta-blockers to help control hemodynamics rather than opiate mediations. I would have the surgeon infiltrate the laparoscopic port insertion wounds with a maximum allowable dose of local anesthetic. I would then add PONV prophylactic medications such as 5-HT3 antagonists (ondansetron) and a steroid (dexamethasone). Given the previous extreme reaction to anesthesia, I would also place a scopolamine patch during the anesthetic and have her keep this in place for 2 days after surgery. To the greatest extent possible, I would limit the administration of opiates. Finally, during the procedure, I would be sure to replace any fluid deficit and be sure to administer copious fluids to facilitate optimal hydration during recovery and in the immediate postoperative time frame [2].

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# Chapter 29
# Anesthesia Outside the Operating Room


**Robert S. Holzman**

An 18-year-old, 130 kg autistic boy is scheduled for an MRI of the brain to evaluate new onset grand mal seizures. He is developmentally delayed and requires assistance with activities of daily living but is conversational and loves to talk about trains. He has always had mask inductions for anesthesia and becomes very agitated and confrontational when you even mention the words "IV," "shot," or "a little drink." He takes omeprazole daily for reflux, and although he has been instructed to wear a CPAP mask at night, he refuses to do so because "it doesn't feel good."



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## Preoperative Evaluation

### Questions

1. How would you approach the preanesthetic evaluation of an autistic patient? What are your important considerations with regard to his mental status? How would you begin your conversation? What is the role of his parents in your preanesthetic discussion?

2. How do you assess his aspiration risk?

3. Would you insist on an IV prior to induction or is it OK for him to undergo a mask induction with a volatile agent? Is a parent-present induction all right, or is this "too high a risk?"

## Intraoperative Care

### Questions

1. You have decided to go ahead with a mask induction because the patient is becoming increasingly agitated with the placement of routine noninvasive monitors; is it all right to go ahead without them? What can you do to make his anesthetic induction as safe as possible?

**Answers**

1. From the autistic patient's viewpoint, the deluge of new faces, sounds, smells, icebox-like room temperatures, and other sensory novelties can be overwhelming. An insistence on "sameness" is a common feature, impossible to provide in the context of the perioperative experience when routines are interrupted and when patients are placed in unfamiliar environments, particularly those that involve separation from caregivers [1]. Broadly, 75% of autistic patients are nonverbal and either don't communicate or use specialized devices; about 25% are verbal. In this regard, the ability to converse with this patient is a great advantage. In addition, trusted caregivers, whether parents or others, are crucial to include regarding patient management, thorough communication with the surgeons and nursing staff for consistency, and appropriate modification to the perioperative environment (e.g., quiet, minimize traffic, low lighting when appropriate, etc.) based on specific patient needs. It would be good to engage him in a conversation about his favorite topic.

2. Aspiration risk even with a history of GERD does not exist in isolation. He and his parents should be asked about the effectiveness of his omeprazole, whether he wakes up at night with indigestion, whether he wakes up with regurgitated food on his pillow in the morning, whether he sleeps in an antireflux position, etc.

3. While it is always worthwhile to consider pre-induction placement of an IV with local anesthesia or EMLA cream, if it results in a struggle it may not be worthwhile. Preparations should be made for an inhalation induction, sitting up on the stretcher, and the availability of a videolaryngoscope to facilitate intubation. Preinduction airway assessment is crucial with regard to evaluating anticipated ease or difficulty of intubation. He can be induced in a semi-sitting position, maintaining spontaneous breathing. The drawback is the length of time for an inhaled induction in a large adult; however, the trade-off may be worthwhile if all goes smoothly and no struggles ensue.

**Answers**

1. With a history that is strong for untreated obstructive sleep apnea, a body habitus that is likely to impair gas exchange, and a procedural requirement for minimizing movement, I would plan for a general endotracheal anesthetic.

2. What anesthetic technique will you choose following the mask induction? Why? Would it be acceptable to transition to the total intravenous anesthesia technique once an IV is established? If you continue with a volatile anesthetic, would you choose a laryngeal mask airway or should the trachea be intubated? Why? Would it be safer to use a muscle relaxant or avoid one (assume the brain MRI will take 45 minutes)?

3. The medical student working with you asks about the use of dexmedetomidine because of its lack of effect on the minute ventilation/$CO_2$ response curve. What are your considerations in favor of/against its use?

## Postoperative Care

### Question

1. As the patient is emerging, he develops rhythmic tonic-clonic movements that begin in one arm and progress to generalized tonic-clonic activity. What to do next? Why did this happen? What if the anesthetic technique was continuous infusion propofol without an endotracheal tube? What are his risks of aspiration? What would you do to treat the seizure? How long would you keep the patient in the PACU? Should he be admitted? Assume that he had no seizure—what are your criteria for hospital discharge?

2. A TIVA technique may work, but for the previous reasons, it may not, and depending on how much time you would like to invest in identifying the pharmacological "sweet spot" for TIVA infusion, controlling the airway and ventilation will enable you to get the study done in a safe and timely manner. A laryngeal mask may, ironically, ensure a patent communication between regurgitated esophageal material and the airway, so I would plan to intubate the trachea.

   Neuromuscular blockade, while not required for the procedure itself, may well allow a lighter anesthetic, with quicker recovery, and motionlessness, essential for MRI.

3. Dexmedetomidine is a very acceptable alternative regardless of whether one goes down the pathway of intubation or natural airway. There is sparse data available on the safety and efficacy of dexmedetomidine in adults with mild OSA; that data suggests a better margin of safety than propofol. However, in this circumstance, when it is not simply about dexmedetomidine's effect on the $CO_2$ response curve but also the amount of residual somnolence, and effects on heart rate and volume requirements, I would still favor a general endotracheal anesthetic and the fastest possible emergence so that patient can resume his normal and usual mental status.

**Answer**

1. This situation is not very surprising; the emergence from a general anesthetic produces an abrupt change in the patient's anticonvulsant status, and this shift can especially make a more poorly controlled patient have an increased incidence of seizures. Preparation is the key to treating this problem. If the airway is protected with an endotracheal tube, then the seizure can be treated with small intravenous doses of propofol or a benzodiazepine. If the airway is unprotected or if an LMA is in place, then the risk/benefit ratio of leaving it in or protecting the patient's airway has to be decided upon. If there have been no preceding difficulties with the airway, incremental doses as described above can be given with the patient receiving supplemental oxygen. If it seems that the patient may progress on to regurgitation, then he will be at increased risk of aspiration because the gag reflex will undoubtedly be less competent during the ictal and immediately postictal phase following the seizure. The duration of PACU stay will depend on how many seizures the patient ordinarily has per day, the competence of the parents in dealing with these seizures, and to some extent the risks of driving home (e.g., distance home, distance to closest hospital, etc.). I would probably keep the patient for 4–6 seizure-free hours in the PACU, in consultation with the patient's neurologist.

## Additional Topics

### Questions

1. A 7-year-old boy is scheduled to undergo esophagogastroduodenoscopy (EGD) for symptomatic reflux disease. Is there a safe way to administer nitrous oxide or some other sedative strategy or must he just have an IV placed? Is a rapid sequence induction with succinylcholine necessary? After EGD, biopsies, and much air insufflation, the endoscopist declares he is done and ready for you to wake the patient up. Your considerations? Is all the air *ever* out of the upper GI tract? The patient is now light, bucking on the tube, and burping—what is your next move?

   Would it have been better to do a "deep" extubation?

2. A 3-year-old is receiving weekly intrathecal methotrexate for a brain tumor. His oncologist requires motionlessness and his parents require lack of recall and comfort. What will your approach be? What does it depend upon? If he has a pansinusitis from his immune suppression, (assume his oncologist and parents feel strongly that his treatment must go on), how does this influence your anesthetic choices?

3. A patient with a prior history of hives following contrast injection for IV pyelography is returning for a repeat pyelogram with "anesthesia standby." What is his risk of subsequent reaction? How can he be treated to lower this risk? Why does this occur? How should you be prepared?

## Answers

1. Chances are a 7-year-old will not be particularly enthusiastic about having an IV prior to induction, but unless this is an insurmountable problem, good preparation will probably enable you to place the IV. EMLA cream, the use of local anesthetic (once the EMLA has taken effect), and possibly an oral premed are all reasonable for this child, while carefully explaining to the parents that a mask induction may not be the best strategy. That is not to say that it can't be done, however, and the risk/benefit ratio has to be evaluated. If it were necessary to go ahead with a mask, then I would have him sit up or be placed in an antireflux position, with cricoid pressure, and then place an IV as soon as practicable. It is important to make conditions as safe as possible for intubating the trachea, and muscle relaxation would be very helpful. Succinylcholine will provide the most rapid intubating conditions. There is emerging experience with pre-induction evaluation of gastric contents (volume as well as density) using point of care ultrasound; however this experience is primarily in adults with the "risk range" identified as 1.5–2 mL/kg. The data is far more sparse in children, and the significance of this data is unclear [2]. With the amount of insufflation typically necessary for EGD, I would have the patient wake up completely rather than consider a deep extubation.

2. Much would depend on the underlying disease being treated and the patient's current medical state. He may be quite debilitated from chronic chemotherapy or doing relatively well, and therefore, this will influence the anesthetic approach. Most 3-year-olds (and their doctors) will require motionlessness, and if the patient has an indwelling central line, I would probably start a propofol infusion, administer a low induction dose (so as not to cause apnea but simply allow unconsciousness) then turn him on his side for the bone marrow and lumbar puncture, and encourage the use of liberal amounts of local anesthetic. An intercurrent infection would not be too surprising, and this situation arises frequently because children with prolonged hospitalizations or community exposure will often acquire various opportunistic infections or even URIs from other patients, family members, or visitors. Most of the time this does not interfere with the progression of anticancer therapy because of the urgency involved, unless the patient is significantly symptomatic, showing high fevers, constitutional lethargy, or other signs and symptoms that may make it worthwhile to wait for 24–48 hours (but hardly longer because of the context).

3. Approximately 5% of radiological exams with radiocontrast media (RCM) are complicated by adverse reactions, with 1/3 of these being severe and requiring immediate treatment. Reactions occur most commonly in patients between 20 and 50 years of age and are relatively rare in children. With a history of atopy or

4. A 12-year-old girl with cerebral palsy and spasticity is scheduled for Botox® treatment of her upper and lower extremities. What can you anticipate with regard to her anesthetic management? Does she require airway protection with an endotracheal tube? Does she need an IV for induction and continuation of a TIVA technique? Does she need an IV at all for this 5-minute procedure?

allergy, the risk of a reaction is increased from 1.5- to 10-fold. Reactions may be mild, subjective sensations of restlessness, nausea, and vomiting to a rapidly evolving, angioedema-like picture. Low osmolar RCM are relatively safe with regard to life-threatening reactions. The treatment of severe allergic reactions, whether anaphylactoid or anaphylactic, is no different than for any other allergic reaction. Epinephrine, aminophylline, atropine, diphenhydramine, and steroids have all been employed in order to control varying degrees of adverse reactions. A patient who requires RCM administration and who has had a previous reaction to RCM has an increased (35–60%) risk for a reaction on re-exposure. Pretreatment of these high-risk patients with prednisone and diphenhydramine 1 hour before RCM administration reduces the risk of reactions to 9%; the addition of ephedrine 1 hour before RCM administration further reduces the rate to 3.1% [3].

4. Cerebral palsy patients are often veterans of the operating room and numerous office medical visits, and it is crucial to understand the patient's level of communication and cognition. While it is ideal to place an IV before the procedure, this will not always work out because the patients are frequently fearful and "veinless." Premedication may blunt the anxiety, but an effective dose will likely be much larger than the time it will take to recover from the anesthetic and procedure. A concern about reflux will usually prompt serious consideration for a rapid sequence induction and endotracheal intubation, but most of these patients, even with a history of reflux, undergo these procedures with TIVA, even following an inhalation induction and IV placement. The induction can be accomplished in a sitting position as an antireflux adjunct. If all went smoothly (and the past anesthetic records can help you here), then I would place an IV and continue with a TIVA technique. It is safer to place an IV if at all possible [4].

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# Chapter 30
# Vascular Anomalies


**Robert S. Holzman**

A 4-day-old with a vein of Galen malformation is scheduled for embolization. The diagnosis was not known prenatally. She was noted to have tachypnea with feeding, which led to a subsequent workup revealing cardiomegaly on chest X-ray and dilated cardiac chambers with systolic failure on echocardiogram. Her HR is 172 bpm with a blood pressure of 54/32. Respiratory rate is 64/ min. She is cool, clammy, and peripherally cyanotic, with a hyperactive precordium and what sounds like a continuous murmur over the heart, and coarse breath sounds diffusely. She is on a high-flow nasal cannula with an oxygen saturation of 92%.



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## Preprocedural Evaluation

### Questions

1. What accompanying conditions and findings should you anticipate?

2. How would you plan for effective anesthetic management?

3. What are the consequences of success of the procedure? What are the procedurally related risks?

## Answers

1. Newborns with large vein of Galen malformations will typically present in high-output congestive heart failure, which in this age group will be characterized by tachypnea and sweating with feeding with signs of adrenergic stimulation like tachycardia. I would anticipate the cardiovascular findings to be characterized by right ventricular strain including RVH on ECG, an elevated RV pressure, and perhaps impairment of RV shortening fraction. There may be tricuspid insufficiency and elevated pulmonary artery pressures as well. The ductus arteriosus may well be patent and with elevated pulmonary artery pressures may be shunting left to right. The baby may be intubated and mechanically ventilated for respiratory support.

2. Safe anesthetic management would consist of minimizing myocardial depression, minimizing volume loading, support of effective gas exchange, and normocarbia with the lowest tolerable peak airway pressures. Access preparations should include adequate intravenous access (at least two IVs) and an arterial line. It is important to remember that the radiologist's sheath also represents access if needed. Motionlessness is key to the procedure—the baby is small, and the blood vessels tiny. Any slight movement is also magnified considerably on the image intensifier equipment in the radiology suite.

3. The procedure will hopefully result in successful embolization of the AV malformation, with a reduction in high-output failure and improvement in the "melting brain" phenomenon seen on imaging. This will ultimately result in some cardiac remodeling, lowering of the elevated PA pressures, and improvement in gas exchange, work of breathing, and ultimately growth and development. Nonetheless, the risks are high in this setting—neonatal presentations accompanied by intractable cardiac failure have a poor overall prognosis. Death may result, in the short term, from intractable cardiac failure or procedurally related complications, and the longer term from neurological injury or neurodevelopmental abnormalities. In referral centers, with this constellation of findings, good quality survival approaches 50% [1].

## Intraoperative Course

### Questions

1. What anesthetic technique will you choose? Why? How will you know that it is effective?

2. What ventilation management strategy will you choose? Why? How can mechanical ventilation affect cardiac performance in this neonate? How will you assess these effects?

3. Discuss your IV fluid management? What are your fluid management goals with regard to the cardiopulmonary system? Is this management strategy likely to affect the imaging characteristics for the radiologist?

4. What are your specific clinical considerations for injection of the embolization material?

## Answers

1. I would try to minimize myocardial depression from high doses of volatile agents and provide attenuation of stress responses and pulmonary vasoreactivity with relatively high doses of fentanyl. The sympatholysis from a stress-attenuating strategy is also important in blunting the vasospasm response in the small intracranial vessels when they are mechanically stimulating by the microcatheters used for embolization. Neuromuscular blockade is crucial in order to provide motionlessness for imaging and embolization.

2. There are complex interactions in the cardiopulmonary system and the cerebral circulation. Positive pressure ventilation with high peak inspiratory pressures, perhaps necessary when the lungs are suffused with pulmonary edema, will worsen venous return and further impair myocardial contractility on both sides of the heart. Low tidal volume ventilation, so-called "lung protective" strategies, may not provide sufficient alveolar ventilation to promote gas exchange and eliminate CO2 to normocarbia. The correct balance of positive end-expiratory pressure with defend the FRC and promote gas exchange, but it must be carefully titrated so as not to impair cardiac performance in this patient with heart failure. The overall adequacy of systemic perfusion can best be assessed biochemically by looking at lactate levels. It would be very reasonable, in addition, to employ point-of-care ultrasound to evaluate cardiac performance.

3. Likewise, fluid management can be challenging in this situation. Volume overload would be disastrous for this patient, yet an adequate or even generous circulating blood volume provides optimal imaging for the radiologist and is an important consideration for the success of the embolization. Here again, it is important to use physiological markers of adequacy of perfusion like lactate levels. There should also be a background infusion of a dextrose-containing solution so that hypoglycemia (which should be measured frequently) is avoided.

4. The embolization material is important. In the past, the most commonly used embolic agent was the fast-polymerizing liquid adhesive n-butyl cyanoacrylate (n-BCA). The intranidal flow and polymerization of n-BCA was typically quick and unpredictable. The new liquid embolic agent of choice, once it was introduced, is Onyx, which is less adhesive and polymerizes more slowly than cyanoacrylate glue. Onyx contains ethylene-vinyl alcohol copolymer, dimethyl sulfoxide (DMSO)), and tantalum. The polymer is dissolved in DMSO and is prepared in three different concentrations: 6.0%, 6.5%, and 8.0%. Micronized tantalum powder (35% wt./vol) is added for radiopacity. Because Onyx polymerizes more slowly, there are some specific considerations which may affect anesthetic management, particularly with regard to ventilation. The microcatheter is

## Postoperative Course

### Question

1. What are your perioperative recommendations for the NICU staff with regard to care of this baby? Do you have any specific mechanical ventilation recommendations? Cardiovascular recommendations? Anything they should be on the lookout for post-embolization?

## Additional Topics

### Questions

1. A 4-month-old with stridor is scheduled for a diagnostic laryngoscopy and bronchoscopy. She has 3 cervicofacial cutaneous hemangiomas. What will the surgeons likely find on examination? What will they probably recommend? How would you manage her anesthetic if she was started on propranolol therapy and her resting heart rate was 74 bpm? What is the basis for propranolol therapy for hemangiomas anyway? Is it likely she will need more airway exams?

initially flushed with normal saline and then a small amount of DMSO is injected to fill the dead space. Onyx is then injected slowly over about 40 seconds to fill the microcatheter and replace the DMSO in the dead space. Slow injection of the Onyx is then continued under fluoroscopy. Long injection times often require a pause in order to obtain an angiogram to assess nidus occlusion and the status of draining veins. Because positive pressure ventilation can significantly affect cerebral blood volume as well as flow, repeated pauses in ventilation or apneic oxygenation may be required.

**Answer**

1. Despite the intense desire to evaluate this baby neurologically following the VGAM embolization, I would first ensure the stability of the cardiovascular system and look for improvement in the high-output congestive failure state and improvement in gas exchange. With regard to the respiratory system, I would look for improved gas exchange and carefully evaluate the work of breathing before extubating. Nevertheless, the focus once those systems are clearly improving and stable would be on obtaining a neurological exam and waking the baby up. Improvement in ventilation mechanics can follow the usual metrics—improved gas exchange, lower PEEP requirements, improved compliance, etc. Effective VGAM embolization is likely to result in significant improvement or even normalization of the CHF. Routine post-procedural care will be to evaluate the puncture site for hematoma and examine the distal circulation in the leg and foot.

**Answers**

1. Hemangiomas are hypercellular vascular tumors during their proliferative phase and then decrease in cellularity with increasing fibrosis during their involution phase. The usual clinical course is rapid proliferation for a few months followed by regression that can take several years. Primary therapy is medical, currently with propranolol; surgical intervention is rare unless vital structures are threatened [2, 3]. Cervicofacial hemangiomas should be investigated for subglottic lesions when infants have stridor accompanied by cutaneous hemangiomas.

2. What are the radiation safety considerations for you as an anesthesiologist in the interventional radiology suite? What measures are routinely taken to protect the anesthesiologist?

3. At what point in the procedure should you be concerned about airway swelling following sclerotherapy near the airway? For how long?

4. A patient with Sturge-Weber syndrome is scheduled for a head MRI. Why? What are they looking for?

2. The IR suite is designed to protect the radiologist; anesthesiologists incur ionizing radiation exposure at a rate 4× greater than the radiologist does while at the same distance from the patient. Enhanced radiation protection consists of increased shielding, decreased time of exposure, and increased distance from the radiation source [4, 5].

3. Swelling after sclerotherapy does not peak until several hours after the procedure. Patients with lesions near the airway may need to remain intubated post-procedure for several days, with extubation in the operating room following an airway exam. Tube exchangers are useful adjuncts, and close collaboration with ORL service may be needed.

4. Six to 10% of patients with capillary malformations in the facial V1 dermatome have Sturge– Weber syndrome, which includes the facial CM, ipsilateral leptomeningeal vascular malformation, and choroidal vascular malformation of the eye. These patients can have seizures, developmental delay, hemiparesis, and glaucoma. Facial hemihypertrophy with overgrowth of soft tissue and bony structures is present in up to 60% of patients, many of whom will require surgery for cosmetic and functional improvement.

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# Chapter 31
# Dental


**Joseph P. Cravero**

You are asked to provide anesthesia for a 4-year-old patient undergoing full mouth dental rehabilitation. The patient has a history of a behavioral disorder that has very generally been described "oppositional" and has been diagnosed with autism spectrum disorder. He is difficult to direct, likes to do as he likes, and does not like to be touched, and he is non-verbal. He is on no medications but his mother gives him ginkgo biloba on a daily basis along with a homeopathic "soothing" remedy. History is also significant for a VSD that was repaired at 4 months of age.



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## Preoperative Evaluation

### Questions

1. What are "oppositional" disorder and autism? What are the implications for your anesthetic? What are the strategies available to calm this child or begin the induction process in the preoperative area?

## Answers

1. Behavior and psychological disorders are over-represented in the dental population undergoing anesthesia and full mouth rehabilitation. Oppositional defiant disorder (ODD) describes an ongoing pattern of uncooperative, defiant, and hostile behavior toward authority figures. It seriously interferes with a child's day-to-day functioning. Symptoms can include temper tantrums and excessive arguing with adults. Autism is a condition that is present early in childhood that is characterized by difficulty in communicating (language) and forming interpersonal relationships. These patients tend to be inflexible in their adherence to routines.

   Changes in their daily schedule imposed by fasting, new environments, and new people can result in significant agitation and severe anxiety. There is a wide range of severity of autism. Some individuals are communicative and interactive, but many are unable to communicate their concerns effectively. In light of these issues all attempts should be made to work with parents to bring in familiar objects or toys to their surgical encounter. Nursing personnel in the preoperative and postoperative areas should be made aware of a patient with ODD or autism so that special arrangements can be made for private, quiet space and extra personnel to help with management. Often, the patient may have a preference for a specific video or musical recording and that may be helpful in creating a reassuring environment. Information on what techniques have been successful should be solicited and accommodated to whatever degree it is possible. In spite of these efforts, it can be very difficult to gain cooperation from this population of patients, and premedication is often necessary. Oral midazolam (0.5–0.75 mg/kg) is the most widely used premedication in pediatric anesthesia and generally produces a relaxed state that aids in promoting cooperation, Unfortunately, it may be associated with paradoxical agitation in approximately 10% of patients. Intranasal dexmedetomidine 2–3 mcg/kg has similarly been shown to have sedative and calming effects plus a minimal degree of analgesia. For patients who are completely uncooperative, IM ketamine 4 mg/kg is used to produce dissociation that will allow a quiet state with minimal movement for IV placement and anesthetic induction. The "dissociated state" refers to the functional dissociation of the thalamus from the cortex. The child's midbrain is not anesthetized—so airway reflexes and respiratory drive are preserved.

   Onset of ketamine sedation is heralded by lateral nystagmus with eyes open and breathing/airway tone intact. It is important to warn parents and family members about what the sedated state will look like, or it can be alarming to those present during this process. At this point, an inhaled or IV induction can take place without requiring excessive physical restraint. It is particularly important to involve the parents in management of these patients and allow them to give insight into which techniques have worked in the past. In addition, I would

2. What are the implications of the history of a VSD on your anesthetic management of this child? How would you determine this? Does this child require antibiotic prophylaxis for this procedure?

## Intraoperative Course

### Questions

1. How would you induce anesthesia? How would you prepare and secure the airway? Are there special concerns for this patient? What are the options for airway management? How would you maintain anesthesia in this patient?

involve the Child Life Specialists to assist in finding comfort items and a plan that will be agreeable to the family and result in the best possible behavioral outcome.

2. The history of a repaired VSD would generally not affect the planning of anesthesia to a great degree. The most important issue would be the presence of any residual defect and continued intracardiac shunting. If there were a residual shunt, in this case it would usually be left to right and result in relative pulmonary overcirculation. If the shunt was significant, it would be critical to limit physiologic changes that would encourage further increases in pulmonary circulation—such as hyperventilation, systemic vasoconstrictors, or excessive oxygen tension. These changes could result in lower systemic blood flow or the potential for pulmonary congestion. On the other hand, acute changes that result in profound increases in pulmonary pressures (prolonged and severe Valsalva maneuvers) could result in right to left shunting and lower O saturations. For any patient with an ASD or VSD, it is important to be sure that all lines have been de-aired and/or add air filters to the intravenous lines. A detailed exercise history from the family is important to detect any evidence of significant shunting or poor ventricular performance. Questions should be asked about any episodes of cyanosis, unexpected shortness of breath, or other signs of inadequate activity tolerance. Any patient with this history will have had multiple visits with a pediatric cardiologist and is likely seen yearly for follow-up unless the issue is considered completely resolved. The most accurate assessment of residual shunt and ventricular performance would come from an echocardiogram.

   Antibiotic prophylaxis is only required for those with unrepaired congenital heart disease, repaired defects with prosthetic material or device in the first 6 months after the procedure, and any lesion with a residual defect adjacent to a patch or graft. With this in mind, it is extremely unlikely that this patient would need perioperative antibiotics.

## Answers

1. As stated above, an IV or inhaled induction is acceptable. If the patient were sedated with ketamine, I would place an IV (since patients sedated with ketamine tolerate IV placement very nicely). Inhaled induction could be performed but the anesthesiologist needs to be aware that laryngospasm is reported in a small portion of patients who have been sedated with ketamine. I would administer a small dose of opiate along with propofol for induction. Muscle relaxation is not required for this procedure but is also not contraindicated. For full mouth

2. During the case the patient coughs and there is a sudden decrease in the oxygen saturation. Is this likely due to the residual effect of the VSD? How would you manage this problem?

dental rehabilitation, a nasotracheal tube is preferred by most dentists since it allows easy access to all quadrants of the oral cavity without having to reposition the tracheal tube (or LMA if that was chosen). Preparation of the nose can be performed in many ways. Several drops (sprays) of a vasoconstrictor such as phenylephrine (Neosynephrine®) to the nasal mucosa are helpful in minimizing bleeding with nasal tube placement. It should be noted that nasal spray bottles are intended to be used in the upright position. When they are turned upside down and used as a dropper bottle, excessive doses can be delivered, and significant overdoses have been reported. In this case the dose should be carefully limited secondary to cardiac concerns.

Nasal passages are often asymmetric. Dilation of the nasal passage with progressively sized, lubricated, nasopharyngeal airways is often helpful in determining the appropriate side for tube placement and for dilating the orifice on that side. After the tracheal tube is passed through the nose into the posterior pharynx, it can be placed under direct visualization with a Magill forceps or it can be placed over a nasally placed fiberoptic scope. Anesthesia may be maintained with an inhaled agent but in this case I would opt for a total intravenous anesthesia technique utilizing propofol and fentanyl with the goal to minimize the emergence delirium. Emergence phenomena are less common after a TIVA (propofol based) when compared with sevoflurane-based inhaled anesthesia. A dose of dexmedetomidine (1 mcg/kg) given over 5–10 minutes after induction would also be helpful to maximize comfort and minimize agitation postoperatively.

2. If the patient coughs and bucks violently creating massively increased pulmonary pressures, a residual shunt or subclinical defect could be opened and flow may be reversed from right to left, thus resulting in an intracardiac shunt and decreased oxygen saturations. While this is possible, it is more likely that the decrease in saturation would be due to problems with the ET tube (mainstem intubation, plugging, or kinking) or lung pathology such as bronchospasm or lobar collapse. In response to this incident the anesthetic depth should be deepened with additional inhaled agent, opioid, or a significant dose of sedative hypnotic. The lungs should be auscultated for equal breath sounds or evidence of wheezing. It would be appropriate to suction the ET tube and assure appropriate $FiO_2$ and ventilatory parameters are set and being met. Bronchodilators should be given if indicated.

## Postoperative Course

### Questions

1. On emergence, the child is crying and thrashing about for 30 minutes. He does not make eye contact; he cannot be quieted regardless of the efforts that are made by family and nurses. What is the likely cause? Could this have been avoided? What would you do to treat this problem? Are there certain patients that are more likely to have this problem? What would you tell the parents about the likely effect on the child in the long term?

2. After the procedure the child has prolonged bleeding from the gingiva. The mother thinks this may have happened to the child's sibling as well—and treatment was needed. What could be the possible causes of this issue and what would you do to treat it? Is there any chance that the child's intake of herbal supplements (from mother) are a cause of the problem?

## Answers

1. The child is likely to be experiencing emergence delirium—likely not emergence agitation. Although these terms are used interchangeably, "delirium" refers to a state of disconnection from reality while agitation refers to an unhappy, irritated state. The delirious child does not make eye contact and cannot be soothed by parents using food or entertainment. An agitated child often calms if appropriate distraction is employed. The exact cause of emergence delirium is not known but is likely similar to the type of state that is present when children have night terrors or partial awakening behaviors. Young children and toddlers are most at risk for this phenomenon. As mentioned above, patients who receive pure inhalation anesthesia are more likely to have delirium than those who receive *tiva*. This episode *might* have been avoided if pure *tiva* was chosen over inhaled anesthesia. Ear, nose, oral, and throat procedures are accompanied by more of this type of behavior than peripheral orthopedic procedures. This problem can be treated by "resedating" the patient with propofol or dexmedetomidine; however, delirium and agitation generally do not last longer than 30 minutes in the postanesthesia setting. Gently restraining the child while reassuring parents is an option. I would tell the parents that studies have shown that patients with emergence delirium or severe agitation can have behavior changes that last up to 2 weeks postoperatively, but no one has connected this phenomenon with permanent behavior changes.

2. Prolonged bleeding after a dental procedure in a child who has a possible family history of this kind of bleeding, but no other serious bleeding history, is most likely related to a platelet issue such as von Willebrand's disease. Approximately 1% of the population is affected by this disorder but the bleeding tendency is usually mild and diagnosis may be very delayed. This is actually a heterogeneous disorder with three primary subtypes characterized by a quantitative or qualitative abnormality of the von Willebrand factor (VWF) protein. This protein is made in the endothelium and has two primary roles: (1) it binds platelets to collagen at the sites of vascular injury, and (2) it binds and stabilizes factor VIII. The most common forms of this disease are autosomal dominant in inheritance and involve a quantitative lack of normal protein. The primary treatment for the disorder is with desmopressin (DDAVP) which can be administered IV, subcutaneously, or intranasally. DDAVP is synthetic and poses no infection risk. Administration is associated with a six- to eight-fold increase in the concentration of VWF and is effective for most mild/moderate episodes. Ginkgo biloba is thought to produce neuroprotective effects as an antioxidant, free-radical scavenger, membrane stabilizer, and an inhibitor of platelet-activating factor. It is the inhibition of platelet-activating factor that is thought to have the greatest likelihood of effect on bleeding tendency in patients. The drug should not be used in

## Additional Topics

### Questions

1. A 17-year-old needs her severely impacted wisdom teeth extracted. Her past medical history is significant for Treacher-Collins syndrome. She is moderately developmentally delayed and has been difficult to intubate in the past. She has no current breathing problems and does not snore according to her mother. You are asked if she needs to have a pregnancy test prior to anesthesia—and why? Does she need to be intubated for this case? Would deep sedation with a natural airway or an LMA be sufficient for this case?

conjunction with warfarin, aspirin, or other antiplatelet agents. Ginkgo should be stopped 36 hours to 14 days prior to surgery. Depending on the dose given and the presence of other medications, Ginkgo could add to a bleeding tendency, but is unlikely to be the primary cause for bleeding in this case. Other herbal medications that could be implicated in bleeding include feverfew, garlic, ginseng, dong quai, and red clover.

## Answers

1. There is no definitive answer to the question of whether or not the patient needs a pregnancy test. The American Society of Anesthesiologists allows physicians and hospitals to implement their own policies with regard to pregnancy testing. The overall rate of incidental positive pregnancy testing is between 0.3% and 2%. Studies have demonstrated an increased probability of spontaneous abortions, congenital anomalies, and low birth weight in infants born to mothers exposed to anesthesia and surgery during pregnancy; however, the nature of the surgery and the duration of exposure can greatly affect this risk. Specific data on the risk posed by tooth extraction as in this case are not available. Nonetheless, it is important to identify individuals who are pregnant and avoid exposure that is not absolutely necessary. Some institutions prefer a detailed history of sexual activity, menses, and contraception—with testing only for those who are at risk for pregnancy. Most children's hospitals opt for universal pregnancy testing for females past menarche since the procurement of this detailed history is (at times) difficult and (often) inconsistent. The need for airway management will depend on the nature of the impaction of the wisdom teeth. Impacted wisdom teeth (also known as third molars) have not fully erupted because of blockage from other teeth. If not removed, pain, inflammation, and infection can result. Severe impaction requires extensive work to break up and extract the teeth. Less severely impacted teeth can be managed relatively quickly with much less surgery. A conversation with the oral surgeon is in order. If significant surgery is required, general anesthesia with a nasal or oral endotracheal tube is appropriate. For less involved cases, deep sedation with copious local anesthesia can suffice. It would be important to establish that this patient did not have a history of severe sleep apnea before moving forward with natural airway sedation. There is no indication (with this patient) that bag-mask ventilation would be difficult. As such, if the teeth were severely impacted, I would secure the airway with a videoscope or fiberoptic scope after induction of anesthesia to avoid the chance of an emergent airway obstruction and need for intervention in the middle of this case. These cases can be performed with an LMA as the primary airway—if the provider is comfortable with the child's status and the oral surgeon is willing to work around the device.

2. A 6-year-old with relapsed ALL is sent to you for dental rehabilitation prior to repeat bone marrow transplant. What are the implications of prior treatment with an anthracycline for ALL?

2. Good dental hygiene is important for patients undergoing bone marrow transplantation or other transplantation due to the immune compromise that is part of these procedures. Anthracyclines include doxorubicin (Adriamycin), daunorubicin, and epirubicin. Collectively these are the commonest agents implicated in the development of cardiotoxicity after chemotherapy. This effect can be acute or chronic in nature. Acute effects include ST-T changes, decreased QRS voltage, and QT prolongation. These changes are present in as many as 30% of patients and resolve with time. The chronic effect is usually manifested as a cardiomyopathy resulting in decreased contractility in a smaller percentage of patients. These effects are dose dependent being present in 7% at 550 mg/m$^2$ and 35% at 700 mg/m$^2$. At less than 400 mg, the incidence is 0.14%. Essentially all of these patients are followed with serial echocardiograms, and this information should be sought prior to administering any anesthetic agent to a patient who has received one of these drugs. It is important to recognize, however, that previous treatment with anthracyclines may enhance the myocardial depressive effect of anesthetics even in patients with normal resting cardiac function. Anthracycline agents can also cause primary dysrhythmias (supraventricular tachycardia, complete heart block, ventricular tachycardia, and prolonged QT interval) unrelated to the cumulative dose. These problems can evolve hours or days after administration.

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# Chapter 32
# Trauma I

**Robert S. Holzman**

A 12-year-old, 52 kg. boy, previously healthy, was attending a gun show with his family. After trying an AR-15 at the range, he ran downrange to retrieve his target when his younger brother picked up the rifle, with its safety off, and accidentally discharged the weapon, hitting his brother in the upper right leg. In the ER, the 12-year-old was clutching his stomach, in a lot of pain, and was pale and short of breath. HR 150 bpm, RR 62/min and crying, BP 75/50 mmHg, HCT 27%. There is a 22 ga. IV in place in the left saphenous vein and seems to be running well.



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## Preoperative Evaluation

### Questions

1. Why is he clutching his stomach and in pain there when his entry wound is his leg? What further details do you want to know about the injury? Why is that important?

2. What is your interpretation of his vital signs? Is there anything else you want to know?

3. What if there were decreased breath sounds on the ipsilateral side to the leg injury?

4. The patient's neck veins are distended and don't appear to be varying with respiration; what is the significance?

## Answers

1. The assumption should never be made that an exit wound can be predicted from the trajectory of the entrance wound; many bullets are flattened or carved (dum-dum bullet) and therefore their exit wound can be very remote from the point of entry, or they can expand and not have an exit wound at all. Detailed knowledge about the missile and weapon informs an understanding of the type of injury to expect. High-velocity missile injury is caused by missiles (bullets or fragments) traveling at a rate >750 meters/second [1, 2]. While low-velocity missiles, such as 0.38 caliber, 0.45 caliber, and 9 mm pistols typically carried by police, will result in laceration and crushing of surrounding tissue, high-velocity missiles form a temporary cavity due to energy transfer from the missile to the tissue and create a tissue shock wave that compresses tissue in front of the missile. The physics of this phenomenon are such that it is especially destructive against solid tissue. Fragmentation bullets, for example, are engineered to break apart after a certain traveling distance following penetration.

2. The vital signs are concerning for significant volume depletion. A heart rate of 150 is markedly elevated and the blood pressure is low. Even more concerning, the pulse pressure is narrowed which supports a presumptive diagnosis of significant volume depletion. He is also very tachypneic which may reflect lung injury. The hematocrit is moderately decreased, but this could also be artifactually high if he was hemoconcentrated or artifactually low if he was aggressively fluid resuscitated with crystalloid. A chest X-ray would be helpful for rib fractures, cardiac size and contour, as well as pneumo- or hemothorax or abnormalities in the cardiac silhouette. A KUB of the abdomen might also reveal free air and could be obtained at the same time as the chest X-ray in the ER.

3. This finding warrants immediate intervention with a chest tube if the patient's respiratory situation deteriorates. There may be enough time to confirm a pneumothorax, for example, with a chest X-ray. Alternatively, the absence of a pleural slide on ultrasound along with the absence of breath sounds and respiratory deterioration should prompt immediate chest evacuation for a presumed pneumothorax.

4. With distended neck veins, the patient probably has obstruction of venous drainage to the central circulation. Likely causes include pericardial tamponade, intrathoracic obstruction with impaired venous drainage, or pneumo-/hemothorax with mediastinal shifting. While in most circumstances with adults it would be fitting to place a chest tube prior with only local anesthesia, it may be more practical with conscious children to induce anesthesia first and then be prepared to place a chest tube immediately.

5. Does he have all the intravenous access you want? What do you think about the placement of the IV?

## Intraoperative Care

### Questions

1. Does this patient need an arterial line? Why/why not? Prior to induction, or after? What about stat mode on automated blood pressure cuff? Is it the same?

2. Does he need a central line? If so, when? Prior to induction? Just after induction? Can it wait until after the surgeons get started?

3. Does the patient need a chest tube prior to induction of anesthesia, or can one be placed after rapid sequence induction and intubation? What if there was subcutaneous emphysema? Clinical significance and implication for anesthesia plan?

4. Because the abdomen is so large, the surgeon is planning an exploratory laparotomy first. What are your considerations with regard to the size of the abdomen? The blood bank tells you it will be 45 minutes until crossmatched blood is available. What are your plans? What issues are involved with proceeding emergently? Urgently? Waiting?

5. Following rapid sequence induction and endotracheal intubation, additional vascular access should be secured, primarily above the level of the diaphragm assuming that a superior vena cava syndrome existed. Again, for practical purposes, insertion of the IV above the diaphragm, assuming that veins were visible, could be done immediately following induction. He will need adequate access to support major transfusion requirements if needed.

**Answers**

1. The patient definitely needs an arterial line, which should be anticipated from the number of systems potentially involved as well as the likelihood of postoperative ICU support. He may not require it pre-induction (and you may not be able to get one in anyway) but it should be placed as early as possible. Stat mode on an automated blood pressure cuff will work for blood pressure assessment until the line is established, although it provides less information (discontinuous, no waveform analysis).

2. A central line may not be a bad idea, especially if peripheral veins are collapsed, but it is not likely to be necessary, based on the previous information. With distended neck veins, the patient probably has obstruction of venous drainage to the central circulation; likely causes include pericardial tamponade, intrathoracic obstruction with impaired venous drainage, or pneumo-/hemothorax with mediastinal shifting. Placement of a central line prior to induction would entail supine or Trendelenburg position, likely worsening the respiratory embarrassment.

3. This would depend very much on the clinical picture. Fortunately, at 12 years old, it is possible to contemplate sedated chest tube placement with local anesthesia. The circumstances, however, may render this strategy impractical because the acute trauma setting also involves a lot of fear and anxiety with the patient, and the luxury of titrating an adequate amount of sedation along with the administration of adequate local anesthesia may not be possible. With an adequate pre-induction diagnosis of pneumothorax, the patient can be expeditiously moved to the OR, through a rapid sequence induction, and immediate placement of a chest tube, probably in a much shorter time.

4. A KUB of the abdomen might reveal a hepatic hematoma and/or free air to account for the abdominal pain and worrisome vital signs and could be obtained at the same time as the chest X-ray in the ER. Realizing that an abdominal compartment syndrome may account for the clinical picture, blood needs to be prepared and in

5. The course of the bullet, upon examination of the surgical field, appears to be an entry wound at an angle just below the right inguinal ligament, creation of several traumatic large and small bowel enterotomies in the right lower and right upper quadrant, several liver lacerations, an intact porta hepatis, a diaphragmatic perforation, and embedding in the pleural surface of the right tenth rib, without an exit wound. How does this knowledge help you understand what the surgeons will likely be doing?

## Postoperative Care

### Questions

1. The surgeons have completed an exploratory laparotomy with repair of several small enterotomies, a colostomy, repair of several liver lacerations, repair of the diaphragmatic perforation, and removal of the bullet with rib resection and chest tube replacement. What are your plans for perioperative management?

2. Would the patient benefit from regional analgesia in the perioperative period?

the room at the time of surgery. The clinical context will dictate how expeditiously the patient will need to be moved to the operating room and whether, if vital signs are stable, the decision to proceed with surgery can wait long enough for a crossmatch or whether the more conservative approach would be to operate earlier and have O negative or immediate-spin crossmatched blood available in order to avoid waiting 45 minutes. The overall risk of hemolysis following the transfusion of uncrossmatched erythrocytes to patients needing an emergency transfusion is 0.1%. Nonetheless, uncrossmatched blood should not be used for otherwise stable patients who can wait until crossmatched units become available [3].

5. After examining for vascular injury, it is likely that the surgeons will be [1] exploring and repairing the superficial and deep soft tissue wounding at the entry site and [2] exploring and possibly repairing some of the bowel enterotomies and likely small and/or large bowel ostomies, exploring the liver with repair of lacerations, repairing the diaphragm, and removing the embedded bullet from the rib. Anesthetic planning includes a relatively long procedure with likely transfusion and likely postoperative mechanical ventilatory support in the ICU and close postoperative monitoring until recovery.

**Answers**

1. The overall strategy for perioperative management includes the critical care support of all vital organ systems. In all likelihood this patient should remain intubated, if for no other reason than to keep the respiratory system in balance during the acute phase of recovery. The usual criteria for extubation include adequacy of tidal volume, minute ventilation, and bellows strength. In addition to that, one would have to anticipate the perioperative course; massive transfusion, for example, might influence a conservative approach to weaning and extubation. The massive tissue trauma will require aggressive nutritional support in order to provide an effective milieu for tissue healing. Liver function and synthetic ability will need to be carefully monitored as the liver heals from trauma. In addition, changes in the splanchnic circulation should be anticipated; therefore, renal function may be at risk.

2. Regional analgesia may very well be of benefit not only for pain relief but also in order to assist pulmonary function and the weaning process. There are a variety of choices that may provide analgesia to the multiple surgical areas involved, but this would not become clear until surgery is completed. Even then, with uncertain outcomes of the potential for sepsis as well as the effects of the trauma on liver function, it may be prudent to wait until the postoperative period to place a block, which may take the form of an epidural, a paravertebral, or erector spinae catheter with a continuous infusion.

3. The surgeons want to re-explore the abdomen in 48 hours; should the patient remain intubated, ventilated, and sedated in the ICU until that time?

## Additional Topics

### Questions

1. You are preparing a patient with a chest tube for Medevac transport via nonpressurized fixed wing aircraft. What are your critical care considerations for this flight? How will you monitor? Does it matter if he is intubated or not? Is it any different for helicopter transport?

2. A knife fight between two gangs results in one member (the lucky one) arriving in your OR with a stiletto in place, piercing his trachea at approximately the level of the cricoid cartilage. He was brought directly to the OR and skipped the ER entirely. How will you proceed?

3. The timing of extubation should be determined by patient readiness rather than the convenience of leaving the endotracheal tube in for subsequent procedures unless the procedure is just around the time of anticipated extubation, when it wouldn't make much practical sense to extubate and then reintubate.

## Answers

1. Nonpressurized aircraft will have a lower ambient cabin pressure and lower atmospheric oxygen tension as they rise in altitude. If chest tubes are clamped as the aircraft leaves the ground, the increase in altitude and lower atmospheric pressure will result in expansion of the pneumothorax and respiratory compromise. This will be aggravated by the lower oxygen tension in the cabin atmosphere if the patient is not receiving supplemental oxygen (assuming he is breathing on his own). Respiratory compromise has to be treated by decompressing the clamped chest tube and readjusting it if necessary. Helicopters fly at a lower altitude than fixed wing aircraft.

2. The biggest risk in this scenario is that the knife is physically interfering with the passing of an endotracheal tube, which must be accomplished before the neck can be explored. The knife may also be tamponading any blood vessel through which it has passed, so that the ideal airway management would be an awake intubation, with the surgeon already having prepped the neck for exploration. As the patient is undergoing laryngoscopy and intubation, the surgeon should be prepared to remove the knife and exert pressure for local vascular control until the neck can be incised (following intubation) and the structures identified. This calls for a patient who is cooperative and topically anesthetized—in an ideal circumstance. Otherwise, speed counts, and after the neck is prepped, the patient must be rapidly induced, the trachea intubated, and the surgeon prepared immediately to explore the neck. Positive pressure ventilation must be gentle and kept to a minimum in anticipation of accumulating subcutaneous emphysema.

3. A 5-year-old 22 kg. boy, previously healthy, was brought to the emergency room following a terrorist bombing at his school. He was in a remote corner of the gym when the bomb went off, also in the gym. He is writhing in pain in the ER, clutching his stomach, short of breath, and bleeding from his left ear. He has a broad bruise across his chest where he was hit by a volleyball pole. He seems to have difficulty hearing you in between his crying. HR = 150 bpm; RR 42/min and crying, BP 75/50 mmHg, and HCT 27%. There is a 22 ga. IV in place in the left saphenous vein and seems to be running well. What is your immediate assessment in the ER, where you were called to see him urgently before he gets in the elevator to go to X-ray. If the EMT's reported that he lost consciousness en route and then improved on arrival, would that affect your plan? What is the significance of the bleeding from the ear? What if he had a clear discharge from his nose? On arrival in the OR, you note ST segment elevation on lead II—significance? What are the possibilities and what would you do?

3. Starting from the top down, you would have to be concerned about any secondary head and cervical spine injuries, although his mental status seems all right [4]. The hearing impairment and bleeding from the ear suggest tympanic membrane rupture from the shock wave injury, which would also rupture other gas containing structures like the lung and bowel [5]. The stomach pain suggests that might be the case with intestinal disruption. The shortness of breath and tachypnea may reflect lung injury, but could also reflect myocardial contusion and impaired myocardial performance as a result of the chest wall trauma [6]. The blood pressure is lower than expected and the heart rate higher than normal, which could represent volume depletion or myocardial impairment which might include a hemopericardium with tamponade. The hematocrit is moderately decreased, but this could also be artifactually high if he was hemoconcentrated or artifactually low if he was aggressively fluid resuscitated with crystalloid. The mental status exam is probably the most important "monitor" in the ER, followed by the cardiovascular system. Supplemental oxygen would be advisable; at the very least, it would provide a margin of safety should further interventions be required on an urgent basis. If there is time, a head and body CT scan and C spine series might reveal additional areas of injury, but the yield might be low. A chest X-ray would be helpful for rib fractures, cardiac size and contour, as well as pneumo- or hemothorax or abnormalities in the cardiac silhouette. A KUB of the abdomen might also reveal free air and could be obtained at the same time as the chest X-ray in the ER; this strategy would probably have a higher yield than the CT scan. An ECG is necessary to look for myocardial injury. The fundoscopic examination might reveal retinal hemorrhages, which reflect the severity of the blast injury as well as retinal detachment. A loss of consciousness should prompt more aggressive investigation for closed head injury and the C spine. A clear discharge from the nose, in the absence of recent symptoms of an URI, should be suspicious for a CSF leak from the skull base. ST segment elevation indicates injury and should prompt a more geographically oriented investigation such as a 12-lead ECG; in this setting, one would be suspicious for an anterior wall pattern of injury because of the volleyball pole's path of chest injury. Nitroglycerin is not likely to be of any benefit in this setting and may actually make the patient worse because of the reduction in preload. A transesophageal echocardiography (TEE) examination would help in terms of assessing regional wall motion abnormalities and areas of hypokinesis; a transthoracic echocardiogram would also be helpful to look for the same problem.

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# Chapter 33
# Trauma II


**Thomas J. Mancuso**

A 4-year-old who fell into an empty swimming pool headfirst is brought to the hospital for evaluation and treatment. A cervical spine fracture is noted on the X-rays taken in the ED. At the pre-op visit in the ICU, the child is awake and alert in a hard collar, breathing spontaneously but with slight tracheal retractions. He wiggles his fingers and toes. He is scheduled for fixation of his cervical spine fracture.VS: HR 65/min; RR 22/min; BP 100/60 mmHg; RASpO2 96%.



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## Preoperative Evaluation

### Questions

1. Will you evaluate him for other injuries?

## Answers

1. This child should have a head-to-toe evaluation for other injuries [1]. In this case, there is not a neurosurgical emergency necessitating an immediate trip to the OR. There is time to complete a thorough evaluation for other injuries to the child. The hard collar must not be removed, but nevertheless a physical exam looking for signs of other injuries can be done. With major pediatric trauma, the head is the most frequently injured body area. The neurologic exam should assign a Glasgow coma scale score and evaluate the child for the presence or absence of raised intracranial pressure. The presence of raised ICP will have important impact on the overall management of the child as well as the conduct of an anesthetic [2]. The Glasgow coma scale was initially developed to provide an organized, uniform measure to the level of consciousness [3]. Using the scale, CNS injuries are categorized as mild (13–15), moderate (9–12), or severe (<8). In addition, cranial nerve function should be assessed and focal signs in the motor exam noted. During the evaluation of the cranial nerves, signs of basilar skull fractures such as hemotympanum and "raccoon eyes" should be noted. The Glasgow coma scale is used to assess cortical and brainstem function. The activity, best verbal and motor response and total score for pediatric patients are presented below.

*Infants*

- Eye opening
  - Spontaneous 4
  - Opens to verbal stimuli 3
  - Opens to painful stimuli 2
  - No response 1
- Verbal response
  - Coos and babbles 5
  - Irritable cry 4
  - Cries to pain 3
  - Moans to pain 2
- Motor response
  - Spontaneous purposeful movements 6
  - Localizes to pain 5
  - Withdraws to pain 4
  - Flexion to pain (decorticate) 3
  - Extension to pain (decerebrate) 2
  - No response 1

*Children*

- Eye opening
  - Spontaneous 4
  - Opens to verbal stimuli 3
  - Opens to painful stimuli 2
  - No response 1
- Verbal response
  - Oriented 5
  - Confused 4
  - Inappropriate words 3
  - Incomprehensible words 2
  - No response 1
- Motor response
  - Obeys commands 6
  - Localizes to pain 5
  - Withdraws to pain 4
  - Flexion to pain (decorticate) 3
  - Extension to pain (decerebrate) 2
  - No response 1

*Adults*

- Eye opening
  - Spontaneous 4
  - Opens to verbal stimuli 3
  - Opens to painful stimuli 2
  - Non response 1
- Verbal response
  - Oriented to person, place, time 5
  - Confused 4
  - Inappropriate words 3
  - Incomprehensible words 2
  - No response 1
- Motor response
  - Obeys commands 6
  - Localizes to pain 5
  - Withdraws to pain 4
  - Flexion to pain (decorticate) 3
  - Extension to pain (decerebrate) 2
  - No response 1

2. What is the significance of the neurologic exam?

3. Which laboratory tests are important prior to going to the OR?

4. Is there a role for the immediate administration of steroids?

In addition to the neurologic exam, the child should be evaluated for injuries to the thorax and abdomen. An initial FAST exam followed by CT has become the primary tool for the evaluation of pediatric trauma patients for abdominal damage. A child who is the victim of a major trauma should first have a CT of the head, and then, if clinically stable, the scan should include the thorax, abdomen, and pelvis. When deciding about the importance of these scans, the clinician must balance the time needed and the IV and enteral contrast administration required for the scans against the urgency for surgical intervention for other injuries. It is important not to ignore the long bones during the evaluation of a pediatric trauma victim. Fractures of the femur can be associated with significant blood loss in the absence of clinical signs.

2. It is encouraging that the child is awake and alert. The GCS score of the child as described would likely be at least 13 (eye opening: spontaneous, 4; verbal, oriented, 5; motor withdraws to pain, 4). With a higher score on the motor portion of the scale, the GCS could be as high as 15. The fact the child is moving his fingers and toes is an indication that there is some neurologic function below the level of the cervical spine fracture, but the residual function appears to be incomplete.

3. Additional radiologic studies are indicated unless the child requires emergent surgical intervention. In addition to scans of the head, chest, abdomen, and pelvis, several additional tests are indicated prior to going to the OR. The X-rays taken that showed the spine fracture should be reviewed. It may be that additional studies are needed to rule out additional fractures including a limited CT of the neck. Blood should be sent for a CBC, type and cross, and coagulation studies.

4. A prospective study done in the 1990s demonstrated that spine-injured patients given large doses of methylprednisolone very soon after the injury had a slightly improved neurologic deficit. The study was not designed to assess long-term neurologic function. The doses of methylprednisolone used in the investigation were 30 mg/kg IV followed by 5.4 mg/kg/h for 23 h. There have been three trials of this drug and dose, one conducted in North America (NASCIS), one in Japan, and one in France. A meta-analysis of these trials indicated that significant recovery of motor function occurred when the therapy outline began within 8 h of the injury. A more recent trial showed that continuing the therapy for an additional 24 h led to additional improvement in motor function particularly if the therapy is not begun within the 8-h period. Significant controversy still exists regarding the use of high-dose steroids for spinal cord injury (5).

## Intraoperative Care

### Questions

1. Which monitors and vascular access are necessary for this case?

2. How should general anesthesia be induced? Which muscle relaxant should be used to facilitate laryngoscopy?

3. Are there particular concerns regarding prone positioning for the repair of the cervical spine fracture?

## Answers

1. Prior to the administration of any medication, the child should have at least one well-functioning IV and should have had fluid resuscitation with normal saline. If additional injuries had been uncovered in the workup, the location of IV placement may be affected. If there is also significant abdominal or lower extremity trauma with potential for hemorrhage, IVs should be placed in the upper extremities. The usual ASA monitors should be used. In addition, once induction is completed and the airway secured, an arterial line and central venous pressure line should be placed. If the preoperative evaluation revealed any signs of raised ICP, consideration should be given to placement of an intracranial pressure monitor. Since the child will be in the prone position for the approach to the fractured spine, placement of a precordial Doppler and/or monitoring of end-tidal nitrogen will be important for detection of venous air emboli.

2. Given the X-ray findings and clinical exam, it is imperative that the child not move his neck during induction and when the airway is secured. An IV antisialagogue such as glycopyrrolate should be given prior to any airway manipulations. It should be assumed that the child has a full stomach during the induction even though a complete RSI would not be indicated in this situation. The child has retractions with respirations. If the CXR does not reveal any possible etiology, it is possible that the child has traumatic damage to the airway. The child may have aspirated during the traumatic event and the CXR has not yet shown abnormalities. There could be partial or complete paralysis of the vocal cords or edema or hematoma formation within the airway. Given these considerations, securing the airway with fiberoptic bronchoscopy is a prudent choice. Following administration of a drying agent, the child is gradually anesthetized with an IV agent such that spontaneous respiratory effort is maintained. Topical lidocaine, in the proper dose and concentration, is applied to the nasal and pharyngeal mucosa prior to instrumentation of the airway.

3. Once the airway is secured, and if no further neurologic exams are to be performed, the child can be completely anesthetized and muscle relaxants administered. If the vital signs are satisfactory and ventilation adequate, the child can be safely positioned prone for the procedure. The hard collar should be kept in place, and the neurosurgeon should be present and control the head during the turn from supine to prone. Fortunately, the child is small so that he/she can be easily lifted and rolled to the prone position. In the prone position, the abdomen should be free to minimize effects on ventilation. With the abdomen free, the loss of FRC is minimized. If the head is above the level of the heart, the risk for venous air embolism is increased, but if the head is below the level of the heart, venous congestion of the face, neck, and CNS can become problematic. Visual

4. Are there particular maintenance agents best suited in cases where neuroprotection is important? What is the optimal BP for this procedure?

5. During the procedure, the patient has bradycardia, with HR decreasing to the 60s. What might be the etiology? Your response?

impairment has been reported in patients who were kept prone for long periods and who also had impaired oxygen delivery to the retina from hypotension, anemia, and/or excessive pressure on the eye, limiting blood flow.

4. Although barbiturates have the reputation for cerebral protection, other commonly used induction and sedative agents such as propofol, etomidate, and benzodiazepines also lower cerebral blood flow (CBF), cerebral metabolic rate for oxygen (CMRO), and intracranial pressure (ICP) while maintaining autoregulation. Ketamine causes an increase in CBF as well as ICP. Opioids have no direct effect on these parameters but, as respiratory depressants, will lead to hypercarbia and the resultant cerebral vasodilation in the spontaneously breathing patient. All inhaled anesthetics, nitrous oxide included, dilate cerebral vessels to varying degrees. This effect is mitigated by hyperventilation. Isoflurane and sevoflurane are both appropriate for neurosurgery. Both do increase CBF to a modest degree. Both decrease CMRO. Blood pressure management is directed toward maintaining adequate cerebral perfusion pressure (CPP) [6]. Cerebral perfusion pressure is equal to mean arterial pressure (MAP) minus ICP or CVP, whichever is higher.

$$CPP = MAP - ICP \ (\text{or } CVP \text{ if } CVP > ICP)$$

Since in this case we do not know the ICP or CVP, the blood pressure should be kept at or within 20% above the BP measured preoperatively.

5. The sudden occurrence of bradycardia in a neurosurgical procedure can be the result of raised ICP or a venous air embolism (VAE) [7–9]. If associated with VAE, the bradycardia is accompanied by hypotension, the "mill-wheel" murmur from the precordial Doppler, and lowered ETCO2 indicating a decrease in pulmonary blood flow. Bradycardia caused by raised ICP is part of the Cushing's triad of bradycardia, hypertension, and abnormal respirations. Bradycardia in the absence of significant hypertension may be more likely due to a VAE. In the anesthetized patient receiving mechanical ventilation, only bradycardia and hypertension will be seen. The treatment of a suspected VAE includes notification of the surgeon, positioning the head below the level of the heart, flooding the operative field with saline, discontinuing N2O, treatment of the hypotension and bradycardia with epinephrine, and if needed chest compressions. Once the VAE has been pushed through the heart, the vital signs and ETCO2 normalize. Raised ICP severe enough to cause Cushing's response is indicative of imminent herniation and requires immediate treatment. Maneuvers to lower ICP include lowering the PaCO2 with judicious ventilator management, assuring free venous return from the head, administration of additional sedation, muscle relaxants, raising the head of the bed, and administration of mannitol or hypertonic saline [10, 11]. Decompressive craniectomy may be needed if these maneuvers are not successful in reversing the vital sign abnormalities [12, 13].

6. The surgeon expects that the case will be 45–60 min. Would you plan to extubate the child at the end of the procedure?

## Postoperative

### Question

1. Following extubation, the child is hoarse. What are possible etiologies? What is the appropriate management for this?

## Additional Topics

### Questions

1. A previously healthy 16-year-old male arrives in the emergency department 15 min after he was an innocent bystander in a convenience store robbery, held hostage, and shot in the mandible with a pistol, type unknown. Are there further studies you would like? Additional concerns? What if the surgeons want to go directly to the operating room? What are your extubation considerations?

6. The length of the procedure is only one of a number of factors that affect the decision whether or not to extubate this child. The degree of difficulty with airway management preoperatively is a consideration. In addition, the child is likely to remain in some sort of fixation device postoperatively and will now have just had a stabilization procedure on the cervical spine and might benefit from a period of complete immobility. Also, if during the case the vital sign instability was due to raised ICP, it may be important to continue with intubation until that problem has resolved. If, after consideration of the factors listed above, extubation is thought to be in the child's best interest, then it should be done with the child awake and responsive.

**Answer**

1. Hoarseness following intubation has several possible etiologies in this case. Many causes of respiratory distress in this child may also cause hoarseness without necessarily involving edema of the vocal cords themselves. If the endotracheal tube was too tight against the tracheal mucosa, there may be postintubation edema in the subglottic area, leading to both hoarseness and inspiratory stridor. It is possible that, during the fiberoptic intubation, the arytenoid cartilages were damaged or dislocated leading to hoarseness. In the preoperative assessment, some respiratory distress is noted. The cervical fracture itself may have damaged cervical roots that innervate the diaphragm (C3–C5), and the respiratory insufficiency is leading to hoarseness. Spinal cord injury without radiographic abnormality (SCIWORA) is a well-described entity in children. MRI and electrophysiologic testing such as SSEPs are used to determine the presence and extent of this type of spinal cord injury [14]. In addition to hoarseness, neurogenic pulmonary edema may occur.

**Answers**

1. Facial trauma of any type is of particular concern since, in addition to the problems that result from hemorrhage, there is significant likelihood of airway compromise. When a child or teenager is brought to the ED for evaluation following a gunshot injury to the jaw, evaluation of airway patency is essential. Radiographic evaluation is a very important part of the initial survey. The gunshot bullet or

2. What are your concerns about the patient with rhabdomyolysis? In what clinical setting does this occur? How would you treat? Are there specific anesthetic considerations?

3. How is a triage area organized to effectively deal with trauma? Are there commonly accepted triage categories? Are there special roles for anesthesiologists in triage care?

pellets not only will cause bleeding but, as foreign bodies, can lodge anywhere in the airway and inside the skull, orbits, or sinuses. If the patient is unstable, an airway must be established immediately, and the best option in this case would be a tracheostomy with local anesthesia. If the patient is stable and able to breathe with relative comfort, further evaluation can be done prior to securing the airway and inducing anesthesia. A brief survey for other injuries can be done as the patient is prepared for surgery and anesthesia. Portable radiographic studies can be done, and if the patient shows no signs of deterioration, an urgent CT scan of the head can be performed. If a CT is done, the anesthesiologist and ORL surgeon should accompany the patient to radiology, prepared to intervene with emergency airway support if needed. Additional IV access can be secured and an arterial line placed, an antisialagogue administered, and appropriate analgesic and anxiolytic administered as the patient is taken to the OR. The airway should be secured with either a tracheostomy or fiberoptic bronchoscopy based on the result of the evaluation and consultation with the surgeons. If it is likely that the jaw will be wired shut postoperatively, this will affect planning for extubation at the conclusion of the case if a tracheostomy was not done. Another consideration that will affect airway management will be the degree of airway edema anticipated in the postoperative period. If extubation is planned, it will be important that the patient be awake and following commands prior to extubation. Antiemetics should be given and the stomach emptied before removing the endotracheal tube.

2. Children who have suffered extensive musculoskeletal injuries are at risk for myoglobinuria. If this occurs, renal dysfunction is possible. The patient should have the urine alkalinized and urine flow maintained with vigorous IV fluid administration and administration of diuretics if necessary. Another complication occasionally seen in children who have had extensive long bone trauma with multiple fractures is fat embolism, although this problem is not generally seen immediately following the injury.

3. Organization of the triage area for trauma depends upon the type of patients expected by the facility. Trauma centers are given one of three designations depending upon the medical services available [3]. Trauma centers devoted specifically to children were first created in the 1970s at several major pediatric medical centers. The organization of a triage area at a particular center will depend upon the resources available at that location. Level I trauma centers have the largest range of services and medical personnel available, while Level III centers provide stabilization of trauma victims prior to transport to centers with more extensive resources.

    - Level I: These centers have immediate availability of trauma surgeons, anesthesiologists, nurses, physician specialists, and resuscitation equipment. These centers treat 240 major trauma patients/year.

4. Describe the advantages/disadvantages of various locations for vascular access.

- Level II: These have similar requirements for the availability of personnel as Level I trauma centers, but there are no requirements for the number of cases/year.
- Level III: These centers can evaluate and stabilize trauma patients prior to transfer to Level I or II centers. Emergency surgery is available.
- Trauma centers for children have been characterized in the University of Michigan Pediatric Trauma Classification System into three levels:
- Level I: These centers treat children with single or multisystem injuries, unstable vital signs and respiratory distress, shock, neurologic injury, or gunshot wounds or burns.
- Level II: These centers treat children with multisystem injuries and stable vital signs such as children with open fractures, less severe burns, or less severe neurologic injuries with stable GCS.
- Level III: These centers treat children who are conscious with an isolated injury and low potential for multisystem injury.
- Individuals are assigned to triage categories as follows, in decreasing order of urgency:
- *Urgent*. Life-saving intervention is indicated if death is to be prevented. If initial resuscitative interventions are successful and some degree of stability is achieved, then the urgent casualty may revert to a lower priority.
- *Immediate*. Severe, life-threatening wounds that require procedures of moderately short duration; high likelihood of survival. They remain temporarily stable while undergoing replacement therapy and further evaluation.
- *Delayed*. Able to be supported and evacuated. May go without surgery for several hours, after which there will be a direct relationship between time lapse and complications.
- *Minimal or Ambulatory*. Superficial wounds requiring no more than cleansing, minimal debridement under local anesthesia, tetanus toxoid, and first aid type of dressings.

4. Vascular access is always an important consideration in trauma patients but particularly so in pediatric trauma victims [3]. Central access is important in patients who have suffered more severe trauma. In children, the internal jugular or subclavian veins are not good choices for a variety of reasons. Pediatric trauma victims are generally in cervical collars, limiting access to these sites. Even if the cervical spine was not involved in the accident and there is no worry about its stability, there remains a significant risk of pneumothorax or hemothorax from placement of CVLs into the internal jugular or subclavian veins. Cannulation of a femoral vein is a good option for central access in pediatric trauma patients. However, the risks of delay in establishing access in the unstable patient are real. Intraosseous lines can be readily placed even in very small trauma victims. The safest location is the medial surface of the proximal tibia. There are a variety of needles in different sizes for placement in all ages of children.

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## *Annotated*


Head TRC, trauma s c. Chap. 56. In: Nichols DG, editor. Rogers' textbook of pediatric intensive care. Philadelphia: Wolters Kluwer Lippincott Williams and Wilkins; 2008. p. 887–911.

This chapter reviews the epidemiology and pathophysiology of various types of CNS traumatic injury such as blunt and sharp trauma and compression injury. Also included is a discussion of clinical, laboratory, and imaging assessment of patients with CNS trauma, management of raised ICP and spinal injury, and review of the available outcome data

Baird JS, Cooper A. Multiple trauma.Chap. 27. In: Nichols DG, editor. Rogers' textbook of pediatric intensive care. Philadelphia: Wolters Kluwer Lippincott Williams and Wilkins; 2008. p. 384–407.

This chapter is a good survey of trauma in children with emphasis on the initial assessment and stabilization of trauma victims. The emphasis is on non- neurologic injuries

# Chapter 34
# Burns


**Joseph P. Cravero and Robert S. Holzman**

A 3-year-old is rescued from a burning apartment after hiding under the bed. He has a 55% burn, primarily below the knees and above the waist, including the face, and around the chest. He is short of breath, tachypneic, with a blood pressure of 130/90, a heart rate of 160, and a temperature of $39.4^0$ C. He has a headache, is restless, and somewhat confused. You are his ICU doctor.



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## Initial Evaluation

### Questions

1. How do you determine whether this patient should be intubated? Are there physical signs that would help in this evaluation? Chest X-Ray? Arterial blood gas? Would steroids be of any help in decreasing airway edema?

2. What toxic products of combustion would you be concerned about? How can you evaluate the clinical significance? What will you do? Would prophylactic antibiotics be of any help in this case? Why? What are the important considerations for the management of carbon monoxide poisoning?

## Answers

1. Because the child was burned in an enclosed environment, sustained facial burns, and is tachypneic, he probably has thermal injury to the airways and alveoli from inhalation of smoke, noxious gases, and heated air. This child's trachea should be intubated prophylactically in anticipation of rapid swelling of the upper airways and respiratory tract. With time, edema and secretions/blood in the airway can make laryngeal access difficult or impossible.

   The chest X-ray may not be very helpful in the early stages of the pulmonary thermal injury. Arterial blood gas analysis may show lactic acidosis. It is not useful in diagnosing carbon monoxide poisoning because it measures dissolved oxygen and thus overestimates the oxygen saturation of hemoglobin. Unless the analyzer is specifically measuring carbon monoxide (CO), it will estimate the saturation based on the PaO2. Partial pressure of oxygen (PaO2) levels remain normal; oxygen saturation is accurate only if directly measured but not if calculated from PaO2, which is common in many blood gas analyzers. Similarly, the pulse oximeter overestimates the oxygen saturation because it cannot differentiate oxyhemoglobin from carboxyhemoglobin. Saturations will generally read in the 88–90% range even if the true saturation is much lower. Carbon monoxide poisoning is suspected in burn victims when symptoms of headache, dizziness, restlessness, and confusion are present. A co-oximeter or a handheld breath analyzer can be used to confirm the diagnosis of carbon monoxide poisoning. Steroids have not been shown to be helpful with the massive inflammatory process of burns.

2. Carbon monoxide gas is of most concern because it is colorless and odorless and is produced in large quantities during incomplete combustion of carbon or carbon products and fuels. The clinical significance is that carbon monoxide poisoning can produce severe hypoxemia and long-term neurological impairment. Carbon monoxide gas has 200–250 times the affinity of oxygen for binding with hemoglobin. Carboxyhemoglobin shifts the oxyhemoglobin dissociation curve to the left. As a result, oxygen delivery is seriously compromised. The primary treatment of carbon monoxide toxicity is administration of 100% oxygen by a non-rebreathing system. Elimination of carboxyhemoglobin is dependent on alveolar oxygen pressure rather than alveolar ventilation. The idea is to provide a huge amount of oxygen so that it can compete with CO for binding sites on Hb. In severe cases (carboxyhemoglobin >30%), oxygen can be provided via positive pressure ventilation or (preferably) in a hyperbaric chamber. The carboxyhemoglobin half-life can be reduced from 4 hours in room air to 90 minutes with the administration of 100% inspired oxygen. Prophylactic antibiotics can be helpful to treat opportunistic infection but should not be given in indiscriminately. They are not part of the primary treatment for this problem.

3. What are your initial considerations in volume support? Is it safe to give so much fluid initially? What formula would you use to calculate volume replacement? Is there a need to modify this formula in this case? Is it likely that this hemodynamic picture will change? Over what period of time will change occur? Why is this patient hyperdynamic?

## Initial Critical Care Management

### Questions

1. What special considerations do you have for mechanical ventilation of this patient? Metabolic considerations? Mechanical lung considerations?

2. What are the nutritional needs of the burn patient? When should hyperalimentation begin? Should it be central, peripheral, or enteral? Why choose one over another?

3. Large volume resuscitation is necessary initially and can be guided by the Parkland formula [crystalloid 4 mL/kg × percent burn × wt. (kg)] or the Brooke formula [crystalloid 0.5 mL/kg + colloid 1.5 mL/kg × percent burn × wt. (kg)]. "Moderate" and "major" burn criteria require less of a total body surface area burn in the very young and very old when compared to normal adults. In addition, the percentage of body surface area for the head and trunk areas is different for infants and toddlers vs. adults. These formulae are useful guides to the replacement of massive fluid loss but the overall requirement is determined by clinical monitoring of the patient's mental status, hemodynamic parameters, acid-base balance, and urine output. Nevertheless, these formulae may underestimate fluid requirement in infants under 10 kg. There is emerging interest in using dynamic hemodynamic parameters to guide volume resuscitation, such as pulse pressure variation when an indwelling arterial line is in place, transesophageal Doppler estimates of stroke volume, or transesophageal echocardiography to assess left ventricular cavity size in order to determine ventricular filling [1]. Some burn centers use hypertonic saline or colloids particularly in the very young and elderly to minimize the potential of edema. To date the controlled trials have not demonstrated a difference in outcome or mortality among the different types of solutions. Usually, the fluid losses from inflamed surfaces continue for days and weeks depending on the extent and severity of the burns. The hyperdynamic circulation is due to a massive surge in catecholamines and corticosteroids, 10–50 times that of a normal patient. Injury-induced cytokines and endotoxins released into the circulation further perturb the hemodynamics of these patients. Elevated metabolic rates also compensate for the large amounts of heat and water lost through disrupted tissues.

## Answers

1. The minute ventilation requirements are greater in order to eliminate the increased carbon dioxide production and meet the increased oxygen demand resulting from the hypermetabolic state. High positive end-expiratory (PEEP) and inspiratory pressures may be necessary for effective oxygenation in the presence of pulmonary edema from the thermal burn of the alveoli and airway passages. In addition, a restrictive lung defect with impaired ventilation can result from circumferential burns to the chest wall or, eventually, from scar restriction [2]. Chest wall escharotomies may be necessary.

2. The hypermetabolic state is associated with increased utilization of glucose, fat, and protein and that in turn leads to greater oxygen demand and increased carbon dioxide production. Proteins and amino acids are mobilized to meet the immense metabolic demands and energy requirements resulting in significant loss of lean body mass that can impair immune function and wound healing. Hyperalimentation

3. What are the consequences of a 55% total body burn on thermoregulation? Of what importance is that in your ICU management? How do you compensate for increased thermogenesis and increased heat loss? Will occlusive dressings or raised ambient temperature alter this caloric loss? How do you compensate for increased free water loss? To what extent is electrolyte loss a component? How do you compensate for that?

4. If you were a nephron, how would your physiology be altered as a result of a burn injury? What is the effect of decreased cardiac output, increased endogenous catecholamine activity, and decreased splanchnic blood flow on renal sodium conservation? Why? Are there any consequences to antidiuretic hormone elaboration? What are they? How do they affect renal function?

5. Twelve hours after admission the patient develops diffuse petechiae associated with bleeding at line sites. How would you evaluate? Why do you get this picture? Would fibrin split products analysis be of any help? You do a factor analysis and factors V and VII are actually elevated—does that matter? Why? What kind of support is necessary in this circumstance? Would whole blood help?

is started soon after the initial fluid resuscitation and stabilization of the hemodynamics. The hyperalimentation fluid is administered via a central line. Central access allows the administration of higher concentrations of glucose and other nutrients than peripheral access. In addition, it is difficult to secure peripheral venous access when all extremities are involved in the burn injury and for a prolonged infusion time. Patients with severe burns develop gastric stress ulcers and ileus. As a result, enteral absorption is ineffective for meeting the high nutritional requirements of burn patients.

3. A 55% skin burn produces extensive destruction of body surface and significant loss of body heat and regulatory function, leading to serious hypothermia. To prevent hypothermia, it is crucial to cover the patient's body, elevate the ambient temperature, use radiant warmers, and warm the inspired respiratory gases and intravenous fluids. Occlusive dressings and insulating blankets are more effective in preserving heat, minimize evaporative losses, and reduce caloric requirement. Nursing of open wounds in a raised ambient temperature will enhance evaporative water and electrolyte loss.

4. Nephron function is adversely affected immediately after the burn by hypovolemia, hypotension, hypoxemia, myoglobinuria, and hemoglobinuria leading to acute tubular necrosis. Other contributing factors are stress-related release of catecholamines, angiotensin, vasopressin, aldosterone, and endothelin-1 that cause systemic vasoconstriction, hypertension, and impairment of renal function. Therefore, fluid retention occurs during the first 5–7 days after the burn injury with diuresis thereafter.

5. The diffuse spontaneous bleeding is a presumptive diagnosis of disseminated intravascular coagulopathy (DIC) until proven otherwise. Extensive burns and intravascular hemolysis can introduce tissue phospholipids into the bloodstream resulting in activation of the "extrinsic" clotting system. To confirm the possible diagnosis, I would send a sample of the patient's blood for coagulation studies. A PT of >15 seconds, fibrinogen concentration less than 160 mg/dL, and a platelet count of less than 150,000 $mm^3$ raise concern for a consumptive coagulopathy. Platelet consumption is the most sensitive indicator of DIC. The presence of high circulating levels of fibrin split products and D-dimer concentration is indicative of a secondary fibrinolytic process that accompanies the thrombosis during DIC. Initially, factors V and VIII could be elevated in response to burn stress. The most important component of the therapy is to maintain an adequate circulating volume. Since the primary cause of the DIC cannot be reversed, the consumed blood factors (factors V, VIII, fibrinogen, and others) should be replaced by transfusion of cryoprecipitate, FFP, and platelets. The use of whole blood depends on its shelf-life. Fresh whole blood contains a significant amount of coagulation factors, but un-refrigerated whole blood usually does not contain adequate coagulation factors and platelets to replace the high requirement during DIC.

## Anesthesia Consultation

### Questions

1. When called to intubate this patient, an anesthesia resident asks you if he should do a rapid sequence induction—what is your answer? What about using rocuronium? Any special considerations for burn patients with regard to nondepolarizing neuromuscular blocking agents? What if this is the first day post burn and the patient is going for escharotomy?

2. How can you keep a burn patient warm in the OR? Any special considerations? Is this different from a non-burn patient?

## Additional Topics

### Questions

1. An electrical worker reached for a capacitor and it shorted through his arm. Other than excruciating pain, he actually looks like he only has a little superficial puncture wound. The surgeon is very worried and wants to rush him to the OR for exploration, fasciotomy, angiography, and debridement. Why is he making such a big deal out of it?

## Answers

1. I would discourage the resident from performing a rapid sequence induction without carefully assessing the airways. Inhalation of smoke and toxic fumes may cause severe swelling of the oral mucosa, tongue, and glottis that can make intubation very difficult. Tightness of the submandibular space, a reflection of swelling of the tongue base, can make laryngoscopy difficult. Although succinylcholine is generally safe to use within the first 24 h after the burn injury, even a small acute rise of serum potassium in burn patients can produce serious arrhythmias and cardiac arrest because skeletal muscle burns and intravascular hemolysis produce a rapid rise of baseline serum potassium. Rocuronium is safe to use provided there is no evidence of direct oral and respiratory tract burns, e.g., inhalation of live steam. A severe intrathoracic injury will make ventilation that much more difficult when you switch to positive pressure ventilation. Concerning the use of nondepolarizers, it is well recognized, for reasons not well understood, that burn victims are resistant to nondepolarizing muscle relaxants and hence larger doses must be used to achieve adequate intubation conditions. This resistance develops about a week after the injury and is proportional to burn size.

   From 24 hours after the burn until the wounds are completely healed, succinylcholine should not be used because of upregulation of cholinergic receptors at the neuromuscular junction and extrajunctional receptors on the skeletal muscle membrane. Therefore, administration of succinylcholine may produce exaggerated muscle contractions and a rapid rise in serum potassium.

2. Burn victims tend to develop hypothermia in the OR because of the hypermetabolic state with rapid evaporative fluid losses from the exposed burn surfaces. To minimize heat loss in the OR, the room should be heated to 28–30 degrees C, intravenous solutions should be heated, and the inhaled anesthetic gases should be humidified and heated.

## Answers

1. Electrical injuries produce minimal skin injury but massive internal tissue injury. The current spreads through the neurovascular bundles and produces extensive depolarization and thermal burn of the nervous tissue, cardiac and skeletal muscles, and all soft tissue in general. As a result, there is a massive fluid shift, myonecrosis (rhabdomyolysis, compartment syndrome), and hemolysis (vascular

2. A 14-year-old boy was standing in a cornfield when he was hit by lightning. He suffered a cardiac arrest, but he fortunately was resuscitated from successfully because an EMT was on site. What kind of burn injury is he likely to suffer? How is this different from other electrical burns? Which is more lethal?

3. A high school chemistry student is brought in with burns on his arms from white phosphorus powder. How do you treat this chemical burn? What is your general approach to chemical burns? How do you assess when the airway is of significant concern?

spasms, thrombosis). The electricity involves the flow of energy (electrons) along the path of least resistance toward a natural ground. All tissues are either resistors (skin and bone) or conductors (neurovascular bundles, soft tissue). Therefore, deeper soft tissues such as the nervous system, cardiac and skeletal muscles, gastrointestinal tract, and vascular system are at high risk for electrically induced depolarization and thermal burns.

2. Lightning may cause complete cardiac arrest by inducing either asystole or central apnea because of the delivery of massive macroshock. Extensive depolarization of the heart muscles leads to asystole and massive depolarization of the brain can result in apnea. This patient may have had transient asystole, which may account for prompt response to resuscitation. Therefore, lightning burn is more detrimental than standard household current. The lightning burn involves a single massive current impulse that is approximately equivalent to a DC burst of 2000 to 2 billion volts of an extremely short duration of 0.1–1 ms.

3. Remove the patient's clothes and footwear to avoid further contact with white phosphorus. Irrigate or apply saline-soaked or water-soaked pads to the affected skin to wash off the offending agent and stop further oxidation of the phosphorus. Phosphorus is a lipophilic agent and therefore tends to bind to fatty tissue, and upon oxidation, it produces an exothermic reaction resulting in second and third degree burn injury. Avoid the use of any greasy solutions to irrigate and cleanse the affected area because it will enhance binding it to the tissue. Avoid contact with the patient's clothes contaminated with white phosphorus because it is ignitable and can cause chemical burn injury to the patient or the healthcare provider. Debride lacerated, devitalized, or contaminated tissue. Visualization of phosphorus is aided with a Wood's lamp (ultraviolet light) by fluorescing the white phosphorus [3, 4].

   The general approach to all chemical injuries is to remove the offending agent because the injury becomes progressively more severe as long as the offending agent is in contact with the skin. Victims must promptly remove clothing and footwear in contact with the chemical, must brush away dry chemicals, and must apply water lavage to dilute and remove the chemical. Some chemicals are insoluble in water, and other chemicals create exothermic reactions when combined with water. For example, dry lime contains calcium oxide, which reacts with water to form calcium hydroxide, an injurious alkali. Therefore, dry lime should be dusted off the skin prior to washing with water. Phenol is water insoluble, and it should be wiped off the skin with sponges soaked in a 50% concentration of polyethylene glycol. Concentrated sulfuric acid often causes an exothermic reaction when combined with water; therefore, soap or lime should be used first to neutralize the agents. The use of neutralizing agents is the exception to the rule because the key to effective treatment is dilution. Some antidotes may produce exothermic reactions and thereby increase toxicity. Some have advocated the use of turpentine for white phosphorus burns.

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## *Suggested Reading*

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# Chapter 35
# Behavioral Issues

**Thomas J. Mancuso**

A 5-year-old with autism spectrum disorder (ASD) is scheduled for a dental exam, cleaning, and possible extractions under anesthesia. He is uncooperative at the pre-anesthetic visit. You have records from his pediatrician documenting his generally good health with no cardiac, renal, pulmonary, or liver insufficiency. He is in main-streamed kindergarten at the local public school and has an Individualized Education Program (IEP). He has been treated with risperidone for the past year.



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## Case 1 Management

### Questions

1. In this situation, can the routine preanesthetic evaluation be shortened in order to minimize troubling the child? Is a physical exam necessary other than observation? Are preanesthetic vital signs necessary?

2. Would you prefer that the child receive his usual morning dose of risperidone?

## Answers

1. Autism disorder (AD), or autism, is the most common pervasive developmental disorder (PDD) with a recently estimated prevalence of 13 per 10,000. The etiology of AD is still unknown, although genetic factors are probably involved, and in 5–10% of cases, there is an identifiable associated known medical condition. The onset of autism disorder has been set at before the age of 3 years, and other autism spectrum disorders (ASDs) may have a later onset. The prevalence of Asperger's disorder is approximately 3 per 10,000, and childhood disintegrative disorder is very rare, estimated at 0.2 per 10,000. Rett disorder prevalence is 1 per 15,000. The prevalence of all PDDs in recent surveys is about 60 per 10,000. Behavioral disturbances are fairly common in these disorders and are very often challenging to treat. Tantrums, aggressive behavior, and overactivity/hyperactivity are frequent from the early phases and may last throughout adulthood, causing serious problems in adaptation. The severity and the development of the various symptoms and their clinical features vary on an individual basis [1].

   Records from the primary pediatrician are sufficient for the preanesthetic visit. The vital signs obtained with the child resisting and struggling will not reflect his usual state of health in any case. If these records are not available, it still may not be useful to restrain the child and force him to have BP, HR, and RR documented. With patience and engaging in play with the child, it may be possible to obtain an oximeter measurement.

2. Risperidone is generally given BID. Unless this presents an excessive burden to the family, usually the morning dose should be given to the child. Moderate efficacy and safety of risperidone for treating maladaptive behaviors, including aggression, hyperactivity, self-injury, and irritability, have been documented in the available studies. Two studies also found some degree of improvement in some of the core features of ASD. Risperidone was promising in preschoolers with ASD also combined with behavioral interventions. Efficacy and tolerability of risperidone in the various types of PDDs, including the different degrees of core symptoms, from mild to severe, are still undetermined and should be appropriately addressed. At present much caution is therefore warranted in this vulnerable population that raises additional concerns and that needs continuous care, especially when receiving pharmacotherapy [1].

   Risperidone doses varied from 0.1 to 0.5 mg/day, and all the studies started with low doses that were increased slowly. The initial dose was 0.25–0.5 mg/day once or twice daily, with increments of 0.25 mg or 0.5 every 3–7 days until a therapeutic response was reached. Therefore, a flexible schedule of dosing is advisable to coincide with characteristics of the child and to minimize unwanted side effects.

3. What premedication would you choose? Why?

4. What is your induction plan?

5. What are your plans for maintenance of general anesthesia?

The Cochrane database review reported possible benefit but noted the generally limited amount of data [2].

Weight gain was the most frequent adverse event, ranging from 1 to 10 kg. Weight increase usually stabilizes over time, but it is more pronounced during the first 2–3 months of therapy. Several potential long-term health risks arise with weight gain, such as hypertension, heart disease, diabetes, and dyslipidemia. Sedation is another common side effect, but in most studies, it is usually referred to as transient [1].

The US Food and Drug Administration (FDA) has approved risperidone and aripiprazole for treatment of irritability associated with autism in children and adolescents. Despite their efficacy, the use of these medications is limited by their side effects. In individuals with severe irritability, the first-line treatment is often risperidone. Because of its relatively lower risk of weight gain and metabolic side effects, aripiprazole may be used initially if there is a personal or family history of obesity or diabetes. Monitoring of body mass index and metabolic profiles is indicated with both medications [3].

3. This is based on the situation at the bedside in the preanesthetic area. The child is almost certainly going to be very anxious and would benefit from an effective and generous premedication. Oral administration is preferred, but, of course, that requires the child's cooperation. If she or he is willing to take a PO medication, then midazolam with the possible addition of ketamine will help alleviate the child's anxiety and facilitate the induction of general anesthesia. If, on the other hand, the child refuses PO medications and a premedication is considered essential, the IM route of administration is an option. Midazolam, ketamine, and possibly glycopyrrolate as an antisialogogue can be given. Prior to the administration of ketamine, it is important to be ready to manage the patient's airway, in the unlikely event that airway obstruction occurs, as well as to have the OR completely ready for the induction of general anesthesia.

4. An inhalation induction is very likely to be the safest and easiest technique, even after administration of an IM premedication. For safety, only sevoflurane and oxygen should be used. As soon as the child is safely anesthetized, a peripheral IV should be started. The first IV should be of any size. If the case demands large and/or additional IVs or invasive monitoring lines, these can be placed later once induction is complete and the airway secured.

5. Maintenance should be planned to assure excellent conditions for the surgeon/operator while simultaneously assuring stable, normal vital signs and unconsciousness of the child. A combination of inhaled agents and neuromuscular blockade is the basis of the anesthetic. Analgesia, as needed, can be provided with acetaminophen; nonsteroidal medications, if considered safe; and opioids. Additional medications that will help with emergence include antiemetics such as dexamethasone and ondansetron and alpha 2 antagonists such as dexmedetomidine and/or clonidine.

6. The dentist performs two extractions and there is some bleeding, controlled with pressure. She does not think overnight admission is indicated for the child. Once the case is completed, how will you manage emergence from anesthesia?

7. In the PACU, he immediately pulls out his IV. What would be safe post-op analgesia for the child?

## Additional Questions

### Questions

**Case 2** You meet an anxious 15-year-old young lady in the pre-op holding area. She is scheduled for laparoscopic surgery for endometriosis and has never undergone anesthesia. During your interview she is tearful and reports great anxiety about awareness under anesthesia. Her father reported to his family that he recalls the sternal saw during his CABG several years ago, and he still is troubled by that memory.

1. Are her worries justified? How will you reassure her? Are there specific techniques associated with a greater chance of awareness than others?

6. Even if there were no surgical troubles such as the bleeding noted here, the goal should be the smoothest emergence possible. In this particular case, with unexpected bleeding, it must be assumed that the child is at significant risk for aspiration. With this consideration, a deep extubation is not as safe as an awake extubation. The inhaled agent desflurane, with its significantly shorter elimination half-life, has that advantage in allowing a rapid return of the awake state prior to extubation. An infusion of dexmedetomidine, begun at least an hour prior to the conclusion of the procedure, will help keep the child calm and therefore safe at extubation.

7. Prevention of this mishap is, of course, the best option. This can be done with the use of "welcome sleeves" and/or wrapping the limb with the IV loosely with a Kling conforming dressing. Second would be replacing the peripheral IV, but if that is considered too stressful for the child, the only routes available for the child are oral and parenteral. Subcutaneous MSO4 is an option that will likely be less painful than IM administration. If IM administration is chosen, in view of the bleeding noted, it is best to avoid ketorolac and administer an opioid by this route. If the child is willing and able to take oral medications, an option is PO acetaminophen ± an opioid.

## Answers

### Case 2

1. Her concerns are important to her and should be validated. At the same time, she should be reassured of the rarity of such intraoperative awareness and of your diligence in caring for her. In general, adolescents can understand abstract reasoning. Taking time to hear her concerns and address them with knowledge and sensitivity can result in alleviation of her fears.

   A review of reported cases of intraoperative awareness compared the data of 271 cases of awareness with 19,504 patients who did not suffer it. The authors of the review performed an electronic search of the literature in the National Library of Medicine's PubMed database for case reports on "awareness" and "anesthesia" for the time period between 1950 and August 2005. The authors made the following comments: aware patients were more likely to be females ($P < 0.05$), to be younger ($P < 0.001$), and to have cardiac and obstetric operations ($P < 0.0001$). Patients who suffered intraoperative awareness received fewer anesthetic drugs ($P < 0.0001$) and were more likely to exhibit episodes of tachycardia and hypertension during surgery ($P < 0.0001$). Nearly one-fourth of the patients suffered late psychological symptoms [4].

2. Will you premedicate her? Is PO midazolam as effective in providing amnesia as IV midazolam? Would ketamine be a more effective choice in this situation?

3. If you had seen her the day prior to surgery in a preanesthetic clinic, would you have prescribed PO diazepam to be taken prior to her departing for the hospital?

**Case 3**

You are a director of the PACU at a children's hospital and, along with the head RN, are developing a management plan for children with emergence delirium. Define emergence delirium. Is this phenomenon more common in children compared to adults? How would you evaluate a child in the PACU who you think may be exhibiting emergence delirium?

2. Midazolam is the most commonly used benzodiazepine in pediatric anesthesia. It can be administered orally, nasally, and rectally, as well as intravenously and intramuscularly. Nasal administration is quite uncomfortable, however, while IM administration is not as painful as IM administration of diazepam. The clinical effects include anterograde amnesia in approximately one-half of patients as well as sedation and anxiolysis. Midazolam also produces a calming effect with some minimal sedation. When using midazolam in combination with other potent CNS depressants, significant depression of respiratory drive may occur. IV midazolam depresses the response to hypoxemia, an effect that is exaggerated in the presence of a potent opioid. Peak CNS effect with IV midazolam is approximately 5 min. This means that additional doses of midazolam if given too rapidly lead to excessive effects after 10 min or so have passed. Children with sleep-disordered breathing who were premedicated with 0.5 mg/kg PO midazolam experienced only a small incidence (1.5%) of transient decreased SpO2. In contrast administration of 0.1 mg/kg midazolam through an IV has resulted in central apnea as well as upper airway obstruction. Ketamine has many disadvantages in this situation. In addition to increased oral secretions, ketamine can cause dysphoria, unpleasant sensory experiences, and, over the subsequent 24 h, troubled sleep.

3. Administration of sedative/anxiolytic medications should only be undertaken when the patient is directly in your care. Administration of midazolam, as safe as it is, should not be done when the patient is not in the care setting. If you chose to undertake administration of an oral premed in the hospital lobby, it would be of the utmost importance to have obtained a very recent negative pregnancy test. It may be that the family would appreciate it if you were to meet them in the lobby and, with carefully hidden resuscitation equipment and an O2 tank, administer a dose of PO midazolam and then escort the family to the preanesthetic area.

**Case 3**

Although it is difficult to quantify the aspects of the diagnosis such as vital signs, VAS, etc., emergence delirium is a state recognized by pediatric anesthesiologists and pediatric PACU RNs. It is characterized by inconsolability, thrashing, and incoherence. Prior to making the diagnosis, however, caregivers must do their best to assure adequate analgesia for children exhibiting these behaviors.

No specific technique has been shown to minimize the incidence of emergence delirium. As mentioned, these behaviors have numerous etiologic factors including specific anesthetic agents, pain, hypothermia, hyperthermia, hypoglycemia, hyponatremia, prior neurologic disturbances, a behavioral response to sudden awakening in a strange environment, separation anxiety, airway obstruction with resultant hypoventilation and hypoxia, and combinations of these factors. The difficulty in making the diagnosis makes ascertaining the relationship of these behaviors with anesthetic techniques, the conduct of the induction of anesthesia, and/or patient characteristic challenging.

New less soluble, inhaled agents such as sevoflurane do allow for faster emergence from anesthesia, but unfortunately emergence excitation or delirium seems to be more common than the incidence seen with the use of the older (and less safe) agent halothane. Again, in the context of the difficulty with diagnosis, various agents have been used with some success, including opioids, serotonin antagonists, and α-adrenergic agonists.

Regarding the relationship between patient characteristics and emergence delirium, anxious children have been shown to experience significantly more pain both during their hospital stay and over the first 3 days at home and also had a greater incidence of emergence delirium (9.7 % vs. 1.5 %) [5, 6].

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Costi D, Cyna AM, Ahmed S, et al. Effects of sevoflurane versus other general anesthesia on emergence agitation in children. Cochrane Database Syst Rev. 2014;12:CD007084. https://doi.org/10.1002/14651858.CD007084.pub2.

A review of the prevalence of agitation after a general anesthetic as well as evidence relating to a sevoflurane general anesthetic compared with other general anesthetics. Also reviewed is evidence that other treatments or parental presence at emergence may affect agitation.

Raviola GJ, Trieu ML DeMaso DR, Walter HJ. Chapter 30 Autism spectrum disorder. In: Kliegman RM, Stanton BF, St. Jeme III JW, Schor NF, editors. Nelson textbook of Pediatrics. 20th ed. Philadelphia: Elsevier; 2016.

A concise review, in 8 pages, of this challenging problem in pediatrics.

# Chapter 36
# Adolescence


**Robert S. Holzman**

Three teens arrive in the emergency room after being transported by ambulance from the same party.

*Patient 1*. The first, a 17-year-old male, captain of the football team, reeks of alcohol, is stuporous and has a left-sided ankle fracture, tib-fib fracture, and Colles' fracture. He is banged up and bruised throughout, including his face, but has no other fractures. He vomited twice in the ambulance on the way to the ED. VS: HR = 110, BP = 140/90, RR = 28. SpO2 = 97%. Height 6 feet; weight 85 kg.

*Patient 2*. His best friend, a 17-year-old offensive lineman on the football team, was shot in the abdomen at close range by a party crasher. He is not complaining of abdominal pain but does have pain in his neck and right arm. He is in a Miami J-collar. He is also actively hallucinating when he is not in pain. VS: HR=123, BP = 180/110, RR = 26. SpO2 = 97%. Height 6 feet 3 inches; weight 135 kg. He was given 3 mg of hydromorphone in the ambulance on the way. A tox screen in the ED is positive for opioids, cannabinoids, and amphetamine. His blood alcohol level is 0.15.

*Patient 3*. The football captain's 16-year-old girlfriend was punched in the face when she tried to break up the fight. She has a displaced mandibular fracture and an ipsilateral orbital fracture. Her HCG just came back positive in the ER. VS: HR = 120, BP = 140/92, RR = 42. Height 5 feet 4 inches; weight 45 kg. She is in a soft cervical collar. She admits to taking an unknown amount of methylenedioxyamphetamine (ecstasy) 2 hours earlier.



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## Preoperative Evaluation

### Questions

*Patient 1*. How would you like to work this patient up further? What are his risks right now? Is it important to determine his blood alcohol level? Why? How will you decide whether he can protect his airway? Do you have to wait until he is sober to go to the operating room? Is there anything about his situation for which he needs immediate surgery? What if he needs a head CT scan or MRI—what are your considerations?

*Patient 2*. The emergency medicine physicians would like to administer ketamine 1 mg/kg (the upper limit of the dose they are allowed to administer as procedural sedation) in order to obtain imaging studies of the head, chest, and abdomen. They call to give you a "heads-up" but don't want to bother you in the operating room because "you will probably be seeing this kid later." Your thoughts? Your advice? Why is he hallucinating? What is the significance of his vital sign abnormalities? Do they have any specific implication for his "sedation?" What is your anesthetic plan, assuming you would like to anesthetize him for these diagnostic and possibly therapeutic procedures? Does he need a rapid sequence induction of anesthesia? What are your concerns? What is the typical intoxication level for blood alcohol? Is this a national standard? How does it influence your anesthetic plan? How do you interpret his pain complaint with regard to his gunshot wound entry?

## Answers

*Patient 1*. Concurrent injuries are a significant concern so a thorough primary and secondary survey is warranted, including imaging studies to rule out intracranial, intrathoracic, or intra-abdominal injury. His stupor may be attributable to his intoxication but in order to be sure, it is prudent to obtain a blood alcohol level and a toxicology screen for other substances as well. However, his stupor can also represent a head injury, especially in the presence of vomiting. With attendance at a party, he is likely to have a full stomach, and if protective airway reflexes are impaired for any reason, it would be prudent to protect his airway during any diagnostic studies. If he has straightforward fractures without any other injuries, he will likely have an optimal surgical result if the time to the operating room is minimized because of local edema following injury; otherwise, he will have to wait at least several days for the swelling to decrease. His vital signs seem reasonable for someone in pain.

*Patient 2*. The disparity between the gunshot wound point of entry and the patient's symptomatic complaint (although perhaps incoherent because of drugs or other factors) is immediately worrisome. Referred pain in the neck and right arm could represent diaphragmatic injury and therefore, with an abdominal entrance wound, liver injury, which can be life-threatening. In addition, he is hypertensive and tachycardic, so hemoperitoneum as well as intrathoracic and major vascular injuries need to be considered. The rigid J collar is a challenge for airway maintenance and tracheal intubation because by design it is intended to provide firm cervical spine fixation. It has a hole in the tracheal area, but that said, it does not make intubation of the airway any easier. If the collar needs to be kept in place, then a videolaryngoscope would seem like a good option in order to improve the chances of visualizing the airway while maintaining neck neutrality. The other possibility would be removal of the J collar with in-line stabilization and then replacement of the collar, if possible and compatible with the scheduled procedure, following tracheal intubation. The hallucinations suggest a significant psychotropic effect from the cannabinoids and amphetamine, although the patient could be delirious for a variety of reasons including intracranial injury and hypoxia. He is hypertensive, tachycardic, and tachypneic which may be drug effects combined with the very considerable stress response, increase in oxygen consumption and carbon dioxide production, and any degree of pain. He already received a large dose of hydromorphone, the equivalent of over 20 mg of morphine, without much of an effect. However, with the induction of anesthesia, the effect of the hydromorphone may be magnified. His blood alcohol level is over twice the legal limit in most states. It is unlikely that 1 mg/kg of ketamine will be effective in providing motionlessness for the imaging studies required, and the lack of airway protection is a major consideration for patient safety as well. Similar anesthetic considerations for securing the airway apply to this patient as well; he is combative and should be treated as a full stomach,

*Patient 3*. The maxillofacial surgeon and ophthalmologist both agree that the patient should go to the OR expeditiously to have the mandibular fracture put into fixation before it gets too swollen to obtain a good repair and that the orbit should be stabilized to prevent any chance of ocular injury or visual impairment. How will you proceed? How should this patient be counseled for her procedure? If she is intoxicated with ecstasy, can you/should you involve her parents in the consent? Is there an optimal anesthetic strategy in this circumstance? How would you counsel her for the risks of fetal wastage with a general anesthetic? Does it vary with the gestational age?

## Intraoperative Course

### Questions

*Patient 1*. The patient proceeds to the OR 1 ½ hours after arrival at the ED for a closed reduction of the Colles' fracture, an ORIF of the ankle fracture, and placement of an IM rod to the tibial fracture. Following an uneventful RSI, the closed reduction is accomplished and the repair of the tibial fracture is underway. The patient has developed some diffuse wheezing and his baseline SpO2, which was 97%, is now 94%. Your evaluation? After bronchodilator administration, his SpO2 has worsened, to 90%. His compliance is unchanged. After some more medications, you wonder about fat embolism. How can you evaluate this possibility? What are the symptoms? Signs? Under anesthesia? Therapeutic interventions?

coming from a party. His hallucinations are probably from polypharmacy with the strong possibility that whatever he ingested likely had an admixture of hallucinogens, although hallucinations are common with this combination of substances anyway. A toxicology screen would be helpful to identify specific substances [1].

*Patient 3*. This patient also has a full stomach plus an abnormal airway, with trismus probably due to muscle spasm from the mandibular fracture. Nevertheless, a rapid sequence induction with securing of the airway is likely to be successful because the muscle relaxation will eliminate the trismus. If there are concerns about airway visualization, an initial orotracheal intubation can be followed by changing the tube to a nasotracheal intubation. The issue of consent is tricky in this circumstance. The patient is pregnant and in many states can consent for the procedure; however, she is also inebriated/intoxicated, and a determination has to be made by the clinicians as to whether she is able to consent to the procedure by acknowledging the risks, benefits, alternatives including no treatment, and particularly with regard to her pregnancy. This should be carefully examined with the institution's legal counsel as well. The issues may have to be separated, i.e., the parents' consent to be obtained with regard to the oral-maxillofacial and ocular surgery and the patient with regard to the pregnancy. Fetal wastage is generally acknowledged to be higher in the setting of pregnancy and surgery/anesthesia, but the etiological factors are unclear [2]. In general, no special anesthetic strategy is indicated and there are no medications that are particularly favored or restricted. Vital signs should be kept as normal as possible, and consideration should be given to monitoring the fetus when feasible via fetal monitoring, generally after the first 16 weeks of gestation [2].

## Answers

*Patient 1*. There are several possibilities for worsening shunt and insidious hypoxia in this situation. The patient could have aspirated prior to the induction of anesthesia or during induction. An occult pneumothorax and/or airway disruption may be present. If the hypoxemia evolves progressively, it may also be a fat embolism accompanying instrumentation of the tibia with an intramedullary rod. It would be relatively straightforward to evaluate the lung and pleural space, even during surgery, by X-ray. In the absence of findings, the diagnosis of fat embolism should be entertained, although the diagnosis is difficult to confirm and often difficult to treat acutely.

*Patient 2*. This patient's hypertension has improved to 120/85 but he has remained tachycardic. He was intubated uneventfully in the emergency room using etomidate and succinylcholine and securing the airway with a videolaryngoscope. The surgeon would like to go directly to the OR for an exploratory lap, and in fact, the patient is en route to the OR in the elevator. What would you like to have ready? Anything you wish to order in preparation for the case? Can you anticipate what the surgeon is likely to find, based on the patient's history and symptoms? What are your considerations for access (the patient had a 16-gauge saphenous line placed by the paramedics prior to transport)? What are your monitoring considerations? Does this patient need an arterial line?

*Patient 3*. Following a rapid sequence induction of anesthesia with easy placement of a nasotracheal tube, the surgery proceeds uneventfully. An axillary temperature probe shows a temperature of 38.4 degrees; the heart rate is 134 BPM and the blood pressure is 110/70. Your thoughts? What is your differential diagnosis? The bloodwork from the ED comes back: the serum sodium is 124; other electrolytes are normal. The temperature is now 38.9 degrees. Further thoughts? Anything else you wish to do? Why? Will your intravenous fluid management change? In what way? What is your working hypothesis for this change in management? How can you rule out malignant hyperthermia? Further lab work?

Under general anesthesia, several of the most sensitive signs such as altered mental status and hyperthermia are absent. Pulmonary hypertension is difficult to diagnose in the routine case but may manifest, if severe enough, as right ventricular dysfunction. An upper-body petechial rash occurs in up to half of patients, but again, may be difficult to diagnose in the operating room.

Care is typically supportive with regard to mechanical ventilatory support and monitoring of improvements in shunt fraction [3]. For severe fat embolism, high-dose steroid therapy (methylprednisolone) has a role in decreasing the inflammatory response [3].

*Patient 2*. This is very concerning because of the uncertain path of the bullet; there could be a significant hemoperitoneum. Blood in the chest would probably result in significant respiratory embarrassment, but blood in the abdomen, especially in the presence of an altered mental status, may be very subtle in its presentation. In an urgent situation, I would make an assumption that there is liver damage and would anticipate large volume transfusion, therefore the need for large-volume supradiaphragmatic access. The blood bank needs to be called in anticipation of a large-volume transfusion and the massive transfusion protocol should be initiated. A reasonable reserve would be one blood volume with additional FFP and platelet support. Hopefully there would have been time to have a FAST (focused assessment with sonography in trauma) done in the ED prior to transporting to the operating room. An arterial line, if there is time, would be a reasonable additional monitor, although not absolutely necessary in a healthy patient.

*Patient 3*. While it is reassuring that the airway management was uneventful, there is a mystery evolving. The patient's temperature is elevated and there is a marked tachycardia. This is compounded by hyponatremia and the ongoing trend for hyperthermia, which may suggest evolving malignant hyperthermia, particularly if the muscle exam reveals contracture or cogwheel rigidity. If there is evidence of an increased end-tidal CO2 in the presence of a normal minute ventilation (i.e., approximately 100 mL/kg/min at normothermia, which needs to be increased by 7% for every degree centigrade), then a malignant hyperthermia crisis may be evolving. Typical minute ventilation requirements during an evolving MH crisis are several-fold above baseline, and these usually precede a temperature elevation, rather than follow the temperature elevation. Nevertheless, it is possible. The other possibility is a central effect of the MDMA, which has been associated with hyponatremia and hyperthermia. [1, 4].

## Postoperative Course

### Questions

*Patient 1*. How will you determine this patient's fitness for extubation? What are your goals for respiratory mechanics? What are your goals for oxygenation? How will you assess his neurological exam? Is this possible in the setting of his acute intoxication? Would it be "safer" for him to remain intubated and sort things out later in the ICU?

*Patient 2*. His liver laceration was repaired with good hemostasis and excellent surgical technique, and you administered 3 units of packed red blood cells without any additional products. He has 4 peripheral IVs and an arterial line and is hemodynamically stable with a heart rate of 62 and a blood pressure of 114/80. What are your plans for emergence? Should he go to the PACU? To the floor? Remain intubated? What are your indications for admission to the ICU for this patient?*Patient 3*. You have decided to leave the patient intubated and admit her to the ICU because she is in fixation and had an altered mental status at the start of the case anyway, and you wish to keep her airway secured. The ICU calls 3 hours later wondering when she will wake up; they have not had to give her anything with regard to further sedation. Any concerns? How will you evaluate her prolonged sedation? Are there any medications you would administer as reversals, or does she need something else? Why?

## Answers

*Patient 1*. My initial assessment will be the patient's mental status, to determine if he is easily arousable and responsive to commands. There can certainly be residual party drug or alcohol effects which will impair his mental status and potentially impair his ability to protect his airway. Surgically, it does not appear that he has any reason to require mechanical ventilation postoperatively, but if his ability to oxygenate is impaired as a result of fat embolism and he shows ineffective oxygenation, then he should be supported, at least overnight. Typical pulmonary mechanics at the end of surgery would be a forced vital capacity maneuver of 5–7 mL/kg and a negative inspiratory force of about 25 cm H2O, to command (to assess his ability to follow directions).

*Patient 2*. The mental status following emergence is key in this patient as well. He is large, and a combative, dysphoric emergence will be dangerous for the patient as well as all surrounding personnel. That said, he might be fine because his most threatening problem, the liver laceration, was repaired. Depending on the need for postoperative immobility, and following discussion with the surgeon, I would be influenced by the need for perioperative sedation vs. extubation and careful monitoring on the floor. Of course, this is predicated on his ability to cooperate and self-report. If that is not possible, then he should go to the ICU, sedated with a protected airway, until he recovers from his acute intoxication.*Patient 3*. This is a worrisome situation because part of the differential includes cerebral edema from hyponatremia superimposed on an altered sensorium due to the illicit drugs and a general anesthetic. She should have an immediate neurological exam including fundoscopic exam to evaluate her for papilledema, and an imaging study of the brain, preferably an MRI, should be emergently scheduled. Neurology consultation should be obtained, and if there is a question of any residual anesthetic medications that could be reversed, they should be naloxone for opioids, flumazenil for benzodiazepines, and physostigmine as a nonspecific central anticholinesterase.

Electrolytes and glucose should be immediately evaluated, and hyponatremia, which should have already been on its way to normalization because of the change in fluid management, should be re-evaluated.

## Additional Topics

### Questions

1. What is the epidemiology of adolescent anorexia? What about this disorder is life-threatening? What role does the anesthesiologist play in the context of the physiological and emotional issues?

2. Patients with attention-deficit hyperactivity disorder (ADHD) can be on a variety of stimulants (Adderall® (amphetamine/dextroamphetamine), Ritalin® (methylphenidate), Concerta® (methylphenidate), Strattera® (atomoxetine), Vyvanse® (lisdexamfetamine)) for treatment. How do they work? Do you find this somewhat counterintuitive? Why/why not? What preoperative preparation does a patient on chronic stimulant therapy need prior to anesthesia and surgery? What are the concerns? Theoretical or real?

## Answers

1. Anorexics die at a rate of 10–20% from complications of starvation or from suicide. All ages, races, and cultures are affected. About 15% of anorexics are men. The disorder is more about control and emotion than it is about food and many clinicians avoid probing medical questions during their preoperative evaluation of these often intelligent, organized, driven, and often "perfectionistic" patients, typically high-achieving female adolescent athletes such as gymnasts or dancers. These children often exist at the knee of the metabolic and electrolyte curve and deteriorate rapidly [5, 6]. Their suicide rate is 57 times greater than a similar population. Sudden death has been related to ventricular tachyarrhythmias, QT prolongation, and torsade de pointes—all of which may be affected by the antidromotropic properties of volatile anesthetics. Hypokalemia due to vomiting, starvation, and the use of diuretics is common. It is particularly tempting to not order pre-op lab tests in athletic (albeit "thin") adolescent patients who seem otherwise well. In a tipoff to a deeper underlying problem, these patients are often angry and defensive when you ask more probing questions and may refuse laboratory testing because they are well aware that abnormalities may accompany their underlying disorder.

2. Stimulants, as a general category, increase endogenous levels of dopamine and norepinephrine in the central nervous system. The way this works for ADHD patients is that these increased levels of catecholamine neurotransmitters exert a "permissive" effect on cognition and particularly executive function. When levels of these neurotransmitters rise, children with ADHD no longer have to "self-stimulate." Their efficacy is remarkable; about 2/3 to 3/4 of pediatric patients are significantly improved. Chronic stimulant medication does not seem to alter anesthetic requirements although there is some evidence that sedation may be more difficult and emergence may be hastened with methylphenidate [7, 8].

3. A male adolescent rugby player, 92 kg, undergoes an ACL repair under general anesthesia with an LMA. Following removal of the LMA, he coughed and experienced an episode of laryngospasm which lasted about 30 seconds. After several minutes of emergence, his SpO2 remains around 90% with a simple face mask. What do you think is going on? How would you assess him further? Physical diagnosis? Chest X-ray? Arterial blood gas? What can you do to improve his current status?

3. It is likely that in a muscular male adolescent (a very typical demographic), this represents negative pressure pulmonary edema, a result of forced inspiration against a closed glottis, the Muller maneuver. Because of the obstruction, a very large, negative, intrathoracic pressure is generated by the patient's increased effort to breath. The large, negative pressures can be upward of −100 cmH2O. The negative pressure causes an increase in left ventricular preload and afterload and a decrease in extramural hydrostatic pressure. The right ventricle dilates, and the interventricular septum shifts to the left and results in left ventricular diastolic dysfunction. Pulmonary edema then develops. In the setting of the ongoing post-laryngospasm hypoxia, the pathognomonic sign by physical diagnosis is the presence of rales at the lung bases (often difficult to hear in a noisy operating room or PACU). Chest X-ray findings may lag behind the physical diagnosis findings, although by the time the chest X-ray is obtained, one can often see diffuse patchy haziness and frequently cephalization of flow in the pulmonary circulation. With this history and the accompanying physical findings, an arterial blood gas is not necessary for the diagnosis but may be a good idea as a baseline if advanced interventions are needed. At that point, I would place a high-flow non-rebreather mask on the patient and administer furosemide which will not only have a diuretic effect but will also directly decrease left ventricular diastolic pressure and mean pulmonary artery pressure, both unloading the right heart [9].

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# Chapter 37
# Obesity and Surgery


**Robert S. Holzman**

A 7-year-old girl is scheduled for cystoscopy and bilateral ureteral reimplants for grade IV reflux, recurrent UTIs. She weighs 55 kg (120 lbs) and is 4 feet (48 inches, 122 cm) tall (BMI = 36.6). She has a history of asthma and uses albuterol, ipratropium, and budesonide (Pulmicort®) intermittently. She has depression, treated with Prozac® (fluoxetine). Her blood pressure is 142/88 measured with an obesity cuff; HR = 110 and SpO2 = 94%. Her HbA1c = 5.1, which is part of a screening study for the surgery. She takes no other medication.



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## Preoperative Evaluation

### Questions

1. How common is obesity in children? Is it getting worse? What seem to be the relevant factors? How will you judge the family context with regard to causation?

2. Does this patient need any specific additional laboratory evaluation? How do you interpret her HbA1c? What are the implications? What is the significance for your anesthetic plan?

3. Would pulmonary function testing help? Why? What would you expect it to show? Are there any additional comorbidities you should expect?

## Answers

1. The prevalence of overweight children doubled between 1976–1980 and 1999–2002.

   Although the prevalence of overweight among blacks, Mexican-Americans, and Native Americans exceeds that of other ethnic groups, overweight has increased among both sexes and among all racial, ethnic, and socioeconomic groups. The risk for overweight is increased among persons with high birth weight (4000 g or more) and parental obesity. The family history of obesity and obesity-related diseases and the dietary and activity patterns should be part of the evaluation.

   Identifiable endocrine abnormalities or syndromes account for less than 1 percent of cases of overweight. This child, according to CDC age and gender-adjusted criteria, would be considered severely obese (BMI > 35).

2. Type 2 diabetes now accounts for up to 45 percent of all newly diagnosed diabetes in pediatric patients. Conditions associated with overweight, such as sleep apnea and gallbladder disease, tripled in children and adolescents between 1979–1981 and 1997–1999 [1]. A search through the medical records may reveal previous profiles of lipoprotein, insulin, and glucose levels. Hepatic steatosis occurs in approximately 10 percent of overweight children and adolescents and may be reflected in elevated liver function studies. This patient's HbA1c is normal; so far, she hasn't developed type 2 diabetes, characterized by insulin resistance.

3. Pulmonary function testing, if she can cooperate, is unlikely to be helpful unless there are reversible components to reactive airway disease that are undertreated. That said, while no statistically significant differences in FEV, FVC, or FEV1/FVC have been found, there are significant reductions in PEF and FEV25–75 in overweight, obese, and morbidly obese children. Others have noted adverse effects on FRC and diffusion capacity, in proportion to the degree of obesity [2, 3]. If the history of reactive airway disease is of long duration or the signs and symptoms of obstructive sleep apnea are moderately severe or severe, then further cardiology evaluation is warranted, with specific examinations for pulmonary hypertension and its consequences such as right ventricular hypertrophy and/or right-heart failure. Even with an unremarkable exam, I would still be concerned about "inducible" pulmonary hypertension if the OSA symptoms were severe.

4. What counseling will you provide the patient and her family for her anticipated perioperative course? ICU? Mechanical ventilation? Regional anesthesia? Awake vs. sedated vs. asleep placement of a regional block?

## Intraoperative Course

### Questions

1. What are your considerations for monitoring? Will a noninvasive blood pressure cuff be accurate in this child? Does this patient need an arterial line? Why? Is it likely that an arterial line will make a difference in quality of care/perioperative outcome?

2. You select placement of an epidural catheter for perioperative analgesia. What landmarks will you use? What imaging would be helpful, if any?

4. Parents, and patients when appropriate, should be carefully counseled that obese children are at increased risk for perioperative adverse respiratory events including upper airway obstruction, bronchospasm, and desaturation [4, 5]. Length of stay in the postanesthesia care unit (PACU) and unplanned hospital admissions are also higher in obese children. Continuous pulse oximetry should be employed in the perioperative period, especially in obese children with associated OSAS. A multimodal analgesic regimen is advantageous for control of pain. If opioid administration is necessary, smaller doses of short-acting opioids should be used. Regional anesthesia does not necessarily decrease the perioperative risk of disordered breathing since the underlying pathophysiology is well established in everyday (every night) life. If a block were to be utilized, it should be established with a controlled airway under general anesthesia.

**Answers**

1. The upper arms of obese patients are often conically shaped and may make blood pressure measurement inaccurate. Blood pressures are often obtained on the forearm in these patients [6]. The risk/benefit ratio of placing an arterial line for a procedure that doesn't typically require one should be measured against the underlying medical comorbidities of the patient and their likely postoperative destination. With signs and symptoms of moderate to severe OSAS and a strong possibility of postoperative mechanical ventilation, it may be an excellent idea to place an arterial line not only for intraoperative monitoring but for postoperative care. If the patient has significant systemic or pulmonary hypertension, these findings would also weigh in favor of an arterial line. Adequacy of ventilation and gas exchange should be examined at the end of surgery; to this end, regional analgesia may be very beneficial as an adjunct. The ventilatory mechanics, adequacy of gas exchange and subjective sensations of dyspnea, and findings of respiratory distress can all be assessed clinically, but again, the severity of associated respiratory and cardiac disease may inform the level of monitoring.

2. A continuous caudal or epidural block can be accomplished following the induction of general anesthesia, to ensure good gas exchange without a level of sedation that would have adverse respiratory effects. Bony landmarks will be difficult to palpate for caudal or epidural placement. Ultrasound guidance or nerve stimulation may be utilized to facilitate placement when bony landmarks are obscure [7].

3. Does this patient need a rapid sequence induction? Would you anticipate a difficult airway? How would you evaluate and predict the airway difficulty? Is there anything you can do to optimize patient positioning for airway success?

4. Do these patients require drug dosing based on ideal body weight, total body weight, or lean body weight? Do these differ with regard to specific drugs? Should succinylcholine for RSI be dosed on ideal body weight? Lean body weight? Total body weight? Why? Is dosing adjusted based on volume of distribution? Elimination kinetics? And if so, how?

5. Choice of inhalation agents—desflurane? Sevoflurane? Nitrous oxide?

6. Choice of opioids—fentanyl, sufentanil, remifentanil?

7. Choice of analgesic adjuncts—acetaminophen, ketorolac, dexmedetomidine, clonidine?

3. Signs and symptoms of reflux should be evaluated carefully; the majority of obese patients are *not* at increased risk for pulmonary aspiration [8]. Nevertheless, induction of anesthesia with a 30 degree "head-up" position improves airway visualization and allows the diaphragm to descend, providing a more effective total lung volume for preoxygenation. If a difficult airway is anticipated for the usual anatomic reasons (small mandible, limited mouth opening, high-grade Mallampati score), then advanced airway management strategies and equipment are needed. The majority of patients can be intubated by conventional or videolaryngoscopy without the need for fiberoptic laryngoscopy or "awake" intubation. They will, however, have a tendency to desaturate more rapidly.

4. Obesity is associated with an increase in fat mass and lean body mass, the ratio of which (LBW/TBW) decreases with increasing obesity. The volume of distribution of lipophilic drugs increases in obesity; thus, dosing is more influenced by total body weight. Water-soluble drugs show very little change in volume of distribution and dosing is therefore more influenced by ideal body weight. Drug clearance is increased in obesity because of increased cytochrome P450 activity, phase II conjugation, and increased renal clearance. Succinylcholine should be dosed based on total body weight because of the patient's larger volume of distribution and also increased levels of plasma cholinesterase. Dose adjustments are made as above, depending on the lipid characteristics of the drug as well as its anticipated clearance.

5. Desflurane is the least soluble potent inhaled agent and is the least lipophilic; therefore, it should have the best profile with regard to rapid emergence. That said, any agent can be utilized effectively as long as solubility characteristics are taken into account with regard to duration of elimination. Caution about desflurane, notwithstanding its appeal as the least soluble of the currently available volatile anesthetics, is its notoriety for airway irritability, especially in children. For that reason, the majority of the general anesthetics delivered to obese children continue to be sevoflurane-based. Isoflurane is rarely used in morbidly obese patients because of its solubility characteristics and likely prolonged emergence.

6. Same considerations apply to the opioids. Remifentanil has the additional advantage of biotransformation via nonspecific plasma esterases, which, along with the cytochrome P450 system, are likely to be elevated in the morbidly obese. Fentanyl clearance is higher in obese patients, although comorbidities such as obstructive sleep apnea may affect dosing.

7. All of these drugs may reduce the amount of opioid required, which would be beneficial for this patient; this should include the use of local/regional anesthesia. Care must be taken to evaluate any associated liver or renal dysfunction, especially when using acetaminophen or ketorolac.

8. Following induction and easy intubation of the trachea, you initiate mechanical ventilation. Describe your choices for tidal volume, rate, peak inspiratory pressure, use of PEEP (or not), and optimal I/E ratio. Conservative volumes are selected with a minute ventilation approximately 150 mL/kg/min. Are you surprised?

## Postoperative Course

### Questions

1. Would you leave this patient intubated? Why/why not? What criteria will you use to decide about extubation? Why? What technique would you use for extubation? At the end of the procedure, following extubation, desaturation recurs while the patient is struggling, bearing down, and breath-holding. Your management? Why?

8. Mechanical ventilation must compensate, to some degree, for the chronic loss of lung volume as a result of poor diaphragmatic excursion as well as chest wall restriction. The intraoperative decrease in functional residual capacity will be aggravated by the supine position, general anesthesia, and the use of muscle relaxants as well as surgical retractors. Airway manipulation should begin with preoxygenation in the head-up position for several minutes, whether a rapid sequence induction is chosen or not [9]. The use of continuous positive airway pressure (CPAP) during preoxygenation and spontaneous ventilation or positive end-expiratory pressure (PEEP) during mechanical ventilation is an important defense of the FRC and should be administered by titration, looking for optimal PEEP with regard to oxygenation. I would select a moderately low lung volume strategy, perhaps 6–8 mL/kg, while assessing compliance with a tidal volume/compliance curve in order to keep peak inspiratory pressures reasonable, realizing that the manometer that typically measures PIP is often located near the anesthesia machine ventilator, so pressure readings that are supposed to reflect intrathoracic pressures are not necessarily accurate. Also, the patient's respiratory compliance will change with the start of surgery, abdominal exposure and retraction, and the various positional changes required for the surgery. I am not really surprised at the minute ventilation requirement for this age and this body habitus, and this finding is actually encouraging. It illustrates two principles—the adequacy of the metabolic stress response and the long-known direct relationship between VO2 and body mass, even for obese and morbidly obese subjects. There may be some further metabolic changes with regard to the respiratory quotient in morbidly obese patients, and their minute CO2 production may be increased beyond the normal respiratory quotient of 0.8.

## Answers

1. Unless there is recognized difficulty with ventilation and gas exchange during surgery, the patient may not need to remain intubated postoperatively, although it may be prudent to use cardiorespiratory monitoring in the perioperative period if indicated. If postoperative intubation and ventilatory support are warranted, then the patient should probably have invasive monitoring placed and plans made for the intensive care unit postoperatively.

   The usual criteria apply for mechanics and gas exchange, but mental status is a critical component, so at the end of surgery, following reversal of neuromuscular blockade and establishing the ability of the patient to respond to command, I would ask if the patient is getting enough air to breathe, and judge the response. Tidal volumes and maximal inspiratory force should meet or be close to the

2. What is your strategy for providing segmental analgesic blockade? Which agents would you choose? What effects on the CO2 response curve would you expect these epidurally administered narcotics to have? Will it be worth it for this procedure?

## Additional Topics

### Questions

1. What is the mechanism of insulin resistance in childhood obesity? Are there race or gender considerations? Family history? Prematurity? What percentage of obese adolescents in the United States are insulin resistant? What other endocrinopathies are associated with obesity in girls?

usual criteria of a vital capacity maneuver of 10 mL/kg (lean body mass) and negative inspiratory force of greater than -20 cm H2O. While it is difficult to judge the peak expiratory flow rate (PEFR) with an anesthesia machine spirometer, clinical judgment should be applied with regard to the likelihood of patient fatigue as an additional factor for needing perioperative mechanical ventilation.

No special equipment should be required for extubation if intubation was routine; the same equipment (e.g., videolaryngoscope) used at the beginning of surgery should be available at the end of surgery. As a trial, particularly for patients with difficult airways, an airway exchange catheter with the ability to attach a 15 mm connector to deliver a continuous flow of oxygen can be utilized.

2. A weak solution of ropivacaine ± fentanyl will be effective for perioperative analgesia. The CO2 response curve has been shown to be blunted with neuraxially administered narcotics, and a patient like this, who will probably do very well with neuraxial analgesia, should nevertheless be monitored with an apnea monitor during the time of the infusion because her ventilatory drive is not normal at baseline.

   It will be worthwhile if there are no adverse effects and it aids in the maintenance of gas exchange, analgesia, and defense of the FRC. If it helps the patient breathe and cough better postoperatively, that is a significant plus to the strategy. Other techniques in lieu of an epidural would include local infiltration at the end of surgery, transversus abdominis plane (TAP) or rectus sheath blocks as a single shot or with a catheter technique, a quadratus lumborum block, or bilateral lumbar paravertebral blocks. Even with ultrasound guidance, placement may be challenging and results variable.

## Answers

1. Insulin resistance is the inability of insulin in an appropriate concentration or dose to increase glucose utilization peripherally in muscle, liver, and adipose tissue as a result of impaired sensitivity of insulin receptor substrates and glucose transport. Risk factors for the development of insulin resistance include obesity, visceral adiposity, puberty, race/ethnicity, family history of type 2 diabetes, female sex, small for gestational age, and premature children [10–13].

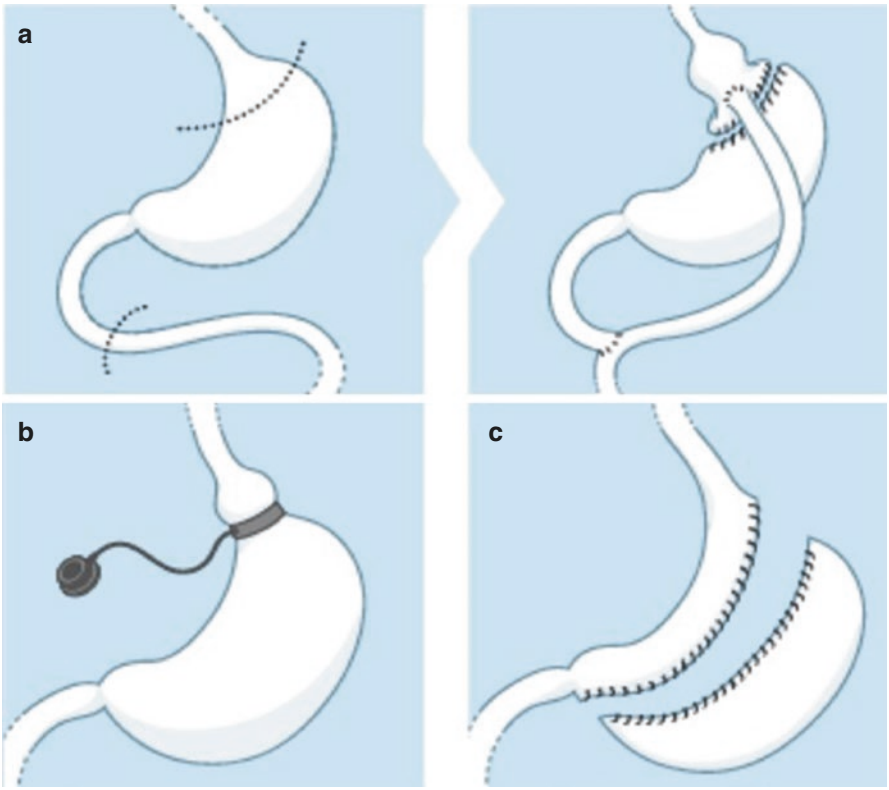
2. A severely obese 17-year-old female is scheduled for laparoscopic sleeve gastrectomy. She is 5′9″ and weighs 310 pounds (BMI = 45.8). She has a history of asthma and uses albuterol, ipratropium, and budesonide (Pulmicort®) intermittently. She has depression, treated with Effexor® (venlafaxine). Her blood pressure is 142/88 measured with an obesity cuff; HR = 110 and SpO2 = 94%. Her HbA1c = 9.2. She takes no other medication. The surgeons begin to insufflate; the peak inspiratory pressure increases to 48 cm H2O and the rate is 24, for an ETCO2 of 46 mmHg. Is this optimal? Is it the best it can be? What can you do to make things better? Does it matter? If so—why?

3. What are the differences between sleeve gastrectomy, Roux-en-Y gastric bypass, and lap band procedures? Are there outcome differences? Are there endocrine differences?

2. Mechanical ventilation must compensate, to some degree, for the chronic loss of lung volume as a result of poor diaphragmatic excursion as well as chest wall restriction. The decrease in functional residual capacity will be aggravated by the supine position, general anesthetic, use of muscle relaxants, and abdominal insufflation. This peak inspiratory pressure would be very concerning if we were sure that all of it was being seen in the chest, which may not be the case. The best way to verify that the number displayed actually has physiological relevance would be to place a catheter within or alongside the endotracheal tube so that intrathoracic pressure can be measured. Indirectly, the influence of very high peak inspiratory pressures can be assessed noninvasively by palpating the pulse or examining the plethysmograph trace of the pulse oximeter.

   A significant and palpable depression in the pulse that varies with positive pressure ventilation (pulsus paradoxus) suggests a significant cardiovascular effect of positive pressure ventilation, whether due to a relative depletion of intravascular volume or a direct effect on cardiac output. Ventilation mechanics can be altered by decreasing tidal volume when the ventilator is used in volume mode, decreasing peak inspiratory pressure when the ventilator is used in pressure mode, changing the I/E ratio, typically shortening the inspiratory time to decrease the total peak minute positive intrathoracic pressure, and changing patient position (i.e., taking the patient out of Trendelenburg position) [14].

3. The most commonly performed bariatric surgical procedures in adolescents are laparoscopic Roux-en-Y gastric bypass (RYGB) and laparoscopic adjustable gastric band (LAGB), though the latter is fading in popularity. Sleeve gastrectomy is also increasingly being performed in obese adolescents. RYGB is the most commonly performed bariatric procedure. A small proximal gastric pouch is created with an anastomosis to a food collection pouch (Roux limb) of jejunum that bypasses 75–150 cm of small bowel. This arrangement restricts intake and limits digestion and absorption. In so doing, it limits gastrointestinal hormone secretion. The adjustable gastric band is a restrictive procedure that results in a tight, adjustable prosthetic silicone band around the gastric inlet. Sleeve gastrectomy is also a restrictive procedure that involves a partial gastrectomy and creation of a tubular stomach from a portion of the greater curvature.

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# Chapter 38
# Gastrointestinal Disease

**Joseph P. Cravero and Thomas J. Mancuso**

A 5-day-old, 3.5 kg female with tracheoesophageal fistula and esophageal atresia (TEF/EA) is scheduled for ligation of the fistula and direct anastomosis of the esophagus. She is on 2LPM NC O2 and has intercostal retractions. There is an oral-tube/drain in place that is helping to drain oral secretions.
Vital signs: HR = 120 bpm, NIBP = 85/50, RR = 42/min, SpO2 = 93%, T = 37° C. Hgb = 14 gm/dL.



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## Preoperative Evaluation

### Questions

1. Is it likely that there are other anomalies associated with the TEF/EA? What anomalies are associated with this disorder? What additional tests or information would be critical to obtain? Why?

2. Of what importance is pulmonary insufficiency on the conduct of this case?

3. How would you counsel the parents of this child about the likelihood of further operations? Is this likely to be the only operation or procedure this child needs? Are there likely to be more procedures in the future—if so, what might they be and how might you predict this?

## Answers

1. TEF/EA has an incidence of 1:2500–3000 and is associated with other congenital anomalies in 50% of cases. The most common form of the EA/TEF anomaly is esophageal atresia associated with a distal fistula (C/IIIb). The diagnosis is rarely made prenatally but may be suspected due to the associated findings such as polyhydramnios and an absent or small stomach bubble. Cardiac anomalies occur in 29% of EA/TEF patients including ventricular septal defect (VSD), patent ductus arteriosus (PDA), tetralogy of Fallot (TOF), atrial septal defect (ASD), and right-sided aortic arch. Gastrointestinal anomalies including duodenal atresia, imperforate anus, intestinal malrotation, pyloric stenosis, and omphalocele are associated. Genitourinary issues including renal agenesis, hypospadias, horseshoe/polycystic kidney, and ureteral/urethral abnormalities occur in 10%. Musculoskeletal anomalies including radial limb abnormalities, polydactyly, lower limb defects, hemivertebrae, rib defects, and scoliosis are found in 10% as is the confluence of these findings designated as VACTERL syndrome. Overall, genetic syndromes such as trisomy 21, trisomy 18, and 12Q deletion are present in 4%. An echocardiogram and ECG are important to obtain prior to starting anesthesia. An abdominal ultrasound to determine renal anatomy is also helpful as is a plain film of the chest and abdomen, which would reveal vertebral anomalies. Presence of air below the diaphragm in any of these tests confirms the presence of a fistula in addition to the esophageal atresia.

2. The presence of pulmonary insufficiency could be caused by prematurity (30% of EA/TEF patients are premature) or aspiration due to the fistula itself. Most often these patients are actually doing well on room air when you meet them. If this is the case, induction and intubation usually proceed without incident although there are specific issues to be addressed. Poor lung compliance changes this situation significantly. Poor compliance increases the chance that air will be forced into the stomach on mask ventilation and will increase gastric distension which in turn will make ventilation much more difficult. Any positive pressure ventilation (by mask or ET tube) that occurs without isolation of the fistula is very likely to continue to distend the stomach in these cases.

3. The prognosis for further procedures in patients with TE fistula is such that one would predict the majority will need several further procedures. The exact likelihood depends on the nature of the fistula and the trachea prior to surgery. Longer "gaps" between the segments of atretic esophagus portend more procedures. Patients with particularly "long gap" esophageal atresia cannot be primarily repaired and require staged procedures. Even patients with relatively favorable "shorter" gaps frequently require dilations of the esophagus where the anastomosis was made. When the anomaly occurs in patients with VACTERL syndrome, it is assured that the patient will need many surgeries during childhood and parents should be given appropriate counseling prior to beginning this process.

## Intraoperative Course

### Questions

1. How would you proceed with the induction of anesthesia? Why would you choose this technique? What is the advantage of keeping the child breathing spontaneously vs. controlling ventilation?

2. What monitors are required for this procedure? Is a central venous catheter required?

## Answers

1. This child has an open tracheoesophageal fistula. It is advisable to avoid significant positive pressure ventilation without having the fistula isolated. Several strategies are possible, and it is critical to understand the intentions of the surgeon before undertaking intubation. Traditionally, preoperative gastrostomy has been described, but this procedure has the potential to further compromise ventilatory stability and create a very low-pressure bronchocutaneous fistula that can render positive pressure ventilation impossible. It is rarely performed at this point. The possibility of other forms of airway anomalies exists. In order to confirm the anatomy plan for the surgery, many surgeons prefer to perform a rigid bronchoscopy prior to beginning the surgery. In this case, it is best to provide general anesthesia via a slow inhaled induction with a spontaneously breathing patient for the bronchoscopy. After confirming the location and size of the fistula, the surgeon may place a Fogarty catheter in the fistula followed by tracheal intubation. Alternatively, the endotracheal tube may be placed distal to the fistula if it is far enough proximal to the carina to allow this placement. If the fistula is at or below the carina, the tube may be placed in a main stem bronchus for the fistula ligation and then brought back into the trachea for the esophageal anastomosis. For most cases where pulmonary compliance is not compromised, the use of a standard induction technique including neuromuscular blockade is also successful as long as care is taken to minimize the peak airway pressures. In cases where thoracoscopic repair of the EA/TEF is planned, the ET tube must be placed below the level of the fistula or in the left main stem bronchus, thus allowing the surgeon to collapse the right lung with carbon dioxide using a pressure of 5–8 cm H2O for surgical exposure. In emergent cases, some clinicians prefer a rapid sequence induction and placement of the tube into a main stem bronchus—then withdrawing until bilateral breath sounds are auscultated. However, the airway is secured; the use of a fiberoptic bronchoscope to confirm the anatomy and assure that the endotracheal tube is in proper position (preferably ventilating the lungs but blocking the fistula) is appropriate and helpful.

2. As a general rule, an arterial line is indicated for TEF repair. The arterial line allows the detection of acute changes in blood pressure, which are not uncommon when surgeons are operating in the mediastinum of small babies. Obstruction of venous return or arterial outflow from the heart occurs frequently. In addition, since ventilation will be impaired at several points during the surgery, the ability to obtain blood gases is helpful. Central venous access is not usually required but may be desired if adequate IV access is not available peripherally, or if the child's status is so tenuous that the use of vasoactive agents is likely.

3. What will your maintenance of anesthesia technique be? Why? Will you use nitrous oxide? Why/why not? What is your choice of muscle relaxant? Why?

4. During surgery, the patient's SpO2 suddenly decreases to 82% from 99% and the inspiratory pressure increases from 25 to 45 cm H2O. What do you think is going on? What would you do? Why? How would you manage her depth of anesthesia? Why? How can her inhaler be administered intraoperatively? How much would you administer through the endotracheal tube? Why?

5. The previous maneuvers are ineffective, and the patient progresses to worsening saturations accompanied by bradycardia and widening of the QRS complex. What is your differential diagnosis? How would you confirm your leading diagnosis? What interventions would you make at this time? Why?

3. Anesthesia can be maintained with inhaled or intravenous drugs or both. During the procedure, the right lung will be retracted away, necessitating the use of a high FiO2. In addition there will be manipulation of the bowel in this lengthy procedure, another relative contraindication to the use of nitrous oxide. The child likely has normal renal and hepatic function for age, making the choice of muscle relaxant easier. Histamine release should be avoided. When choosing a muscle relaxant, it is important to remember that the use of short-/intermediate-acting nondepolarizing relaxant offers no advantages and a possible disadvantage since frequent dosing will be needed. Vecuronium has a good combination of characteristics for this patient for this case.

4. Increased inspiratory pressure accompanied by hypoxemia can have several causes in this case. Although unusual in a newborn, the child could be wheezing, or she could be resisting ventilation due to inadequate anesthesia and neuromuscular blockade; there could be problems with the endotracheal tube and/or ventilator circuit. In this case, where the surgeon is manipulating structures in a very small chest, mechanical obstruction or kinking of the tube is not uncommon. The surgeon may also have kinked the trachea or the trachea may be obstructed with blood or secretions. The FiO2 should be increased to 1.0 and ventilation by hand begun while the surgical field is quickly scanned. The tracing on the capnograph can give useful information about the quality of ventilation. If albuterol is administered, several puffs should be given since much of the drug will adhere to the inner walls of the endotracheal tube and not reach the lungs. It may be necessary for the surgeon to release the retracted lung in order to decrease the shunt and improve oxygenation.

5. Severe bradycardia could be due to a sudden and marked vagal stimulus, drug effect, or hypoxemia. It would be appropriate to assess the surgical field to assure that no vagal stimulus is being applied, and medication infusions should also be reviewed. It is most likely that this problem is due to hypoxia, and the primary treatment should be aimed at restoring oxygenation and ventilation.

## Postoperative Course

### Questions

1. At the end of the surgery, during planned "awake" extubation, the bronchospasm recurs. Your management? Why?

2. How would you provide postoperative analgesia for this patient?

3. You are called to the PACU 1/2 h later to evaluate this patient's stridor; she is breathing 40 times per minute and retracting. How else will you evaluate the patient? Why? What is causing the stridor? How will you treat it? Why? Should you reintubate? Why/why not?

## Answers

1. Plans for extubation must be made in concert with the surgical team. Depending on the nature of the esophageal anastomosis, some surgeons prefer to have the patient sedated with muscle relaxation for a period of days after this repair. In other cases, rapid extubation to remove the irritation of the endotracheal tube is preferred. Extubation could be considered after careful review of the intraoperative course of events: fluid and blood product administration, the length of the procedure, the extent of the surgical incision, and the plan for analgesia. If extubation is planned, an awake extubation offers a greater degree of safety than deep extubation. Management of bronchospasm at this point is difficult. Albuterol should be administered through the endotracheal tube. IV lidocaine could be given. It is possible that the best course of action would be extubation. The wheezing is in response to the tracheal foreign body, and once the endotracheal tube is removed, the child may no longer wheeze.

2. The plans for postoperative analgesia depend on the operation and the postoperative disposition of the child. The thoracotomy involved in this procedure requires significant analgesia. If the child is to be extubated at the end of the case, most centers will attempt to provide regional anesthesia in the form of an epidural catheter. Often this is accomplished by threading a catheter from the caudal space up to the appropriate thoracic dermatome. If the child is to be kept intubated and ventilated for several days postoperatively, then most centers will sedate and provide muscle relaxation for that period of time—providing opiates as needed for comfort. Other forms of regional block such as paravertebral blocks (catheters) have been described for this procedure as well.

3. Stridor can occur during inspiration, exhalation, or both depending on the location of obstruction. The obstruction can be fixed, meaning the caliber of the airway does not change with breathing, or variable. Inspiratory stridor is due to extrathoracic airway obstruction such as subglottic airway narrowing (croup), while expiratory stridor is due to an intrathoracic obstruction that may be present with tracheomalacia. In this case, the causes most likely are subglottic edema following intubation or vocal cord paresis as a result of surgical trauma.

## Additional Topics

### Questions

1. How do you assess free water and electrolyte deficits in babies with gastrointestinal obstruction preoperatively? How is a hypochloremic, hypokalemic metabolic alkalosis generated? Maintained? How do you gauge the calculations for your therapy? How do you judge the efficacy of therapy? In what setting would hypertonic and hypernatremic dehydration present? What are the risks of treatment of this condition?

2. What is the pathognomonic radiographic finding in duodenal atresia? Are there any known syndrome associations? When do you expect this lesion to present? How do you expect it to present? Will there be bile staining of the vomitus?

## Answers

1. Dehydration in children is generally divided into mild, moderate, and severe. In mild dehydration, the child has lost approximately 5% of body weight and has a fluid deficit of 50 mL/kg and generally appears alert and thirsty, perhaps restless. Vital signs are within normal limits, urine output is 1–2 mL/kg/h, and specific gravity of the urine is approximately 1.020. In cases of moderate dehydration, the child has lost 10% of body weight and has a fluid deficit of 100 mL/kg. Heart rate is increased, pulse is weak, and the blood pressure is low. An infant will be lethargic but arousable, and an older child will be thirsty and exhibit postural hypotension. Urine output will be <1 mL/kg/h with a specific gravity of 1.025–1.030. With severe dehydration, the child has lost 15% body weight and has a fluid deficit of 150 mL/kg. The child has the clinical appearance of shock. The heart rate is very high, and the pulse is very weak with accompanying tachypnea. The child is very lethargic or comatose and has cold clammy skin with very decreased turgor. Urine output is scanty with specific gravity >1.030.

   With GI obstruction, the child vomits fluid, hydrogen, and chloride, which leads to hypochloremic metabolic acidosis. Although serum potassium levels may be normal, there is often total body potassium depletion. The alkalosis is worsened by the production of a paradoxical aciduria. This occurs for two reasons. Depletion of potassium results, in the distal tubule of the kidney, in the exchange of potassium for hydrogen. In the proximal tubule, when sodium is retained to maintain intravascular volume, the only anion present to accompany sodium is bicarbonate due to the severe chloride depletion.

   Hypertonic, hypernatremic dehydration can occur in diabetes insipidus or severe diarrhea. In newborns and infants, excessive evaporative water losses can lead to hypernatremia. The hypertonic state itself can lead to CNS damage in the form of cerebral hemorrhages and subdural effusions. Correction of hypernatremic dehydration is quite difficult. While the patient is hypertonic, the intracellular sodium content of the cells in the brain increases and intracellular idiogenic osmoles (taurine) raise the intracellular osmolality. If the extracellular fluid osmolality decreases too rapidly, water accumulates in the cerebral cells resulting in cerebral edema. Seizures often occur either while the patient is hypernatremic or during treatment of hypernatremic dehydration.

2. Duodenal obstruction is most often due to atresia. Other causes of duodenal obstruction include stenosis, compression by an annular pancreas, or a web. The pathognomonic radiographic finding in duodenal atresia is the “double bubble” that is caused by the gas-filled stomach and proximal duodenum. When the obstruction is complete, there is no gas distal to the double bubble. An important aspect of the clinical presentation of duodenal atresia is bilious vomiting on the first day of life. Associated problems seen in these patients are Down syndrome

3. What is Crohn's disease? What medications are patients usually maintained on, and what should you do about their medications in the perioperative period? What are the primary issues related to pain control in the postoperative time frame?

with associated cardiac defects and low birth weight. Jejunoileal atresia is another cause of bilious vomiting in the newborn, often due to an in utero mesenteric vascular accident. Affected newborns have abdominal distention and radiographs show distended loops of bowel.

3. Crohn's disease is characterized by a chronic transmural inflammation of the intestine. It may occur in any area of the GI tract. The disease may have many extraintestinal symptoms including the eyes, skin, and joints. There is variable severity to the disease, but the relapsing nature may severely affect a patient's quality of life. In some patients, the disease is unrelenting. The primary treatment of Crohn's disease is medical, but at least 75% of patients require surgery after 20 years with the disease. In terms of medication management in the perioperative period, (1) aminosalicylates should be discontinued 1 day prior to surgery and restarted 3 days after surgery (especially important for those with renal impairment); (2) glucocorticoids should be provided in order to assure adequate stress coverage; (3) purine analogs should be withheld on the day of surgery and resumed approximately 3 days after surgery when oral intake is restarted; (4) cyclosporine should be continued throughout the perioperative period, and methotrexate should be discontinued at least 1 week before surgery and restarted 1 week after surgery and wound healing; and (5) immunomodulator therapy with antitumor necrosis factor agents should be continued in the perioperative setting. Pain management is often extremely challenging given the likelihood that these patients have been exposed to multiple opiate medications prior to surgery. The chronic/relapsing nature of the disease also makes these patients particularly emotionally fragile. Regional anesthesia is extremely important when not contraindicated by infection or coagulation issues. Multimodal pain management is a must. Psychological counseling and ongoing involvement are critical.

## Suggested Readings

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# Chapter 39
# Renal Disease

**Joseph P. Cravero**

A 16-year-old male, 48 kg, born with VACTERL syndrome and chronic renal insufficiency. He is scheduled for a bladder augmentation procedure. The patient has a single horseshoe kidney with chronic obstructive uropathy and progressively decreasing glomerular filtration rate. His hematocrit is 28, with electrolytes $Na^+$ 132 mEq/dL, $K^+$ 5.7 mEq/dL, $Cl^-$ 103 mEq/dL, $HCO_3^-$ 19 mEq/dL, BUN 52 mg%, and Cr 2.4 mg%. He is on sulfamethoxazole/trimethoprim once a day and enalapril. VS: HR = 100 bpm, BP = 145/95 mmHg, RR = 20/min.



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## Preoperative Evaluation

### Questions

1. Is this patient hyponatremic? Why? Is he hyperkalemic? Does hyperkalemia need treatment? What does hyperkalemia indicate concerning overall renal function? Why is the $HCO_3^-$ low? Will this patient's urine be concentrated, dilute, or isosthenuric?

2. Is a hematocrit of 28% normal for a patient like this? Should he be transfused prior to the procedure?

3. What is the GFR of a 16-year-old with a creatinine of 2.5?

4. Is the patient likely to have a low or high cardiac output? Is this patient hypertensive? Why? How will you assess this patient's volume status?

## Answers

1. The patient has a sodium level of less than 135, making him hyponatremic, most likely due to sodium wasting as part of his renal impairment. The patient's potassium is elevated (over 5.5) likely due in large part to impaired potassium elimination along with his acidosis that shifts $K^+$ extracellularly. Potassium balance is usually maintained early in renal failure; thus, hyperkalemia is a late sign (a GFR of 8 mL/min is adequate to clear potassium). His total body potassium is *not* elevated. In addition, the use of renin-angiotensin-converting enzyme inhibitors causes further elevation of $K^+$ by decreasing aldosterone and thus diminishing the excretion of potassium. Acute hyperkalemia is dangerous because it lowers the resting membrane potential of myocardial cells. Renal failure patients tolerate chronic hyperkalemia relatively well because they are able to establish a stable, higher, resting membrane potential at their high $K^+$ level (unlike those with acute hyperkalemia). If ECG does not show characteristic findings of hyperkalemia, treatment is not needed. If peaked T waves were present, treatment would be indicated. The plasma bicarbonate is low due to academia. Chronic renal failure results in a loss of renal tubular mass. When it reaches a critically low level, the kidney is unable to excrete adequate hydrogen ions in the distal tubule. The patient's urine will be diluted because of failure of the tubules to concentrate the urine leading to both salt and water wasting.

2. The hematocrit is low for age due to reduction in erythropoietin production in the kidneys. Chronic low hemoglobin is compensated by an increase in 2–3 DPG. As such, transfusion is not needed for a procedure where copious blood loss is not expected.

3. A rough estimation of GFR can be calculated as GFR (mL/min/1.73 $M^2$ = 0.55 × Length (cm)/plasma creatinine).

4. Due to the low hemoglobin, renal failure patients generally compensate with a high cardiac output. The patient is hypertensive. Patients with renal failure have an increased level of angiotensin II which is a potent vasoconstrictor. Its action is mediated through primary activation of renin-angiotensin-aldosterone system and/or depletion of the extracellular fluid due to salt and water wasting. Volume status is best estimated by evaluating clinical signs of dehydration—notably weight loss greater than 3% of body weight, increased thirst, dry mouth and tongue, increased heart rate, fast breathing and cool extremities, sluggish capillary refill longer than 2 s, and impaired skin turgor.

5. Why is this patient on antibiotics? Is prophylaxis effective? Would infection make this problem worse? How so? Any particular implications for your anesthetic management?

6. What would you do to attempt to avoid perioperative acute renal failure in this patient?

5. Patients with horseshoe kidney anomalies and abnormal drainage have urinary stasis and are at risk for urinary tract infection. The use of prophylactic antibiotics is effective for the prevention of recurrent UTI prior to definitive surgical treatment. Chronic infection can worsen obstructive uropathy and hydronephrosis and cause kidney damage because chronic inflammation of the glomeruli and tubules results in scarring. If infection is ongoing during the procedure, it may spread into the bloodstream, causing bacteremia and septic shock under general anesthesia or in the immediate postoperative period.

6. Postoperative acute renal failure most often occurs in the setting of established preoperative renal impairment. It is defined by a threefold increase in creatinine level. There is little exacting evidence on the best methods to avoid further injuring the kidneys that are impaired—particularly in children where data is almost completely lacking. The avoidance of nephrotoxic agents such as nonsteroidal anti-inflammatory drugs and nephrotoxic antibiotics would be sensible. Maintenance of hemodynamics and renal perfusion is important. Filling pressures should be kept as close to normal as possible. In major invasive cases involving fragile patients, cardiac filling should be assured through the use of echocardiography or invasive pressure monitoring. If cardiac performance is inadequate in spite of optimized preload, inotropes should be started in order to attempt to keep the cardiac output in a low-normal range (minimum). It is not clear if intrinsic renal failure (once it has occurred) can be improved with improved renal perfusion through optimization of volume, hematocrit, and cardiac output.

   Prophylactic low-dose dopamine, while improving renal blood flow and output in adult patients with acute renal failure, has not been shown to improve renal function or outcomes and may increase the incidence of bowel ischemia. Data on the use of furosemide is not conclusive. While there is no convincing evidence that it will reverse or prevent ongoing acute renal failure, it has not been shown to be harmful and most practitioners will attempt escalating doses of furosemide as a trial. If not successful, it should be discontinued. Hemofiltration with continuous venovenous hemodialysis (CVVH) or continuous arteriovenous hemodialysis has been established in adults and older adolescents as a means of maintaining acceptable fluid volume and electrolyte balance. In children with acute kidney injury, its effect on overall prognosis is not clear. Across all populations, approximately 30% of patients who experience acute kidney injury after surgery end up requiring hemodialysis.

## Intraoperative Course

### Questions

1. Does this patient need an arterial line? Would a central venous line be helpful? How would you monitor volume status intraoperatively?

2. How will you induce this patient? Why? What metabolic and electrolyte effects will this patient experience if there is laryngospasm on induction, the patient desaturates, and in the meantime, the end-tidal $CO_2$ increases to 65 mmHg? Are these of significance to your anesthetic management?

3. Which muscle relaxant will you choose? Why? What if there is laryngospasm prior to starting the IV? Is there a problem using succinylcholine? What if the $K^+$ was 5.0 mEq/dL? 5.5? 6.0? Is muscle relaxant use influenced by the presence of renal failure? How so?

## Answers

1. Clinical signs of volume status are not easily interpreted during general anesthesia. Since this is a long procedure with a significant chance for fluid shifts, a measure of volume status could direct fluid administration and optimize volume status in a manner that a patient with renal impairment cannot do for himself/herself. As a general rule, an arterial line is not needed for a procedure with relatively small amount of blood loss or major fluid shifts. On the other hand, an arterial line could help with evaluating the overall volume status of this patient through analysis of the change in systolic pressure from beat to beat with respect to the phase of respiration. A significant change in the systolic pressure different with inspiration vs. expiration is indicative of a low volume state. Alternatively, a central line would help with evaluation of volume status in this patient who has no cardiac or pulmonary pathology (that would interfere with accurate CVP measurement). There is no need for both of these monitors.

2. The patient should be induced with intravenous anesthetics because elevation of serum potassium and metabolic acidosis can be aggravated by hypoventilation, respiratory acidosis, and laryngospasm. Laryngospasm could produce severe metabolic and respiratory acidosis, acute hyperkalemia, cardiac dysrhythmias, and cardiac arrest. It should be noted that the outcome of resuscitation in the face of severe acidosis is not favorable since drugs such as epinephrine are not as effective in this milieu.

3. Cisatracurium would be a good choice for muscle relaxation since this medication undergoes Hoffman elimination and is not dependent on renal clearance. If laryngospasm occurs, I would try non-pharmacological maneuvers such are jaw thrust, CPAP, and mastoid process pressure. Succinylcholine (0.25–0.5 mg/kg) could be used to allow ventilation if these methods fail. The use of succinylcholine could be considered risky here since a full dose (1–1.5 mg/kg) of the medication is associated with an increase in serum potassium of 0.5 mEq/L. Studies in renal failure patients with elevated potassium who are given succinylcholine have not indicated a high frequency of cardiac dysrhythmias. It is still wise to use caution and be ready with measures to mitigate the increased potassium by assuring adequate ventilation and preparing to give calcium, insulin, and glucose. In this case however, securing an adequate airway takes precedence.

4. Is a regional anesthetic indicated for perioperative pain management? Specifically how and where would you place the catheter? Is coagulopathy a significant concern? Which drugs would you dose the epidural with? Why? Are your considerations any different with regard to the patient's underlying problems?

## Postoperative Course

### Question

The anesthetic vapor has been off for 30 minutes, the end-tidal concentration is 0, but the patient is completely obtunded. You have just looked at the pupils after waiting in the OR for 1/2 hour after the case was over and the pupils are fixed and dilated. What is your differential? Approach? Concerns?

4. The use of combined epidural and general anesthesia may improve intraoperative pain control, reduce the need for intravenous opioids and inhaled anesthetic agents, and facilitate the immediate postoperative tracheal extubation. I would place the epidural catheter after the provision of sedation or general anesthesia depending on the level of patient cooperation. The catheter should be placed in the lower thoracic spinal interspace with the catheter tip at approximately T8–9. The presence of coagulopathy, particularly platelet dysfunction, related to renal failure and the use of low molecular weight heparin postoperatively are significant concerns. Coordination with the surgical team is critical to be sure that anticoagulants are discontinued for an appropriate period of time prior to, and after, catheter removal. Epidural infusions containing ropivacaine or bupivacaine with fentanyl would be appropriate choices. Fentanyl is primarily metabolized in the liver and does not have active metabolites that require renal excretion so it is ideal for renal failure patients. On the other hand, the amount of morphine in the epidural infusion is considerably smaller than that used for IV infusion and is likely not a major problem until renal function is severely impaired. Alternatively, the placement of a TAP block with continuous infusion via a catheter or a rectus sheath block could provide pain control for the abdominal wall where the procedure is performed. This option would be preferred if renal function is severely compromised and coagulation is a critical issue.

**Answer**

My differential diagnoses are total spinal anesthesia, cerebral edema, intracranial hypertension from a space-occupying lesion such as a subdural or intracranial bleed, cerebral stroke, brainstem ischemia/anoxia, or drug-induced effects, e.g., anticholinergics and sympathomimetics. If the condition does not resolve within the expected duration of total spinal anesthesia and drug-induced pupillary dilation is ruled out, neurology consultation is obtained and brain MRI and/or EEG is considered. The major concern is brain edema, brainstem ischemia/anoxia, or an intracranial space-occupying lesion that may need immediate neurosurgical intervention.

## Additional Questions

### Questions

1. What is the role of the kidney in vitamin D metabolism? Calcitonin metabolism? Parathormone metabolism? Why is this more of a problem in children than adults?

2. A 2-year-old with nephrotic syndrome is scheduled for ultrasound-guided kidney biopsy. What is the basic ultrastructural lesion? How does this account for the manifestations of the disease? What are the anesthetic implications? Is albumin administration of any help?

3. What is the difference between proximal and distal renal tubular acidosis?

4. What are the primary problems encountered with prune belly syndrome?

## Answers

1. The kidney synthesizes calcitriol, the most active metabolite of vitamin D, which is the rate-limiting factor in promoting intestinal absorption of calcium. Impaired calcium absorption from the gut will result in bone demineralization. Calcitonin hormone antagonizes the effect of parathormone by reducing the tubular reabsorption of calcium and phosphate and inhibition of osteoclast activity. In CRF as the GFR deteriorates, phosphate excretion is reduced and hyperphosphatemia stimulates the release of parathormone, which in turn will mobilize the calcium and phosphate from the bone to maintain normal serum calcium at the expense of bone demineralization. Renal osteodystrophy during skeletal growth in childhood can result in significant skeletal growth failure, fractures, and deformities as opposed to skeletal demineralization of the fully matured skeletal system in adulthood.

2. The basic ultrastructural lesion on biopsy is the absence of glomerular inflammation and the active sediments. On electron micrographs the glomerular basement membrane appears normal, and there are no immune deposits and characteristic widespread fusion of the epithelial cell foot processes. Focal glomerulosclerosis is seen in some patients. There is abnormally increased permeability of the glomerular basement membrane filtration of large particles including serum albumin that results in proteinuria of greater than 50 mg/kg/day (hypoalbuminemia with serum albumin less than 3 g/dL) and water and salt retention. Clinically, patients present with fluid and salt retention manifested as generalized edema, pleural effusion, irritability, fatigue, and hypertension. The anesthetic implications in these patients are increased total body fluid by 3–5% or higher, pleural effusion, pericardial effusion, hypertension, reduced cardiac output due to fluid overload and hypertension, impaired renal function, and potential for cerebral edema due to reduced or absent cerebral autoregulation with volatile anesthetics. Nitrous oxide could cause hypertension by activation of the podocyte receptors. The administration of albumin may not be of help in these patients because it will be filtered out in urine by the impaired glomeruli but it can be harmful if acute rise in oncotic pressure expands the intravascular fluids and potentially leading to acute heart failure and cerebral or pulmonary edema.

3. The difference between the two types of renal tubular acidosis is that in proximal tubular acidosis the tubules fail to reabsorb bicarbonate, resulting in bicarbonate wasting. In distal tubular acidosis the renal tubules fail to filter or excrete hydrogen ion and acids in urine, resulting in systemic acid load accumulation.

4. In patients with prune belly syndrome, the lungs are hypoplastic and fail to expand and develop because of oligohydramnios during early fetal development. Oligohydramnios results from inadequate urine production by the hypoplastic kidneys. In addition, oligohydramnios causes mechanical compression of the chest wall causing deformities and a restrictive chest wall disorder.

# Chapter 40
# Acid-Base Disturbances


**Joseph P. Cravero**

You are asked to anesthetize an 11-month-old who was born with tetralogy of Fallot characterized by severe pulmonary outflow tract obstruction. He underwent a repair as a newborn. He is scheduled for a hypospadias repair. He has clear lung fields but left ventricular function is depressed, and he is maintained on chronic Lasix treatment as well as a salt-restricted diet. ABG reveals a pH = 7.44, $paCO_2$ = 64, $paO_2$ = 62, and $HCO_3^-$ = 44, on room air.



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## Preoperative Evaluation

### Questions

1. Interpret the ABG. How might you explain the findings? Is this patient alkalotic? Is the patient acidotic? Based solely on the $PaCO_2$—what would you expect the pH to be in this case? Explain why this patient's acid/base profile is the way it is. What does chloride loss have to do with the development of alkalosis? Why would the fact that there is a salt-restricted diet add to the problem?

2. Is he ready for the OR? Does his acidosis/alkalosis need to be corrected prior to the OR? Where should you attempt to keep his $CO_2$ during the operation? Is there a problem with normalizing his $CO_2$? How much oxygen should he be on?

3. What is a "normal" bicarbonate level? How does the concentration of $HCO_3^-$ relate to pH?

4. Is it necessary to calculate an anion gap in this case?

## Answers

1. The patient's blood pH is in the normal range between 7.35 and 7.45. He has a significant metabolic alkalosis that is largely compensated by a respiratory acidosis. There are many possible explanations for these findings. Most probably the ABG is due to the fact that this patient has poor cardiac function that has necessitated him being on long-term diuretics to avoid congestive heart failure. This has led to chronic chloride and sodium loss. Since the kidney must maintain electrical neutrality, for every positive ion excreted, a negative ion needs to be absorbed. When chloride is deficient, bicarbonate is resorbed to maintain electrical neutrality—leading to accumulation of bicarbonate in the bloodstream and the resulting alkalosis. On the other hand, when carbon dioxide levels in the blood increase, there is a rise in carbonic acid. A 1 mmHg change in the $PaCO_2$ above or below 40 mmHg results in a 0.008 unit change in the pH in the opposite direction. If there was no alkalosis, we would expect the pH to be $7.4 - 24(0.008) = 7.2$ (approximately). The entire problem might be avoided if the patient was not on a salt-restricted diet (and inadequate Cl in the diet). GI absorption would make up for losses, and alkalosis would be minimal since the kidneys would be able to continue to resorb chloride instead of bicarbonate.

2. Strictly in terms of his acid-base status, the patient is ready for the OR for this procedure. Attempting to correct his metabolic alkalosis prior to the operating room would take a long time and would almost certainly result in adverse effects (likely worsening of his cardiopulmonary status because of volume overload). During the case it would be wise to allow his $CO_2$ to remain moderately elevated to the degree it is at his baseline. Respiratory drive in a child like this is primarily based on the pH of the CSF in the midbrain around the respiratory centers. Normalization of the respiratory acidosis would leave this child very alkalotic and would almost certainly lead to respiratory depression.

3. Bicarbonate is normally about 24. Bicarbonate is related to pH through the Henderson-Hasselbalch equation which in this case would be $pH = 6.1 + \log([HCO_3^-]/.03 \times PaCO_2)$. A change in the $HCO_3^-$ concentration of 10 mEq/L will result in a change in pH of approximately 0.15 pH units in the same direction. In general, an increase in $HCO_3^-$ of 1 mEq/L will result in a rise in the $paCO_2$ of approximately 0.7 mmHg. If the rise is more or less than that, it reflects a coexisting respiratory acidosis or alkalosis.

4. In this case the patient is not acidotic, so the calculation of the anion gap would not be particularly helpful.

5. What is the most common cause of alkalosis in a child coming for surgery? If a child with severe vomiting or an NG tube to suction is given $H_2$ receptor blockade—does that stop the development of alkalosis? Could that treatment cause them to become acidotic?

6. Why does $K^+$ depletion cause alkalosis?

## Intraoperative Course

### Questions

1. You would like to perform a caudal for postoperative analgesia. What are your specific considerations for this patient? Does this patient need a volume load prior to a neuraxial block? What is your choice of IV fluid? If he has a preexisting right bundle branch block from his cardiac surgery, would bupivacaine be as good a choice as ropivacaine? 2,3-Chloroprocaine? Would the pH of the patient affect the onset of local anesthetic? Why?

5. The most common cause of alkalosis is acid loss through vomiting and/or NG suction. Loss of gastric HCl causes alkalosis—loss of acid and volume leads to an increase in mineralocorticoid activity which sustains alkalosis because increased catecholamines and angiotensin II levels increase bicarbonate absorption in the proximal and distal nephrons. Volume repletion with saline restores the ability of the kidney to excrete excess $HCO_3^-$. H2 blockers can decrease $H^+$ secretion in the stomach and stop the process since the fluid lost would lack significant $H^+$ ions. It is unlikely that the patient would become acidotic unless the fluid loss was so great it led to significant hypovolemia and poor perfusion.

6. $K^+$ depletion—which occurs with mineralocorticoid excess—increases bicarbonate reabsorption from the proximal and distal tubule, resulting in alkalosis. Serum aldosterone release from the renal cortex would be suppressed by hypokalemia.

## Answers

1. Hypospadias repair involves penile (sacral) dermatomes that are well covered by a caudal block. In general, caudal block performed after induction of anesthesia would decrease the MAC requirement during surgery by decreasing sensory input from the surgical intervention. The caudal would also supply an effective analgesic for the immediate postoperative time frame. A single-shot caudal or a caudal catheter with continuous infusion could be used depending on the extent of the surgery and the postoperative plans for the patient. In this particular patient we would like to know the current hemodynamic status of the patient. If the repair is completed, is there any residual shunt? Appropriately administered caudal anesthesia does not usually have much effect on the overall hemodynamic profile of patients in the infant and toddler age groups. These blocks have been safely applied for congenital heart surgery patients prior to and after repair. The volume status of this patient should be carefully considered. Prolonged NPO times could lead to volume depletion; however, this patient could be sensitive to volume overload. I would consider the NPO duration and administer a volume of fluid that would replace fluid deficit but it would not be necessary to administer additional fluids to a child in this age group to prepare for the caudal block. If the patient was on any anticoagulants, this could impact the regional anesthesia plan. Regional anesthesia is considered safe in patients taking nonsteroidal anti-inflammatory drugs. On the other hand, if the patient was on other anticoagulants such as low molecular weight heparin or specific antiplatelet agents, the recommended time frames for interruption of these medications prior to regional

2. What type of fluid would be most appropriate for this patient? Do you think lactated Ringer's should be avoided? Why? What would happen if you mistakenly infused an IV fluid containing bicarbonate into this child? Would he/she become dangerously alkalotic? For how long? How is bicarbonate excreted? Why would bicarbonate containing solutions exist? Is there any reason to administer such a solution on purpose?

anesthesia must be observed. Implications of an intravascular injection could be significant in this patient and extra care should be taken to administer a test dose and fractionate the administration of a bolus dose. Local anesthetic toxicity is related to blockade of sodium channels. Bupivacaine is a stronger blocker of these channels than lidocaine or ropivacaine. If the patient has a pre-existing conduction abnormality such as a right bundle branch block, induction of dysrhythmias with high levels of local anesthetic could be an issue. As a result, ropivacaine or 2,3-choloroprocaine would be considered a better choice in a patient with compromised cardiac performance. Local anesthetics are weak bases. The ratio of the forms of the drugs can be expressed as $BH+ = B + H^+$. The pKa of each local anesthetic (the pH at which the drug exists 50% ionized and 50% unionized) will determine the concentration of the cationic and base forms that exist at any pH. This is expressed by the Henderson-Hasselbalch equation as $pKa = pH + \log([BH^+]/[B])$. Since the unionized base form of local anesthetics determines the onset of the drug, the more acidic the patient is, the more the drug will be in the ionized form. Therefore, the onset will be slower.

2. Normal saline contains 154 mM of $Na^+$ and $Cl^-$. It has a pH of 5 and an osmolarity of 308 mOsm/L. Lactated Ringer's solution has an average pH of 6.5, is hypoosmolar (272 mOsm/L), and has similar electrolytes to plasma. The administration of large amounts of NS has been associated with the development of hyperchloremic acidosis. There is some theoretical concern that lactate accumulation could lead to difficulties with high-volume LR administration, but this has not been clinically shown to be an issue. LR is not approved for administration with blood products. In this case, I would choose LR for fluid administration as long as blood products were not co-administered (extremely unlikely in this case). In the kidney, the distal tubule will resorb 100% of $HCO_3^-$ until the blood level reaches 24 mEq/L—at that point it will excrete almost all of $HCO_3^-$ above that level. An inadvertent bicarbonate infusion will increase the $HCO_3^-$ level significantly which will, in turn, lead to spilling of massive amounts of $HCO_3^-$ and alkalinize the urine. This is done purposely in some cases to encourage the excretion of toxins or chemotherapeutic agents. Systemic alkalosis would be very short-lived as the kidney is extremely effective at excreting excess bicarbonate.

## Postoperative Course

### Questions

1. On emergence your patient has developed a significant acidosis. What is the effect of acidosis on the major body systems? What would be the effect of alkalosis on these systems?

2. Your patient is not waking up after completion of the surgery. What possible effects of acidosis could be a cause of delayed emergence?

## Answers

1. Acidosis can have multiple effects. With pH <7.22, myocardial and smooth muscle depression can result in decreased cardiac stroke volume, cardiac output, and systemic vascular resistance. Hypotension that results can lead to poor perfusion and inadequate $O_2$ delivery (in spite of a rightward shift of the $O_2$-Hb dissociation curve) and add to acidosis. The response to vasopressors and inotropes is impaired in this setting. Pulmonary artery pressures will increase secondary to increased pulmonary vascular resistance which leads to greater afterload on the right heart. An increased acidosis will lead to increasing $K^+$ levels which could be dangerous if $K^+$ was elevated to begin with. Alkalosis has several effects as well. (1) Increasing pH leads to a leftward shift of the $O_2$-Hb dissociation curve. This decreases $O_2$ unloading which can be problematic in a patient with marginal cardiac output. (2) Calcium levels will decrease which can lead to decreased cardiac contractility—particularly in newborns and infants. (3) From the pulmonary perspective, alkalosis will lead to an increase in bronchial smooth muscle tone. (4) Respiratory alkalosis is associated with low $CO_2$ and will result in lower cerebral blood flow due to cerebral arteriolar constriction. The effects of pH extend to the coagulation system as well. Coagulation factors are most active at elevated pH levels (pH = 8). While it is not recommended to purposely raise the pH to these levels for bleeding prophylaxis, it is important to appreciate that significant acidosis can impair procoagulant activity.

2. Extremes of $CO_2$ retention can induce a $CO_2$ narcosis due to neuronal intracellular acidosis and intracranial hypertension from increased cerebral blood flow in those patients that are susceptible. In addition, monoquarternary muscle relaxants such as vecuronium and rocuronium will be potentiated and could be prolonged in their action. Finally, acidosis will decrease renal and liver blood flow, thus extending the half-life of most anesthetic agents and opioids—although unionized fractions will be reduced for almost all agents at low pH.

## Additional Questions

### Questions

1. You are asked to provide MRI sedation for a child with a pH of 7.28. The Na is 145, the K is 4.5, the Cl is 111, and the bicarbonate is 18. How would you characterize the acidosis in this patient? High or low anion gap? What kind of problems might lead to a high-gap acidosis? What kind of problems would lead to a non-anion gap acidosis?

2. You are asked to anesthetize a 5-year-old WF patient for renal transplant who is in end-stage renal failure. Her pH is 7.21. Why are renal failure patients acidotic?

**Answers**

1. The anion gap is calculated by subtracting the sum of the serum anions from the sum of the serum cations as follows: ($[Na^+] + [K^+]$) – ($[Cl^-] + [HCO_3^-]$). The normal range is between 3 and 11. In this case, the anion gap is elevated which indicates there is an acidosis. Possible causes include lactic acidosis and diabetic ketoacidosis. Exogenous ingestion of acid such as methanol, ethylene glycol, propylene glycol, or aspirin could account for this. Endogenous (inborn errors of metabolism) leading to an acid load can also account for this. Hyperchloremia resulting in bicarbonate loss is the most common cause of a non-anion gap acidosis. This could be due to gastrointestinal loss or rental tubular acidosis—proximal or distal.

2. If the renal damage affects glomeruli and tubules, the acidosis is a high anion gap acidosis. It is due to failure of adequate excretion of various acid anions due to the greatly reduced number of functioning nephrons. If the renal damage predominantly affects the tubules with minimal glomerular damage, a different type of acidosis may occur. This is RTA, characterized by a normal anion gap or hyperchloremic type of acidosis.

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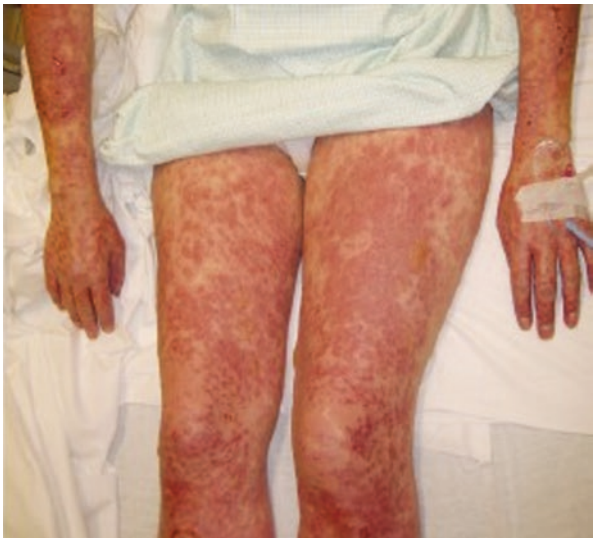
# Chapter 41
# Skin Disorders

**Robert S. Holzman**

An otherwise healthy 5-year-old boy with vesicoureteral reflux was started on Bactrim 2 days earlier for uroprophylaxis. He was admitted to the intensive care unit with malaise, fever, headache, sore throat, vomiting, diarrhea, and extremely painful erosive bullae surrounding the mouth, lips, nares, and conjunctivae. He is not intubated. He needs a central line for better vascular access and has a 24-guage peripheral IV.





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## Preoperative Evaluation

### Questions

1. How will you evaluate and manage his airway?

2. What fluid/electrolyte problems can you anticipate?

3. Nutritional problems?

## Answers

1. Stevens-Johnson syndrome (SJS) is on a spectrum of immune-complex-mediated inflammatory skin reactions characterized by disseminated epidermal necrolysis, commonly related to drug exposure in the case of SJS and toxic epidermal necrosis (TEN), and herpesvirus in the case of erythema multiforme (EM). Drugs more highly associated with SJS/TEN in children less than 15 years of age include sulfonamides, phenobarbital, carbamazepine, and lamotrigine [1]. The differentiating feature is the percent of skin detachment from total body surface area, with SJS at <10% detachment and TEN >30%. The mucosal injury may consist of erythema, edema, sloughing, blistering, ulceration, or necrosis.

   Mucous membranes are affected in more than 90% of patients, particularly at ocular, oral, or genital sites. The vermilion border is almost always involved. Specific considerations for the airway include the amount of patient discomfort, the accumulation of sloughed/necrotic tissue which can worsen dramatically with airway manipulation through the use of oral airways, laryngoscopy and suction equipment, tissue friability, bleeding, and the risk of subsequent scarring, aspiration of necrotic intraoral debris, and edema of intraoral as well as soft tissue structures of the pharynx and neck, making visualization difficult. A central line would typically require endotracheal intubation, but consideration in these circumstances should also be given to avoiding a general anesthetic if possible and considering a TIVA technique for PICC line placement, perhaps in the radiology suite.

2. Stomatitis and mucositis lead to impaired oral intake with consequent malnutrition and dehydration. Other laboratory abnormalities include hypoalbuminemia, electrolyte imbalance, and increased blood urea nitrogen and glucose in severe cases, due to massive transdermal fluid loss and hypercatabolic state. Mild elevations in serum aminotransferase levels (two to three times normal) are present in approximately one-half of patients with TEN, while overt hepatitis occurs in approximately 10%. The generalized epidermal sloughing results in large, raw, painful areas of denuded skin which makes intravascular volume support the equivalent of burn management.

3. Depending on the length of the interval between onset and the time of critical care intervention, the patients have already been in an extremely high metabolic state (e.g., very high fevers up to 400 C) with poor nutritional intake, hence the hepatic injury, hypoproteinemia/hypoalbuminemia, frequently prerenal azotemia, and intravascular volume depletion. There may be insulin resistance as well. The recovery course can be prolonged, with the acute phase lasting 8–12 days followed by reepithelialization lasting 2–4 weeks.

4. What is the nature of this skin problem? How long will it take to resolve? Can it recur? Is this problem different than erythema multiforme?

## Intraoperative Course

### Questions

1. Does this patient need an IV in order to accomplish a "quiet" induction? How will you accomplish this?

2. The medical student suggests an IM pre-induction dose of ketamine and midazolam; do you think this is a good idea?

3. An elderly attending says "in the old days we would use rectal methohexital to get started." What do you think about this idea?

4. Medications are the leading cause (about 2/3) of SJS/TEN in adults and children. In 25–33% of cases, a drug etiology cannot be identified. In children, the most commonly associated drugs are sulfonamides, phenobarbital, carbamazepine, and lamotrigine. *M. pneumoniae* is the next most common trigger, especially in children. HIV and malignancy are recognized comorbid risk factors.

   Following initiation by whatever mechanism, a cell-mediated cytotoxic reaction against keratinocytes leads to massive apoptosis. Cytotoxic T cells are direct against the native form of the drug which then stimulates the immune, resulting in the clonal expansion of a population of drug-specific cytotoxic T cells that kill keratinocytes directly and indirectly through the recruitment of other cells that release soluble death mediators. The severe reaction is similar to acute graft-versus-host disease.

   The differentiating feature is the percent of skin detachment from total body surface area, with SJS at <10% detachment and TEN >30%. The mucosal injury may consist of erythema, edema, sloughing, blistering, ulceration, or necrosis.

**Answers**

1. A calm induction is best, no matter how one accomplishes it. A collaborative approach among the OR team and parents is best; generous premedication may help considerably if needed. This can be accomplished orally with midazolam, rectally with methohexital, or even parenterally (spray the skin prep rather than rubbing the skin). If an IV option is chosen, especially in patients who are wrapped in bandages, it will be wise to have the parent remove the bandages, for patient comfort. The need for restraint must be avoided, however. Securing of an IV can be accomplished with tape over nonadhesive dressings; they may also be sutured in place.

2. Within the general guidance above, it is an acceptable alternative as long as restraint is avoided and the area to be injected is not rubbed with a prep pad but rather sprayed with alcohol, Betadine, or chlorhexidine.

3. Again, a perfectly acceptable idea as long as the perianal area is carefully examined with regard to blistering and skin irritation [2]. The usual induction dose (methohexital) of 25–30 mg/kg (10% solution) may have to be increased if the patient is on concurrent enzyme-inducing drug therapy.

4. You proceed with a gentle mask induction with a well-lubricated mask. What airway management considerations do you have? Do you anticipate having any difficulty visualizing the airway?

5. What are your equipment considerations?

6. Can this case be done without intubation of the trachea? What are your choices? What are your risk-benefit considerations for those choices?

4. As outlined above, the first consideration is with application of the mask; a "tight" mask fit is risky because of the inevitable friction it creates and subsequent bleeding. Liberal lubrication should be generously applied between the cuff of the mask and the skin of the face. As a result of the EB, the small mouth and any intraoral lesions will restrict access of any visualizing device except a fiberoptic scope. Limitation of head and neck movement could make visualization more difficult, and any intraoral scarring could result in a loss of laryngeal or tongue mobility. Most of this is assessable with a careful physical examination. The mouth opening has to be large enough to accommodate a rigid blade, whether a conventional laryngoscope blade or a videolaryngoscope. Laryngoscopy must be gentle, especially close to and around the areas of mucosal fragility. Supraglottic airway devices such as a laryngeal mask airway can be used but are probably ill-advised because of the trauma with blind insertion.

5. Almost all routine monitors will involve adhesives or shearing forces to the skin, which must be eliminated. Pulse oximeters can be applied via clip probes or can be wrapped after the adhesive is cut off. Blood pressure cuffs can be applied over cotton; inflation applies direct pressure rather than a shearing movement, although the "compression" of the underlying skin can be minimized by lengthening the cycle interval. ECG can be monitored with needle electrodes (which present personnel risks of their own) or by cutting off the adhesive circumference [3]. Older patients should position themselves while awake in order to confirm comfort and pressure points (elbows, heels, ischium, occiput, etc.) should be well lubricated/padded. The entire operating table should be padded as well.

6. This depends on the kind of line planned. A peripherally inserted central catheter (PICC) can easily be accomplished with local anesthesia and sedation. A cutdown approach for the internal jugular vein can be accomplished with spontaneous breathing as well, although there is a greater chance of venous air embolism during the final vein cannulation after the initial cutdown. A laryngeal mask can be used with the same considerations, but may be contraindicated depending on the extent of the mucous membrane lesions. A total intravenous technique can be utilized with various combinations of propofol, propofol-remifentanil, dexmedetomidine, or ketamine; however, the patient will be covered up with a general anesthetic in the absence of a protected airway, itself a significant risk when the patient is in a fragile, hypermetabolic state.

## Postoperative Course

### Questions

1. When would you extubate? Deep? "Awake?"

2. You extubate, and the patient has inspiratory stridor in the OR. What will you do? How will you evaluate? Should you do a rigid bronchoscopy?

## Additional Topics

### Questions

1. Anesthesiologists usually encounter skin as the surface upon which monitoring electrodes and devices are placed, and through which vascular catheters are inserted. However, skin can and should also be viewed as one of the largest body organs. As such, can you describe the functions of the skin in the context of:

   - Protection
   - Immunology
   - Fluid and protein balance
   - Thermoregulation
   - Sensation
   - Metabolism
   - Social interaction
   - Storage

**Answers**

1. Minimizing struggling and the need for restraint is key for emergence and recovery. While a deep extubation would provide immediate satisfaction as far as calmness, emergence through stage 2 remains hazardous and might require, as it would with anyone else, application of a tightly fitting mask, continuous positive airway pressure, and manual ventilation. I would prefer a sedate, stage 1 emergence with easy arousability, which would provide reassurance about the patient's ability to protect his airway but at the same time not struggle. This could be accomplished with narcotics or dexmedetomidine.

2. Depending on the degree of stridor, management will likely be medical, with standard nebulized racemic epinephrine. Pre-existing tracheal stenosis might predispose to postintubation croup, but unless the patient was in extremis, one should be reluctant to re-instrument the airway. A diagnostic bronchoscopy might be indicated if the patient had to return to the operating room for reintubation, but the more trauma and manipulation, the higher the likelihood of long-term sequelae with regard to laryngeal/tracheal stenosis.

**Answers**

1. Functions of the skin [2]:

   - Protection: guards against chemical, thermal, or mechanical injury, including environmental injury from solar radiation and weather.
   - Immunology: prevents entry of microorganisms and provides an antiseptic layer of lipid secretions from sebaceous glands.
   - Fluid and protein balance: reduces loss of fluid and moisture; skin loss results in a proteinaceous exudate.
   - Thermoregulation: insulates to decrease heat loss; also allows for rapid cooling through sweating and vasodilation.
   - Sensation: nerve endings, thermo- and mechanoreceptors enable the nervous system to process and interpret information (pain, touch, heat, and cold).
   - Metabolism: supports the production of vitamin D3.
   - Social interaction: facilitates behavioral, interpersonal, and social development.
   - Storage: skin is the largest iron-storing organ in the body.

2. A 4-year-old boy is scheduled for a circumcision for recurrent balanitis. He has junctional epidermolysis bullosa with hypoplastic dental enamel and intraoral blistering. He has normal vital signs including oxygen saturation and weighs 13.2 kg. He functions normally in preschool, with restricted physical activity. His medications include prophylactic antibiotics and anabolic steroids. His hematocrit is 34.7. On physical exam, there are areas of moderate blistering on the trunk and severe blistering on all contact surfaces of the extremities, without scarring or digit fusion. There are mild nasal excoriations, multiple scalp blisters with areas of alopecia, circumoral irritation without scarring or contracture, and several small buccal blisters; the tongue is spared.

   What will you discuss with your surgical, OR, and PACU nursing colleagues about your anesthesia plan and perioperative care plan? What is the significance of his treatment with anabolic steroids? Is a penile block a good plan for postoperative analgesia?

3. A 14-year-old patient with Ehlers-Danlos syndrome is coming to the OR for posterior spinal fusion. Pre-op considerations? Special anesthetic requirements? Laboratory evaluation? How much blood would you set up for him?

   You find a 3/6 holosystolic murmur at the cardiac apex—any special thoughts? Are there any consequences to invasive cardiovascular monitoring in this patient? How would you judge the risk/benefit ratio of invasive monitoring when you explain it to the patient and parents? What contribution can an anesthesiologist make to the special perioperative considerations for wound healing in these patients?

2. The avoidance of blistering is the single most important consideration for pre, intra-, and post-op care. That will require minimizing struggling and handling (particularly through the creation of shearing forces on the skin) and maximizing protective padding and positioning. Good preoperative rapport between every member of the OR team, the patient, and the parents is crucial in order to smooth the induction of anesthesia. Adequate premedication may be crucial to ensure a smooth induction. Specific induction methods such as intramuscular or rectal medication may be needed to forestall struggling. Prevention of secondary infection is crucial; therefore, the use of antibiotics, meticulous wound care, and the use of sterile synthetic nonadhesive hydrocolloid dressings (e.g., DuoDERM®) are a must. Anabolic steroids (testosterone derivatives) are typically given to stimulate appetite and increase muscle mass because it is almost impossible for children to keep up with the constant metabolic challenge of chronic skin injury and repair. Regional anesthetics are typically well tolerated in EB patients because the injection does not result in shearing forces to the dermal/epidermal junction. Skin prep should be with a spray skin disinfectant, and care must be taken not to inject into subcutaneous (dermal-epidermal) junctions. Ultrasound guidance will help but probe dragging on the skin surface should be minimized (ultrasound jelly notwithstanding).

3. Ehlers-Danlos syndrome (EDS) actually consists of several connective tissue disorders characterized by joint hypermobility, skin laxity, and vascular and soft tissue fragility. "Classic" EDS is characterized by joint hypermobility and skin fragility, with easy bruising and impaired wound healing. "Vascular" EDS patients have thin skin and vascular fragility. They also have fragile blood vessels in all organ systems; the GI tract and pregnant uterus as well as liver and spleen have a tendency to rupture spontaneously. Other types of EDS are rarer [4]. In addition to the usual physical exam findings, particular attention should be paid to a history of bleeding and prior difficulty with intubation. Skin fragility may inform the choice of dressings or herald the difficulty of intravascular access. With specific reference to the cardiovascular system, valvular incompetence with mitral or aortic insufficiency should be evaluated on physical exam, with a low threshold for echocardiographic examination. Invasive monitoring is indicated, but specific complications should be outlined to the patient including difficulty with placement or thrombosis/aneurysm formation/wall dissection due to connective tissue incompetence. Extra attention must be paid to padding, positioning, and support because of the laxity of joints and the tendency for subluxation. With a significant history of easy bruising and bleeding, coagulopathy should be ruled out in the preoperative laboratory evaluation. Setting up additional blood is a good idea because of the increased risk of bleeding in the setting of the vasculopathy.

   A loud pansystolic murmur is a pathological murmur that needs to be investigated with an echocardiogram. The collagen abnormality may result in AS/AI or MS/MI. Although connective tissue integrity is impaired in all forms of EDS, the

vascular form is more frequently associated with ectatic vessels predisposing to aneurysms, rupture, etc. There is a more significant risk of such events at the site of intravascular monitoring. The risk/benefit ratio should be clearly outlined to the patient. That said, the risk of a big operation with perhaps more volume loss than in a patient without EDS would suggest a strong need for invasive intravascular monitoring.

Wound healing is often impaired under the best of circumstances and is aggravated by patient movement and struggling, so a carefully planned perioperative analgesic strategy is important. For this patient, the primary strategy would probably involve patient-controlled analgesia with close monitoring by a pain service if available, intensivists, as well as surgeons and nurses, in order to minimize stress and tension across suture lines. Antiemetics in order to prevent nausea and retching are important. Challengingly, early mobilization is key to preserving muscle strength in the perioperative period [3].

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# Chapter 42
# Allergy and Immunology

**Robert S. Holzman**

An 8-year-old boy with DiGeorge syndrome is scheduled for functional endoscopic sinus surgery. He has a history of recurrent sinus and respiratory tract infections and was seen in the emergency department 2 weeks ago requiring an epinephrine injection for moderate respiratory distress and wheezing. He has a constant productive cough and bronchiectasis by X-ray. His medications include montelukast (Singulair®), ipratropium (Atrovent®), and albuterol (Ventolin®) when needed. His vital signs are BP 90/60 mmHg, P 100 bpm, R 18/min and non-labored, and T 370 C. His mom says he is better today than he usually is.



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## Preoperative Evaluation

### Questions

1. How does the embryology of DiGeorge syndrome form the basis for understanding the anesthetic risks?

2. Does his airway reactivity require further treatment prior to surgery? Why?

## Answers

1. The 22q11.2 microdeletion syndrome eponymously known as DiGeorge syndrome is a subset of the "CATCH-22" syndromes which also include velocardiofacial syndrome and Shprintzen syndrome. They are composed of a classic triad of conotruncal cardiac abnormalities, thymic hypoplasia leading to cell-mediated immunodeficiency, and hypocalcemia from the absence of the parathyroid glands. It is the result of abnormal development of the third and fourth pharyngeal pouches and fourth branchial arch. Other sequelae include seizures, abnormal facies, palatal dysfunction, feeding problems, congenital gut abnormalities, behavioral disturbances, and developmental delay. Clinical problems include a high likelihood of congenital heart defects, airway abnormalities, aspiration risk, immunodeficiency, metabolic abnormalities, and neurocognitive delay that can complicate induction and make perioperative recovery challenging. Conotruncal cardiac defects occur in about 80% of patients. Dysmorphic facies occur in 80% of patients, often accompanied by abnormalities of the palate, pharynx, larynx, and trachea and a cleft palate in about 10% of patients. Pierre-Robin Sequence is seen in about 15% of patients. T cell immunodeficiency is typical of the DiGeorge syndrome and is the result of thymic hypoplasia. It is present in about 17% of patients. These patients tend to have repeated infections, especially infections of the upper airway, which may result in reactive airway disease and tracheabronchomalacia. Hypocalcemia, typically presenting with jitteriness, tetany, and/or generalized seizures, develops in the newborn period in up to 60% of patients. Low serum calcium, elevated serum phosphorus concentrations, and low serum parathyroid hormone levels are seen. However, the presence of one feature does not predict the presence of any other feature [1, 2].

2. It may or may not. He may be the best he can be, on a chronic basis. It will ultimately be important to decide whether he is indeed the best that he can be, on multimodal treatment, and simply be prepared for intraoperative interventions, or whether wheezing along with other signs and symptoms represents an acute exacerbation superimposed on chronic illness. It is equally important to remember that the chronic sinusitis, which may be contributing to his ongoing impairment, will likely not get better without a surgical intervention, so there may be some degree of urgency to the procedure.

## Intraoperative Course

### Questions

1. Premed or no? Use of anticholinergic? Require rapid sequence induction? Succinylcholine? Specialized airway equipment? Planning for what kind of potential airway problem?

2. Can this case be done with routine noninvasive monitoring? Why? Despite a parent-present induction without a premed, just after mom left the OR, he coughed, and you could not ventilate him. His $O_2$ saturation rapidly decreased to 84%. What do you think is likely to be going on, and what do you want to do about it? Is it safe to give succinylcholine?

## Answers

1. Here again, the correct answer is "it depends." Many 8-year-olds are articulate and easy to converse with, so he may be absolutely fine going into the operating room for an inhalation induction; sometimes, with chronic respiratory illness, they hate masks and prefer an intravenous induction. On the other hand, he may be developmentally delayed or have substantial fears that developed during his numerous encounters with the medical system, and therefore would benefit from premedication—the full range of premedication, from doses designed to be anxiolytic to doses intended as pre-induction doses. Clinical judgment about the patient's level of maturity is crucial. An anticholinergic such as glycopyrrolate can be administered intravenously once access is established in order to attenuate bronchoreactivity mediated at the level of the conducting airways by cholinergic receptors A rapid sequence induction would be indicated if the patient had GERD and was symptomatic. Succinylcholine, however, is a potent releaser of histamine and, in itself, a potentiator of cholinergic reactivity when the dicholine moiety cleaves into two monocholines. Depending on the airway assessment and the comorbidity of dysmorphic facies or prior history of difficult airway, advanced airway equipment such as a videolaryngoscope or fiberoptic scope may be necessary, as well as consultation with an otolaryngologist for possible assistance during induction and intubation. Exposure of the larynx may be difficult if the pharyngeal antero-posterior diameter is foreshortened and even mask fit may be difficult if there is significant midfacial dysmorphism.

2. Invasive monitoring should not be necessary for this surgery. The comorbidity is chronic disease, not an acute decompensation, and the procedure will not significantly affect the patient's physiology during surgery. That said, if the patient decompensates in some way during surgery, one needs to be prepared to escalate the level of monitoring, such as an arterial line if wheezing worsens or gas exchange becomes significantly impaired. The inability to ventilate after a cough probably represents laryngospasm following some airway irritability such as secretions in the oropharynx. Patients with reactive airway disease have a much greater (i.e., an order of magnitude) risk of laryngospasm [3]. This laryngospasm can be treated in several ways—deepening the anesthetic with inhalation agent and positive pressure; intravenous propofol, or administration of a muscle relaxant. The risk-benefit ratio of the administration of succinylcholine, notwithstanding the concern expressed above about possibly contributing to bronchospasm, would favor succinylcholine as the most rapidly acting muscle relaxant we routinely use. A nondepolarizing muscle relaxant would also be effective albeit a little slower in onset. Vecuronium releases less histamine than rocuronium and should be kept in mind for the child prone to bronchoreactivity [4, 5].

3. Now that the endotracheal tube is in, the peak airway pressure has gone up from 20 to >50 cm $H_2O$ and there is audible wheezing on exhalation and inspiration. How could this have happened? What will you do about it? Should the case be canceled? Why?

## Postoperative Course

### Questions

1. Discuss your extubation sequence if compliance is normal without wheezing. Awake vs. "deep"? Why?

2. In PACU, after extubation child becomes combative. How will you proceed? Why?

3. This boy is susceptible to bronchospasm and manipulation of the airway is certainly a strong precipitant. He may be experiencing light anesthesia, which is a common reason for susceptible children. Also, it would not be surprising in the enthusiasm for having secured an endotracheal tube, the zealous practitioner may have placed the tube in proximity to or just past the carina, stimulating potent protective reflexes. More mundane considerations like tube obstruction (e.g., kinking) or secretions within the tube blocking free inflow and egress of gases more typically occur later in the case. My strongest suspicion for etiology would be light anesthesia ± airway stimulation from the endotracheal tube. In rapid order, I would assess the depth of the tube (simple calculation 12 + (age/2)) and listen to the chest. I would also inspect the chest for symmetrical excursion with inflation. Ultrasound can be used to look for the absence of pleural sliding. A chest X-ray can be obtained when in doubt. Pharmacologically, the depth of anesthesia should be adjusted to ensure an adequate depth; intravenous lidocaine can be utilized to attenuate airway reflexes, bronchodilators can be administered through the endotracheal tube (e.g., albuterol puffs), or epinephrine administered in small doses (e.g., 0.1 mcg/kg) intravenously to decrease bronchoreactivity in the lung parenchyma. Depending on the rapidity of resolution and, in hindsight, the etiology (e.g., light anesthesia, accidental endobronchial intubation), the case can continue because reversible causes have been addressed. With inadequate resolution, the procedure should be canceled, the patient recovered, and the surgery rescheduled.

**Answers**

1. Here the data is controversial. You can make a theoretical case to extubate deep, supported by the notion that emerging with an endotracheal tube in place may recreate all of the unstable airway conditions seen at the beginning of the case, with “light” anesthesia (i.e., going through the excitement stage, or stage 2). I would prefer to extubate awake, reasoning that if, at the end of surgery and with emergence, the patient was lightening and therefore bronchospasm worsened, I would remove the endotracheal tube with the expectation that the bronchospasm would resolve. The alternative would be to extubate deep and hope that the patient passes through the excitement stage uneventfully, which seems more hazardous, although both methods have been used successfully. The additional consideration, a syndromic patient with the potential for airway difficulty (even if not encountered during induction) would push me even further in the direction of an “awake” extubation.

2. The combativeness can be difficult to deal with and dangerous for the patient because he can pull out IVs, etc. A rapid assessment must be made for hypoxia

## Additional Topics

### Questions

1. The mother of a 5-year old spina bifida patient scheduled for tendon lengthening tells you that the child's lip swelled at the dentist recently. What else would you like to ask? How likely do you think it is that this patient is truly latex allergic? How would you test for this allergy? Is RAST testing as sensitive/specific as patch testing? What precautions will you take?

   If the patient develops wheezing and urticaria, how much epinephrine is effective?

2. How should blood be prepared for a patient with severe combined immunodeficiency syndrome? Why? What if the blood was a month old? Other than the risks of infection, are there any other risks of transfusion in this patient?

or any other cause of instability, pain, and/or delirium. If the patient is not conversing or answering questions, it is reasonable to try some pain medication first (morphine, fentanyl) before going on with the differential; however, if they seem delirious rather than in pain, a watch-and-wait attitude seems safest. Small doses (or continuous infusions) of propofol have been used to get through this stage of emergence, particularly following sevoflurane anesthesia. Dexmedetomidine will also be useful for its analgesic, sedative, and behavior control properties.

**Answers**

1. Children with spina bifida are at high risk for developing latex allergy. There is increasing evidence for a genetic predisposition, and in addition, patients who have catheterized for bladder emptying with natural rubber latex catheters have been chronically exposed to the latex antigen. Because dentists often use natural rubber latex dams, more than 50% of latex allergic patients will have their initial allergic presentation at the dentist's office. The suspicion for true latex allergy should be very high in spina bifida patients for the above reasons as well as for their typical history of multiple surgical procedures. Testing for confirmation can consist of patch testing or epicutaneous (intradermal) testing, but probably the most common initial test is a latex radioallergosorbent (RAST) test, which is an in vitro test of the patient's IgE-specific antibody for latex. It is almost as sensitive and specific as patch testing and is a good approximation for intradermal testing, which is riskier because of its in vivo nature. Epinephrine in doses of 0.1 mcg/kg intravenously can be used to initially treat anaphylactic reactions, with a continuous infusion administered if the initial treatment is effective [4].

2. Severe combined immunodeficiency syndrome is a primary T cell abnormality that also results in the failure of formation of the thymus. Patients are vulnerable to overwhelming viral infections and often die in the first few years of life. The risk of infection is constant, and because of chronic anemia, transfusions are often required. However, graft-versus-host disease is a risk of transfusion. Lymphocytes in blood are viable for 3 weeks; therefore, blood must be irradiated prior to transfusion in order to kill lymphocytes. If the blood was a month old, then theoretically, lymphocyte viability should not be a significant clinical consideration, but other problems of using month-old blood should be considered. Furthermore, the blood should be negative for CMV virus and it should be depleted of leukocytes in order to prevent interference with future bone marrow transplant [6].

3. A hemostatic sealant (FloSeal®) has just been delivered into a pedicle site during pedicle screw insertion for a spine fusion. Within a minute, the patient developed hypotension, tachycardia, and elevated peak airway pressures. No wheezing was noted and there was no obvious rash. What do you think is going on? What immediate measures can you take to treat this situation? Is there anything the surgeons can do? Would flushing the field help? Can you explain the absence of wheezing and a rash? Would a tryptase level be of any benefit? What about further testing?

3. Hemostatic sealants like FloSeal® contain gelatin typically derived from animal sources (bovine, porcine) which can be allergenic and precipitate anaphylaxis. This has especially been reported in association with prior exposure to aprotinin [7, 8]. Immediate steps the anesthesiologist should undertake are support of effective circulation (volume, blood pressure, contractility if required) and gas exchange (treatment of bronchoreactivity with epinephrine administration in order to stabilize the ongoing degranulation of mast cells, basophils, and biochemical mediators of severe anaphylaxis). Administration of steroids can help. It is unclear whether the surgeon's attempt to remove the already coagulating sealant will be helpful in the sense of decreasing antigen exposure; flooding the field or aggressive flushing may have the opposite effect and enhance uptake in decorticated areas of bone. Tryptase is a trypsin-like proteinase that is most abundant in human mast cells and basophils. The rise in tryptase levels starts to be detected in serum within minutes of anaphylaxis but the level will gradually revert to normal over the next 6–24 hours depending on the height of the increase. It often correlates with the severity of the anaphylaxis [9]. Further testing should be accomplished for the specific agent (i.e., FloSeal®) which may demonstrate specific allergy to animal-derived gelatin.

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# Chapter 43
# Inborn Errors of Metabolism


**Robert S. Holzman**

An 8-year-old boy is scheduled for muscle biopsy, brainstem auditory evoked potentials, echocardiogram, and eye exam under anesthesia. He has mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes (MELAS) syndrome.



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## Preoperative Evaluation

### Questions

1. What would you like to know about him *before* he comes to the operating room for his anesthetic and diagnostic tests? What problems do you anticipate?

## Answers

1. Symptoms of MELAS syndrome usually begin between the ages of 2 and 15 years, but delayed onset cases have also been reported between 15 and 40 years and late onset cases after 40 years. In approximately 75% of cases, onset of the disorder is before the age of 20 years [1]. At 8 years of age, it is likely that much is known about this patient qualitatively but not quantitatively. It should be anticipated that he might have some cardiac functional abnormalities like cardiac myopathy as well as cardiac conduction abnormalities like varying degrees of heart block. It would not be surprising if he had some degree of respiratory insufficiency on a chronic basis; therefore, a careful history should be solicited for airway obstruction, sleep apnea, and breath-holding episodes. The likely chronic nature of these problems, if present, should foster a high index of suspicion for pulmonary hypertension, which can be confirmed by the echo study. Sensorineural hearing loss has also been reported in MELAS patients; for this reason, the BAEP is included in the EUA. Patients may undergo successful cochlear implant surgery [2, 3]. Visual problems such as ophthalmoplegia, acute and intermittent vision loss with exacerbations, and retinal changes occur.

   The underlying defect of accumulating lactic acid can lead to vomiting, abdominal pain, fatigue, muscle weakness, and difficulty breathing. This accumulation of lactic acid has also been noted in the spinal fluid and in the brain. In some cases, affected individuals will experience a slow deterioration of intellectual function (dementia) and/or a diminished ability to communicate by speech, writing, and/or signs (aphasia). Individuals with MELAS syndrome may also have episodes of confusion and hallucinations often due to a preceding fever (febrile illness) and/or headache. Less common symptoms include involuntary muscle spasms (myoclonus), impaired muscle coordination (ataxia), cardiomyopathy, diabetes mellitus, depression, bipolar disorder, gastrointestinal problems, and kidney problems [1].

   Depending on his degree of impairment, it may be possible to evaluate some of these manifestations prior to his anesthetic, such as biochemical profile (electrolyte derangement, degree of lactic acidosis, hepatic function, scalar ECG if possible for conduction defects) and overall level of function, because of the pleiomorphic nature of the disease onset and spectrum. Laboratory investigations of glucose and lactate levels, electrolytes, liver enzymes, and creatine kinase are specifically performed perioperatively. Assessment of the degree of musculoskeletal and neurological impairment including respiratory and swallowing functions should be made. Measurement of forearm muscle oxygenation responses during and following arterial occlusion provides an easily performed screening test that detects impaired oxygen use. During and following arterial occlusion, which can be performed by a tourniquet, deoxy[Hb+Mb] and oxy[Hb+Mb] are registered. Patients with mitochondrial myopathy display altered oxygenation

2. How will you "design" your anesthetic technique?

## Intraoperative Course

### Questions

1. How will you monitor this patient? Only noninvasive monitoring? Arterial line? Why or why not?

2. Does this patient need an additional line for biochemical monitoring?

3. What is your choice for IV fluid? Lactated Ringer's? Normal saline? Glucose-containing fluid or nonglucose fluid?

responses during and following arterial occlusion which might help to evaluate the patient's individual impairment. Lung function evaluation (spirometry, MIPs, MEPs) and blood gas analysis help to evaluate pulmonary function. A 12-lead electrocardiograph should be performed to exclude pre-excitation syndromes and conduction defects (especially Wolff-Parkinson-White).

2. Similar to many other metabolic disorders, prolonged fasting will tend to make MELAS patients worse, so the NPO time should be minimized or supplemental glucose provided. Lactated Ringer's is not the best IV fluid to use because of the baseline lactic acid elevation. The use of succinylcholine is controversial because of the risk of hyperkalemia, but exaggerated responses to nondepolarizing muscle relaxants have been reported. Acid-base status must be followed closely. Five percent of patients have diabetes. Various anesthetic techniques have been reported, but it is important to keep in mind that most anesthetic drugs depress mitochondrial function. Perioperative hyponatremia and hyperkalemia have been noted [4, 5]. Finally, patients with mitochondrial disorders often suffer from general muscle weakness that restricts cardiopulmonary capacity. Due to impaired energy supply, peripheral nerves are at risk of positional damage.

**Answers**

1. Inasmuch as these are mainly diagnostic procedures with very little tissue trauma, surgically related stress will be minimal, but the amount of time to accomplish all of these coordinated services will be significant, on the order of several hours. That being the case, routine noninvasive monitoring should be accompanied by biochemical monitoring for lactic acidosis, adequacy of glucose levels, and overall acid-base status. This can be accomplished with a second IV of sufficient size to permit blood withdrawals for venous blood gases. If there is any significant concern based on clinical deterioration and unreliability of venous blood gases, then an arterial line should be placed for frequent metabolic monitoring.

2. One IV would be sufficient if an arterial line will be placed anyway; otherwise, two IVs would be my plan.

3. Glucose-containing fluid should be utilized; at least D51/2NS would be reasonable or D5NS. I would avoid D5LR solution for the above reasons.

4. What are your metrics of providing a stress-free anesthetic for this case? Why is it important?

5. How will you ensure normothermia? What temperature range would you be willing to tolerate? Why?

6. What is your antiemetic strategy? Would that be rational if the patient had QT prolongation? How much?

## Postoperative Course

### Questions

1. Post-emergence delirium

2. Shivering

3. Weakness

4. Other than the muscle biopsy, there is not likely to be a lot of stress associated with these procedures. A well-controlled general anesthetic can be delivered with many choices of agents—volatile agents, a combination of narcotic and muscle relaxant (carefully monitored because of the possibility of unmasking underlying muscle weakness), or a combination of both. Total intravenous anesthesia with continuous infusion propofol has been reported, but controversy remains about this choice for children with mitochondrial disorders and adverse effects on the electron transport chain and possible association, even with short-term exposure, with propofol-infusion syndrome [6].

5. MELAS is not associated with malignant hyperthermia, and I would direct attention to maintaining normothermia in order to optimize enzyme system function.

6. Antagonists of serotonin and dopamine can affect the QT interval and for this reason should be given carefully to patients with known QT prolongation. Often patients with minimal QT prolongation may receive these medications with continuous monitoring, as in the operating room, and demonstrate no ill effects. Steroids are also commonly used as an antiemetic, but may have deleterious, increased metabolic effects that increase lactic acidosis. The risks and benefits must be weighed.

**Answers**

1. Since organ systems with inherently higher metabolic rates tend to be affected more in MELAS syndrome, it would not be surprising that in response to a stressful event, the accompanying encephalopathy along with alterations in vision and hearing may be affected, worsening the susceptibility to post-emergence delirium.

2. Tight control of body temperature is important; autonomic regulation including temperature is often impaired in mitochondrial disease. Postoperative shivering increases energy requirements and therefore should be controlled by keeping the patient normothermic (important in the PACU as well). All fluids should be warmed to body temperature. Meperidine in small doses may be used for its beneficial antishivering effects.

3. Because patients with MELAS have a high risk of respiratory failure and aspiration due to muscle weakness, care must be taken to dose muscle relaxants very cautiously and monitor their effect. I would avoid succinylcholine because of the risk of hyperkalemia in deconditioned patients.

## Additional Topics

### Questions

1. A 5-year-old with Hurler syndrome needs a recurrent umbilical hernia repaired. He is not yet in school and weighs 24 kg. What is the basic defect? Are there any significant comorbidities? Do they affect your anesthetic plan? What is the significance of the protuberant abdomen and enlarged liver and spleen? How does this disease affect the cardiovascular system?

2. A 3-month-old presents for open liver biopsy with a suspected diagnosis of glycogen storage disease type I (von Gierke). He weighs 4.2 kg and was a term birth without any complications. VS: BP 86/54, HR 124, RR 36. What problems would you expect? Why didn't this show up earlier? What physical characteristics will you see on exam that are of concern to you as an anesthesiologist? What physiological derangements can you expect? Any particular considerations for intravenous fluid management?

## Answers

1. Mucopolysaccharidosis IH, called Hurler's disease, is one of a group of inherited disorders resulting from defects in degradation of complex mucopolysaccharides (now called glycosaminoglycans). Affected patients lack the lysosomal hydrolases responsible for degradation of these compounds. The lysosomes become engorged with mucopolysaccharides. The compounds dermatan and heparan, formed in excess as a result of defects in degradation of the glycosaminoglycans (formerly known as mucopolysaccharides), accumulate in virtually all tissues of the body. Because you have to assume that glycosaminoglycans infiltrate all tissues, morbidity will be related to immobility. This especially includes the airway but also affects cardiac contractility and electrical conduction, the neck, and all joints including the cervical spine. The enlarged chest with limited rib excursion makes performing tracheostomy, especially in an emergency, very difficult. The liver and spleen are enlarged as a result of these accumulated glycosaminoglycans as well, similar to the tongue and other connective tissues. The liver and spleen and are enlarged as a result of accumulation of incompletely degraded mucopolysaccharides, similar to the tongue and other connective tissues. Physical as well as physiological impairments may result. There is distortion of the valves and coronary artery deformation, again from accumulation of incompletely degraded glycosaminoglycans. In addition, the walls of the coronary arteries are thickened in these patients. Cardiac function is impaired in these children as a result of both coronary artery disease and deposition of glycosaminoglycans in the myocardium. The ribs are flared and these patients have frequent respiratory infections. Accumulation of material within the chest wall may lead to a restrictive pattern of disease while airway narrowing due to accumulation of the same by-products may give an obstructive pattern. This child will not cooperate with measurement of pulmonary function. If the serum bicarbonate is elevated, measurement of blood gases will help quantify the degree of preoperative respiratory insufficiency.

2. In glycogen storage disease type I (von Gierke's disease), there is a deficiency of glucose-6-phosphatase and glycogen accumulates in the liver, kidneys, and intestines. Excess glucose-6-phosphate enters the anaerobic glycolysis pathway, leading to accumulation of lactic acid. The first sign that typically shows up is an increase in abdominal girth. Their first symptoms related to hypoglycemia may occur at about this age because they have been fed so frequently previously, but as they start sleeping more and going for longer periods of time without eating, symptomatic hypoglycemia may occur. This is typical. Findings may be very generalized, like heat intolerance, poor growth, or low muscle tone. More specific symptoms may relate to hypoglycemia, including seizures. Easy bruising may occur. Accompanying lactic acidosis may result in a compensatory respira-

3. Why are infants tested for phenylketonuria? What are the consequences of a deficiency of phenylalanine hydroxylase? Can this be managed adequately by reduction of phenylalanine? Let's say that you had to do a gastrostomy in a patient with PKU who was 5 years old with an IQ of 20. Any special anesthetic considerations?

4. A community pediatric dentist calls to ask your advice about a 4-year-old he has scheduled for office-based dental rehab with sedation. The patient has OTC syndrome (ornithine transcarbamylase deficiency, a disorder of the urea cycle). He typically uses a 50% fixed mixture of nitrous oxide and oxygen (Entonox®) and occasionally gives some PO midazolam for "difficult" children. What are your thoughts?

tory alkalosis and hyperpnea. A large belly is often the initial focus. This is of concern with regard to gastric emptying and adequacy of respiration, all of which will get worse with the induction of general anesthesia. Muscle does not normally contain the enzyme glucose-6-phosphatase so neither cardiac nor skeletal muscle is involved in this disease. These children also may have impaired function of platelets and neutrophils and so may be subject to bacterial infection or have a prolonged bleeding time [7]. Children should not be fasted for more than 3 hours and blood glucose should be checked. The blood glucose level is often less than 40 if children have been fasted longer. Triglycerides and cholesterol are typically elevated. Special diets enriched with complex carbohydrates are common and must be included in the preoperative preparation of the patient. Cornstarch has been used for decades. Alternatively, patients may be admitted the night before for intravenous glucose therapy and enteral carbohydrate therapy with close monitoring; nevertheless, the blood glucose is important to check prior to surgery. Some of these patients may have severe hypoglycemia and not show clinical signs and symptoms, perhaps because the brain can utilize the lactate as an energy source when glucose availability is limited. Intra-op, glucose-containing solutions should be administered and glucose checked frequently.

3. PKU is caused by the absence of the enzyme phenylalanine hydroxylase, which degrades the essential amino acid, phenylalanine, via the tyrosine pathway. In PKU, the excess phenylalanine not used in protein synthesis is transaminated to phenylpyruvic acid or decarboxylated to phenylethylamine. These and other metabolites as well as phenylalanine itself disrupt normal metabolism and cause CNS damage. Affected newborns are normal at birth but untreated may lose as much as 50 IQ points in the first month of life. Severe vomiting occurs early on and the condition can be misdiagnosed as pyloric stenosis. Treatment is dietary, with rigid control of phenylalanine intake for at least the first 6 years of life and some lifelong control as well. An older child with PKU should not be seen in countries with screening, but a 5-year-old with untreated PKU will likely have severe mental retardation, microcephaly, increased tone, growth failure, a prominent maxilla, and widely spaced teeth. The airway may be difficult. These children generally have fair skin with seborrhea and blue eyes. Clinically, an anesthetic should take into account the interactive pharmacology of current anticonvulsant medications, the use of proconvulsant anesthetics, the effect of prolonged fasting on hypoglycemia and therefore the creation of a catabolic state of elevated endogenous stores of phenylalanine precursors, and the potential effect of prolonged use of nitrous oxide on methionine synthetase in case the patient is also vitamin B12 deficient.

4. Many of the urea cycle disorders have similar clinical presentations due to the hyperammonemia, respiratory alkalosis, and neurological symptoms (encephalopathy, seizures, signs and symptoms of cerebral edema such as vomiting and headaches). Sedation or anesthesia may precipitate an acute metabolic encephalopathy. Extreme care to evaluate pre- and post-procedure acid base status and

ammonia levels should be taken. Fasting must be minimized in order to avoid hypoglycemia and catabolism, and facilities for administering intravenous glucose must be readily available. Particularly with regard to oral procedures, gastrointestinal absorption of blood may worsen the encephalopathic picture or precipitate seizures. Seizures are common and should be anticipated. For clinically significant hyperammonemia, intravenous nitrogen scavengers such as sodium benzoate or sodium phenylacetate should be available. Steroids (typically used for antiemetic prophylaxis) should be avoided because they increase catabolism [8, 9].

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# Chapter 44
# Infectious Diseases

**Thomas J. Mancuso**

A 6-month-old, 3.5 kg HIV-positive boy is scheduled for bronchoscopy, washings and brushings, and possible open lung biopsy to confirm the diagnosis of pneumocystis jirovecii infection.
VS: HR 150/min; RR 50/min with retractions; BP = 76/55 mmHg; T = 38.5 °C. Lab HCT = 25%, WBC 3500, and platelets 35,000.
CXR: bilateral perihilar, fine, reticular interstitial opacification. Increased cardiac silhouette.
ECHO: mild to moderate pericardial effusion. Ejection fraction = 48%.
Medications: protease inhibitor (PI) atazanavir (Reyataz) with ritonavir (Norvir) and trimethoprim-sulfamethoxazole (Bactrim).



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## Preoperative Evaluation

### Questions

1. Is this patient in respiratory distress? Is this acute or chronic? Why? What difference does it make? How will you evaluate the patient's oxygenation and work of breathing? Would those assessments be of any importance to your anesthetic plan? Interpret the CXR findings.

2. Of what importance is the anemia? What additional information would you like to evaluate the anemia? Does this patient need a platelet transfusion? Are any other laboratory tests indicated? Which ones specifically?

## Answers

1. This child exhibits signs of respiratory distress such as tachypnea and retractions. Infection with HIV in this case was almost certainly through vertical transmission from mother to child, meaning that he has had HIV/AIDS for 6 months. Retractions are evidence of increased resistance to inspiratory flow, and the tachypnea, in the presence of a smaller tidal volume, indicates decreased pulmonary compliance. Measurement of room air $SpO_2$ will give an indication of oxygenation, but if the child is receiving supplemental oxygen, the oxygen saturation measurement can be normal in the face of significantly impaired pulmonary function. This child certainly has pulmonary disease. In addition to an infectious pneumonia, he also may have lipoid interstitial pneumonia (LIP), which can present with bilateral CXR infiltrates, wheezing, tachypnea, and cough [1–3]. The incidence of LIP in HIV-infected children is 20–30%. The most common opportunistic infection seen in children with AIDS is pneumocystis carinii [4].

2. Children infected with HIV often have lowered counts of all the formed elements of the blood. As is the case with other chronic diseases, the anemia seen in these children is hypochromic and microcytic with low reticulocyte counts. The causes for the anemia are the disease itself, poor nutrition due to poor appetite, and side effects of the medications used to treat AIDS. Based on the weight of 3.5 kg, it is likely that the infant is failing to thrive. A comparison with the birth weight will give information about the rate of postnatal growth. Thrombocytopenia is also seen commonly in these children. Both impaired production and increased destruction have been seen in HIV/AIDS patients. In addition, a lupus-like anticoagulant has been noted in up to 20% of HIV children undergoing coagulation testing. Blood products must be available for this child undergoing this procedure. Transfusion should be undertaken after discussion with the child's primary physician. Only CMV-negative, leukocyte-depleted RBCs should be given to AIDS patients. The need for additional platelets for this case depends upon the exact nature of the procedure. It may be prudent to have platelets available and to use them if the open lung biopsy is performed but withhold them if the bronchoscopy alone is done. Renal dysfunction is common in children with HIV/AIDS. A screening urine analysis will detect proteinuria and hematuria. Given the child's poor nutritional status and failure to thrive, it is worthwhile to check the serum electrolytes, total protein, and albumin prior to inducing anesthesia. Abnormal sodium or potassium values would be a reason to delay proceeding, and knowledge of low serum protein would affect dosing of medication during the anesthetic.

3. How should his cardiac function be evaluated? What additional information on the echo report will be of importance in planning the anesthetic? What might an ECG show?

## Intraoperative Course

### Questions

1. What monitors will you choose? Is an arterial line indicated for this case? What are the risks of central line placement in this patient? Which lead would you choose to monitor on the ECG? Would a transesophageal echocardiogram (TEE) be of any help? Why/why not?

2. The bronchoscopist requests "a little sedation only." Do you agree? Why? Why not?

   Your colleague stops by and suggests total intravenous anesthesia technique because the "lungs are so sick, and the heart is too." What do you think? You select an intravenous technique with small incremental doses of propofol; upon withdrawing the needle from the latex hub, you stick yourself and draw blood. What do you do next? Should you continue with the case? Ask someone to take over? Wash your hands in bleach, alcohol, or Betadine? Should you request to be started on AZT?

3. Approximately 10–12% of children infected with HIV have significant cardiac involvement [5]. The infant's resting tachycardia may be due to poor cardiac function from HIV infection. The parent or caregiver should be asked about prior treatment for congestive heart failure (CHF), and signs and symptoms of CHF should be sought when the history is taken. A cardiac echo will demonstrate LV hypertrophy and/or systolic dysfunction if present, but diastolic dysfunction may not be apparent on a routine echo. An ECG will show sinus tachycardia often seen in these children.

**Answers**

1. Routine ASA monitors are sufficient for the bronchoscopy, washings, and brushings. During these cases, drapes are ordinarily not used, the child is available to the anesthesiologist, and the procedure can stop at any time. If the open lung biopsy is done, an arterial line is important in allowing assessment of blood gases. In this infant with pulmonary compromise, development of a pneumothorax during CVL placement would be very dangerous. If a CVL were planned, placement should certainly be done by the most experienced person, using ultrasound guidance. Lead II of the ECG should be monitored since that lead gives a good indication of the rhythm. TEE would not be particularly helpful in this case. If there is significant cardiac dysfunction and the open lung biopsy is undertaken, placement of a CVP and measurement of filling pressures will give adequate information about cardiac performance.

2. Sedation is not a good option for this child for several reasons. The infant has respiratory insufficiency; the bronchoscopist will obstruct part of the airway with the scope and will then instill saline into the child's lungs after which he/she will suction out part of that saline, along with much of the FRC. With administration of general anesthesia through an LMA, a high concentration of oxygen and controlled ventilation can be delivered, if needed. Occupational exposure to the HIV virus is an important consideration in this case. Studies of hundreds of household contacts have confirmed that the risk of transmission from passive contact with an HIV-infected child is practically zero. Seroconversion is not a common occurrence following needle stick exposure. Hollow-bore needles used in drug administration give a much larger inoculum of blood than the solid needles used for suturing. The current risk for seroconversion for healthcare personnel after accidental percutaneous exposure to blood is 0.3%. After a parenteral exposure to a patient with HIV, the healthcare worker should undergo postexposure prophylaxis, postexposure treatment, and follow-up [6]. While determining which agents and how many to use or when to alter a postexposure prophylaxis (PEP) regimen is largely empiric (two- or three-drug regimen), the timing is not [7].

3. What will be your primary technique be? Why? Will you use nitrous oxide? Why/why not? What is your choice of muscle relaxant? Why?

4. Assume intubation. During surgery, the patient develops increasing difficulty, and the $SpO_2$ decreases from 93% to 87%. The breath sounds become even more coarse throughout the lungs. What would you do? Why? Blood pressure is 60/40 mmHg with a heart rate of 160/min. How would you manage the vital signs and maintain anesthesia?

Drugs currently used include nucleoside reverse transcriptase inhibitors (NRTIs), non-nucleoside reverse transcriptase inhibitors (NNRTIs), and protease inhibitors (PI). The wound should be immediately and thoroughly washed with saline and the institutional "stick" team called. As prophylaxis is begun, the exposed person should be tested to document the HIV status. This testing should be repeated at 6 and 12 weeks after the exposure.

3. There are several possibilities for maintenance for this case. The airway management is limited to the use of an endotracheal tube or an LMA. If an endotracheal tube is used, the size of the bronchoscope will be limited to a greater degree than if an LMA is used. The infant does not have a contraindication to an inhalation induction with oxygen and sevoflurane nor does he have a contraindication to an IV induction. Once anesthetized, an LMA can be placed. Once good air entry is assured, the adapter can be connected to the LMA and the procedure started. Anesthesia can be maintained with IV or inhalational agents. Since the procedure is of indeterminate length, an infusion or frequent small doses of a short-acting nondepolarizing relaxant would be a good choice. An infusion of remifentanil would also be a useful adjunct in the maintenance phase of the procedure. If an LMA is used, the anesthesiologist must be certain that adequate ventilation is possible with a peak inspiratory pressure <20 cm $H_2O$ prior to the administration of a muscle relaxant in this patient.

4. The clinical deterioration could very well be a result of administration of excessive lavage fluid by the pulmonologist. Further attempts at suctioning the fluid from the lungs may not yield much and may worsen the hypoxia as the gas in the lung (FRC) is suctioned from the lung along with any residual lavage fluid. Hypotension in this case could be a result of impaired venous return as higher inflating pressures are used to combat the hypoxemia. If higher inflating pressures are needed to maintain gas exchange, increased preload with IV fluid administration may help increase the blood pressure. Another possibility is the development of a pneumothorax. Auscultation of the lungs and/or CXR will confirm or rule out this possibility. If a pneumothorax is the problem, it must be evacuated quickly. It may be necessary to administer vasoactive amines to support BP while the pneumothorax is being evacuated.

## Postoperative Course

### Question

1. Would you perform a deep extubation with this infant? Why/why not? At the end of the procedure, as the patient is emerging, the hypoxemia recurs while the patient is struggling, bearing down, and breath-holding. How do you manage this situation?

## Additional Topics

### Questions

1. What adjustments do you make in your anesthetic care plan for patients with increased body temperature? Does fever change MAC requirements? Should atropine be given prior to a rapid sequence induction in a febrile patient?

2. A 3-month-old is admitted to the ICU in septic shock with meningococcal sepsis. There is considerable difficulty establishing an arterial line, and the intensivist calls you for help; the parents want you to explain the risks and benefits of placing the line in their baby.

**Answer**

1. Prior to extubation, a suction catheter should be passed through the endotracheal tube to remove any lavage fluid. Once suctioning is complete, the infant should be kept on 100% oxygen for several minutes before extubation is considered. This infant should be extubated awake for several reasons. He should have a strong cough to clear residual saline lavage fluid, he should have as strong a respiratory drive as possible given the degree of pre-procedure respiratory distress he exhibited, and he should be able to protect his airway. If the patient appears awake enough for extubation but is coughing and bearing down such that oxygen delivery is impaired, the best course of action may be to extubate and observe the child's degree of distress once the endotracheal tube is removed. Often, in such situations the child begins to breathe more comfortably.

**Answers**

1. With temperature increases, oxygen consumption increases. The increase in metabolic rate is approximately 15%/°C. Thus, a child with a temperature of 40 °C will have an oxygen consumption of more than 150% of normal. In animal studies, hyperthermia has been found to increase MAC. Atropine has many pharmacologic effects mediated through blocking of the effect of acetylcholine: increased heart rate through its effect on the SA node, bronchodilation through its effect on muscarinic receptors in the bronchi, antagonism of gastric hydrogen ion secretion, and also inhibition of the activity of cutaneous sweat glands. This last effect can increase temperature although, in adults, hyperthermia is generally seen only with overdose of anticholinergic drugs. Increased body temperature and a cutaneous rash are sometimes seen in children who received only a therapeutic dose of atropine. Whether or not atropine should be given as part of a rapid sequence induction depends upon the clinical situation, and the patient's body temperature should have very little, if anything, to do with that clinical choice.

2. Nearly all peripheral arteries have been used for direct monitoring of blood pressure in children. Common problems associated with arterial catheters include emboli, distal ischemia, thrombosis of the artery, and infection. In this setting, in which the child has sepsis, complications are more likely. The sepsis and vascu-

3. A baby with congenital rubella syndrome is scheduled for a percutaneous endoscopic gastrostomy. What problems can be anticipated?

4. A child with a rash arrives in the pre-op clinic for evaluation. How will you differentiate between measles and chicken pox? What implications are there for this particular child with regard to an electively scheduled procedure? If you go ahead, are there risks to other patients in the operating room and the PACU?

litis of meningococcemia will lower blood pressure generally, and the inflamed intima of the cannulated artery will make the likelihood of thrombosis or embolism greater. On the other hand, direct, instantaneous blood pressure monitoring is very important for these critically ill children. Considerations prior to placement include the patient's condition, whether or not the vessel has been damaged, and the amount of collateral flow. It is important to remember that the peripheral arteries of the lower extremity, the dorsalis pedis, and posterior tibial may exhibit pressure-wave amplification and may show a pressure higher than the aortic pressure.

3. The congenital rubella syndrome involves virtually all organ systems of the body. Intrauterine growth restriction (IUGR), often with associated microcephaly, is the most common finding. Manifestations of importance to anesthesiologists are myocarditis, patent ductus arteriosus, and pulmonary arterial stenosis. Assessment of cardiac function should be done prior to going to the operating room. Other findings are blueberry muffin skin rash, cataracts, sensorineural hearing loss, and hepatosplenomegaly.

4. Chicken pox or varicella is a disease caused by human herpes virus. In the USA, most people acquire the disease during childhood. The AAP recommends vaccination for children >12 months who have not exhibited the clinical syndrome. There is a 10- to 21-day incubation period, but most children develop a rash 2 weeks after exposure. The rash is often preceded for 1–2 days by fever, malaise, and headache.

   The typical rash of fluid-filled vesicles often begins on the trunk. The vesicles appear in crops and may be quite extensive or may be few in number. Varicella is contagious for 2 days prior to the appearance of the rash and until the lesions have crusted. Transmission is up to 90% in household contacts and 30% for classroom contacts. Measles, or rubeola, is another common infectious viral illness. It is also very contagious with 90% of household contacts becoming infected. The MMR (measles, mumps, rubella) vaccine is given at 12 months and again in early adolescence. The clinical course of the disease has three phases: a 10- to 12-day incubation period; a prodrome of 3–5 days characterized by Koplik spots on the buccal mucosa, moderate fever, cough, and conjunctivitis; and the final stage of 3–5 days with maculopapular rash on the neck, arms, and legs and high fever. This child does not have any particular increased risk if he were to undergo general anesthesia and a DSU procedure. However, varicella and rubeola are both quite contagious, and infection can be quite serious in immunocompromised patients.

5. You have difficulty waking up an otherwise healthy 8-year-old after a 2 h exploratory laparotomy for a ruptured appendix with significant peritonitis. Your anesthetic technique was impeccable and could not possibly account for this situation. Triple antibiotics were used at the beginning of the case, which included ampicillin, gentamycin, and metronidazole (Flagyl). What is your differential diagnosis? Would calcium be of any help? Is another dose of neostigmine indicated? Another anticholinesterase? What are the disadvantages of using additional doses of anticholinesterases? Does calcium have a direct effect on presynaptic portions of the neuromuscular junction? How long would you expect this problem to last?

6. Tuberculosis—what do you need to know as an anesthesiologist?

5. Antibiotics can enhance neuromuscular blockade. There has been evidence of both pre- and postjunctional effects attributed to antibiotics. It is likely that there is no common mechanism by which antibiotics cause neuromuscular blockade, and thus there is no recommended standard therapy. If the recommended standard dose of neostigmine is not effective in reversing neuromuscular blockade, one option is to provide mechanical ventilation until recovery has occurred. Another option is the administration of sugammadex. Sugammadex has been effective in reversal of profound blockade by rocuronium in both children and infants [8]. Administration of calcium is not recommended since the antagonism to neuromuscular blockade is temporary and calcium may inhibit the antibacterial activity of the antibiotics.

6. The incidence of tuberculosis is increasing worldwide. Recent immigrants to the USA, the homeless, and children with HIV/AIDS are specific groups of concern. Most children infected with *Mycobacterium tuberculosis* acquired the organism from close contact with an individual with active disease. Children with TB can have various presentations. Many are asymptomatic and are detected when a skin test is positive. Progressive pulmonary tuberculosis can occur with the development of lobar or bronchopneumonia. Children with pneumonia or pleural effusions may come to the operating room for placement of thoracostomy tubes and video-assisted thoracoscopic surgery (VATS) for drainage of empyema or bronchoscopy [9]. *M. tuberculosis* is a 1–5 μ-sized particle, and disease transmission is by airborne spread. When children with the diagnosis of tuberculosis come to the operating room, prevention of infection of the caregivers and contamination of the anesthetic equipment are of paramount importance. The CDC recommends protective masks be used that meet stringent filtering requirements. Filters are recommended for use in breathing circuits used on patients known to have TB [10].

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## *Annotated*

McGowan CW Jr. Congenital infections. Chap. 66. In: Marcdante KJ, Kliegman RM, editors. Nelson essentials of pediatrics. 8th ed. Philadelphia: Elsevier; 2015. p. 259–64.

The author, in 5 pages, reviews the clinical characteristics of congenital infections included in the TORCH designation and other perinatally acquired infections, *Toxoplasmosis gondii*, cytomegalovirus (CMV), herpes simplex type 1 or 2, *Treponema pallidum*, parvovirus, HIV, hepatitis B, *Neisseria gonorrhoeae*, *Chlamydia trachomatis*, and *Mycobacterium tuberculosis*.

van der Walt JH, Jacob R, Zoanetti DC. Infectious diseases of childhood and their anesthetic implications. Paediatr Anaesth. 2004;14:810–9.

The authors review the immunization schedule for children in Australia and the common infections affecting children. Incubation and infectious periods for measles, mumps, rubella, pertussis, varicella, and other common infections are listed. In addition, management issues of infected children in the perioperative period such as isolation and equipment disinfection are reviewed.

# Chapter 45
# Neuromuscular Disease


**Joseph P. Cravero**

A 14-year-old girl with juvenile myasthenia gravis has been treated with steroids and pyridostigmine for the past year; she now presents for thymectomy via median sternotomy. BP = 110/72, HR = 100 RR = 28/min, RA SpO2 = 96% and Hgb = 12.6.



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## Preoperative Evaluation

### Questions

1. What is myasthenia gravis? Is this patient at risk of pulmonary aspiration of gastric contents? How would you assess this preoperatively? Should she receive pharmacologic prophylaxis for aspiration? Which drugs? Are there any interactive effects with her current medications?

2. Is it important to know which steroid she is on? Should she be "covered" for surgery? Should she receive her pyridostigmine preoperatively? Would you base your management on an assessment of her pulmonary function tests?

## Answers

1. Myasthenia gravis (MG) is a neuromuscular disease that is caused by circulating antibodies that block acetylcholine receptors at the postsynaptic neuromuscular junction, inhibiting the excitatory effects of acetylcholine on nicotinic receptors. The disease leads to fluctuating muscle weakness and fatigue.

   Yes, patients with myasthenia gravis can experience fluctuating weakness including pharyngeal, laryngeal, and respiratory muscles. Swallowing and effective cough effort are impaired. She should be considered at risk.

   Her actual risk could be evaluated by history. How much weakness is she experiencing now? Has she had issues with passive regurgitation or aspiration while sleeping? The patient's ability to protect and maintain a patent airway postoperatively should be assessed. The general muscle strength or weakness is assessed by physical examination including the presence of ptosis, double vision, dysphagia, and rapid fatigue with repetitive movement such as opening and closing her hands. Pulmonary impairment can be defined by a vital capacity less 40 mL/kg, impaired expiratory effort (maximum static expiratory volume), and flow-volume loops showing decreased flow on expiration in supine and sitting positions.

   Yes, a histamine-2 blocker such as ranitidine and an antacid (Bicitra®) are indicated.

   We would not use metoclopramide as the extrapyramidal effects that could occur with that drug would make interpretation of muscle strength and eye findings difficult.

2. It is important to know what type of steroid she is on and how much she has been taking. The hypothalamic-pituitary-adrenal axis is often suppressed by exogenous administration of a glucocorticosteroid. The effect depends on the maintenance dose of steroid that the patient is on. The patient will need stress dose coverage when the maintenance daily dose is greater than 10 mg of prednisone (or equivalent). There are arguments against the need for stress dose steroids if patients have been on daily maintenance doses of less than 10 mg of prednisone (or equivalent) or if the child has had very minor surgery where the physiological stress to the patient is minimal. If one chooses not to give a stress dose, the patient should be carefully monitored for lethargy or hypotension in the postoperative period—both of which could be signs of glucocorticoid deficiency. I will administer steroid coverage for this patient because she is scheduled for a thoracotomy—which is a major surgery. Ideally, the patient should not take her pyridostigmine preoperatively. Omitting the morning dose of pyridostigmine will actually make the patient slightly weaker and potentially obviate the need for intraoperative muscle relaxants. Added weakness may provoke the patient's anxiety so the issue should be carefully discussed prior to the day of surgery. If pulmonary function is impaired despite optimal medical therapy, the morning

3. She is very anxious; how would you counsel her about preoperative sedation? She wants a mask induction; is that OK? What endpoints would you look for to determine the onset of anesthesia? What if she goes into laryngospasm during induction—how will you manage it?

## Intraoperative Course

### Questions

1. What are your monitoring considerations? Does this patient need an arterial line? Why? Should the patient have central access? Why? Are there circumstances in which a transesophageal echocardiogram would help? Would a pulmonary artery catheter ever be indicated?

dose of pyridostigmine would be indicated. Standard pulmonary function tests should be normal when the myasthenia is well controlled with medical therapy. If myasthenia is not well controlled, the forced vital capacity (FVC) and forced expiratory volume in 1 second (FEV) are decreased due to weakness. If FVC is normal and the FEV is decreased, it may indicate intrathoracic airway obstruction due to an enlarged thymus and may portend difficulty when the patient undergoes positive pressure ventilation.

3. Depending on how weak the patient is, preoperative intravenous anxiolytics may be administered with caution and close monitoring for respiratory depression. There is no contraindication to an inhalation induction, provided the patient is appropriately NPO and there is no history of reflux-related aspiration pneumonia in the past. Due to weakness of the extraocular muscles, the loss of response to eyelash stimulation is not as helpful as general stimulation to determine the onset of general anesthesia in this patient. Similarly, testing of the muscle twitch may be falsely absent or diminished around the eyes as opposed to ulnar nerve stimulation. If the patient develops laryngospasm, it can be overcome with simply applying positive pressure, particularly in the presence of weak pharyngeal muscles. Alternatively, due to the deficiency of acetylcholine receptors, a small dose of nondepolarizing muscle relaxant (or a large dose of succinylcholine) is administered to relax the glottic muscles.

**Answers**

1. Standard monitors should be placed including ECG, pulse oximeter, blood pressure cuff, end-tidal carbon dioxide, and body temperature. In this case—where muscle strength is a major issue—a neuromuscular monitor is helpful. I would place an arterial line. An arterial catheter is useful for monitoring intraoperative blood pressure, perioperative arterial blood gases, and intravascular volume status (through respiratory variation in systolic pressure analysis). Central access is not necessary because this particular procedure is not associated with significant blood loss or fluid shift and it is not likely that vasoactive infusions will be needed. A transesophageal echocardiogram is indicated if cardiac dysfunction is present, and monitoring of filling pressures and cardiac ejection fraction is needed. A pulmonary arterial catheter may be helpful if a TEE is contraindicated (due to esophageal pathology) and there is a need to monitor cardiac performance and systemic vascular resistance.

2. Assuming an intravenous induction, which agents will you choose? Any advantage for propofol? Etomidate? Explain your choice. Should the patient undergo a mask induction with a volatile agent and breathe spontaneously? If you were to have the patient breathe spontaneously for any period during this case, explain your choice of inhalation agent.

3. Assume that you have administered a dose of propofol and started sevoflurane; the patient coughs and has stridor. Mask ventilation becomes difficult. Her saturation is 85%. There is no $ETCO_2$ displayed on the monitor. What would you do? Would succinylcholine be safe to give? How do myasthenic patients respond to succinylcholine? Should the patient receive a nondepolarizing muscle relaxant? How much should be given? Which nondepolarizing agent would you choose?

4. The patient is successfully intubated and is oxygenating well. Your blockade monitor indicates no twitches, yet the patient moves when the median sternotomy begins. The surgeon insists that you have to relax the patient. Which relaxant would you give and how much would you give? How will you judge the efficacy of your muscle relaxant? How do you use a blockade monitor for a myasthenic patient?

## Postoperative Care

### Questions

1. During the thymus dissection, the patient begins wheezing and you note a prolonged upstroke to the exhalation $CO_2$ curve on the capnograph. What are your considerations? How do you diagnose the problem? Albuterol inhalation through the endotracheal tube does not help. With an $FiO_2$ of 1.0, the $O_2$ saturation is 92% and the $ETCO_2$ waveform is small and prolonged. What would you do now? The wheezing is better when the surgeons stop operating. Would endobronchial intubation help? Would a double-lumen tube be indicated? What would you tell the surgeons?

2. I would choose propofol over etomidate. Propofol is a longer-acting agent and results in longer respiratory depression than etomidate, but etomidate (even a single induction dose) can potentially suppress adrenal steroid synthesis and may precipitate adrenal crisis (or unpredictably increase the requirement of a steroid stress dose). Propofol also causes less emesis than etomidate. I would not attempt to have the patient breathe spontaneously throughout this case because volatile agents produce muscle relaxant effect and spontaneous ventilation is almost certain to be inadequate for sternotomy. For any case where spontaneous ventilation was used, sevoflurane would cause the least respiratory depression.

3. Yes, succinylcholine is safe. Because of the paucity of receptors, patients with myasthenia are relatively resistant to succinylcholine; they may require a larger dose than non-myasthenic patients. Caution is appropriate since if a large dose is given, these patients are at risk for phase II block with prolonged muscle relaxant effect. Nondepolarizing agents can be used with the expectation that the onset action could be delayed and the duration of relaxation could be very prolonged. It is reasonable to start with one-half the normally expected dose. Any of the nondepolarizing agents could be used, but I will choose cisatracurium because of its generally predictable duration of action.

4. I will administer cisatracurium, 0.1 mg/kg, in a stepwise fashion, to the desired clinical effect after ensuring an adequate depth of anesthesia. Myasthenic patients have variable involvement of different body muscles, and monitoring multiple sites may be useful to monitor a nondepolarizing agent's effect. Sites might include facial muscles or posterior tibial nerve stimulation. I will obtain a baseline $T_1/T_4$ ratio before the administration of a nondepolarizing agent and titrate the dose of the selected agent to maintain a visible T response. There are many surgical cases where muscle relaxant is not necessary. The need for relaxation should be discussed with the surgeon prior to surgery.

## Answers

1. The wheezing could be due to airway obstruction by surgical manipulation. I will ask the surgeon to stop manipulation of the thymus and the airways and remove any retractors. If the obstruction does not resolve, it may indicate bronchospasm due to light general anesthesia. I will prefer placement of a double-lumen tube to unilateral endobronchial intubation to bypass the surgical obstruction and allow ventilation of both lungs because unilateral ventilation can produce unacceptable shunting and ventilation-perfusion inhomogeneity.

2. Should you reverse the neuromuscular blockade in this patient? Why? How would you reverse the patient? Your colleague will not reverse with neostigmine because of the risks of a cholinergic crisis. What are the symptoms and signs of cholinergic crisis? How is it treated?

3. Are pulmonary function criteria helpful for deciding about extubation? Which ones? What would you do if PFTs are OK but there is a weak gag reflex while the patient is still intubated? Would you extubate?

4. How should pain relief be managed? With a thoracic epidural or a morphine PCA? Why? What are the advantages and disadvantages of each?

## Additional Topics

### Questions

1. A child was playing in a commercial tomato-growing field. She is brought to the ED where she is weak, breathing quickly, delirious, and smells of stool. You are asked to help with intubation and management. What do you think is the diagnosis? Are there specific pharmacologic agents you should use? Is there anything other than atropine? How much atropine should be used?

2. I would reverse the neuromuscular blockade with sugammadex. This drug is a cyclodextrin derivative that reverses the effects of steroidal muscle relaxants. The drug works by encapsulating the muscle relaxant and causing it to dissociate from the nicotinic receptor. It is advisable not to administer anticholinesterase drugs (such as neostigmine) to reverse the neuromuscular blockade in myasthenic patients as this may precipitate a cholinergic crisis. A cholinergic crisis is defined as excessive accumulation of acetylcholine at the nicotinic and muscarinic cholinergic receptors in the CNS and in the periphery. The symptoms are salivation, lacrimation, nausea, vomiting, urinary incontinence, diaphoresis, rhinorrhea, bronchorrhea, muscle fasciculation, weakness and paralysis, laryngospasm, bronchospasm, respiratory failure, miosis, agitation, convulsion, and coma. Treatment is to support the respiratory and cardiovascular systems and offer symptomatic treatment. Benzodiazepines will control seizures and atropine will treat the bradycardia.

3. Pulmonary function criteria are helpful; pulmonary function parameters that are useful to guide extubation of the trachea are a vital capacity greater than 10 mL/kg, maximum negative inspiratory pressure greater than −25 cm of water, a respiratory rate below 20 breaths/minute, an inspired oxygen requirement less than 50%, and PEEP of 5 cm of water or less. Clinically, the patient's bulbar strength (gag and cough reflexes) should be normal. I would not extubate until reflexes were present.

4. Pain control is best managed with thoracic epidural analgesia. The use of thoracic epidural analgesia may minimize the PCA morphine-induced depression of central respiratory drive. The advantage of epidural analgesia with local anesthetic alone is to avoid central respiratory depression caused by neuraxial and systemic opioids. The disadvantage of epidural analgesia with local anesthetics is the potential for intercostal muscle weakness that can impair ventilation and may be confused with inadequate treatment of myasthenia or myasthenic crisis.

**Answers**

1. My preliminary diagnosis is pesticide toxicity as the child was likely present when spraying of the field took place. The pesticides are acetylcholinesterase enzymes that have two components. An acetylcholine molecule, bound at both ends to both sites of the enzyme, is cleaved in two to form acetic acid and choline. In organophosphate poisoning, an organophosphate binds to just one end of the

2. A 13-year-old boy with Duchenne muscular dystrophy (DMD) comes to the operating room emergently for an incarcerated umbilical hernia. What are your considerations for induction? Do these patients have difficult airways? What are your considerations for performing an awake intubation in a DMD patient? Is this stressful? Does it matter? If the patient cannot be intubated and he must have general anesthesia for the procedure, is a mask anesthetic reasonable? Is a laryngeal mask airway with spontaneous breathing a reasonable alternative?

acetylcholinesterase enzyme (the esteratic site), blocking its activity and causing an over-abundance of acetylcholine. I would offer cardiorespiratory support and symptomatic treatment.

If organophosphate toxicity is suspected, decontamination is initiated. The patient's clothing is removed and the intact skin cleansed gently with soap and water and ethyl alcohol (for intact skin), eyes are irrigated with saline, and clothing is disposed of as hazardous waste. Medical personnel decontaminating the patient should self-protect against accidental exposure to the pesticide dust by wearing protective gear (waterproof gowns, gloves such as neoprene, and eye wear protection). Yes, pralidoxime is a specific antidote to organophosphates. Pralidoxime is able to attach to the other half (the unblocked, anionic site) of the acetylcholinesterase enzyme. It then binds to the organophosphate, and changes its conformation, and loses its binding to the acetylcholinesterase enzyme. The conjoined poison/antidote then unbinds from the site and thus regenerates the enzyme, which is now able to function again. It is effective when administered within 48 hours of exposure. Diazepam is used to control CNS excitation and seizures. CNS outcomes are improved if seizures and excitation are controlled. If a plan is made to administer pralidoxime, the patient should receive atropine by repeated administration of 50 mcg/kg q 10–30 minutes to maintain a heart rate above 100 beats/minute.

2. A major concern in adolescents with DMD is difficulty swallowing which may predispose them to aspiration during induction. Some patients may develop an enlarged and stiff tongue as a result of tongue muscle degeneration and replacement with fibro-fatty tissue. Awake tracheal intubation is not a preferable strategy for several reasons: (1) most children at this age are anxious and will not be cooperative; (2) stress-induced tachycardia and hypertension could be detrimental in DMD adolescents since cardiomyopathy is common and tachycardia may lead to decreased cardiac output or ischemia; and (3) gagging and coughing induced by awake intubation are likely to cause respiratory decompensation. No, a mask anesthetic is not indicated because a mask airway may be very difficult to maintain for the duration of surgery. Laryngeal mask airway should be used to secure ventilation and I would attempt tracheal intubation via the LMA with a fiberoptic scope. Patients with DMD are at risk for acute rhabdomyolysis in response to succinylcholine and (occasionally) with exposure to vapor inhalation anesthetics. It is wise to absolutely avoid any exposure to succinylcholine and vapor agents. Adequate depth of anesthesia should be maintained throughout the procedure to avoid myocardial decompensation and cardiac arrhythmias. No, laryngeal mask airway with spontaneous breathing is not a reasonable plan because the patient will likely not be able to maintain adequate tidal exchange due to severe restrictive chest wall disorder. This is particularly true with the use of IV anesthetics.

3. A patient is scheduled for a muscle biopsy to confirm suspicion of "central core disease" (CCD). What is this disease? What are the anesthetic concerns? Is a spinal anesthetic likely to aggravate the condition? Why? Is this similar to multiple sclerosis? Are these children developmentally delayed?

4. An infant with a familial history of familial periodic paralysis is scheduled for a hernia repair.

   Why is the family history important? How should this patient be evaluated preoperatively? Would an ECG help? How about serum electrolytes? What if the potassium is normal? Should you include glucose in the IV? What kind of IV fluid would you use? What about muscle relaxants? Does the muscle relaxant effect of volatile agents potentiate the weakness of familial periodic paralysis?

5. You are asked to sedate a patient with spinal muscular atrophy for a spinal tap. What is SMA? How is SMA treated?

3. CCD is a myopathy, inherited as an autosomal dominant disease. It is a congenital myopathy, often with a mild presentation early in life with variable severity. It arises from defects in the calcium channel that results in the release of calcium into the myoplasm, leading to muscle damage and weakness. Many patients with CCD may also carry the defective gene of the malignant hyperthermia syndrome (ryanodine allele on chromosome 19q13.1) on the same locus as the CCD gene. CCD is associated with congenital muscle weakness that causes scoliosis and spontaneous hip dislocation. Spinal anesthesia may aggravate an existing muscle weakness and hip dislocation. Yes, like multiple sclerosis, patients with adult onset CCD may experience transient worsening with intense activity. However in general, unlike other myopathies, exercise improves muscle strength. Normally, these children have normal intelligence.

4. Familial periodic paralysis is an autosomal dominant inheritable disorder. Preoperative evaluation includes a family history of the disorder and symptoms the infant may have that would indicate symptomatic familial periodic paralysis. Usually the onset of the disorder is in the second decade of life. Symptoms include periodic swallowing, breathing, and limb weakness. ECG can be helpful during the acute episodes of the disorder and may change because of hypokalemia, prolonged PR interval >0.32 s, ST segment depression, T wave inversion, and a prominent U wave. Serum electrolytes are helpful during the acute episode and may show a low serum potassium. There are cases of normokalemic periodic paralysis. Normal serum potassium does not exclude the disorder because it is a self-limited disorder. Probably, glucose administrated should be limited because glycemic stimulation produces an insulin surge, with intracellular shifting of glucose and potassium, resulting in hypokalemia. I will use a nonglucose-containing solution such as normal saline or lactated Ringer's solution for maintenance fluid therapy. It is advisable to avoid muscle relaxants unless necessary for the optimizing surgical exposure. Yes, volatile agents can potentiate skeletal muscle weakness.

5. The disease is caused by degeneration of the anterior horn cells. The infantile form manifests within the first 3 months of life and is usually a severe form of the disease. Severe muscle weakness is associated with difficulty swallowing, secretion handling, and breathing. These infants are at risk for aspiration and may require postoperative ventilatory support. The Kugelberg-Welander disease is a milder form of the disease and progresses slower than the infantile form. Yes, this syndrome is associated with hypoplasia or agenesis of the cranial nerve nuclei in the brainstem. The cranial nerves are primarily motor nerves, and hence this syndrome is analogous to SMA syndrome because it involves lower motor neuron degeneration. Both conditions affect motor neurons. Amyotrophic lateral sclerosis specifically affects the motor cortex, spinal motor neurons, or both. Unlike SMA, amyotrophic lateral sclerosis affects both the upper and lower motor neurons. It may manifest as spastic weakness as opposed to flaccid weakness in SMA syndrome. These patients have weak and atrophic muscles of res-

6. You are called to intubate an 18-year-old in the ICU. He has been in the ICU for a week following a flu shot, which resulted in progressive weakness—beginning with walking and now progressive respiratory failure. What is your diagnosis? NG tube feedings were stopped 1 hour ago. Should he be intubated awake? Would you perform?

piration and may require ventilatory support postoperatively. They are at risk for aspiration pneumonitis and postoperative pneumonia due to inability to cough effectively from weakened bulbar reflexes. They are intolerant to sedative, hypnotics, and opioids due to reduced respiratory muscle reserve. Genetic counseling of the family is important to determine whether the disorder is a result of mutation or genetic deletion of survival motor neuron (SMN), which occurs in approximately 90–94% of SMA patients. Counseling is also necessary for prenatal screening with subsequent pregnancy; the screening test has 98% reliability. This information makes this SMN gene test very useful for the diagnosis of SMA. Anesthetic agent choices are made bearing in mind that patients with generalized muscle wasting are unable to protect airways and have limited respiratory system reserve. Therefore, these patients have increased sensitivity to nondepolarizing muscle relaxants and are unable to compensate for hypoventilation and decreased central respiratory drive following administration of CNS depressants such opioids for postoperative pain control. In the past several years, a promising treatment regimen involved the use of the drug nusinersen. This antisense oligonucleotide acts as a splicing modifier and changes the relative concentration of nonfunctional proteins that cause the disease. The drug must be delivered intrathecally. This can be challenging given the severe scoliosis that often characterizes these patients as they age.

6. Possible etiologies include post-viral syndromes and metabolic or autoimmune disorders. Following a flu shot, the most likely etiology for this adolescent is Guillain-Barré syndrome. No, the patient should be anesthetized with ketamine or propofol if hemodynamically stable. If hemodynamically unstable, intubation can be achieved with IV midazolam and a low dose of etomidate. Yes, rapid sequence induction can be performed without the use of a muscle relaxant because the bulbar muscles are either paralyzed or weak enough from the disease to allow adequate rapid intubation condition. Succinylcholine should be avoided because in the acute phase of Guillain-Barré disease, there is an active demyelination process that likely predisposes to serious hyperkalemia.

## Suggested Reading

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# Chapter 46
# Endocrinopathies

**Thomas J. Mancuso**

An 8-year-old girl, 22 kg, previously healthy, presents with diffuse abdominal pain most pronounced in the right lower quadrant, lethargy, weakness, and recent weight loss. She is nauseous and has vomited twice in the ED and several times at home over the preceding few hours. She has a "strange" odor to her breath. Her urine is dipstick + for glucose and ketones. You are called to the emergency room to evaluate her in preparation for appendectomy.

VS: HR 140/min, RR 38/min, BP 82/66 mmHg, T 38.2 °C.



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## Preoperative Evaluation

### Questions

1. Is this patient's intravascular volume depleted? How would you assess her volume status? What factors would you address in correcting her volume status preoperatively? Which intravenous fluid would you choose?

2. What metabolic studies are important to evaluate preoperatively? How will this influence your preparation for surgery? Should the case be delayed? If so, why? How long? Is this problem likely to resolve with improvement of her metabolic status? What organ system consequences are there of uncontrolled diabetic ketoacidosis in the acute phase and chronically?

## Answers

1. It is very likely that this child has a depleted intravascular volume. Her heart rate is elevated more so than would be due to a low-grade fever. In addition, with fever, insensible losses increase and she has been vomiting. Her blood pressure is low but within normal limits for an 8-year-old girl. Since blood pressure is preserved in hypovolemic children until compensation fails, a "normal" measurement is not reassuring. Dehydration is generally classified according to percentage decrease in body weight. A child who is 3% dehydrated has a 30 mL/kg deficit and clinically has an increased HR, dry mucous membranes, and concentrated urine. A child with 6% dehydration has a fluid deficit of 60 mL/kg and a significantly increased HR, very dry mucous membranes, and oliguria. In a child with 9% dehydration, the fluid deficit is 90 mL/kg, the blood pressure is decreased, and there is poor capillary refill, Kussmaul breathing, and obtundation. Fluid replacement should be with isotonic solution such as normal saline or lactated Ringer's and should be given relatively rapidly.

2. Serum electrolytes (Na, K, Cl, $HCO_3$) and, in addition, phosphorus and calcium, anion gap, glucose, and perhaps blood gases should be evaluated preoperatively [1]. If the child is in diabetic ketoacidosis (DKA), surgery should be delayed at least until intravascular volume has been replenished and control of her DKA is underway. It is entirely possible that her abdominal pain is due to DKA and not any surgical problem [2]. Conversely, acute appendicitis can be the insult that precipitates DKA in a child who has diabetes mellitus but has not yet come to medical attention. While a big part of the problem in DKA is dehydration, overly aggressive replenishment can lead, in some cases, to the development of cerebral edema [3]. Cerebral edema has been documented in many patients during DKA, but most patients remain asymptomatic. Fluid administration should be kept to $<4$ L/m$^2$/day. In addition, insulin infusion should be tailored to keep the decrease in glucose concentration to 100–180 mg/dL. During therapy for DKA, frequent measurement of serum osmolality is important in preventing a worsening of the cerebral edema.

## Intraoperative Course

### Questions

1. What are your monitoring considerations? Does this patient need an arterial line? Should the patient have a central venous catheter? A urinary catheter? How would these monitors help your management? Are there any confounding issues?

2. What agent would you choose to induce general anesthesia? What effects of the various induction methods are particularly important for diabetic patients? For maintenance, which inhalation agent would you choose? Is there a place for a regional anesthetic in this case?

3. Is succinylcholine safe to give to this patient? What if the pH is 7.25? What is the effect of ketoacidosis on pharmacokinetics of nondepolarizing neuromuscular blockers? Would you administer bicarbonate?

## Answers

1. Standard ASA monitors are indicated, and given the frequency with which serum glucose, electrolytes, and pH will be checked, there is a strong case for adding an arterial line as well. The line will also be very helpful in postoperative management. The case for a central venous catheter is not as strong. If two adequate peripheral IVs and a Foley catheter are in place, she can be managed without a CVP. Urine output will not be a good measure of preload since she will have an osmotic diuresis due to glycosuria. However, the quality of the arterial waveform as well as the improvement of her metabolic acidosis is indicative of the adequacy of her intravascular volume. If there is a question of access or she remains unstable despite what is thought to be adequate fluid replacement, a CVP catheter should be placed after the induction of anesthesia.

2. The induction of anesthesia should proceed with the assumption that she is not fully fluid resuscitated. This patient should have an intravenous induction. Given her nausea and vomiting, she should have full-stomach precautions. Any IV agent can be used if dosed appropriately. Propofol will lead to hypotension if given in the usual doses to a hypovolemic patient. Etomidate will suppress the adrenal cortex in this child with new-onset diabetes mellitus, DKA, and possible appendicitis. The choice of muscle relaxant to facilitate intubation presents difficulties. A nondepolarizing relaxant will take longer to provide intubating conditions in this child who would do better without mask ventilation, while succinylcholine will cause an increase in the serum potassium in a patient who may already have an acidosis-related elevation of serum potassium; it is important to check electrolytes preoperatively.

3. Succinylcholine can be used but there may be problems. In patients with metabolic acidosis, hypovolemia, and/or hemorrhage, the administration of succinylcholine causes a greater increase in serum potassium that it does in healthy patients. If succinylcholine is used and hyperkalemic arrhythmias occur, treatment with hyperventilation, calcium chloride, and bicarbonate should be started immediately. Nondepolarizing relaxants are also affected by the presence of metabolic acidosis. Metabolic acidosis may augment the neuromuscular blockade from a nondepolarizing relaxant. The antagonism of blockade is not impaired by metabolic acidosis as it is by respiratory acidosis; however, bicarbonate administration should be reserved for severe acidosis (pH <7.2). There are several possible adverse outcomes from bicarbonate administration. Alkalosis will increase potassium entry into cells; bicarbonate may worsen the CNS acidosis. $HCO_3^-$ combines with $H^+$ to form $H_2O$ and $CO_2$ and $CO_2$ diffuses rapidly into the CNS while $HCO^-$ does not.

4. Intra-op ABG: pH = 7.22, $PaO_2$ = 160 mmHg, $PaCO_2$ = 34 mmHg, K = 5.5 mEq, Na = 127 mEq, Cl = 97 mEq, glucose = 990 mg/dL.

   How should this be managed? What implication does this have for your anesthetic choice and technique? Should this be more of a "stress-free" anesthetic? Can this be accomplished with an inhalation anesthetic, or would a narcotic anesthetic technique be preferable?

## Postoperative Care

### Questions

1. Should this patient go to the ICU postoperatively? What are you particularly concerned about? How frequently should the patient be metabolically monitored postoperatively?

2. You are called to the PACU for a urine output of 7 mL in the first 2 h postoperatively. How do you evaluate this? Is a fluid bolus indicated? If so, what type of IV fluid? Is placement of a CVP catheter indicated? Would a urine analysis help understand this situation?

4. The ABG shows a metabolic acidosis and partial correction with a respiratory alkalosis. The serum glucose is markedly elevated, at 990 mg/dL. The patient's DKA is not being treated effectively at all. Therapy should include further fluid resuscitation and an additional regular insulin dose of 0.5 U/kg while the insulin infusion continues. The patient has hyponatremia and at the same time hyperosmolality. Serum osmolality can be calculated from the electrolytes as follows:

$$(\mathrm{Na} + \mathrm{K}) \times 2 + \mathrm{glucose}/18 + \mathrm{BUN}/3$$

If we assume the contribution of BUN to osmolality is 10, then the equation reduces to

$$S_{osm} = (132.5 \cdot 2) + \mathrm{glucose}/18.(132.5 \cdot 2) + 10 + 990/18 = 330$$

The hyponatremia may only be apparent, not real, if the water content of the plasma is reduced by the presence of excess lipids.

**Answers**

1. This patient will be better off in an ICU overnight so that she can have frequent monitoring of her metabolic condition. Even if she had not undergone surgery, her condition was serious enough to warrant an ICU admission. She has arrived with a very large fluid deficit and severe metabolic acidosis. She is at risk for cerebral edema during her resuscitation, and the initial signs of raised ICP might easily be missed if she were not in an ICU. This patient should have hourly determinations of serum glucose, potassium, sodium, pH, and $HCO_3$. Her urine output, urine ketones, and glucose also should be monitored very often.

2. Depletion of intravascular volume is a major part of the pathophysiology of DKA. The low urine output noted in the patient in the PACU can likely be attributed to that problem. Assessment of the degree of dehydration in the postoperative period is similar to what was done preoperatively, namely, history (anesthesia and ED records), physical exam (vital signs, skin turgor, and mental status), and laboratory (urinalysis, electrolytes, and glucose). An abnormal mental status can be due to one or more of several factors: hypo-/hyperglycemia, electrolyte disturbances, acidosis, raised ICP, residual anesthetic medications, and hypovolemia. Urinalysis is also complicated in this situation. Glycosuria will affect the

3. Should this patient's pain relief be managed with an epidural or morphine PCA? What are the advantages and disadvantages of each?

## Additional Topics

### Questions

1. A 14-year-old male presents with growth failure, increased intracranial pressure, and visual field cuts (binasal hemianopsia). His imaging workup so far is consistent with craniopharyngioma. Are there any endocrinopathies you would suspect in this patient? What if he had short stature? What if he was obese? Was still a Tanner stage 1?

2. An 18-year-old with acromegaly presents for sagittal split osteotomy to correct mandibular prognathous. Would you expect airway difficulties? Are there any metabolic abnormalities to be prepared for?

specific gravity determination and will also cause an osmotic diuresis. A fluid bolus is indicated. There is now uncertainty about the role fluid IV rehydration in these children plays in the neurologic outcomes. Nevertheless, in the absence of hypovolemic shock, care should be taken with the speed of administration [3]. A CVP catheter would be a good guide to fluid administration and help minimize the risk of cerebral edema. Clinically, cerebral edema is not present when the child presents with DKA but develops during therapy for DKA, often as biochemical measures are actually improving.

3. Pain management for this 8-year-old child following an appendectomy incision should be easily accomplished with IV opioids and IV PCA. Generally, these patients begin oral intake within 24 h of surgery and are easily switched to oral analgesics. Regional analgesia offers little additional benefit for the risks, albeit low, involved in placement of an epidural catheter. Provided she has not suffered any damage to her kidneys during her DKA episode, ketorolac can be added to her analgesic regimen.

**Answers**

1. Craniopharyngioma, a benign suprasellar tumor, is one of the most common supratentorial tumors in children. Signs and symptoms are due to the location of the tumor. It may be confined to just the sella turcica or it may extend through the sellar diaphragm and compress the optic nerve with resulting visual field cuts as in this case. Pituitary-hypothalamic involvement leads to short stature, and if the tumor extends into the third ventricle, hydrocephalus may result. Most craniopharyngiomas have calcifications, and these are visible on plain films or CT. Adrenal and thyroid dysfunction also are possible in these children. The preoperative assessment of these patients should include an evaluation for the various endocrine abnormalities discussed [4]. Hypothyroidism and/or adrenal insufficiency can cause problems if not managed appropriately. Although DI may be part of the initial presentation, it is more often seen postoperatively [5].

2. Acromegaly is the result of oversecretion of growth hormone (GH) in a person with closed epiphyses. If excess GH is due to a pituitary adenoma, it is possible that the tumor growth will compromise other anterior pituitary function. Secretion of gonadotropins, thyrotropin, or corticotropin may be impaired. The airway may be

3. What is the relationship of Uosm to Posm in the syndrome of inappropriate antidiuretic hormone (SIADH) secretion? Why? How does furosemide work to correct this problem? Should furosemide be utilized intraoperatively if this diagnosis can be made, or should fluid restriction be the treatment of choice?

4. A 3-month-old presents for a cleft lip repair; she has a funny, puffy face, with widely separated fontanelles and a flattened nasal bridge, consistent with congenital hypothyroidism. She breathes through her mouth that has a large, protruding tongue. Her skin is cool to the touch and "marbleized" (cutis marmorata). What anesthetic concerns does this patient present?

5. A 15-year-old presents with a posterior mediastinal mass extending from T3 to T8. What is your differential diagnosis? What would you specifically attempt to evaluate by history in order to differentiate? Preoperative lab work? Radiological studies? The 24 h catecholamines are still pending; as you are placing the epidural in the thoracic space (patient awake, sedated, and comfortable), you notice that the blood pressure is 220/130. What do you do? (You recycle the cuff and it is about the same result.) Should the case be delayed, or can you proceed safely, with your new knowledge?

involved in this disorder [6, 7]. In addition to growth of bone, excessive GH secretion causes enlargement of the tongue and epiglottis. Cases of difficult intubation have been reported, as has laryngeal stenosis. The patient should be questioned about dyspnea and examined for stridor. Peripheral nerves can become trapped by overgrowth of bone and connective tissue leading to various neuropathies.

3. In SIADH, urine osmolality > serum osmolality, Uosm > Posm with serum Na <135 mEq/l, and urine Na concentration >40 mEq/l. In SIADH, levels of antidiuretic hormone are inappropriately high for the osmolality of the blood and do not decrease with further dilution of the osmolality. There are many causes of SIADH such as CNS disorders, pneumonia, and the use of positive pressure ventilation. SIADH is a known side effect of vincristine administration. The signs and symptoms of SIADH are due to the hyponatremia that results from water intoxication. Common signs and symptoms of hyponatremia include weakness, fatigue, confusion, headache, nausea and vomiting, seizures, and coma. In general, treatment of the underlying disorder will correct the problem. Treatment of the hyponatremia itself with fluid restriction and administration of maintenance sodium often will correct hyponatremia. In severe cases, administration of a diuretic such as furosemide will induce a diuresis and eliminate some of the excess water [8].

4. Congenital hypothyroidism can occur sporadically or in a familial pattern. Newborn screening programs are in place throughout the country, and most children with this disorder are detected this way. The overall prevalence is 2.5:10,000 (0.5:10,000 in African-Americans and 5:10,000 in Hispanics), and it is seen twice as often in females compared with males. Although there are many causes, most are due to thyroid dysgenesis, either aplasia or rudimentary ectopic thyroid tissue. In cases where the diagnosis is missed by the neonatal screen, clinical manifestations may not be evident at birth due to the presence of transplacentally acquired maternal thyroxine (T4).

5. Mediastinal masses can have deleterious cardiovascular effects on affected patients, and these effects are significantly worsened during the induction of anesthesia [9]. The differential diagnosis of a mediastinal mass depends upon the location in anterior, middle, or posterior division of the mediastinum:

   - Anterior mediastinum: lymphoma, lymphangioma, teratoma, and thymoma
   - Middle mediastinum: bronchogenic cyst, granuloma, lymphoma
   - Posterior mediastinum: enteric cysts, neuroblastoma, ganglioneuroma

   While all locations can cause airway obstruction, anterior mediastinal masses also often decrease cardiac output by impairing filling of the right atrium (RA) and right ventricle (RV). Affected children will present with a superior vena cava (SVC) syndrome in addition to any signs and symptoms of airway compromise [10]. History and physical exam can give the anesthesiologist clues to the degree

6. An 8-month-old girl with ambiguous genitalia and virilization is scheduled for urological reconstructive surgery. Any metabolic derangements you should expect in the preoperative evaluation? What if her $Na^+$ was 121 mEq and $K^+$ 5.9 mEq? What problems would you expect? How could this be managed medically? What problems would you expect with surgery? How should her fluids be constituted? Should she be treated with fludrocortisone (Florinef) and hydrocortisone? Hydrocortisone only?

of airway and cardiovascular impairment. The presence of stridor and/or difficulty breathing in various positions should be reviewed and progression of these complaints over recent time evaluated. Dilated veins on the face and upper extremities may be present. The preoperative laboratory evaluation of children with anterior mediastinal masses should include, in addition to as thorough an evaluation of the airway as possible, an echocardiogram [11, 12]. The mass may not impinge on the trachea but impair RA filling, a very dangerous situation. Older children with mediastinal masses should undergo imaging studies as well as flow-volume loops [9]. A CT scan of the chest and airway will give important information about tracheal size and/or deviation caused by the mass. Younger children often do not cooperate with the positioning and immobility needed for a CT scan or the more demanding pulmonary function testing. In these cases, the clinical assessment is even more important. General anesthesia for children with anterior mediastinal masses presents many challenges to the entire OR team, anesthesiologists, surgeons, and OR RNs [13]. One of the possible diagnoses for a posterior mediastinal mass is neurogenic tumor including pheochromocytoma. The elevation of blood pressure noted during placement of the epidural may be the result of excess catecholamine release by the pheochromocytoma. Anesthetic care of patients with pheochromocytoma requires careful preoperative evaluation and preparation. Once the diagnosis is confirmed with measurement of metanephrine, a metabolite of norepinephrine in the urine and in plasma, a thorough search for the full extent of the tumor is undertaken. In addition, prior to going to the OR, the patient should be treated with alpha-blocking medications until orthostatic blood pressure and heart changes are evident. Only when this is accomplished can beta blockade be administered. Even once satisfactory alpha and beta blockade has been accomplished, the procedure can be safely undertaken provided the anesthesiologist is prepared for significant vital sign swings. Invasive hemodynamic monitoring is indicated. Induction and maintenance should be planned to minimize the response to surgical stress. Epidural analgesia is a useful adjunct for these patients.

6. Ambiguous genitalia in females are the result of a defect in the enzymes responsible for the synthesis of cortisol. The most common form is due to deficiency of 21-hydroxylase, an enzyme located in the adrenal cortex in the pathway to production of cortisol and aldosterone from cholesterol. The precursor 17-OH- progesterone increases in concentration, leading to excessive production of androgen. This is the cause for the virilization seen clinically. In addition to impaired cortisol production, aldosterone production is also affected. Decreased levels of aldosterone, another result of the 21-hydroxylase deficiency, lead to hyponatremia and hyperkalemia. One of the clinical variants of 21-hydroxylase deficiency, salt wasting, can present as an Addisonian crisis with severe sodium loss. Perioperative treatment of 21-hydroxylase deficiency patients involves the administration of hydrocortisone, 2 mg/kg, every 6 h. If salt wasting is present, IV fluid replenishment with NaCl-containing solutions and administration of an IV mineralocorticoid such as fludrocortisone are indicated [14, 15].

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Anghelescu DL, Burgoyne LL, Liu T, et al. Clinical and diagnostic imaging findings predict anesthetic complications in children presenting with malignant mediastinal masses. Paediatr Anaesth. 2007;17:1090–8.

This retrospective review of the records of 118 pediatric patients with mediastinal masses was undertaken in an effort to identify specific historical, physical exam, and laboratory findings that predict complications when anesthesia is induced. In this series, 11 patients did experience anesthesia-related complications. Orthopnea, upper body edema, great vessel compression, and mainstem bronchus compression were significantly associated with anesthesia-related complications.

Connery LE, Coursin DB. Assessment and therapy of selected endocrine disorders. Anesthesiol Clin North Am. 2004;22:93–123.

This review includes discussion of general anesthetic principles of perioperative care, primarily of adult patients, with diabetes mellitus, thyroid disease, adrenal insufficiency, and pheochromocytoma. Even though the discussion includes etiology and pathophysiology for adults with these disorders, many management issues included have some relevance in the care of children.

Hack HA. The perioperative management of children with phaeochromocytoma. Paediatr Anaesth. 2000;10:463–76.

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Management of the cardiovascular changes expected during surgical removal is also reviewed.

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Rhodes ET, Ferrari LR, Wolfsdorf JI. Perioperative management of pediatric surgical patients with diabetes mellitus. Anesth Analg. 2005;101:986–9.

This paper reviews the epidemiology of diabetes mellitus, current outpatient management options for children with this condition, the effects of surgery and anesthesia on glycemic control, as well as perioperative management of affected children.

# Chapter 47
# Equipment and Monitoring

**Robert S. Holzman**

A 13-year-old who developed abdominal pain and claudication was diagnosed with severe midaortic syndrome. She is very hypertensive (initial presentation 210/142), with calcification and vessel wall thickening of the aorta, bilateral iliac arteries, renal arteries, and the right subclavian artery. She has a severe ascending aortic aneurysm with near-complete stenosis of the abdominal aorta at the level of the renal arteries without a distal abdominal aorta nor common iliac arteries. In addition, the left common and internal carotid artery is severely narrowed and the intercavernous extent of the right internal carotid artery is also narrowed. She has claudication with walking half a city block. Meds: amlodipine, metoprolol, minoxidil, and clonidine. VS: 142/92, 86, 16. T 370 C. $SpO_2$ = 99%.

She is scheduled for left thoracotomy, partial left heart bypass, proximal anastomosis, exploratory laparotomy, aortobiiliac bypass with bifurcated graft, reperfusion and reimplantation of the celiac, and superior mesenteric and left and right renal arteries.



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## Preoperative Evaluation

### Questions

1. How will you evaluate the severity of her ischemic disease? Standard Bruce protocol stress test? Dobutamine stress scan? MRA? Cardiac angiogram? Persantine scan? Echo? Can you do a CT angio and accomplish the same findings in one study? What are you most worried about?

2. How would you evaluate the severity of her peripheral vascular disease?

3. The adequacy of her renal circulation with regard to the end point of preservation of function?

## Intraoperative Care

### Questions

1. Does this patient need an arterial line? Why/why not? Can you get the same information from a pulse oximeter + end-tidal $CO_2$ analysis? What is the difference?

2. How does the pulse oximeter work?

**Answers**

1. She will not be able to do a standard treadmill stress test because of her claudication. The results of dobutamine stress testing evaluation by echocardiogram or dipyridamole scanning are acceptable in this circumstance, although exercise stress testing is more accurate. Stress (exercise or pharmacologic) transthoracic echocardiography can also be used to detect hemodynamically significant coronary artery disease. Coronary artery anatomy can be defined by magnetic resonance angiography although she will likely have CT angiography to delineate her vascular anatomy and the coronary arteries will be imaged at the same time.

2. The severity of her claudication will provide a major clue to the degree of circulatory impairment while the imaging studies will confirm the anatomy. The more severe her claudication, the worse her lactic acid level including the rate of rise of lactic acid intraoperatively, especially when she is being revascularized or during bypass.

3. The renal circulation is of particular concern because it is one of the revascularization sites, which means that it will have to be cross-clamped for a period of time. Susceptibility to injury is greater because of the underlying disease.

**Answers**

1. The patient definitely needs an arterial line for careful monitoring of blood pressure, blood gases, and additional laboratory data. Although a pulse oximeter and $ETCO_2$ analysis will provide excellent feedback about the quality of oxygenation, ventilation and circulation, dynamic changes in the circulation as a result of blood loss, exposure requirements with retraction and its effects on contiguous structures, and metabolic monitoring make arterial access mandatory.

2. A pulse oximeter measures the ratio of the concentration of oxyhemoglobin to the combined concentration of oxy- plus deoxyhemoglobin, taking advantage of the fact that blood absorbs light differently depending on the level of oxygen it contains. Pulse oximeters direct beams of red and infrared light across a finger, earlobe, etc. and a detector on the opposite side calculates the ratio of red to infrared light, comparing the calculation to standard algorithms to create an

3. How does a transducer work?

4. Is $ETCO_2$ equivalent to $PaCO_2$?

5. Would a TEE add any additional information not otherwise obtainable with the monitors you have in place?

oxygen saturation level. The arterial (pulsatile) fraction is calculated by subtracting peak from trough levels; otherwise, other structures through which the light passes, such as skin, bone, and connective tissue, would also be part of the final saturation.

3. As a general statement, transducers modify one form of energy into another. Pressure transducers, typically used for intravascular monitoring in the operating room, measure liquid (or gas) pressures. Most modern pressure transducers at this point work by altering resistance across a Wheatstone bridge, thereby altering voltage. This results in an electrical pattern in proportion to the pressure changes. The key physics concepts involve frequency and damping. Most transducers have frequencies of several hundred Hz, because the natural frequency of the measuring system must exceed the natural frequency of the arterial pulse (16–24 Hz). If the frequency of the monitoring system is too low, the monitored pressure waveform frequency will approach the natural frequency of the measurement system, and the system will resonate. The result will be amplification of the true intra-arterial pressure, such as overshoot, or ringing, which is what happens with tachycardia and steep systolic pressure upstrokes—the higher frequency of these waveforms approaches the resonant frequency of the measurement system. A transducer system must also have an appropriate damping coefficient because the addition of tubing, stopcocks, and air all decreases the frequency of the system, leading to overdamping and underestimation of systolic pressure. An overdamped pressure waveform has a slurred upstroke, absent dicrotic notch, and loss of fine detail. An underdamped pressure waveform displays systolic pressure overshoot as well as additional artifacts, leading to false conclusions about an elevated blood pressure.

4. The $ETCO_2$ is not equivalent to $PaCO_2$ because it reflects the ratio of dead space to tidal volume, with a typical difference of 5–7 mmHg. In small infants and children, the accuracy of the $ETCO_2$ depends on the percent of the exhaled breath actually measured (which can vary with the use of an uncuffed endotracheal tube in pediatrics, for example) and the maximum expiratory flow rate of the patient relative to the sampling rate of the capnograph. The greater the patient's MEFR and the more rapid the sampling rate, the more accurate the $ETCO_2$ due to minimal "slurring" of the end-tidal trace. The gas sample should be taken as close to the patient's airway as possible; ideally, it should be sampled from within the endotracheal tube, but most typically, it is sampled from the elbow connector.

5. A TEE will be helpful diagnostically as well as a monitor to assess myocardial contractility particularly in response to cross-clamping of major vascular structures. It will also aid in assessing left ventricular volume and the potential for aortic dissection following the arch reconstruction. Caution is warranted for placement and manipulation in the anticoagulated or coagulopathic patient.

6. Any special monitors you would like to have with regard to partial left heart bypass? How does near-infrared spectroscopy work? Would it be useful for brain monitoring? Might it be useful in any other organ systems?

7. She has had a series of cross-clamp times that add up to about 5 hours in various splanchnic circulations, in accordance with her surgical plan outlined above. What consequences would you expect? How would this likely be reflected in your available monitoring devices? Anything additional monitoring techniques or instrumentation that you would like to use? Treat with?

8. Is this patient at risk for hypothermia? What is this patient's ideal intraoperative temperature? Optimal temperature for coagulation and hemostasis? Optimal temperature for organ system function? Optimal temperature for cerebral preservation? What are your concerns about choosing a temperature maintenance system? What about her ischemic extremities? Are there risks of hyperthermia?

6. Near-infrared spectroscopy should be strongly considered for this procedure. As a monitor of cerebral oxygenation, it will be particularly useful for the aortic arch reconstruction which will involve clamping and altering blood flow into a compromised carotid circulation. Cerebral NIRS works by passing infrared light through the scalp, skull, and cranial contents and measuring cerebral tissue oxygenation. This is different than pulse oximetry because it measures an uncertain mix of arterioles, capillaries, and venules. Nevertheless, it is a venous-weighted signal (larger venous hemoglobin mass) because the entire returned signal is measured rather than just pulsatile measurements. Because there is significant variability between individuals, it is crucial to establish baselines. As a general guideline, values less than 45–50% are associated with increased anaerobic metabolism and lactate production. Multisite oximetry can monitor multiple organs, often brain plus another organ system such as kidney, liver, intestine, or muscle, and may detect hypoperfusion in regional circulations during shock.

7. The consequences of clamping and unclamping in various regional circulations are similar hemodynamically and physiologically but vary in accordance with the size of the vessels involved. Major (e.g., aortic) arterial vascular cross-clamping results in hemodynamic as well as physiologic changes. The hemodynamic changes are characterized by increases in central venous and pulmonary artery pressure, left ventricular wall tension, segmental wall motion abnormalities, arterial blood pressure, and coronary artery blood flow. There are decreases in ejection fraction and cardiac output as well as renal blood flow. Physiologic consequences include a decrease in total body oxygen consumption and oxygen extraction, a decrease in total body carbon dioxide production, and an increase in mixed venous oxygen saturation with an accumulating metabolic acidosis. Invasive hemodynamic monitoring as well as arterial blood gases with lactate monitoring should reflect the severity of these consequences. Unclamping results in decreased myocardial contractility and cardiac output as well as blood pressure, with an increase in total body oxygen consumption and a decrease in mixed venous oxygen saturation. This can be treated with a decrease in inhaled anesthetics and vasodilators, increased fluid administration, and increased vasoconstriction. Sodium bicarbonate should be considered, but it will also result following biotransformation into an increased $CO_2$ load as the bicarbonate volatilizes; therefore, THAM (tris(hydroxymethyl)aminomethane), especially in this patient with severe vascular disease and pre-existing lactic acidosis, is an important alternative since it doesn't result in the production of a $CO_2$ load.

8. Yes, she is at risk for hypothermia. Given the planned surgery, ideal intraoperative temperature for this patient should probably be directed toward moderate hypothermia in order to decrease organ system oxygen demand in a varying perfusion environment. I would try to keep her at 35.5–36 degrees centrally. PTT increases below 340 C and coagulation factor activity and platelet function

9. About 3 hours into the case, the serum lactate has increased from 2.4 to 5.2 and the temperature is now 34.5°. What do you think is going on? What would you need to confirm the diagnosis? What will your strategy be? Let's say the pH is 7.22. 7.15? Is bicarbonate the best choice? Would you hyperventilate? Use THAM? Wait awhile and get another ABG?

10. The patient is now 33.8 °C. How did this happen? Where did it happen? What mechanisms of heat loss are most significant? Is monitoring of temperature a standard of practice? Is it worthwhile putting in a heat and moisture exchanger in the breathing circuit at this time? Earlier?

decline below 330 C. The room should be sufficiently warm (approx. 210 C) to allow reasonable maintenance of a thermal neutral environment. The thermal neutral environment is that environmental temperature that minimizes the temperature gradient from the patient to the environment, resulting in shivering and heat loss to the environment with the resultant thermal stress to the patient. Other mechanisms for heat loss include convection and conduction heat loss. A blanket warmer will specifically address conduction heat loss when applied under the patient but will address convection and radiation heat loss (the majority of heat loss) when applied over the patient. A Bair Hugger, or forced warm air warmer, will also address convection and radiation loss when placed above the patient and is also a good idea. With impaired circulation, caution has to be exercised with regard to heat delivery; however, direct inspection of extremities periodically should suffice for monitoring skin integrity. Hyperthermia is not a likely possibility because cross-clamping and massive tissue exposure are far more likely to predispose to hypothermia. Hyperthermia would pose the risk of increased oxygen consumption in all organ beds for this patient, in the setting of impaired perfusion and oxygen delivery, so it should be avoided.

9. This is a very substantial increase and worrisome trend, although it is hard to say exactly the clinical significance, since she is already starting out abnormal. Severe lactic acidosis is generally considered greater than 5 and hers is now 5.2. Levels of 6–10 are associated with high mortality. This represents a Type A lactic acidosis where tissue oxygen deliver is inadequate and not a Type B acidosis where there is no evidence of reduction in tissue oxygen delivery, such as in an inborn error of carbohydrate metabolism. The diagnosis of Type A lactic acidosis due to ischemia is context-dependent, so with severe anemia, blood loss, and hypoperfusion, it is much more likely to be Type A when an abnormally elevated lactate comes back from the lab.

   Supporting evidence beyond the clinical context would be multisite NIRS measurements that were consistent with regionally impaired perfusion in major vascular beds like bowel, liver, or kidneys. Treatment with vasoactive amines often results in an increased lactate production due to the stimulation of glycolysis. Treatment with sodium bicarbonate can lead to an increase in lactate production because the intracellular acidosis strongly inhibits phosphofructokinase, the rate-limiting enzyme in glycolysis. Creation of an alkalotic intracellular milieu disinhibits phosphofructokinase, worsening the acidosis and making it appear as if more bicarbonate is required. Hyperventilation is likely to result in worsening of hemodynamics because of the rise in minute mean airway pressure. THAM may be a reasonable option to put the patient back into a more physiological (pH > 7.2) range.

10. This temperature decrease is very worrisome and is not only a consequence of getting into trouble because of inadequate perfusion but will also produce its own set of problems. The hypothermia is a result of dangerously decreased metabolic activity because of ischemic injury with ever-increasing lactic acid

11. You decide to transfuse for a HCT of 23 and no surgical end in sight. The nurse brings you cold blood from the refrigerator, which she checks with you to your satisfaction. Now what? How to warm? Why do you warm blood anyway? What different methods are available? What about filters? How large? What particles? How important? How does a countercurrent blood warmer work? Is there a warmer available on the rapid infusion system? What is a dry heat warmer?

## Postoperative Care

### Questions

1. The patient remains intubated after 18 hours of surgery and is brought directly to the ICU.

   The respiratory therapist inquires: "How do you want the vent set up, doc?" Your answer?

2. What is the difference between volume, pressure, and pressure support ventilation?

levels reflecting this. Warming should be undertaken cautiously, lest it obligate the patient to metabolic activity she is incapable of. Heat loss should address all three mechanisms: convective heat loss should be addressed by covering the patient insofar as possible; conductive heat loss should be addressed by warming contact surfaces (i.e., an underbody Bair Hugger), and radiant heat should be applied in the surgical field, at a safe distance, via heat lamps. The room temperature can be turned up. If not done already, circuit fresh gas flows should be reduced to those that approach closed circuit flow, approximately 250 mL/minute for an adult (at normothermia; 7% less for every degree C decrease). Temperature monitoring "shall be continually evaluated" according to ASA standards. An HME works basically as an airway insulator by passively trapping heat and water vapor loss from the airway. As it is, heat loss from the airway is a very minor source of overall heat loss, and in an already hypothermic patient, the addition of an HME is hardly worthwhile. It would be a little more helpful with a normothermic patient.

11. There are a variety of fluid/blood warming systems. Routine and rapid infusers are the broad breakdown. Routine fluid warmers will deliver approximately 150 mL/minute compared with 750–1000 mL/minute with rapid infusion systems. The methods of heating may be dry heat, countercurrent heat exchange, water immersion, or passing through or around a heating device, such as a Bair Hugger. Particulate filters (170–260 microns) are routinely used to prevent the administration of clots or particulate debris. They are different than microaggregate filters which trap smaller particles and are not generally used in the operating room.

**Answers**

1. Mechanical ventilation has to provide a minute ventilation adequate for the elimination of carbon dioxide to approximately 5% of an atmosphere ($PaCO_2$ approximately 40 mmHg, or ETCO2 approximately 34 mmHg, if the dead space to tidal volume ratio is normal). For adults, this is typically around 80 mL/kg/minute. The settings of the ventilator will ultimately depend on the pulmonary compliance of the patient (a greater or lesser tidal volume, with appropriately adjusted respiratory rate to provide a constant minute ventilation).

2. Volume control ventilation relies on delivering a preset tidal volume. The inspiratory flow and volume are constant but the peak inspiratory pressure can vary.

3. What are the similarities and differences between “ICU” ventilators and “anesthesia” ventilators?

4. What determines your settings for pressure support as a weaning strategy?

## Additional Topics

### Questions

1. The PACU nurse attaches an automated blood pressure cuff to a baby but has difficulty obtaining consistent readings. What makes this technique difficult in babies? What can you do to make it better? How do you select the appropriately sized cuff?

Pressure control ventilation delivers an accelerating inspiratory flow to reach a set target pressure and then the flow decreases as airway pressure approaches the target. The pressure is constant with every breath delivered but the volume may vary from one breath to another. Intraoperatively, the tidal volume and minute ventilation can vary significantly depending on the surgical site and manipulation. Pressure support is similar to pressure control except the patient must trigger every breath and the breath is terminated when the flow decreases to a predetermined level. In this relationship with the ventilator, the patient is active rather than passive and determines the inspiratory time and tidal volume. If no breaths are initiated by the patient, then no breaths will be delivered by the ventilator. However, there is a backup mode if the patient becomes apneic.

3. The essential difference between anesthesia and intensive care ventilators is that ICU ventilators function in an open circuit configuration. The need to deliver inhaled anesthetics compels a closed or semi-closed circuit configuration. The drive gases required to power the bellows or piston have to be separated from the gases used to vaporize the volatile anesthetic.

4. The strategy of pressure support ventilation is the use of varying degrees of pressure support to reduce the work of breathing. The tidal volume is determined by the amount of patient effort, the set inspiratory pressure, and the patient's lung compliance. For example, the ventilator will detect the onset of a spontaneous breath by measuring inspiratory flow which exceeds a set trigger threshold. The ventilator then provides sufficient flow to achieve a set inspiratory pressure. If the trigger threshold is set too low, auto-triggering may occur. If the trigger is set too high, the ventilator will not detect an inspiratory effort. As a weaning strategy, the patient has to be challenged with the defense of $CO_2$ elimination as mechanical support is weaned. This can be accomplished by deciding the desired minute ventilation at a particular point during the anesthetic and then titration of the trigger set point to ensure mechanical support sufficient to achieve this desired minute ventilation.

## Answers

1. Automated oscillometric blood pressure cuffs rely on incremental reductions in the pressure in the cuff and require at least two cardiac cycles for their measurements. Patient movement, irregularities of cardiac rhythm, or external influences such as someone pressing on the cuff can affect the accuracy of the reading, and

2. Now she is having problems with the pulse oximeter on this baby. What can make it better?

3. What methods exist to compensate for the loss of energy when a liquid is vaporized? What happens to the vapor pressure as the temperature of the liquid decreases?

4. How does a Clark polarographic oxygen electrode work? Given a choice of where you could place it in the breathing system, where would that be? For what purpose?

successive cardiac cycles will continue to be compared (and the blood pressure determination will be prolonged) until two comparable cycles are recorded at a given cuff pressure. This prolonged cycling duration may cause a great deal of discomfort for awake children and prolong the cycle even more. Sometimes repositioning the cuff to a lower extremity is a successful strategy. Other times, supplemental sedation or analgesia in order to relieve the surgical pain will ultimately make it easier to monitor the patient during their PACU stay. The proper width for the bladder of the blood pressure cuff should be 04 to 0.5 times the circumference, or 140% of the diameter, of the extremity. The length of the cuff's bladder should be twice the width of the extremity.

2. A pulse oximeter rarely functions well when it is placed on an active child. Algorithms for improving the signal-to-noise ratio are built into most pulse oximeters, but they interfere with the response time and accuracy. The operating mode can be changed in order to increase the accuracy of reading with different levels of patient activity. This works by changing the averaging time; a 5–7 second averaging time is typical for an inactive patient, while a 2–3 second averaging time is useful for sleep studies but is more affected by patient motion. Lengthening the averaging time up 10–15 seconds will enhance accuracy during patient movement. Most pulse oximeters are not affected by the presence of fetal hemoglobin, nor by the color of the skin or the bilirubin level (important for hyperbilirubinemia during infancy). To improve the signal of a poorly perfused patient, local warmth, a digital nerve block, or vasodilating cream can be applied. For arterial vasospasm, intra-arterial vasodilators may be administered.

3. As the temperature of the liquid decreases due to heat energy being lost, the vapor pressure decreases as well. A constant vapor output can be maintained only by compensating for this heat loss. One method is by altering the splitting ratio so that the percent of carrier gas coursing through the vaporizing chamber is changed via thermal compensation, either mechanically by a bimetallic strip that responds to changes in temperature or electronically. As an alternative, heat can be supplied to the vaporizer by an electric heater.

4. A polarographic electrode works by displaying the percent concentration of oxygen detected as a change in current across a gas permeable membrane on one side of which is an anode and the other side a cathode, as well as electrolyte solution. There is also a power source in order to induce a potential difference between the anode and the cathode. When oxygen molecules diffuse across the membrane and the electrolyte, the oxygen molecules are reduced to hydroxide ions. The probe may basically be positioned on the inspiratory or expiratory limb of the circuit. The advantage of positioning the probe on the inspiratory limb is that it serves to verify the inspired oxygen concentration, is not subject to moisture as it would be on the expiratory limb (where the moisture can affect the membrane permeability and electrolyte solution stability, therefore rendering the reading less reliable), and is not subject to any carbon dioxide (which can also affect the electrolyte solution and potential difference, therefore the accuracy of the reading).

5. How can you safely perform surgery in a patient with an implanted cardioverter defibrillator (ICD)?

6. In an effort to evaluate the volume of gastric contents prior to a rapid sequence induction, your anesthesia technician asks whether you would like the phased array probe or the linear probe. Your answer? Why?

5. The danger with the ICD is that the current from the electrocautery may be detected as a tachyarrhythmia and actuate the device causing shocks. The tachyarrhythmia detection should be deactivated before the procedure and then reactivated at the conclusion of the procedure. In addition, the grounding pad should be positioned in such a way as to minimize the flow of current through the ICD, which may damage the electrodes of the ICD. For ICDs that also function as pacemakers, they are more sensitive to electromagnetic interference, resulting in the inhibition of the pacing function. A temporary pacing electrode should be placed for external transthoracic pacing. Finally, bipolar as opposed to monopolar cautery should be used whenever possible, and this should be discussed with the surgical team beforehand. If not possible, then the current should be limited to short bursts with long (10 second) intervals at the lowest effective cutting or coagulation settings possible [1].

6. I would prefer the phased array probe, because it works at a lower frequency, and although the resolution is worse, it is better for tissue penetration. The linear probe has better resolution, but because it works at a higher frequency, it has worse tissue penetration. It is better for vascular and lung applications. The phased array probe is better for cardiac and abdominal applications. Gastric volume is best revaluated in the right lateral decubitus as well as supine positions—if contents are visible only in the right lateral decubitus position, it suggests a small fluid volume, but if contents are apparent in both supine and right lateral decubitus positions, it suggests a larger gastric fluid volume.

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# Chapter 48
# Regional Anesthesia


**Joseph P. Cravero**

## Case 1

A 13-year-old is scheduled for a prolonged penile reconstruction to correct a long-standing cosmetic issue related to severe hypospadias. He is otherwise healthy. He would like optimal pain management for the surgery and the immediate perioperative period.

## Case 2

A 15-year-old broke her right forearm 1 year ago. She now has reflex sympathetic dystrophy for which you have been asked to give the first of a series of stellate ganglion blocks.

## Case 3

You administer a Bier block to a 16-year-old having a plastic surgery procedure on her left hand. Within a minute of injecting 30 mL of 0.5% lidocaine into a vein on the dorsum of her left hand, you notice a dramatic wheal and flare reaction with swelling of the entire extremity distal to the tourniquet.



R. S. Holzman et al., *Pediatric Anesthesiology Review*,
https://doi.org/10.1007/978-3-030-60656-5_48

## Case 4

You are asked to provide anesthesia for a 15-year-old female soccer player who requires reconstruction of a left anterior cruciate ligament. The surgery is planned with a graft taken from the hamstring of the same side.

## Case 5

A 20-year-old male requires a Bankart repair for chronic habitual right anterior shoulder dislocation. This is to be done as an outpatient.

## Case 1

### Questions

1. Is it appropriate to do this surgery under just regional anesthesia? What form of regional anesthesia would you choose? Would your choice be different if the patient was 1 year old? Does the block need to be performed while the patient is awake in either case? What agents would you choose to use for this block?

## Answers

1. Depending on the length of the case, it would be possible to perform this surgery under spinal or epidural anesthesia alone. On the other hand, it is exceedingly rare that a 13-year-old would want to lay supine for 3–5 hours while this case is performed – so regional anesthesia alone is really not a viable option. Psychologically, operations of the perineum are difficult to perform on an awake adolescent. The logical choice would be to accomplish the case with regional anesthesia plus monitored anesthesia care or provide a general anesthetic in addition to the chosen block. Either method would be acceptable. I would choose general anesthesia with the regional block. There are many potential regional anesthesia options for this case. While natural airway sedation would be possible, for a case of this duration, I would choose to provide GA with an LMA in addition to the nerve blockade. I would choose epidural anesthesia to allow analgesia for the duration of the surgery and to provide the option of using a catheter infusion in this block for postoperative pain control. If the patient were only 1 year old, I would choose a caudal block with a catheter insertion largely because the caudal space is generally much easier to access in infants than in adolescents. There is excellent data to support the safety of "asleep" nerve blocks in pediatric age patients. While it is helpful to be able to receive feedback on paresthesias, etc. during the block placement, several large series of peripheral and neuraxial nerve blocks in pediatric age patients have failed to find a significant incidence of injury from blocks performed while patients are anesthetized. I would place an epidural catheter at L4–L5, and I would choose 0.2% ropivacaine or 0.25% bupivacaine for this block. To maintain the block, an infusion of bupivacaine could be continued at 0.4 mg/kg/h during the case. Peripheral nerve blocks can also be used to provide analgesia in this case. To perform this block,

2. On awakening the patient has bilateral foot drop. He is otherwise neurologically intact. What are the possible causes and what is your management?

an ultrasound probe can be used to guide a block needle just medial to the ischial tuberosity, and 0.3–0.5 mL/kg of 0.2% ropivacaine can be injected on each side. Nerve stimulation could be added to assure appropriate location of the needle tip by observing for contraction of the anal sphincter with stimulation. One other option would be the dorsal penile nerve block which could be performed by directing a block needle perpendicularly at the level of the symphysis pubis and inserting local anesthesia just under Scarpa's fascia approximately 1.5 cm lateral to the midline bilaterally. This could be done with ultrasound guidance or by landmarks alone. It should be noted that the peripheral nerve blocks do not provide as complete nerve blockade (in general) as an epidural or spinal block.

2. Bilateral foot drop after an epidural block could result from either epidural hematoma, direct trauma to the spinal cord, or compression neuropathy related to intraoperative positioning. The occurrence of epidural hematoma is less likely in patients who are not anticoagulated and in whom the placement of the epidural catheter is atraumatic. The presence of an arteriovenous malformation of the epidural vessels could also pose a risk. Needle or catheter trauma of the epidural vein can result in excessive bleeding and the development of a hematoma that may compress the spinal cord and cause a neurologic deficit. If epidural hematoma is suspected, the epidural infusion should be stopped to allow complete sensory and motor recovery. If neurologic impairment persists, CT and/or MRI should be obtained immediately because the epidural hematoma should be decompressed within 6–12 hours to avoid permanent neurologic deficit. MRI can rule out direct spinal cord trauma as well. Prolonged intraoperative positioning in lithotomy may predispose to lumbosacral plexus stretch neuropathy particularly in obese and very slender patients or in the presence of subcutaneous edema. This is usually associated with severe pain in the buttocks and legs. Foot drop can also occur from the compression of the common peroneal nerves against the fibula heads due to abnormal positioning of the legs in stirrups rather than straps in lithotomy position. The presence of bilateral foot drop with the preservation of perineal sensation and sphincter function (without severe pain) favors the diagnosis of bilateral common peroneal nerve palsy. Bilateral foot drop associated with urinary and fecal incontinence results from either lumbosacral plexus stretch neuropathy, cord trauma, or cord compression from hematoma. In this case, with isolated foot drop, the patient should be referred for supportive services, physical therapy, and close follow-up.

## Case 2

### Questions

1. How do you perform a stellate ganglion block? Three minutes after your injection of 10 mL of 0.25% bupivacaine with epinephrine, she complains of shortness of breath. What is the differential for her SOB? How would you manage the problem?

2. Fifteen seconds after your injection, she has a seizure. What is the likely cause and what is your treatment? Ten minutes after recovery, she complains of facial flushing, nasal congestion, and a funny feeling in her eye. What is the likely cause?

## Answers

1. The block is performed with the patient supine and the neck slightly extended. The C6 transverse process tubercle (Chassaignac's tubercle) is identified with the index and middle fingers placed at the level of the cricoid ring between the trachea and the sternocleidomastoid muscle. An ultrasound probe can be placed in a transverse orientation to identify the boney and vascular structures. It can also be used to directly observe the needle and the local anesthetic spread during the block. A short beveled 25-gauge needle is introduced perpendicular to the skin and advanced until the needle tip makes contact with the C6 or C7 transverse process. The needle is withdrawn a few millimeters and immobilized. After negative aspiration for blood or CSF, a total of 8–10 mL of a local anesthetic is injected without resistance and incrementally.

   The complaints of shortness of breath after stellate ganglion block could be due to something as simple as a feeling of a lump in the throat as result of block of the recurrent laryngeal nerve. These symptoms could also be due to more worrisome issues such as an intradural (epidural/intrathecal) injection of local anesthetics or (uncommonly) due to paresis or paralysis of the phrenic nerve and pneumothorax. Shortness of breath due to recurrent laryngeal nerve paralysis is best managed by reassurance and offering supplemental oxygen. SOB due to subdural injection, if severe, would be indicated by progressive loss of neurological function and may require ventilatory support and sedation. The management of pneumothorax depends on the severity of the condition. A chest X-ray and close follow-up are indicated. A pneumothorax of 10% or less does not require specific treatment. More severe pneumothorax with impaired oxygenation or cardiovascular changes will require monitoring in the hospital and (likely) placement of intrapleural drain.

2. The vertebral and carotid arteries lie in close proximity to the neural structures at C6–C7. Injection into one of these vessels will lead to an almost immediate seizure. Fortunately, the seizures caused by such an injection will be extremely brief in nature because the drug is eliminated from the brain very quickly. Even when performed with ultrasound guidance, this is a reason to inject the local anesthetic slowly when performing one of these blocks. Treatment other than general support is rarely needed. Facial flushing and dryness are typical signs of Horner's syndrome which is common with this block – particularly when the injection is made at C6. No treatment is needed other than reassurance. It is helpful to warn patients that this is a possibility prior to starting the block.

## Case 3

### Questions

What is your diagnosis? What is your management?

## Case 4

### Questions

1. What nerve block(s) would you plan in order to augment this anesthetic and/or provide postoperative pain control? Explain your choices.

## Answers

The observed manifestation is consistent with a local allergic reaction to either lidocaine or (less likely) due to latex gloves. In either case, all latex-containing products should be disposed. Secure large-bore intravenous access. There should be a call for help, and preparation should be made for full support including intubation/ventilation as needed. The tourniquet should not be released until after prophylactic measures are taken. Prophylactic measures should include rapid fluid administration to assure hydration, diphenhydramine, and epinephrine just prior to gradual tourniquet release. The tourniquet should be deflated gradually and intermittently to avoid severe systemic anaphylaxis and allow effective antagonism of systemically released antigens.

## Answers

1. It is possible to provide analgesia for ACL reconstruction in many ways. A peripheral nerve block is not an absolute requirement. The use of multimodal analgesics such as acetaminophen and nonsteroidal anti-inflammatory drugs, along with a modest amount of opioid medications, could be employed during and after this surgery. In addition, the injection of intra-articular local anesthetic and opioid has been shown to provide a modest amount of pain control in the immediate postoperative time period. Furthermore, local anesthesia injected in the sites of the graft harvest could provide analgesia for a limited time. For an adolescent undergoing a surgery as painful as this, a peripheral nerve block, or a combination of techniques that block sensation from the knee, has been shown to provide improved analgesia and decrease the need for opioid administration. When considering a peripheral nerve block, it should be recognized that analgesia for an ACL reconstruction requires coverage of the dermatomes that involve the anterior knee. In this case, with a graft from the hamstring planned, there is a need to consider pain control for the posterior portion of the knee as well. Coverage of the anterior portion of the knee can be provided by a femoral nerve block or adductor canal block. The femoral nerve block is performed under ultrasound guidance by focusing the transducer on the femoral nerve, artery, and vein on the side of the block. The femoral nerve lies lateral to the femoral artery. It is hyperechoic and triangular in shape. The needle is passed under direct (in plane) visualization from the lateral aspect of the leg. Once the needle tip is adjacent (above, below, or lateral) to the nerve, aspiration should be performed and local anesthetic injected. For the adductor canal block, the transducer is placed transversely on the medial thigh, at the midpoint of the upper leg. At this point, the

2. Three months after the surgery, the patient is weak in the left quadriceps. Could this have been due to the nerve block you provided?

## Case 5

### Questions

1. What regional technique would you use to maximize analgesia during the case and provide pain control for the immediate time frame after surgery? How is it performed? Are there specific contraindications to your block? What medications will you use for rapid onset and long duration? How long is the analgesia likely to last and what will you inform the patient about with regard to transitioning to oral analgesics when the block wears off?

femoral artery is visualized deep to the sartorius muscle. The needle tip is positioned deep to the fascia of the sartorius muscle and anterolateral to the artery. Injection of local is done after aspiration.

As mentioned above, the pain from the graft harvest from the hamstring can be managed with local anesthesia at the site or by providing a subgluteal sciatic nerve block in addition to the nerve block applied for anterior knee analgesia. The addition of a sciatic nerve block has been associated with improved pain control in the immediate postoperative time frame.

2. It has been suggested that because the femoral nerve block includes motor innervation as well as sensory innervation whereas the adductor canal block is predominantly a sensory block, the adductor canal block is preferable since patients lose less of their motor ability with the block in place. In addition, some researchers have suggested that femoral nerve blocks may be associated with long-term weakness of the quadriceps muscle; however, this claim is not supported by studies of large populations of these patients. Long-term weakness is most closely related to the type of surgery performed and the adherence to physical therapy regimens in the postoperative time period.

**Answers**

1. I would perform an interscalene nerve block. The patient should be placed in a semi-upright position with a pillow under the shoulder. The patient's head is turned in the opposite direction of the shoulder to be blocked. The ultrasound probe can be placed along the clavicle where the brachial plexus should be easily visible. The probe can then be moved cephalad and rotated so that the anterior and medial scalene muscles are in view along with the brachial plexus nerves in the groove in between. Local anesthetic can be deposited by a needle that is placed "in plane" with the ultrasound probe. Prolongation of the block can be accomplished by placing a catheter adjacent to the nerves and providing a continuous infusion of local anesthetic for 24 hours. Catheters are not generally available for outpatients, but some centers have created the infrastructure for this as well. Outpatient catheters can only be provided in a well-coordinated system of care for close perioperative follow-up. If a catheter is not placed, the patient should be given multimodal pain medications to manage pain, and these medications should be started before the block wears off to avoid extreme pain and a difficult "catch-up" time where the patient is extremely uncomfortable and systemic medications have not yet taken effect. These blocks are contraindicated in patients who are anticoagulated or who have a neurological deficit on the side

2. After surgery the patient complains of persistent numbness in the fourth and fifth digits of his right hand? What is your differential diagnosis? What do you tell the patient to do?

that you are placing the block. In addition, these blocks are commonly associated with phrenic nerve paralysis, Horner's syndrome, and recurrent laryngeal nerve paralysis. As such, they are not recommended for patients who have severe respiratory impairment or contralateral phrenic nerve paralysis or recurrent laryngeal nerve paralysis.

2. Nerve injury after a brachial plexus nerve block and surgery is rare and could have many sources. In this case, injury in the distribution of the ulnar nerve is particularly unusual since the nerve is often "spared" with this particular block. Nerve injury is more likely to be from the block if there was preexisting nerve injury or if there was a paresthesia or pain at the time of the nerve block. The injury in this case could also have occurred from traction of the arm during the surgery or from direct injury to the nerve during surgery. It is somewhat helpful to determine if there is pure sensory involvement or if there is motor and sensory injury. Motor involvement portends a more guarded prognosis, but most of these injuries improve with time and simple physical therapy. If the symptoms are persistent, it is appropriate to have the patient seen by a neurologist who is familiar with postanesthesia/surgical injuries of this type. It is rare that a specific intervention is undertaken to treat the injury, but nerve conduction and EMG findings can (sometimes) help define the extent and timing of the injury.

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# Chapter 49
# Pain Management

**Joseph P. Cravero**

A 16-year-old, 48 kg female gymnast with a history of complex regional pain syndrome (CRPS) in the *right* foot is scheduled for a *left* ACL reconstruction. She is currently on gabapentin and amitriptyline. Her pulse rate is 95 bpm, BP 108/70 mmHg, and RR 16/min.



R. S. Holzman et al., *Pediatric Anesthesiology Review*,
https://doi.org/10.1007/978-3-030-60656-5_49

## Preoperative Evaluation

### Questions

1. What is complex regional pain syndrome (CRPS)? How do you make the diagnosis? What are the subtypes and how are they defined? What is the most effective treatment for this disorder?

2. What classes of drugs do gabapentin/pregabalin and amitriptyline belong to? What are their mechanisms of action? What impact might they have on your anesthetic? What other drugs or therapies could be used for treatment of CRPS?

## Answers

1. Complex regional pain syndrome is a painful syndrome of unclear etiology and pathophysiology. The clinical diagnosis is made through history and physical examination. It is presumably dysfunction of small fibers of the skin and deeper tissues associated with regional sympathetic nerve dysfunction that manifests with increased cutaneous sensitivity (allodynia and hyperalgesia), skin discoloration, and impaired or excessive sweating. The differential diagnosis may include fibromyalgia, myofascial pain, arthritis, spondyloarthropathy, Fabry disease, peripheral mononeuropathy, and any other condition that can elicit chronic extremity pain. The condition commonly affects distal parts of the limbs in glove-and-stocking distribution (non-dermatomal) and is more frequent in females. Some patients may experience motor dysfunction, weakness, and myoclonus. In the advanced condition, the muscles, bone, and skin are wasted, and the joints become stiff with resultant loss of limb function. CRPS is divided into two types. CRPS type I (also known as reflex sympathetic dystrophy) is usually preceded by some kind of injury, usually minor, such as a sprain, twist, or dislocation. CRPS II is rare in pediatric patients (previously called causalgia) and is precipitated by a nerve injury. CRPS I in pediatric patients is most common among adolescent females. Formal diagnosis is made through application of the Budapest Criteria. These criteria include the following: (1) pain is disproportionate to the inciting event; (2) pain has sensory, vasomotor, edema, or motor/trophic features; and (3) no other diagnosis explains the pain.

   The treatment of these patients is best performed with an interdisciplinary approach. Modalities include intensive physical therapy and rehabilitative occupational therapy, combined with cognitive behavioral therapy.

2. Gabapentin and pregabalin are antiepileptic and pain medications that have been synthesized to mimic the structure of gamma-aminobutyric acid (GABA); however, they are not thought to act on those receptors. Activity is believed to be on voltage-gated calcium channels at which it decreases calcium currents after chronic (not acute) exposure. These drugs also interact with NMDA receptors, protein kinase C, and cytokines, but the exact nature of their action on pain modulation is not known. Pregabalin is a longer-acting version of this drug that has not been thoroughly tested in pediatric CRPS patients. Gabapentin has been shown to inhibit the development of hyperalgesia and C-fiber responsiveness. The anesthesia effects of these drugs have been a lower requirement for intraoperative and postoperative opiates. A small percentage of patients receiving gabapentin have been found to exhibit a prolongation of the Q-T interval when also taking other medications that affect repolarization. Amitriptyline is a tricyclic drug that inhibits norepinephrine and serotonin reuptake at the second-order

3. Are there any techniques for this anesthetic that can reduce her chance of further neuropathic pain?

neuron synaptic transmission. This drug is effective in treating depression and improves sleep and some aspects of pain in patients with CRPS. It has a strong anticholinergic effect and may cause delayed atrioventricular conduction, prolonged QRS and QT syndrome, torsades de pointes, A-V block, lower threshold for seizures, urinary retention, hyperthermia, increased intraocular pressure, extrapyramidal syndrome, and anticholinergic psychosis. Its atropine-like effect may cause sinus tachycardia and ventricular premature contractions particularly when combined with other anticholinergic or sympathomimetic drugs such as atropine, glycopyrrolate, pancuronium, meperidine, succinylcholine, sevoflurane, isoflurane, and desflurane. Numerous other medications could be used in patients with CRPS including acetaminophen, nonsteroidal anti-inflammatory drugs, steroids, opioid medications, other anticonvulsants (phenytoin), local anesthetics for nerve blocks, and intravenous ketamine (for extreme cases). Other therapies include application of heat and cold, topical analgesics, physical therapy (perhaps most important), transcutaneous electrical nerve stimulation (TENS), biofeedback, and spinal cord stimulation.

3. There are many theories for the cause of CRPS including central sensitization, alterations in central nervous system processing, and small fiber changes. It is thought that the brains of these patients respond differently to normal stimuli. A predominant theory of chronic neuropathic pain syndrome is the hyperexcitability of the second-order neuron (i.e., central sensitization) in the dorsal horn caused by the injured nerve. It is reasonable to assume that the use of spinal, epidural, or regional anesthesia with peripheral nerve blocks could prevent the nociceptive impulses generated at the surgical site from reaching the dorsal horn neurons and hence minimize its further excitation and reduce the potential for flare-up of CRPS pain postoperatively. The plan for the anesthetic should be made well in advance of the surgery, with full agreement of the patient and the family. It should include a multimodal pain management strategy, with acetaminophen, nonsteroidal anti-inflammatory drugs, gabapentin (or any other medications the patient is already taking), and opioids as needed. The patient's pain physician should be involved in the planning and postoperative management. Expectations for significant discomfort after surgery and early aggressive physical therapy should be set. In this case, it would be appropriate to administer a regional anesthetic prior to starting surgery.

## Intraoperative Course

### Questions

1. Would you premedicate this patient? Would you provide regional anesthesia? If so, what kind of block would you perform and when would you perform the block? What kind of airway management do you plan? How would you induce anesthesia?

2. How would you maintain anesthesia in this patient?

**Answers**

1. Patients with CRPS have been found to have a higher tendency for recent stressful life events. They also have a greater tendency for difficult family environments. Anxiety and emotional distress surrounding medical interventions are very common in these patients, as are pain amplification and somatic complaints. As such, it would be appropriate to premedicate the patient with small doses of midazolam and fentanyl until her behavior, expression, and interaction are appropriate. Based on the discussion of central sensitization mentioned above, it would be strongly advised to provide a regional anesthetic. There are many choices, but the technique should provide adequate coverage for *all* areas that are directly involved in the surgical intervention. In this case a lumbar epidural or a spinal anesthetic would offer good coverage for the lower extremity surgery. These techniques could be done with adequate sedation in the preoperative time frame or after induction of general anesthesia. Unfortunately, neither of these techniques would offer a significant amount of analgesia in the postoperative time period (several days) during which the patient is likely to be extremely uncomfortable. Operative and postoperative analgesia could be provided by placing a femoral nerve block or adductor canal block and catheter. If the surgery also involved a graft harvest (from the posterior or lateral aspects of the knee), then a sciatic nerve block/catheter would also be required. It is critical that all areas of discomfort are "covered" by the peripheral nerve block technique. These could also be done under significant sedation or while the patient is under general anesthesia as long as ultrasound guidance was available. Regardless of the choice of nerve block, the analgesia should be extended with a catheter infusion for at least 24–48 hours in a case where the patient has a CRPS history. The analgesia could even be extended to 72 hours or more with a home-based catheter technique. It is likely that this patient would need to be deeply sedated or anesthetized even if she was well blocked. For general anesthesia, airway management could be performed with an LMA and spontaneous ventilation or with an endotracheal tube and controlled ventilation. I would choose an LMA and induce the anesthesia with intravenous propofol and fentanyl.

2. I will maintain the anesthesia with air/oxygen, fentanyl, propofol infusion, and a low dose of sevoflurane or isoflurane as tolerated by her hemodynamic and respiratory parameters. The use of a longer-acting opioid to be absolutely sure of pain control on emergence would be advisable as well. A benzodiazepine such as diazepam could also be helpful to prevent muscle spasm in the immediate postoperative time frame.

3. After induction the patient becomes hypotensive with BP = 70/40. How do you treat this?

4. The patient develops ventricular tachycardia after treating her low heart rate with glycopyrrolate.

5. She has prolonged emergence after her anesthetic. Is this unexpected?

## Postoperative Course

### Questions

1. How would you manage the patient's pain postoperatively? What drugs would you use and how do they exert their effect? What monitoring is required?

3. Chronic use of amitriptyline inhibits the reuptake of serotonin and norepinephrine, a noradrenergic catecholamine. In the face of this, the blunting of central sympathetic drive by anesthetic agents can result in hypotension. This hypotension is best treated with intravenous volume. If ineffective, low incremental doses of norepinephrine (1–2 mcg/kg IV) may be necessary to maintain adequate blood pressure. Due to upregulation of receptors, sympathetic nervous system activation and the use of direct or indirect adrenergic agonists may lead to a marked response in terms of tachycardia and hypertension. Ketamine, with its indirect sympathomimetic effects, should be used very cautiously.

4. In the absence of any other reason for ventricular tachycardia in a young patient on tricyclic antidepressant medication, this is likely due to augmentation of anticholinergic effects of amitriptyline. In addition to preparing the standard treatment for VT, incremental administration of intravenous cholinergic drugs such edrophonium (0.5–1 mg) or physostigmine (0.5 mg) may be effective. A major side effect of these drugs is bradycardia, salivation, and nausea on emergence.

5. Gabapentin effect can add to drowsiness on emergence. In addition, chronic tricyclic agent exposure can slow emergence due to depletion of central catecholamine stores. If her nerve block(s) is working well, there may be very little pain immediately after the surgery. The sedating effects of her medications could be enhanced by concurrent administration of opioids and other sedative-hypnotics in the perioperative period.

**Answers**

1. Ideally, pain control should follow a multimodal methodology. She should be given non-opioid pain medications such as acetaminophen and nonsteroidal anti-inflammatory drugs (ketorolac or ibuprofen). These drugs work by inhibition of cyclooxygenase (COX). This results in decreased conversion of arachidonic acid to prostaglandin H2. The reduction of prostaglandin confers pain control in the periphery. Decreases in prostaglandin concentration also lead to activation of descending inhibitory serotonergic pathways that result in analgesia. It is important to note that, while these drugs are not (generally) effective in treating CRPS primarily, they could be helpful in controlling pain in the postoperative time frame. She should continue to receive her gabapentin, which will act by decreasing axon excitability through its effect on NMDA receptors or calcium channels and cytokines. Continuation of epidural analgesia or femoral (+/− sciatic) nerve block(s) would afford the best pain relief. The local anesthetic in these nerve

## Additional Questions

### Questions

1. A 10-year-old male has persistent pain and vasoconstriction after he had an infiltration of his IV in the left hand. A stellate ganglion block is requested. How would you perform the block?

blocks will block sodium channel-dependent nerve fiber transmission and decrease or eliminate pain signals from reaching the spinal cord. Opiates such as morphine, hydromorphone, or fentanyl may also be required. The opiate drugs produce their actions by binding with receptors on neuronal cell membranes. Activation of these receptors results in decreased cAMP production in the cell and inhibits neurotransmitter release. If needed, they are most effectively provided via patient-controlled analgesia (PCA). Patients having significant orthopedic surgery often benefit from the addition of benzodiazepines, which decrease muscle spasm and aid in anxiolysis. Benzodiazepines exert their effect by enhancing GABA activity at the GABA A receptor. Patients should be monitored for excessive sedation, mouth dryness, and paralytic ileus due to enhancement of the anticholinergic effect of amitriptyline. Avoid concurrent use of anticholinergic drugs such as diphenhydramine for treatment of pruritus.

## Answers

1. Stellate block should be performed in a setting wherein resuscitation equipment and drugs as well as personnel trained in cardiopulmonary resuscitation are readily available. The block is performed with the patient supine and the neck slightly extended. The use of ultrasound has made the block more reliable and less risky than previously when the block was done by surface anatomy and bony landmarks. The probe can be placed in a transverse orientation at the level of the cricoid cartilage in order to identify the transverse process of C6 and the nearby vascular structures. At this point, the probe is moved inferiorly to identify the transverse process of C7 as well as the longus colli muscle. A short-beveled 25-gauge needle is introduced perpendicular to the skin and advanced until the needle tip makes contact with the C7 transverse process, avoiding the vertebral artery. The needle is withdrawn few millimeters and immobilized. After negative aspiration for blood or CSF, a total of 8–10 mL of a local anesthetic is injected incrementally. Anxious adolescents and younger children may require general anesthesia to avoid movements during the performance of the block. The patient is anesthetized with an inhalational agent via mask or an LMA and allowed to breathe spontaneously. This technique is suitable for monitoring adverse events such as cessation of breathing or seizure activity in the event of an accidental intracarotid artery or intrathecal injection of the local anesthetic.

2. You wish to perform an interscalene block with the patient asleep. How would you minimize the chance of nerve injury during this block? Is a nerve stimulator helpful for this block – how would you use it? Shortly after infusing local anesthetic, the patient has a seizure. Why does this happen and should it be treated?

3. A patient with metastatic Ewing's sarcoma has metastases to the spinal cord. What options do you have in managing his pain if he sees you as an outpatient? If he is increasingly debilitated and has just entered hospice care? If he is at the end of life?

4. A 16-year-old with sickle cell (SS) disease is admitted with a painful crisis involving his lower back and both legs. What would you choose for pain control in this situation? Would this be different if his pain was located in his mid-sternum?

2. It is imperative to use all safety measures to avert neural injury in an anesthetized patient. Such injury could be minimized by the use of a nontraumatic needle (blunt pencil-tip needle). The use of ultrasound should also minimize the likelihood of nerve injury; however, if the sonoanatomy is uncertain, an insulated needle and a nerve stimulator may be used for precise localization of the nerves. The initial current of 1–1.5 mA at 1 or 10 Hz is used to identify proximity to the nerve(s) as the needle is advanced. After identifying the appropriate motor response to the stimulated nerves distal to the elbow, the stimulating current is gradually decreased as the needle is advanced until a maximum motor response is maintained at a current of 0.5 mA or less indicating that the needle tip is within 1–2 mm of the target nerve. The seizure is almost certainly due to an infusion of local anesthetic into the carotid artery or vertebral artery. A 0.1 mg/kg dose of lorazepam (Ativan) could be administered to treat a prolonged seizure. One should be alert for apnea that often accompanies the postictal/sedated state. In most cases, protection of the airway and support of vital functions are all that are required. The seizure is short-lived as the drug will be quickly redistributed to the rest of the body and the concentration that caused the seizure will not be present. If a large dose of drug has been administered, care must be taken that cardiac toxicity does not occur. If ventricular tachycardia or fibrillation results, treatment should follow as quickly as possible with intralipid.

3. Surgery followed by radiation is the most effective treatment for spinal cord compression caused by metastatic cancer. The addition of surgery allows most patients to remain more mobile and to retain bladder control. If the metastatic cancer is not amenable to anticancer therapy, the goal of the hospice care is to improve quality of life by providing pain and symptom relief care. The pain is managed by oral and/or intravenous opioids and adjuvant anti-neuropathic pain agents. If the pain is not adequately controlled or unacceptable side effects occur, then intrathecal opioids and local anesthetics should be considered. At the end of life, pain and distressing symptoms are alleviated by incremental large doses of opioids, adjuvant drugs, and sedatives. Intra-epidural or spinal local anesthetic + opioid combinations should be considered to alleviate suffering.

4. Pain management for SS disease must include an appreciation for the chronic/relapsing nature of pain in this illness. Ideally, pain management should have begun at home with oral medication, escalating from acetaminophen and NSAIDs to oral opiates. In this case, home management has failed, and aggressive pain management is indicated. These patients should be rapidly assessed and treated in the emergency department, ideally within 30 minutes. Appropriate hydration and oxygenation should be assured. Therapy should be multimodal and can include IV ketorolac (not absolutely proven to change the course) and PCA plus continuous opioid (morphine, hydromorphone, or fentanyl). If pain is not controlled with this management, low-dose ketamine infusion could be used to aid management although the effectiveness of this drug has not been proven. For patients with localized pain (such as a specific shoulder or knee), regional

anesthesia with an indwelling catheter should be pursued. All of this management should be undertaken with close cooperation with the medical hematology team. Patients should be maintained on their home doses of hydroxyurea, which aids in blood rheology. Transfusion is only indicated when the patients have symptomatic anemia – not simply for pain management. Alternative therapies such as hot packs or massage are helpful in some patients and should be offered. Treatment of primary insomnia problems with environmental manipulation or drug therapy is indicated. Some of these patients have neuropathic pain that accompanies their acute pain, and this should be treated with neuropathic-specific medications.

Pain in the mid-sternum would be indicative of a possible chest crisis and carries with it the danger of acute respiratory deterioration. In addition to all of the interventions mentioned above, close attention to oxygenation, pulmonary toilet, and anemia should be discussed with the primary hematology team. Escalation of care should be prompt in these cases with some patients requiring maximal respiratory support.

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# Chapter 50
# Postanesthesia Care Unit (PACU)


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## Case 1

A healthy 4-year-old has just undergone open reduction internal fixation of a mid-shaft femur fracture under general endotracheal anesthesia with an epidural in place which appeared to be functioning well during surgery. You are called to the bedside because he is clenching his teeth and crying and he and his parents are very unhappy.

### Questions

1. How will you evaluate this child? How will you determine the reasons for these behaviors?

2. How can you confirm if the epidural is in the epidural space?

## Answers

1. It is important to determine that the child's distress is due to surgical pain, not nausea, a full urinary bladder, emergence delirium, anxiety, an NG tube, or the tightness of the cast. The patient should be fully examined for such common sources of discomfort or behavioral disturbance. If there are no obvious extraneous causes, the assumption should be that the child is experiencing surgical pain until proven otherwise. In a patient such as this, acute postoperative pain can be treated with IV opioids or the administration of an appropriate dose of local anesthetic via the epidural catheter. If the epidural infusion used during the anesthetic in the OR contained an opioid, it is important to note the type and amount of epidural opioid given during the surgical procedure. The duration and onset of the opioid will be largely determined by the pharmacokinetics of the drug, when it was administered, and the dose given. A hydrophobic opioid such as fentanyl will pass through the tissue planes and exit the epidural space rapidly. A dose of hydrophilic opioid (morphine) will stay in the epidural space and spread cranially over several hours.

   Dosing of IV opioids must be titrated carefully in light of the synergy between the neuraxial opioid and intravenous opioid. If IV opioids are administered just as hydrophilic epidural opioids are reaching maximum effect, this could lead to respiratory depression.

2. If the child's discomfort is not extreme, the effectiveness of the catheter can be tested by administering a dose of epidural local anesthetic and evaluating the child's comfort after the bolus. Epidural analgesia is a safe and effective analgesic regimen for children [1–3]. Administration of a rapid-onset local anesthetic such as chloroprocaine will provide a solid sensory block in a relatively short period of time (if the catheter is in good position) resulting in increased comfort and should engender changes in the child's behavior. Chloroprocaine is an amino ester local anesthetic that has a rapid onset, a very short duration, and an excellent safety profile. When administering a bolus dose of any local anesthetic, it is important to consider the timing and dose of local anesthetic that has already been given via the epidural catheter. Local anesthetic toxicity is additive, so that an appropriate dose of a drug such as lidocaine, to a child who has been receiving an infusion of ropivacaine, could result in that child receiving a combined dose above the safe limit.

3. You test it, and it doesn't appear to be working. The patient doesn't have any relief after 20 minutes. What other treatment is appropriate?

4. He pulled out his IV in the meantime; how would you attempt to reinsert the IV? Should he receive IM morphine before you attempt to reinsert the IV? If he was going to need to stay in the PACU for an extended period, how would you provide for ongoing opioid needs?

The position of the epidural catheter could also be determined by visualization with ultrasound. This technique has been shown to be useful in neonates and infants where the immature anatomy allows for adequate images between vertebral bodies. When localization is critical and ultrasound visualization is not possible, an "epidurogram" could be obtained by infusing a small amount of radiopaque dye and taking AP and lateral X-rays of the spine in the area of the catheter.

3. If it appears that the child is receiving no analgesia from the drugs given into the epidural space, that technique should be abandoned and another analgesic modality begun as soon as possible. If, after administration of appropriate doses of IV opioid, the child still is uncomfortable, additional doses should be titrated in with consideration of the respiratory rate. In addition, multimodal adjuncts should be administered if not contraindicated. These include IV or oral acetaminophen and parenteral ketorolac. Although there is some evidence that ketorolac may affect new bone formation, the magnitude of this effect is uncertain and is most likely in cases where many doses are used over time. Many orthopedic surgeons agree with the use of ketorolac for 24–36 hours in the treatment of pain associated with long bone fractures. In addition to other analgesics, antiemetics and/or anxiolytics should be considered for this child. Comfort may be increased by the addition of a benzodiazepine in cases where muscle spasm is a consideration – such as extremity surgery. If benzodiazepines are used, however, the clinician must be aware of the synergistic depressive effect on respiratory drive these drugs have with opioids.

4. If the child now has no IV, venous access should be reestablished as quickly as possible, but it is not an emergency. There is likely time for application of a topical analgesic or intradermal local anesthesia prior to placement of an IV. With use of a topical anesthetic prior to replacement of the IV, IM medications can be avoided. An IV will be needed for fluid and medication administration in any case; thus, IM medication should be used only as a last resort. Opioid administration to children via PCA/NCA is a good option even for young children such as this. Both have been shown to be safe and provide better pain control than intermittent morphine doses [4, 5].

## Case 2

A 15-year-old girl has just undergone laparoscopy for endometriosis. She had a general endotracheal anesthetic. You are called to the bedside because she feels continuously nauseous, has retched a few times, and now has bile-tinged vomitus. She received 4 mg of ondansetron and 10 mg of metoclopramide as part of her anesthetic management.

**Questions**

1. Was she at high risk for nausea and vomiting? What are the most common strategies for prevention of postoperative nausea and vomiting? Is there anything else you can offer? Should she have a nasogastric tube placed? You note that one was not passed in the OR – does this make a difference in perioperative nausea and vomiting? Why?

2. Should droperidol or haloperidol be added at this point?

## Answers

1. Postoperative nausea and vomiting (PONV) is a complex phenomenon. The young lady in this case has several risk factors for PONV. She is female, has undergone abdominal surgery, and has had general anesthesia with endotracheal intubation. She was given appropriate prophylaxis with a serotonin receptor antagonist and metoclopramide, a medication that stimulates gastric peristalsis through the blockade of dopaminergic receptors. It is used clinically as an antiemetic and as an aid to gastric emptying in conditions with delayed gastric emptying. In the PACU, the OR course should be carefully reviewed. Prophylaxis for PONV can be accomplished with the administration of a serotonin antagonist, dexamethasone, droperidol, or haloperidol. Metoclopramide, used in standard clinical doses, has not been effective in prevention of PONV [6, 7].

   Since prophylaxis with a serotonin antagonist has failed, further treatment with a drug in the same class is not likely to yield positive results. Another class of medication, such a promethazine or droperidol, can be given. Promethazine (Phenergan®) is a phenothiazine derivative with antiemetic, antihistaminic, anticholinergic, and sedative effects. Although emptying the stomach with an NG tube might have helped decrease this girl's volume of emesis, it is not as effective in preventing nausea and retching that are due to activation of the chemoreceptor trigger zone that anesthesia is associated with. In addition, the trauma of NG tube placement makes this particular intervention less than desirable at this point. Inadequate fluid resuscitation can contribute to PONV, and if there is any indication that she has not received sufficient IV fluid, this should be corrected. Dexamethasone has been shown to be effective in prevention, but not treatment, of PONV [8, 9].

2. Droperidol and haloperidol are major tranquilizers belonging to the group of drugs known as butyrophenone antipsychotics. It has antiemetic and sedative effects. In 2001, the FDA issued a black box warning about the use of droperidol, describing cases of prolonged QT interval and torsades de pointes in patients given droperidol. Performance of a 12-lead ECG to check for prolonged QTc (>440 msec for females, >450 msec for males) prior to administration of droperidol is recommended. In addition, ECG monitoring for 2–3 hours following administration of droperidol is recommended. No case of prolonged QTc or arrhythmia has been reported after administration of the small doses of droperidol used to treat PONV [10, 11].

3. What part of the brain is responsible for postoperative nausea and vomiting?

4. Are some anesthetic techniques better than others for perioperative emesis control?

5. Are some operations more emetogenic than others?

6. What about total intravenous anesthesia with propofol and avoidance of nitrous oxide? How does that work?

7. Is it safe to discharge this patient home? What are you concerned about?

3. Vomiting is mediated via the vomiting center and the chemoreceptor trigger zone (CTZ) located in the brainstem. The vomiting center is in the reticular formation, and the CTZ is located in the floor of the fourth ventricle. It is activated directly by visceral afferent impulses from the pharynx, peritoneum, bile ducts, coronary vessels, and the cortex. The CTZ is located on the blood side of the blood-brain barrier and cannot cause vomiting without an intact vomiting center. The exact pathways that cause nausea are not known for certain, but it is assumed that they are the same as the pathways described for vomiting. Stimuli associated with nausea include pain as well as labyrinthine stimulation.

4. Various alterations in anesthetic technique have been shown to reduce the incidence of PONV. Patients who receive a primary regional anesthesia are at much lower risk for PONV compared to those who are given general anesthesia. When general anesthesia is administered, avoiding or minimizing the use of nitrous oxide has a similar effect in decreasing the incidence of PONV. When propofol is used as the intravenous anesthetic agent, it is associated with a lower incidence of PONV compared with the potent inhaled vapors. Reversal of neuromuscular blockade with neostigmine is also associated with a higher incidence of PONV. Opioids are well known to be associated with nausea and vomiting, and minimizing their use can decrease nausea in the postoperative time frame [12]. Dexamethasone, given at the start of a surgical procedure (particularly for head and neck procedures), has efficacy in preventing PONV, but it is not an effective treatment of PONV once it has occurred.

5. Certain surgical procedures are associated with higher incidence of vomiting, although the reasons are not always obvious. Some of these procedures are laparoscopy, gynecological surgery, laparotomy, head and neck procedures, breast surgery, and strabismus surgery [8, 10, 13].

6. In large survey reviews of postanesthetic nausea and vomiting, risk factors identified include the use of nitrous oxide, neostigmine reversal of NMB, and opioids. The potent inhaled agents, although also considered medications that increase the incidence of PONV, are not as clearly identified with this problem. Total intravenous anesthesia (TIVA) with propofol, oxygen, regional analgesia or peripheral nerve blocks, and non-opioid analgesia if not contraindicated by the specifics of the surgical procedure or anesthetic requirements can be recommended for patients with a history of severe PONV. A risk score for postoperative nausea and vomiting has been proposed using four factors – duration of surgery; age >3 years; history of postoperative vomiting in patient, parent, or sibling; and strabismus surgery [12].

7. In this patient, concerns with discharge include the possibility of dehydration due to continued emesis without PO intake and also bleeding or other surgical complication from repeated forceful contraction of the abdominal muscles. A trial of 2–3 hours of IV hydration and administration of another antiemetic of a

8. What are the optimal times to administer antiemetics?

## Case 3

You arrive in the intensive care unit with a 13-year-old girl after just finishing a posterior spinal fusion from T6 to L5 with an estimated blood loss of 3,000 mL and a volume replacement of three autologous units of packed red blood cells, 500 mL of Cell Saver blood, and a reasonable (2 L) amount of crystalloid. She looks pale on arrival, with BP = 50/20 mmHg and a heart rate of 96 bpm.

**Questions**

1. What do you think is going on? Why?

2. Why did these findings become apparent now, and not earlier?

different class than serotonin antagonists might improve the situation. If the young lady is then able to tolerate PO clear liquids, she may be safely discharged home.

8. There is general agreement regarding prophylactic versus rescue treatment of PONV [7]. The number of patients who would need to be given prophylactic treatment in order to prevent one case of clinically significant PONV varies with the patient, the anesthetic provided, and the procedure performed. Prophylactic treatment for patients with moderate to high risk for PONV is a sensible course and should include a serotonin inhibitor and steroid [14, 15].

**Answers**

1. This patient has deficient intravascular volume. Significant blood loss is an expected part of scoliosis surgery [16–18]. If we assume that the patient's weight preoperatively was 50 kg, her calculated blood volume would be 3500 mL. Her EBL, therefore, is approximately one blood volume. The volume she has been transfused is approximately 800 mL autologous PRBC, 500 mL Cell Saver, and 2000 mL crystalloid. If we assume a one to one replacement of blood lost with PRBC and Cell Saver, those products have replaced approximately 1300 mL of her 3500 mL blood loss. She also was given approximately 2200–2500 mL of crystalloid as replacement for the approximately 2 liters of EBL not replenished by the blood products. Replenishment of intravascular volume lost from intraoperative bleeding with crystalloid is generally at a 3:1 ratio. In this case, then, adequate IV fluid replacement for the blood loss would be in the neighborhood of 6000 mL, not the 2000 given in the OR. The likely explanation for the pasty appearance and hypotension is inadequate preload.

2. Once the surgery had been completed, there was much less stimulation, and the sympathetic tone that had been partially responsible (presumably) for BP maintenance has now greatly diminished.

3. Why do you think she is not tachycardic? Could this be anything else?

4. She seems sleepy when you try to awaken her? What are you concerned about? What can you do?

3. The hypotension is not accompanied by tachycardia as would be expected. There are several possible explanations for the low heart rate. The anesthetic may have included a significant dose of fentanyl, or, if induced hypotension was part of the technique, beta-blockers may have been given. Alternatively, it may be that, during the procedure, there was significant spinal cord ischemia that has led to loss of sympathetic tone as well as loss of the cardioaccelerator innervation of the heart. If there were abnormalities in the SSEP tracings during the case, this possibility should be given careful consideration [19]. In any case, the lack of a heart rate response to the inadequate preload is only worsening the clinical picture here and increasing the importance of rapid expansion of the intravascular volume [20].

4. Sleepiness can be the result of intraoperative medications, specifically opioids. Opioid effects include somnolence and depression of respiratory drive. If the ABG shows elevated $PaCO_2$ opioid-induced depression of her respiratory drive must be considered. Whether or not to treat this with a reversal agent like naloxone depends upon the degree of hypercarbia. Naloxone administration in this setting must be undertaken carefully, starting with low doses. The starting dose of naloxone for respiratory depression in this case should be 1 mcg/kg IV with additional doses given as needed. An ABG taken at this point would likely show a metabolic acidosis. This is an urgent situation. The child may be at the limit of her ability to sustain an inadequate intravascular volume without suffering a cardiac or respiratory arrest. The ABG obtained will include a hemoglobin measurement, and that number will guide IV fluid therapy. Fluid resuscitation should begin immediately with available crystalloid. Once hemoglobin is available and colloid has become available, the specific fluid given can be tailored to the situation. Consideration should be given to measurement of central venous pressure if there is not a rapid improvement in the blood pressure once intravascular volume has been even partially replenished.

## Case 4

An 18-month-old underwent uneventful tonsillectomy/adenoidectomy for peripheral and central sleep apnea and received 3 mcg/kg fentanyl with a sevoflurane/oxygen/nitrous oxide anesthetic. He awakened in the OR, but now you are called to the bedside for periodic breathing, breath holding, and an $O_2$ saturation of 86%.

**Questions**

1. What might be going on?

## Answers

1. This is an urgent situation. As the delivered $FiO_2$ is increased, the child must be quickly evaluated and the cause determined. The child may have a decreased functional residual capacity (FRC) due to atelectasis or extravascular lung water that has led to the hypoxemia on the basis of V/Q mismatch. If this child has reactive pulmonary vasculature and elevated pulmonary vascular resistance as a result of long-standing obstructive sleep apnea, a brief period of hypoxemia may lead to a longer period of pulmonary artery hypertension with a resulting shunt at the atrial level across a patent foramen ovale. It is possible that long-standing elevated pulmonary artery pressure has resulted in right heart failure (cor pulmonale). It is important to treat the oximeter reading as accurate and begin therapy and evaluation. However, it is possible that the reading of 86% does not represent the true arterial saturation. The pulse oximeter calculates $SpO_2$ using an algorithm based on the differential absorption of red and infrared light by $HbO_2$ and Hb. $HbO_2$ absorbs less light in the red wavelength and more in the infrared wavelength. Absorption of light of 800 nm wavelength is identical by both $HbO_2$ and Hb. In cases of probe malposition or movement artifact, the algorithm may calculate an inaccurate $SPO_2$, and when this happens, the monitor reads an $SPO_2$ of 85–86%.

   If the child had laryngospasm following extubation at the end of the case, another cause for the hypoxemia would be pulmonary edema. Occasionally, after laryngospasm, vigorous inspiration against a closed glottis can lead to the rapid development of negative pressure pulmonary edema. This can be diagnosed on the basis of typical findings on both physical exam and plain CXR.

   The ABG shows a mixed respiratory and metabolic acidosis. There is moderate hypercarbia and a more severe acidosis than would be caused by the $PaCO_2$ alone. It can be assumed that the child has not had chronic hypercarbia, and this can be confirmed by measurement of the bicarbonate. Hypoxemia is also present but the degree cannot be ascertained without knowledge of the $FiO_2$. The hypoxemia is concerning but if the ABG were done while the child's $FiO_2$ was low, then intubation and distending airway pressure may not be indicated. The decision whether or not to reintubate should be made on the entire picture, the condition of the child, and the expected course he will take over the next 30–60 minutes is as important as the laboratory evaluation.

   The baby likely has postintubation stridor. Postoperative, postintubation stridor occurs in approximately 1% of children, particularly in children < 4 years of age [21, 22]. The syndrome is the result of edema of the tracheal wall below the

2. How does dexamethasone work to prevent postintubation edema? Is it effective if administered post hoc? What will you do?

3. Will sedation help? Does cool mist help? In what way?

4. Would the administration of nebulized racemic epinephrine help the child? How much would you give? How would you administer it?

level of the vocal cords. The mucosa in this area is not as tightly joined to the submucosa so that when edema develops there, the lumen is compromised to a greater degree than it would be if the mucosa and submucosa were tightly adherent to each other. Resistance to airflow in the trachea is inversely proportional to the radius taken to the fourth power for laminar flow and to the fifth power for turbulent flow. With subglottic edema and increased respiratory rate and effort, the gas flow will be turbulent. In this situation, the flow is now inversely proportional to the radius to the fifth power. Therefore, the smaller the tracheal lumen prior to the development of subglottic edema, the more likely it is that any edema will lead to clinical symptoms.

Poiseuille equation:

$$P = \frac{8Q\mu L}{\pi R4}$$

$Q$ = volumetric flow rate
$\mu$ = viscosity, lb·sec/in$^2$
$L$ = tube length, in
$R$ = tube radius, in

Some risk factors have been identified for the development of this problem. Tight-fitting endotracheal tubes, movement of the head and neck while intubated, intubation for 1 hour, and the presence of a URI may predispose the child to the development of postintubation croup [21, 23, 24]. Stridor usually develops soon after extubation and can worsen for several hours after it first is clinically apparent. In this case, the airway manipulations, including rigid bronchoscopy, are the causes of the stridor. In the situation where the $SPO_2$ is not reading, it is important to administer a high $FiO_2$. Without a reliable $SPO_2$ measurement, one must rely on the clinical picture in evaluating the patient.

2. Treatment of postintubation stridor includes the administration of dexamethasone (decadron), a potent, longer-acting glucocorticoid. This medication will act to limit the degree of inflammation and the severity of the subglottic edema. Decadron administration will not decrease edema already present, however [25].

3. Sedation should be avoided. Depression of respiratory drive is dangerous in this situation. Nebulized racemic epinephrine may help by actually decreasing the degree of subglottic edema. The duration of the effect is generally 1 hour or less.

4. Nebulized racemic epinephrine will act to decrease the subglottic edema. Once racemic epinephrine has been administered to a child with postintubation croup, the child should be admitted for observation with the expectation that subsequent doses will be needed. The usual dose of racemic epinephrine is 0.5 mL of the 2.25% solution diluted into 3–5 ml of NS and administered via a nebulizer. The mask is held near the child's face with 100% oxygen used to nebulize the solution.

5. Should this baby be reintubated? Why? What tube size would you choose?

## Case 5

You are called to the bedside of a 10-month-old who has just undergone a cleft palate repair. He has a temperature of 41.2 °C.

**Questions**

1. What might have happened that his temperature got to this point?

2. Could this represent malignant hyperthermia? Why/why not?

5. In the event that reintubation is necessary, a smaller than normal endotracheal tube should be used. Intubating conditions should be as good as possible in order that the intubation does not cause additional trauma. The narrowed part of the airway, the subglottis, will not be visible to the laryngoscopist, but if the tube meets resistance once the tip is beyond the vocal cords, a smaller-diameter tube should be used.

**Answers**

1. Postoperative hyperthermia usually is the result of excessive warming and is more common in pediatric patients than adult patients. In cases such as repair of cleft palate, the child is well covered by surgical drapes. If warming is undertaken during the case with devices such as forced hot air mattresses, warming blankets, humidification, and warming of inspired gases, the child's core body temperature could easily rise to the level noted in the cases here. Hyperthermia in the PACU that is the result of excessive warming in the OR generally dissipates rather quickly once the sources of additional heat are removed. If the elevated temperature persists, other causes must be sought.

2. It is possible that the temperature elevation is part of a response to systemic infection. Of course, any case of temperature elevation should bring the possibility of malignant hyperthermia to mind. The anesthetic record should be reviewed to learn the time course of the temperature elevation and to review the medications administered during the anesthetic. It is likely that the child would have been exposed to potent inhaled agents during the anesthetic. The presentation described here would be quite unusual for malignant hyperthermia. Most cases of malignant hyperthermia develop within the first few hours of an anesthetic, but there are reports of MH occurring well after the conclusion of a case. It is also unusual for fever to be the presenting sign of an episode of MH. Often tachycardia, hypertension, and tachypnea are noted first. If the child in this case were developing MH, mottled skin and muscle rigidity would be expected. The most consistent laboratory finding in cases of MH is a combined respiratory and metabolic acidosis.

   If MH is being considered in this child, laboratory evaluation including arterial and venous blood gas analysis, serum electrolytes, lactate, a coagulation profile and creatinine should be done prior to instituting any therapy. Other studies that may take longer to return but that will help to confirm the diagnosis such as CPK, myoglobin can also be sent [26].

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The present guidelines were compiled by a multidisciplinary international panel of indi- viduals with interest and expertise in postoperative nausea and vomiting (PONV). These guide- lines identify risk factors for PONV in adults and children; recommend approaches for reducing baseline risks for PONV; identify the most effective antiemetic monotherapy and combination therapy regimens for PONV prophylaxis; recommend approaches for treatment of PONV when it occurs; and provide an algorithm for the management of individuals at increased risk for PONV.

# Chapter 51
# Critical Care

**Thomas J. Mancuso**

A 4-year-old with gram-negative sepsis being treated with broad spectrum antibiotics is admitted to the PICU for increasing respiratory distress.



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## Case # 1: Acute Respiratory Distress Syndrome (ARDS)

### Questions

1. What do you think is going on? Why?

2. How does endotoxin release promote ARDS? What are some of the vasoactive mediators involved?

3. How does the release of leukotrienes, tumor necrosis factor (TNF), and prostaglandins involve the pulmonary circulation in ARDS? What implications are there for you as a critical care anesthesiologist? Why?

## Answers

1. This child may be developing acute respiratory distress syndrome (ARDS) [1–3]. ARDS is characterized by the following:

   (a) Acute onset of symptoms
   (b) Severe respiratory failure with $PaO_2/FiO_2 < 200$ mmHg regardless of positive end-expiratory pressure (PEEP) levels
   (c) Chest X-ray that shows bilateral infiltrates
   (d) Lack of clinical evidence that left ventricular (LV) failure is the etiology of the respiratory distress

   In addition, the deterioration of pulmonary function often is associated with a non-pulmonary clinical insult [4]. In pediatrics, the more commonly associated conditions are shock, sepsis, or drowning. Other associated conditions include massive transfusion, smoke inhalation, burns, or trauma [5].

2. Since sepsis is associated with the development of ARDS, endotoxins released by bacteria, mediators released by inflammatory cells, and other compounds such as complement, products of disseminated intravascular coagulopathy (DIC), prostaglandins, and leukotrienes have all been evaluated for their role in the development of this clinical syndrome. Although the effects of these many mediators are interconnected, endotoxin has been shown to have several effects itself. Lipopolysaccharide from gram-negative bacteria has been shown to directly affect the integrity of the endothelium and also to stimulate macrophages to release tumor necrosis factor (TNF) and interleukin (IL)-1 [6]. One source of endotoxin in patients with sepsis and hypotension is thought to be the pulmonary capillary endothelium that is damaged. This damage is caused by the release of many of the mediators mentioned above. Once the integrity of the alveolar-capillary membrane is disrupted, a proteinaceous, hemorrhagic fluid enters the alveolar space.

3. Another effect of some of these mediators, particularly products of arachidonic acid metabolism such as the leukotrienes and prostaglandins, is to contribute to the development of pulmonary hypertension. A cycle of pathology ensues. As hypoxia worsens, the pulmonary artery pressure rises further, and with continued release of the various mediators, pulmonary edema worsens, and reactivity of the pulmonary vessels to hypoxia also worsens. Although the role(s) of mediators in ARDS is being more and more well characterized, therapy directed toward these compounds is still investigational. Studies of the use of steroids have been disappointing. Steroid administration has neither reversed the pathophysiology nor decreased mortality in ARDS [7, 8].

4. Is there a role for surfactant instillation in such patients? Does surfactant work normally in patients with ARDS? Why/why not? What can you do about it?

5. The child requires intubation. What ventilation strategy is best for her once intubated? Will PEEP help? Why? What are the adverse effects of high ranges of PEEP? How can these effects be minimized?

4. In ARDS many of the pathophysiologic abnormalities are due to diminished activity of surfactant. Functional residual capacity (FRC) is reduced and lung compliance is decreased. Surfactant acts to stabilize alveoli. Surfactant keeps surface tension proportional to surface area, allowing smaller alveoli to remain inflated at the same transpulmonary pressure as larger alveoli. Without surfactant, smaller alveoli would empty into larger ones, resulting in areas of collapse. In the lab, it has been shown that the surface activity of phospholipids from patients with ARDS is poor. Surfactant administration to preterm newborns with RDS, while not curative, has had some salutary effects on the course of the disease such as decreased mortality, decreased incidence of air leak, and improved oxygenation [5]. Unfortunately, administration of surfactant to patients with ARDS has not affected mortality.

5. The goal of therapy for patients with ARDS is to maintain adequate oxygen delivery while minimizing the harm of the therapy directed toward achieving that oxygen delivery [9]. Loss of FRC is an important part of the pathophysiology in ARDS. Application of PEEP increases the FRC. The likely mechanism for the increase in FRC is recruitment of previously collapsed terminal alveoli. PEEP also improves the static compliance of the lung. The pulmonary effects of PEEP in patients with ARDS, then, are increased FRC and improved compliance, resulting in increased $PaO_2$ and decreased shunt (Qs/Qt). The improved oxygenation is the result of better blood flow to ventilated alveoli. PEEP may also decrease cardiac output, however, primarily by decreasing venous return. At modest levels of PEEP, increasing intravascular volume may compensate for these deleterious cardiovascular effects. At some point, excessive PEEP will actually decrease oxygen delivery since the fall in cardiac output will exceed the increased oxygen content of the blood. Another deleterious effect of PEEP is the development of pulmonary edema. PEEP lowers pulmonary interstitial pressure, increasing the pressure gradient for an increase in extravascular lung water (EVLW). Inhaled nitric oxide (iNO), a pulmonary vasodilator, has been administered to patients with ARDS. Since pulmonary arterial hypertension is a large part of the pathophysiology of the syndrome, iNO could be an effective treatment. Because it would only be delivered to well-ventilated parts of the lung, iNO in theory could improve V/Q matching while it lowered PAP. In adult studies, iNO has indeed been shown to decrease peak airway pressure and Qs/Qt. There is an emerging experience with iNO administration to children with ARDS, but no controlled studies have documented improved survival. Fluid therapy for children with ARDS can have a significant effect on the course of the illness. While both crystalloid and colloid will increase the intravascular volume initially and both will eventually leak into the alveoli, there are differences. In general, less colloid will leak into the alveoli. The amount of colloid that leaks into the alveoli depends on the molecular weight of the colloid. Pentastarch leakage into alveoli was less than hetastarch in an animal model of septic shock. Blood administration has many advantages in these patients. Oxygen delivery is increased immediately, and cardiac output is increased as preload is augmented.

6. Under what circumstances would you consider extracorporeal membrane oxygenation (ECMO)?

## Case # 2: Shock/Multiorgan System Failure

### Questions

A 15-year-old girl is admitted to the ICU with a presumptive diagnosis of toxic shock syndrome. Her blood pressure is 64/20 mmHg, heart rate 142/min; she is intubated and mechanically ventilated. Her arterial blood gas ($FiO_2$ = 1.0) is pH = 7.12, $PaO_2$ = 97 mmHg, and $PaCO_2$ = 32 mmHg.

How will you proceed? What do you think is going on? She has no urine output; Foley catheter is in place. You place a pulmonary artery catheter; pulmonary capillary wedge pressure (PCW) is 3 mmHg, and PA pressures are elevated at 42/22 mmHg. What now? Should you give sodium bicarbonate? Why not? Will it make the dopamine work better? How does that happen?

In addition, the packed red blood cells (PRBCs) are much less likely to leave the vascular space in significant amounts compared with colloid molecules or the ions in crystalloid.

6. While ECMO does not appear to offer advantages in the care of adults with ARDS, it may be of benefit to children with this syndrome. The criteria for institution of this therapy (ECMO is not a treatment) have changed over time, but some underlying considerations remain. Among the considerations for determining the suitability of a patient for ECMO are the severity of the lung disease, the reversibility of the lung disease, and the involvement of other organ systems. The severity can be judged using a variety of measures such as the oxygenation index (OI), the A – a gradient, and Qs/Qt. The OI $(MAP \times FiO \times 100)/PaO_2$) has been used to evaluate possible ECMO candidates for some time. An OI > 40 was believed to represent > 90 % risk for mortality.

## Answers

This patient's condition meets the criteria for shock. She has evidence of circulatory failure and inadequate tissue perfusion [10]. The clinical diagnosis of shock is supported by the following: tachycardia, hypotension, poor capillary refill, oliguria, decreased pulse pressure, and tachypnea. The shock is probably due to both hypovolemia and maldistribution of the circulating blood volume. Abnormal vasomotor tone will exacerbate the effects of pre-existing hypovolemia. She may have inadequate preload due to a variety of factors: recent poor PO intake during the prodrome of the illness and loss of intravascular volume through capillary leak. The distributive aspect to shock in this case results from the endotoxins released in the syndrome of toxic shock. These mediators diminish sympathetic tone, and the resulting lowered systemic blood pressure contributes to impaired perfusion of organs. The patient is responding to decreased stroke volume with an increased heart rate, but with the low systemic blood pressure, perfusion will be inadequate. The first priority in this situation is to improve cardiac output [6]. Increasing preload should be done first.

A rapid infusion of isotonic IV fluid, 20 mL/kg, should begin the improvement. If the patient's perfusion still is inadequate after 40–60 mL/kg of isotonic IV fluid, placement of a central venous line (CVL) should be considered to monitor preload more directly and also to support cardiac function. The arterial blood gas (ABG) shows a metabolic acidosis, severe hypoxemia, and slight

hyperventilation. The patient has an alveolar to arterial (A – a) gradient of >500. The $PAO_2$, the alveolar partial pressure of oxygen, can be calculated with the formula

$$P_AO_2 = \left(FiO_2 \times \left(760 - 47\right)\right) - P_aCO_2 / 0.8$$

$$A - a \text{ gradient} = P_AO_2 - P_aO_2$$

For this child, the calculation is

$$\begin{aligned} P_AO_2 &= \left(1.0 \times 713\right) - \left(32 / 0.8\right) \\ &= 713 - 40 \\ &= 673 \end{aligned}$$

$$\begin{aligned} A - a \text{ gradient} &= P_AO_2 - P_aO_2 \\ &= 673 - 97 \\ &= 576 \end{aligned}$$

For a well person breathing room air, the calculation is

$$\begin{aligned} \mathrm{P_AO_2} &= \left(0.21 \times 713\right) - \left(40 / 0.8\right) \\ &= 149 - 50 \\ &= 99 \end{aligned}$$

$$\begin{aligned} &P_aO_2 \text{ in a well person with } SpO_2 = 100\%, \\ &P_aO_2 = 110 \\ &A - a \text{ gradient} = 110 - 99 \\ &\qquad\qquad = 11 \end{aligned}$$

As preload is replenished and cardiac contractility is improved, there should be improvement in $PaO_2$. If the patient's A-a gradient does not improve as the circulatory disturbances are corrected, the ventilator setting should be adjusted. The patient may have better oxygenation with increased peak inspiratory pressures, a higher PEEP, or a change in the I/E ratio. Increases in mean airway pressure will generally increase oxygenation but will also affect venous return. As ventilator settings are adjusted, the cardiovascular parameters must be carefully observed for deterioration.

The acidosis is most likely due to impaired delivery of oxygen and substrates to the tissues, the pathophysiologic abnormality in shock states. Correction of the circulatory disturbances will lead to correction of the metabolic acidosis. If severe, acidosis will affect the function of many enzymatic systems, including those responsible for myocardial performance. In cases of severe acidosis, drugs such as

## Case # 3: Neurologic Intensive Care

### Questions

A 5-year-old is admitted to the ICU with a small epidural hematoma and closed head injury. The neurosurgeon wants to observe him and will take the child to the OR only if the child demonstrates acute deterioration.

1. Do you agree with this? What is the basis for observing a patient with an epidural hematoma?

2. The patient arrives from the ER with an IV of D5 1/2NS. Would you change it? Why? Under what circumstances would you reconstitute the IV fluids?

dopamine that enhance cardiac contractility have severely diminished effectiveness. With the pH seen in the ABG, dopamine is likely to have some effect and improve contractility. The pulmonary capillary wedge pressure (PCWP) reflects left atrial pressure, which, in turn, reflects LV end-diastolic pressure or preload. A PCWP of 3 mmHg in this case indicates a relatively low preload. The PA pressure of 42/22 mmHg is elevated. These numbers together indicate a lower preload with PA hypertension and increased RV afterload. Further treatment will be guided by the child's clinical response to fluid therapy and dopamine, urine output, ABGs, and mixed venous blood gas analysis. If the patient continues to exhibit clinical signs of shock and continues with a base deficit of >6 mEq/L, administration of bicarbonate is indicated. Initial dosing of bicarbonate can be estimated with the formula mEq $NaHCO_3 = 0.3 \times (\text{weight in kg}) \times (\text{base deficit})$.

## Answers

1. Although most epidural hematomas, a collection of blood between the skull and the dura, are treated with emergency craniotomy and evacuation of the blood/clot, stable patients with this problem can be managed conservatively [11]. Epidural hematomas are not as common in children as adults, possibly because the dura is adherent to the inner table of the skull, especially at the suture lines. Children with an epidural hematoma have had head injuries severe enough to separate the dura from the skull [12]. The collection of blood is often stopped at a suture unless there is an associated skull fracture that crosses the suture line. Children with epidural hematomas who are managed conservatively must be very carefully monitored. The child should have a repeat CT if any neurologic deterioration is noted [13]. Epidural hematomas that result from venous bleeding may continue to enlarge for up to 24 h. Clinical outcome in children with epidural hematomas is related to the speed of evacuation when there is clinical deterioration. If the child is kept in the PICU and carefully monitored and CT is immediately available, he/she may recover without having to undergo a craniotomy.

2. Isotonic fluid should be used in children with neurologic injuries [14, 15]. Glucose should be avoided except when it is needed to treat symptomatic hypoglycemia. Glucose administered IV can quickly enter the brain and increase water content, and hypotonic IV fluid administration can also increase brain water content.

3. The nurse asks you what position you would like the head of the bed to be in. Your answer? Why? What are the considerations in your answer? What is the optimal positioning for a patient with a space-occupying lesion and possible elevated intracranial pressure?

4. During the night, the patient becomes whiny and combative, and the parents ask if he can be sedated; do you agree? What may be going on? How would you evaluate? What would you be looking for in the physical exam while awaiting the CT scan?

5. He has a seizure 1 h after eating dinner. How would you treat? Would you use a barbiturate? Benzodiazepine? Is phenytoin indicated? Would you use phenytoin or fosphenytoin? Should the patient be intubated?

6. What about using succinylcholine in this setting? Would you? What is your differential diagnosis for the cause of the seizure? Assuming the patient is intubated, how will you prevent rises of intracranial pressure during suctioning and noxious procedures? Why is this important? What is the optimal drug regimen to use in order to blunt the elevation of ICP?

3. In this child with an intracranial mass, therapy should be directed to minimizing the volume of the intracranial contents. The patient's optimal head position would be head up, at 30 degrees and in the midline. This position allows the best cerebral venous drainage, keeping the cerebral blood volume low. If mechanical ventilation is instituted, airway pressures used should be kept low since elevated airway pressures decrease cerebral venous drainage [15].

4. A change in the patient's condition, particularly his mental status, is a cause for alarm. He should not be sedated. While a CT is arranged, he should be evaluated for signs of increased intracranial pressure. If the epidural hematoma is enlarging and the intracranial pressure is rising, the child will exhibit altered mental status, hyperventilation, and systemic hypertension [16]. Cranial nerve signs may be noted as well. Palsy of the third nerve may occur as it is pressed between the falx and the expanding brain, leading to asymmetry of the pupils.

5. Posttraumatic seizures occur regularly in children who experience head trauma [17, 18]. The more severe the trauma, the greater the incidence. Following severe trauma, up to 30% of children will have seizures. Children who have hematomas or depressed skull fractures are at higher risk for the development of seizures. Seizures should be treated quickly and effectively. Oxygen consumption is greatly increased during seizures, as is intracranial pressure (ICP). IV benzodiazepines will stop most seizures. An alternative would be an administration of IV barbiturates. In treating the seizures, respiratory drive may be impaired. It may be prudent to protect the patient's airway and begin mechanical ventilation. Another reason to intubate the patient and begin ventilation is that the child will likely soon be taken to the OR. The seizures may indicate that the hematoma has enlarged, and even if it has remained the same size, surgery and evacuation may be indicated. Intubation of this patient presents several problems.

6. He has a full stomach and raised ICP. Even if he had not recently eaten, he should be treated as though he has a full stomach since he had suffered head trauma earlier and that event would have resulted in delayed gastric emptying. During laryngoscopy and intubation, every effort should be made to minimize, if not eliminate, hemodynamic perturbations. Intracranial pressure will increase if blood pressure increases, if the $PaCO_2$ increases, or if $PaO_2$ decreases. The use of an appropriate dose of hypnotic is essential to prevent hypertension and tachycardia. The use of succinylcholine as the muscle relaxant to facilitate intubation is controversial. This relaxant can increase ICP in patients with reduced intracranial compliance. The mechanism is most likely a reflex increase in cerebral blood flow resulting from increased afferent muscle spindle activity. The increase in ICP caused by succinylcholine can be blunted by prior administration of a defasciculating dose of a nondepolarizing relaxant. Hyperkalemia has been seen after administration of succinylcholine in patients with various CNS problems including closed head injury. Although the exact period of vulnerability is unknown, it does not begin until at least 24–48 h after the injury. The advantage

7. He begins to develop “raccoon eyes” and discoloration of the eyelids. Now what do you think?

8. He becomes increasingly difficult to ventilate, with O2 saturations that are slowly but progressively decreasing and coarse-sounding breath sounds, reminiscent of rales. Next step? Diagnostic or therapeutic?

of the use of succinylcholine in this situation is its very rapid onset that minimizes the chance for aspiration of gastric contents or the development of hypoxemia/hypercarbia. Once the child is intubated, it is important to provide the brain with adequate cerebral perfusion pressure (CPP = CVP or ICP – MAP). This is done by maintaining systemic arterial pressure and minimizing intracranial pressure. Often, these patients are best cared for sedated and relaxed. An arterial line is needed and often an intracranial pressure monitor as well. In the absence of an ICP monitor, the patients should be kept in the 30-degree head-up midline position and any signs of excess activity of the sympathetic nervous system treated with IV sedatives, opioids, or barbiturates. When these agents are administered, it is important to prevent excessive lowering of the blood pressure.

7. "Raccoon eyes" (periorbital ecchymoses) are one of the signs of a basilar skull fracture. Other clinical signs of this type of skull fracture are Battle's sign (retroauricular or mastoid ecchymosis), blood behind the tympanic membrane, and cerebrospinal fluid (CSF) otorrhea or rhinorrhea. Since the cribriform plate is disrupted in patients with basilar skull fractures, placement of tubes in the nose is to be avoided. Endotracheal and gastric tubes should be placed through the mouth. With conservative management, many patients with isolated basilar skull fractures do well. The most common morbidity is a persistent CSF leak.

8. Worsening pulmonary function in this setting may be due to the development of neurogenic pulmonary edema (NPE) [19], which can develop anytime from 2 h after the head injury to several days afterward. Although not certain, it is thought that a transient increase in sympathetic tone is responsible for the development of NPE. This sympathetic discharge leads to increased PVR, accompanied by damage to the endothelium allowing leakage of fluid into the alveoli. The presentation includes hypoxemia, CXR findings of diffuse fluffy infiltrates, and normal-to-low cardiac filling pressures. In most cases, neurogenic pulmonary edema resolves on its own. Pulmonary edema can significantly complicate the management of patients with raised ICP, however. Hypoxia must be avoided in all patients, but the increase in cerebral blood flow that results from a lower $PaO_2$ can be very deleterious to those with raised ICP or limited intracranial compliance. The application of high PEEP to improve oxygenation will decrease cerebral venous drainage, again affecting ICP.

## Case # 4: Pancreatitis

### Questions

A 3-year-old is admitted to the ICU following blunt abdominal injury after hitting a wall and falling over her tricycle. She is in intense abdominal pain and is not intubated.

1. She has a sentinel loop in her epigastrium on a flat plate X-ray of the abdomen (KUB). What is the significance of that?

2. Her CT scan obtained via the ED shows a fractured pancreas. What do you think? Anything you would watch out for?

3. What are the consequences of the release of pancreatic enzymes into the retroperitoneal space?

4. Does she need invasive cardiovascular monitoring for this problem?
5. It hurts her to breathe. Is there anything you can do to help? She is developing atelectasis. What will you do to treat this problem?
6. How will you support her intravascular volume? Why is this important?

7. Under what circumstances should surgery be considered?

## Answers

1. The most immediate threat to a child after blunt abdominal trauma is bleeding from injury to the spleen or liver. Other organs may also be injured. The pancreas and duodenum may be injured when a child suffers abdominal trauma from a high-speed deceleration or from child abuse. The hollow viscera may also be lacerated or torn from sites of attachment such as the ligament of Treitz. The evaluation of a child who has suffered blunt abdominal trauma must include a search for injuries to the GU system and the bony pelvis, the ribs, and the lumbar spine and sacrum. The child in this case suffered a relatively high-speed deceleration when she crashed against the handlebars of her bicycle. Her abdominal pain may be due to any of the injuries mentioned or a combination.

2. CT scanning has become an essential part of the evaluation of trauma patients. Not only will this test reveal the presence of injuries to the liver or spleen, but it also can be used to visualize the kidneys and pancreas. The finding of a sentinel loop on KUB indicates the presence of an ileus. It is likely that there is inflammation in the epigastric area as a result of the trauma. If there was damage done to the pancreas in the event and pancreatic enzymes were released, a localized ileus is likely. Another KUB finding seen in acute pancreatic damage when the transverse and descending colon are affected is the so-called colon cutoff sign. The CT findings confirm the diagnosis of fractured pancreas. It is important that damage to the liver and spleen is ruled out by the CT.

3. The release of pancreatic enzymes can be quite harmful in these patients. The laboratory evaluation of this child should include serum calcium, amylase, lipase, and a CBC. The laboratory evaluation can help with prognostication. A poor prognosis is associated with serum calcium <7.5 mg/dL, a WBC count of 20,000, a $PaO_2$ of 60 mmHg, a BUN >20 mg/dL, and a base deficit >4 mEq/L.

4. 5. and 6. If the prognosis appears to be poor, management should be more aggressive [20–23]. It might be prudent to intubate the trachea and also institute invasive hemodynamic monitoring sooner rather than later. She will splint her breathing due to pain and develop respiratory insufficiency. As the inflammation caused by the pancreatic enzymes released into the peritoneum increases, there will be greater and greater fluid transudation into the peritoneum, leading to intravascular volume depletion.

7. There are limited, but relatively clear-cut, indications for surgery for acute pancreatitis: (1) for differential diagnosis, when the disease may be something other

8. Her serum calcium is now 6.4 mg%; is that important? Why did it happen? What symptoms would you look for? What could you do about it?

9. What if her BUN is rising (let us say it is 24 mg% 16 h after admission)? Why is this important? What can you do about it? Why does this happen?

## Case # 5: Hemolytic Uremic Syndrome

### Questions

A 4-year-old with a 5-day history of diarrhea that seemed to be getting better has just been admitted to the ICU from the pediatrician's office with the following findings:

- Pallor
- Anuria
- Tachycardia
- Irritability
- Ataxia
- Tremors

Lab findings:

- HCT. 24 platelet count 76,000 BUN 46.
- The peripheral smear has evidence of microangiopathic hemolysis.

1. What do you think is going on? Whom would you consult?

than pancreatitis; (2) for persistent and severe biliary pancreatitis, when an obstructing gallstone that cannot be managed endoscopic ally is lodged at the ampulla of Vater; (3) for infected pancreatic necrosis; and (4) to drain a pancreatic abscess, if percutaneous drainage fails.

8. The etiology of the hypocalcemia seen in these patients is due to saponification of fatty acids, sequestration of calcium in bones, and hypomagnesemia. Calcium is involved in cardiac conduction, neuromuscular transmission, and muscle contraction, and all these are altered by hypocalcemia. Patients with hypocalcemia can exhibit cardiac arrhythmias, a prolonged QT interval, and seizures. They may experience tingling, numbness, and muscle weakness. Treatment is not only with IV calcium, as gluconate or chloride, but magnesium as well.

9. If this child has an elevated BUN, it indicates prerenal azotemia, likely from fluid loss into the peritoneal space. The response should be to increase her intravascular volume. Pancreatic rest should be undertaken while she is watched and monitored carefully as described. A nasogastric tube should be placed, and IV hyperalimentation should be started soon since these patients often must be kept NPO for several days or more than 1 week while the pancreas heals.

**Answers**

1. This patient has hemolytic-uremic syndrome (HUS). He has had a typical prodrome of several days of diarrhea. In some cases, the diarrhea is bloody. HUS is characterized by microangiopathic hemolytic anemia, thrombocytopenia, and

2. The renal service recommends dialysis. What is the patient's prognosis if dialysis continues for >2 weeks? Is there a risk for chronic nephropathy following 5 days of dialysis with apparent resolution?

renal cortical injury. This condition occurs in children between 6 months and 4 years of age, and it may occur sporadically or in epidemics in the USA. In other countries such as Argentina, HUS is endemic.

The exact etiology is not known, although toxins have been implicated in many cases of HUS [24]. The syndrome is seen following colitis caused by certain *E. coli* or *Shigella* species. Several viruses, such as EBV, influenza, and Coxsackie, have also caused HUS. In most cases, it seems that the initial event in the development of HUS is injury to the vascular endothelium with subsequent platelet activation. Adhesion of platelets to the damaged endothelium leads to thrombocytopenia. Damage to RBCs during passage through damaged vascular beds results in the development of microangiopathic hemolytic anemia. The mechanism of renal damage also involves endothelial injury, specifically the endothelium of the glomerular capillaries. The laboratory evaluation of this patient should include, in addition to those tests noted above, a full set of electrolytes and evaluation of the coagulation system. Children with HUS may be dehydrated due to the diarrhea. In addition there may be significant electrolyte abnormalities such as hyper- or hyponatremia, hyperkalemia, metabolic acidosis, and hypocalcemia. Seizures may be seen in affected children. There is no specific treatment for HUS other than careful supportive management of fluids and blood pressure and transfusion of blood products as the clinical condition dictates.

2. Many children with HUS require dialysis, but most children do recover and have normal renal function. Children who have had HUS should be followed for the development of hypertension or chronic renal failure. The best outcome is seen in children who have epidemic HUS with the typical prodrome. Familial cases or children who develop HUS at older ages do not have as favorable a prognosis.

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This chapter reviews acute respiratory failure from any cause in children including epidemiology, clinical and laboratory evaluation, management, and complications.

Marraro GA. Protective lung strategies during artificial ventilation in children. Paediatr Anaesth. 2005;106:1813–9.

This paper reviews various recruitment and ventilation strategies for patients with ARDS and included CT scans of adults during various respiratory maneuvers. Also discussed are the concepts of barotrauma and volutrauma.

Petersen TL, Lee KJ. Chap 40. Shock the acutely injured child. In: Marcdante KJ, Kliegman RM, editors. Nelson essentials of pediatrics. 8th ed. Philadelphia: Elsevier; 2019. p. 151.

This chapter reviews the following categories of shock: hypovolemic, distributive, cardiogenic, distributive, and obstructive shock. General treatment principles and specific interventions for the various types of shock are discussed.

Smith LS, Srnivasarao B, Hernan LJ. Shock states, Chap. 30. In: Fuhrman BP, Zimmerman JJ, editors. Pediatric critical care. 5th ed: Elsevier; 2017. p. 417–29.

Holzman R. Trauma and casualty management. In: Holzman R, Mancuso TJ, Polaner DM, editors. A practical approach to pediatric anesthesia. 2nd ed. Philadelphia: Wolters Kluwer/Lippincott Williams and Wilkins; 2016. p. 783–805.

This chapter reviews the general principles in the care of pediatric trauma victims as well as management of specific types of traumatic injuries including head injury, spinal cord injury, and abdominal injuries. Also discussed is the organization of trauma centers for mass casualty situations.

Corrigan JJ, Boineau FG. Hemolytic-uremic syndrome. Pediatr Rev. 2001;22(11):365–9.

This paper reviews the epidemiology and management of HUS in children and also the immediate and longer-term prognosis for recovery from this condition.

# Chapter 52
# Resuscitation

**Thomas J. Mancuso and Joseph P. Cravero**

A 4-year-old male is brought to the emergency department unresponsive and without palpable pulses after an incident where he was underwater for "many minutes." You are asked to help with resuscitation.



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## Management of Prolonged Submersion

### Questions

1. How is pediatric cardiopulmonary arrest different epidemiologically from adult arrest? What are the signs of impending respiratory failure? What is shock? Differentiate *compensated* vs. *uncompensated* shock.

2. The ECG shows ventricular fibrillation. When do you attempt defibrillation? What energy level do you use in a child for ventricular fibrillation? What situations call for synchronized cardioversion and what energy level is required?

3. Which should come first when beginning the resuscitation process, ventilation or chest compressions? How should chest compressions be performed? How should rescue breathing be accomplished in a child with cardiac arrest? Where should you feel for pulses in an infant or child?

## Answers

1. In adults cardiac arrest is most often cardiac in origin. Conversely, cardiac arrest in children is usually the result of progressive respiratory deterioration associated with hypoxemia, acidosis, and progressive loss of cardiac performance. The cause of this can be anything from an extrathoracic obstruction of the airway to bronchiolitis, severe asthma, or pneumonia. When arrest in children is not respiratory in origin, it is often due to sepsis, drug toxicity, or hemorrhage from trauma. Notably the arrest state is usually preceded by a period of deterioration during which appropriate intervention can prevent the progression to arrest. Reversible causes of arrest can be remembered by the Hs and Ts—hypovolemia, hypoxia, hydrogen ion (acidosis), hypoglycemia, hyperkalemia, and hypothermia and tension pneumothorax, tamponade, toxins, thrombosis (pulmonary), and thrombosis (coronary). Respiratory failure is defined as the inability to maintain adequate oxygenation or ventilation and is signaled by increases in work of breathing such as tachypnea, grunting, and nasal flaring and the use of supraclavicular or intercostal muscles (appearing as retractions). Shock refers to inadequate circulation (and oxygenation) to meet the metabolic needs of an individual. Shock can be categorized into hypovolemic, cardiogenic, or distributive—although clinically it is rarely easy to completely separate these entities. Compensated shock is characterized by blood pressures at or near normal range, whereas decompensated shock is associated with hypotension.

2. Defibrillation (unsynchronized shock) should be delivered as soon as possible after the child is found to be in ventricular fibrillation or pulseless ventricular tachycardia. CPR should be performed while setting up the defibrillator. The shorter the time interval between the last chest compression and defibrillation—the better chance for conversion to a perfusing rhythm. It is recommended to be less than 10 s. The exact dose for defibrillation in a child is not known. An initial energy dose of 2–4 J/kg is advised. Generally 2 J/kg is given as an initial shock and then 4 J/kg thereafter. Higher doses may be used, but it is not recommended to deliver more than 10 J/kg. Two minutes of CPR should be provided between shock attempts. Synchronized cardioversion is indicated for ventricular tachycardia that has a pulse but is associated with cardiopulmonary compromise. The energy for cardioversion is 0.5–1.0 J/kg and repeated with 2 J/kg if repeat shock is needed. Synchronized cardioversion is similarly recommended for supraventricular tachycardia that is associated with cardiopulmonary compromise.

3. Current guidelines from the American Heart Association recommend that chest compressions should begin prior to ventilation [1–3]. In the fully staffed and equipped emergency department, these two interventions would start almost simultaneously, but it is important to recognize the priority of starting chest compressions as soon as possible for a child without a spontaneous circulation—

4. How would you go about finding IV access? When should you go to intraosseous administration of medications? Which drugs can be given in this manner? What is the indication for intratracheal administration of resuscitation medications? How would you change your dose administration in these cases?

and not wait for ventilation equipment or other advanced resuscitation equipment. Prior to starting CPR, for this child, I would provide compressions that were about 1/3 the AP diameter of the chest at a rate of 100 compressions per minute or greater. The individuals providing compressions should be changed every 2 min. Rescue breathing must be tailored to the age of the patient and the clinical scenario. In this case, I would clear the airway, provide a jaw thrust, insert an oral airway, and begin positive pressure ventilations. The ventilation would be calibrated by the chest wall movement that I observed—only the pressure required to provide good chest rise would be needed. While preparing to place an endotracheal tube, I would provide 2 breaths for every 15 compressions delivered by the other code team members. The timing of breaths to compressions should be 2 breaths for every 30 compressions if you are providing CPR alone. Intubation should occur when all equipment and personnel that are needed are present and (ideally) without interrupting chest compressions. After intubation, I would provide a breath every 6–8 s, and these would not need to be synchronized with the compressions. If an infant or child is unresponsive and not breathing, I would take 10 s or less to feel for a pulse. I would feel for the brachial pulse in an infant and at the carotid or femoral artery for a child. If pulse is not felt or I am uncertain of a pulse, I would start chest compressions.

4. In most cases of pediatric cardiac arrest, a peripheral line will provide adequate access to the circulation to assist in resuscitation. In this case I would use an ultrasound probe to help guide an attempt at peripheral access in the antecubital or saphenous vein while CPR was ongoing. Central access is not required. In addition, acquisition of central access generally requires an interruption in CPR that peripheral access does not require. Possible complications associated with obtaining central access during CPR include vascular lacerations, hematoma formation, pneumothorax, and bleeding. If attempting central access, it would be advisable to obtain an ultrasound view of the vascular structure and interrupt CPR for the *minimum* time required for one access attempt prior to resuming CPR. With a peripheral line, central access of the drug is slightly delayed which requires the drug be given by a bolus injection, continuing chest compressions, and flush with at least 5 cc of isotonic solution after each administration. If IV access cannot be obtained with three attempts, intraosseous (IO) access should be obtained as it offers an effective route for administration of medications and fluids. IO is preferable to intratracheal administration. It is best to use a needle specifically designed for IO access (with a stylet that prevents obstruction of the needle with cortical bone during insertion), but butterfly needles, standard hypodermic needles, and spinal needles have all been successfully employed for this purpose. As a last resort, drugs can be given by the intratracheal route. Lipid-soluble drugs are absorbed through the trachea—this includes lidocaine, epinephrine, atropine, naloxone, and vasopressin. It should be noted that drug absorption would be erratic. The typical drug dose required when given IO is two to three times that of the IV route although with epinephrine, the dose is ten times the IV dose recommendation [1].

5. The child is in ventricular fibrillation with a tibial IO line in place. He remains in this rhythm without a pulse after two defibrillation attempts. What drug would you administer next and in what dose would you give that drug? What would you do if this does not result in return of spontaneous circulation?

6. A blood gas is obtained in the course of the resuscitation. The pH is 7.14 and the $CO_2$ is 45. Should bicarbonate be given—why or why not? A colleague suggests that calcium would be helpful in this situation. Do you agree? When is calcium indicated during resuscitation?

7. The child regains a spontaneous circulation but remains comatose. Is there anything that can be done to optimize chances for survival and minimize the chance of neurological injury?

5. At this point I would administer epinephrine at 0.01 mg/kg or 1 ml/kg or epinephrine 1/10,000 dilution. Epinephrine increases aortic diastolic pressure and coronary perfusion, which is a major determinant of successful resuscitation. I would deliver this every 3–5 min during the resuscitation. High-dose epinephrine administration can cause adverse effects and should be reserved for cases such as beta-blocker overdose. If there were no responses to this intervention, I would prepare to give amiodarone 5 mg/kg immediately after delivering the next shock. Amiodarone has alpha-adrenergic and beta-adrenergic blocking activity as well as effects on ion channels. It slows ventricular conduction. Alternatively I could give lidocaine 1 mg/kg.

6. Bicarbonate is not routinely recommended for patients with cardiac arrest. It may be helpful in specific situations where acidosis is severe and ventilation is adequate. In the absence of adequate ventilation, administration could lead to the accumulation of extra $CO_2$ which may cross the blood-brain barrier and lead to worsening acidosis in the CNS. Bicarbonate may be helpful in cases of hyperkalemia or hypercalcemia where the change in pH will drive ions into cells and out of the available circulation. It can also be helpful when resuscitating patients from tricyclic antidepressant overdose, in which case bicarbonate should be given to target a pH of approximately 7.5. This treatment narrows the QRS complex in these patients, improves systolic blood pressure, and controls ventricular arrhythmias. In the standard formulation (8.4%), bicarbonate is hyperosmolar and could be injurious to newborns. Because of this, there is a 4.2% solution that should be administered to neonates. Calcium is not recommended as it has not been found to improve survival and may be harmful. Calcium is indicated in cases where the serum calcium has been documented to be low or when serum potassium is high enough to create the danger of serious dysrhythmias. Ionized hypocalcemia is not uncommon in children with sepsis in which case it treats the underlying problem and causes potent vasoconstriction that will (at least transiently) increase blood pressure. Calcium should also be considered for cases of calcium channel blocker overdose and hypermagnesemia.

7. Out-of-hospital cardiac arrest in children often results in death or poor long-term neurological outcome. Many interventions have been tried to improve neurological outcome after cardiac arrest. Clinical trials involving treatment with barbiturates, steroids, calcium channel blockers, magnesium, and diazepam have all failed to prove beneficial. Therapeutic hypothermia has a mixed record of success. Several clinical trials *in adults* showed that therapeutic hypothermia (targeting 33 °C) improved neurologic outcomes in comatose survivors after out-of-hospital arrest. On the other hand, other adult trials have not shown benefit—probably owing to the overall management of fever in the non-hypothermic cohorts. While initial trials of hypothermia for hypoxic injury in neonates showed improved outcomes, most recent trials with good control of temperature in the non-hypothermic cohort have similarly failed to show improvement with hypothermia. The one randomized trial of therapeutic hypothermia in children (after

## Additional Questions

### Questions

1. A 2-year-old boy is undergoing a circumcision. The surgeon administers a penile block with 1 cc/kg of 0.25% bupivacaine. Shortly after the block is placed, you notice widening of the QRS complex and then ventricular fibrillation. Why did this happen? What would be your immediate treatment options? Why does this work?

2. The parents of a 9-month-old who presented to the emergency department with their child in extreme respiratory distress are present when the child has a cardiopulmonary arrest. Is it appropriate for them to stay during the resuscitation? How would you decide when to stop resuscitation efforts? What is the role of end-tidal $CO_2$ monitoring during resuscitation?

arrest) failed to show improvement in neurological outcome or survival overall in a hypothermic (33 °C) group [4]. In this case I would not opt for therapeutic hypothermia, but I would not actively warm the patient if the temperature were greater than 33 °C. I would attempt to optimize all other parameters that can affect neurological outcome, including seizure treatment, control of cerebral edema, managing blood pressure, controlling serum glucose, and generally attempting to make the physiological milieu as close to normal as possible [5, 6].

**Answers**

1. Local anesthetics such as bupivacaine block cardiac voltage-gated sodium channels preventing depolarization, blocking repolarization via potassium channels, and blocking the sarcoplasmic reticulum voltage-dependent calcium channels to limit the rise of intracellular calcium available for excitation-contraction coupling. In large doses bupivacaine also causes vasodilation, leaving the patient with a compromised myocardium and a very low vascular resistance. Bupivacaine also decreases the autonomic response to cardiac compromise at the nucleus tractus solitarius. This leads to further hypotension and dysrhythmias. I would immediately begin CPR, and I would administer intralipid 20% at 1.5 mL/kg over 1 min followed by an infusion of 15 mL/kg/h. If spontaneous circulation did not return in after 5 min, I would administer two further boluses of 1.5 mL/ kg at 5 min intervals and I would double the infusion rate to 30 mL/kg/h. The current hypothesis for the effectiveness of intralipid in treating local anesthetic systemic toxicity is the formation of a “lipid sink” that absorbs the lipophilic bupivacaine, thus reducing the amount available to bind to the myocardium. This therapy has clearly been proven in animal models to be superior to treatment with epinephrine or vasopressin. There is evidence of increased washout of bupivacaine in rat hearts in the presence of intralipid. Some have suggested that the lipid may act as a fatty acid substrate to enrich mitochondrial respiration in the heart [7].

2. I would allow the parents to stay during the resuscitation as long as there was an individual from my hospital team that was able to be with the family at all times, address their concerns or questions, and assist them in any way they needed during this event. In the past, families were asked to leave during a cardiopulmonary arrest because it was thought their presence would be traumatic to them and possibly inhibit the performance of the rescuers involved in the care of the child. Many studies have now shown that parents want to be present during resuscitation and that there is less litigation and concern about the care provided when

they are allowed to be present. For them, being present can confirm that everything possible was done to save their child, and it can give them the chance to "say goodbye" during the last minutes of life. Presence during these events has actually been shown to improve the grieving process [8, 9]. The decision to stop resuscitation efforts is complex and should take into consideration the cause of the arrest, the likelihood of the return of spontaneous circulation, pre-existing conditions, possible outcomes, duration of the resuscitation, the availability of ECMO for reversible problems, and the parent's/family's requests for continuing the efforts. In the past the lack of return of spontaneous circulation after two rounds of epinephrine was considered a poor prognostic sign for return of spontaneous circulation, but subsequent reports have been published concerning recovery after prolonged arrest. In particular the American Heart Association recommends prolonged efforts be made for infants and children with recurring or refractory VF/VT, drug toxicity, or a primary hypothermic insult. One potentially useful monitor during CPR is the end-tidal CO2. The determinants of $ETCO_2$ are alveolar ventilation, pulmonary perfusion (cardiac output), and $CO_2$ production. During acutely low cardiac output states such as cardiac arrest, decreased pulmonary blood flow is the primary determinant resulting in abrupt decrease in $ETCO_2$ [10]. Several studies in adults and children have confirmed that persistent low $ETCO_2$ (<10) is associated with an extremely low probability of return of spontaneous circulation. I would use this indicator to attempt to optimize resuscitation, and if it remained below 10 for more than 20–30 min, I would consider halting resuscitation efforts [11–13].

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## *Suggested Reading*

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