Pediatric Anesthesiology Review

Clinical Cases for Self-Assessment

Second Edition

Robert S. Holzman
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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 in our department since 1992 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 mimic the spontaneity achievable through conversation. First of all, a "buddy" system is advisable. Recording your answers is extremely useful when using the questions as prompts; the contemplative reader will listen critically to the responses he or she has offered into the recorder and then hopefully improve with time and practice. When all else fails, you can find the closest 4-year-old, who will gleefully ask you "why" after every response, uncannily similar to a board exam. 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 the patient her- or himself, 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 those diverse audiences.

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.

This second edition has the same purpose as the first – to accompany the reader's journey in attaining proficiency, expertise, and, finally, mastery in 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, and eventually cover the questions on the left, so that probing questions become self-generated. In this very simple, programmed text manner, practice at generating the appropriate breadth and depth of answers, and then questions, can be encouraged.

With this basic guidance, the reader is encouraged to be creative throughout this book, to use imagination as well as 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."

Boston, MA, USA

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Part I
Pediatric Medicine for Pediatric Anesthesiologists

Chapter 1
Newborn Medicine

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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. E. 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?
   A. D5 0.2 NS at 8 mL/h
   B. D10 0.45 NS at 10 mL/h
   C. D5 LR at 10 mL/h
   D. 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/h

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 02 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 NaHCO3, 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 normal perinatal history who has a brief seizure and who is now clinically stable should include:
    1. Measurement of electrolytes, Ca+2, 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, the 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 Sp0 >90 %. If this is not accomplished with an Fi0 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):
    A. Is primarily seen in prematures born between 30 and 34 weeks of gestation
    B. Can progress to chronic lung disease if untreated
    C. Resolves within 24–48 h
    D. 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:
    A. The presence of a shadow at the aortic knob on CXR
    B. The presence of diminished peripheral pulses due to excessive pulmonary blood flow
    C. The presence of pulsus paradoxus
    D. 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. Resolves within 24–48 h

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, tachypnea, and 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 (Qp/Qs). An infant with a ventricular septal defect with a Qp/Qs >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 neonates 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 PaO_2 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 sequleae 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, 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
    2. Because newborns have inadequate stores of vitamin K
    3. Because the newborn lacks sufficient bacterial flora to produce vitamin K
    4. 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 2
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 the rhinorrhea, 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:
   1. Necrosis of the respiratory epithelium.
   2. Edema of the submucosa.
   3. Destruction of cilia.
   4. 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:
   1. Ex-preterm newborns
   2. Infants with seizure disorders
   3. Children with congenital heart disease
   4. 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:
   1. Amoxicillin
   2. Ribavirin
   3. Racemic epinephrine
   4. Oxygen
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

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.

8. 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, and 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 FEV1 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 FEV1 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. Commonly 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 1:3,000 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 transmembrane conductance regulator (CFTR) that is expressed largely in epithelial cells of the airways, GI tract, sweat glands, and GU system.

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, 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 some time 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 super-infection 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 URI's in children:
    1. Mucociliary dysfunction can persist for weeks after recovery from the URI.
    2. Viral URI's 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 URI's.
        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/year decreases, with most adults reporting 1–3/URI's 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 URI's in children:
    1. The incidence is highest between the ages of 6–8 years.
    2. School-aged children normally experience one to two colds/year.
    3. Boys have more URI's than girls.
    4. Among children aged 1–4 years, those in day care have fewer URI's 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 3
Surgery

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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 gonorrhea
   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 4:1,000. 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 to 18 months. The incidence varies from 0.5–4/1,000 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 <1,000 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 1:2,500 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 1:2,500 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/or 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. Extra-intestinal 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 1:600–1,000 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. Extra-intestinal 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 1:2,500 live births. Cleft lip with cleft palate is more common that either is seen in isolation. The frequency is higher than 1:1,000 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.

Chapter 4
Hematology/Oncology

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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. 1, 3
      C. 2, 4
      D. 4 only
      E. 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 a 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.
Stage IV the malignancy is disseminated, involving greater than one 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:
    1. They tend to present with symptoms of raised intracranial pressure.
    2. The most common histology, cerebellar astrocytoma, also has the best prognosis.
    3. Morning headache with associated vomiting may be part of the presentation.
    4. 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 5
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 prominence 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
   E. Coarctation of the aorta (CoA)

4. 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 Qp:Qs >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 Qp:Qs 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 (VSD's):
   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 VSD's 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 ASD's:
   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 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 twenties 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. 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 arteriosCus

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 SpO_2?
    1. A Rashkind-Miller procedure
    2. Administration of milrinone
    3. Prostaglandin E1 administration
    4. Dilation of the ductus arteriosus in the catherization 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 (machine-like) 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 directly 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. PaO_2
    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 $Sp0_2$ 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 (Qp:Qs).

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 bi-directional 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 6
The Musculoskeletal System

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Questions

1. Cerebral palsy, a movement and posture disorder:
   1. Is seen in 1–2/1,000 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 encephalopathyis 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 30–70 % 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:
   1. Is slightly more common than hematogenous osteomyelitis in children
   2. Occurs more often in infants and young children
   3. May present in infancy with fever, poor feeding, and subtle asymmetry of soft tissue folds
   4. 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):
   1. Has an incidence of 1:1,000
   2. May be bilateral or unilateral
   3. May be treated conservatively until the second birthday
   4. 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?
   1. Defects in collagen formation
   2. Bones with thin cortices
   3. Deafness
   4. 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 sclerae, short stature, andthin 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 60–100/1,000,000
    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 7
General Pediatrics

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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
   2. Tonsillitis
   3. Impetigo
   4. 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 non-suppurative complications of group A beta-hemolytic infections?
   1. Rheumatic fever may develop after tonsillitis.
   2. Neither nephritis nor rheumatic fever develops after impetigo.
   3. Rheumatic fever is caused by the same strains of the organism as nephritis.
   4. 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
    4. Exercise
        A. 1, 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, 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. An apparent life-threatening event (ALTE):
    1. Would have previously been called a near-miss SIDS event
    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 ALTEs?
    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 non-immunized 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

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.

28. A. 1, 2, 3

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 2,000 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 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 both 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 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 1–3-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

1. Custer J, Rau R. The Harriet Lane Handbook by the Johns Hopkins Hospital. Philadelphia: Elsevier Health Science Division; 2008.
2. Kliegman R, Marcdante K, Jenson H, Behrman R. Nelson essentials of pediatrics. Philadelphia: Elsevier Saunders; 2006.
3. MacDonald M, Mullett M, Seshia M. Avery's neonatology: pathophysiology & management of the newborn. 6th ed. Philadelphia: Lippincott Williams and Wilkins; 2005.
4. McMillan J, DeAngelis C, Feigin R, Warshaw J. Oski's pediatrics: principles and practice. 3rd ed. Philadelphia: Lippincott Williams and Wilkins; 2006.

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 SpO_2 = 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: FiO_2 = 0.5; RR = 30/min; PEEP = 3 cm H_2O; PIP = 22 cm H_2O.

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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?

Preoperative Evaluation

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 work-up 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 2-D 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 O_2, 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 D_{20} solution? D_{10} 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?

Intraoperative Course

Answers

1. Direct arterial monitoring would be very helpful in this case, but technically difficult and associated with complications. Radial arterial cannulation should preferentially be done in the postductal left-sided location. 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 PaO_2 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 D_{10}, 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 D_{10}, 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 SpO_2 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 postextubation 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.

Postoperative Course

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, postextubation stridor or "croup" is a possibility, but not a certainty. Subglottic stenosis and tracheomalacia are complications of intubation to which newborns are subject to. 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 dam- age, 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

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 2,000 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?

administered to newborns provided there is appropriate 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.

Additional Topics

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 60 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 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:

$$P\left(dyn/cm^2\right) = \frac{2T\left(dyn/cm\right)}{R\left(cm\right)}$$

   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 [13–16]. 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 surgi- cal 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 un- less 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/.

Annotated References

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Chapter 9
Newborn Emergencies

Thomas J. Mancuso

A 14-hr-old male, 2,400 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 = 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?

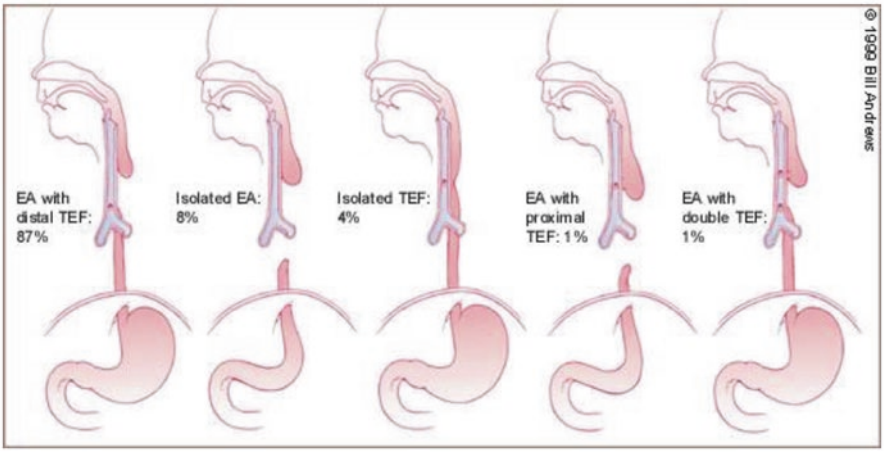
2. What other evaluations of the newborn are needed prior to undertaking the induction of anesthesia? What conditions are likely or associated with EA/TEF?

Preoperative Evaluation

Answers

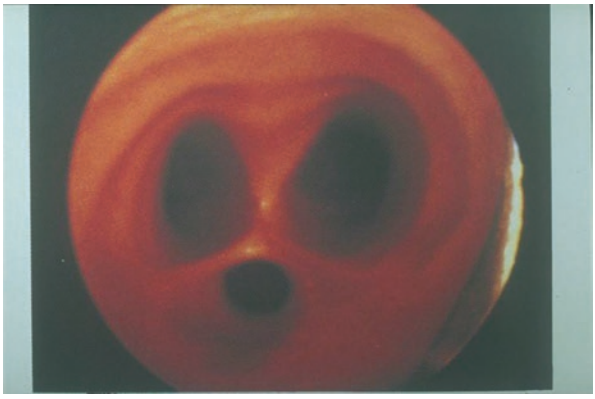
1. While repair of the esophageal atresia (EA) with tracheoesophageal fistula (TEF) may not be a true emergency, it is, at the least, very urgent. The longer the newborn is unrepaired, the greater the risk for aspiration. Surgical correction should proceed very quickly but proper preparation can be accomplished 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 [2, 3].



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 [4]. 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.

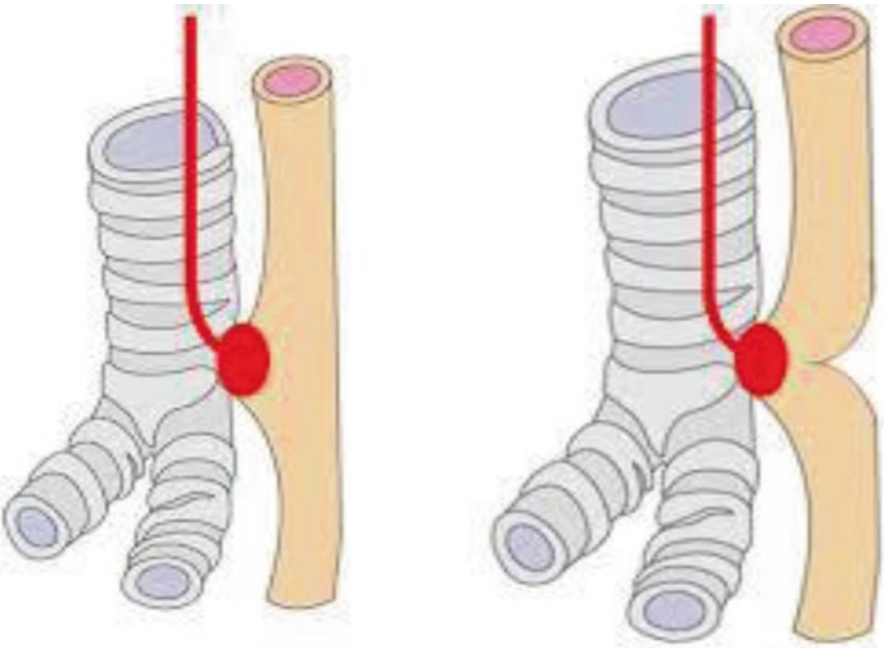


Intraoperative Course

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?



Intraoperative Course

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 if 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 [5]. 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 [6]. 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 [7].

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 [7].

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?

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?

Postoperative Course

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 [8] or via an ultrasound-guided right paravertebral catheter [9]. 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 [10]. If regional analgesia is not undertaken, parenteral opioids can be used to provide analgesia. In either case, cardiorespiratory monitoring must be done [11].

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

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 [12, 13]. 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

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?

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?

often patent ductus arteriosus (PDA) [14]. Poor prognosis is associated with birth weight <1,000 g, gestational age <33 weeks, and an A-a gradient >500 mmHg [15]. Placement of a nasogastric tube may help ventilation by decompressing the stomach. Mechanical ventilation should be done with the lowest possible airway pressures [16]. 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 [17].

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 [1].

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. 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

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?

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 [23]. Epidural catheters (caudal, lumbar, thoracic) have been used to provide analgesia for these procedures [24] and post-thoracotomy analgesia also can be provided with ultrasound-guided paravertebral catheters.

6. Meconium ileus results from obstruction of the distal small intestine by abnormal meconium [25]. 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

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 amnionic 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 Preparation

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?

Preoperative Preparation

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 you 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?

Intraoperative Care

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, Neo-Synephrine, or dopamine to keep blood pressure approximately within 20 % of baseline. When the fetus is exteriorized, intramuscular injection of muscle relaxant and fentanyl 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.

Postoperative Care

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?

Additional Questions

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" with 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.

Suggested Readings

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Chapter 11
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.

Preoperative Evaluation

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?

Intraoperative Course

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.

Postoperative Care

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 brain dead?

Additional Topics

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. Activity Best Response Score (changes pediatric patients)

   - Eye opening
     - Spontaneous 4
     - To verbal command 3 (young child: to shout)
     - To pain 2
     - None 1
   - Verbal
     - Oriented 5 (words, phrases, smiles/coos based on age)
     - Confused 4
     - Inappropriate words 3
     - Nonspecific sounds 2
     - None 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
  - Follows commands 6 (young child: spontaneous)
  - Localizes pain 5
  - Withdraws from pain 4
  - Flexion to pain 3
  - Extension to pain 2
  - None 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 [12]. 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 [13]. 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 12
Central Nervous System/Orthopedics

Robert S. Holzman

A 12-year-old, 50 kg girl with myelomeningocele and T10 paraplegia has a 90° thoracic curve and is scheduled for posterior spinal fusion. She has a functioning ventriculoperitoneal shunt and is allergic to penicillin, vancomycin, milk, and nuts. She self-catheterizes and is incontinent of feces. Her hematocrit is 32 %, and she has 6 units of designated donor blood available.

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Preoperative Evaluation

Questions

1. Of what anesthetic significance is scoliosis? What longer-term comorbidities should you expect by adolescence? What is the significance of the 90° curve? Why? What specific laboratory tests will you order for the respiratory system? Why? With what specific tests will you evaluate the cardiovascular system? Why? What other systems are you concerned about in this patient? Why?

2. Of what significance is the myelomeningocele for preoperative evaluation of the patient? Why? How would you evaluate the functioning of the ventriculoperitoneal shunt? Do you need a shunt series preoperatively?

Preoperative Evaluation

Answers

1. Scoliosis surgery involves prone positioning, extensive tissue trauma, considerable blood loss, and extensive bone dissection which 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° 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 require postoperative ventilatory support. Respiratory function can be assessed by pulmonary function tests (PFT) and a room air arterial blood gas (ABG). PFTs assess the mechanical function of the lungs and chest wall. It is effort-dependent and can be useful in a cooperative patient. ABGs assess the adequacy of gas exchange. I would use an ECG to evaluate rhythm abnormalities and conduction system abnormalities, particularly in patients with significant clinical impairment of the cardiorespiratory system. Long-standing severe scoliosis can increase pulmonary vascular resistance by several mechanisms: reduced pulmonary vasculature, 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. Other organ systems and issues to be concerned about in this patient include chronic urinary tract infection, obstruction, and possible renal insufficiency, patency of the VP shunt, and a Chiari type II malformation, often associated with abnormalities of ventilatory control [1].

2. The preoperative evaluation of a child with myelomeningocele should include an assessment of overall cognitive function, autonomic function (depending on the level of the spinal cord lesion), symptoms of VP shunt malfunction, control of seizures if present, extent of the neurological deficit, lower extremity contractures, and history of latex allergy, to which these patients are often susceptible. Contractures of the lower extremities and obesity due to non-ambulation require extra care to avoid pressure injury. Attention to the VP shunt includes the avoidance of compression and avoidance of the insertion of a central line on the ipsilateral side. Brain stem compression may occur by hyperextension of the cervical spine if a Chiari malformation is uncorrected. In general, there is a decreased

Intraoperative Course

Questions

1. How will you monitor this patient? Is an arterial line necessary? Why/why not? Where would you place a central venous line? Why? When would you consider a pulmonary artery catheter? 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?

requirement for neuromuscular blockade due to reduced muscle mass in the lower extremities and a reduced analgesic and anesthetic requirement for lower extremity and perineal surgery because of impaired sensation. Standard doses of opioids and other CNS depressants may cause excessive sedation and respiratory depression due to an unfavorable shifting of the carbon dioxide/minute ventilation response curve. Brain stem compression by the Chiari malformation produces a rightward shift of the carbon dioxide response curve as well. The patient does not need a sleep study unless there is a history of sleep apnea. Malfunctioning of the VP shunt produces symptoms and signs of increased intracranial pressure such as unexplained abdominal pain, nausea and vomiting, decline in school performance, decreased mental status, and persistent or morning headaches. A head and neck radiograph may be helpful to visualize shunt catheter disconnection and migration. CSF pressure can be measured through the ventricular reservoir. In the absence of an abnormal mental status or other findings of elevated intracranial pressure, a shunt series is of no significant benefit. An MRI with a contrast injection can resolve remaining questions about shunt function.

Intraoperative Course

Answers

1. The patient should be monitored with routine noninvasive monitors as well as an arterial line. A central venous line (peripherally inserted central catheter (PICC) or formal central line) may be placed for the appropriate clinical context (difficult vascular access with anticipated prolonged perioperative course, anticipated significant blood loss, e.g., >1 blood volume). The internal jugular or subclavian veins, using ultrasound guidance, are reasonable choices. Judgment should be exercised when considering this option in patients with a VA shunt. In addition, a central line placed in the superior vena cava close to the right atrial junction may impede cerebral venous drainage and be a conduit for potential contamination. A pulmonary artery catheter should be considered if there is preoperative evidence of severe pulmonary hypertension and/or ventricular dysfunction in order to monitor ventricular filling pressure and guide fluid and inotropic therapy. $ETCO_2$ sampling may not be accurate because of severe ventilation-perfusion mismatch and increased respiratory dead space. It is not likely that somatosensory evoked potentials would be useful for this patient if sensory impairment is severe, and even in the presence of some sensation and motor function, baselines will often not be normal and changes from baseline may be difficult to interpret. Anesthetic technique choices are affected if EP monitoring is chosen because the dose of inhalation anesthetic has to be tailored to minimizing its effect on amplitude and latency of signals on SSEPs and neuromuscular blockade must be avoided in order to allow monitoring of MEPs.

2. Having had numerous prior procedures, the patient is terrified of needles and requests a mask induction. Your response? Why? How do you justify it to the patient?

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

4. During surgery the patient becomes progressively difficult to ventilate, with PIP increasing from 25 to 45 cm. H_2O, blood pressure falling from 100/64 to 64/34, oxygen saturation decreasing to 92 % from 99 %, and wheezing noted. You also note hives on the forearms. Your considerations? Why? What is your differential diagnosis? Specific therapeutic interventions? Why? Would diphenhydramine help? Cimetidine? Why/why not?

5. The patient loses 750 mL. of blood during the first 1.5 h of the procedure. At what point would you begin transfusion? Why? Even with autologous blood? Why/why not? What is the role of hetastarch in this case? Is there a disadvantage to using it? Why? What other efforts can you direct to blood conservation?

2. A mask induction is fine provided that venous access can be readily established and the patient does not have significant gastroesophageal reflux. These patients often have difficult IV access. 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. If reflux is severe, the potential for aspiration pneumonitis may override the 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.

3. A balanced anesthetic consists of a potent opioid such as fentanyl or sufentanil supplemented with a low dose of volatile agent. The presence of a Chiari II malformation may compress the brain stem with its abnormal nuclei and shift the CO_2 response curve adversely. While the patient can be sensitive to opioid-induced respiratory depression, she will also be sensitive to respiratory depression from the volatile agent. A low-dose volatile agent (not > ET of 0.5 % MAC) will reliably produce unconsciousness, while the potent opioid will provide additional analgesia.

4. These are manifestations of an allergic reaction, one possibility of which is latex anaphylaxis. Other possible causes are a drug- or transfusion-induced anaphylactic or anaphylactoid reaction. The clinical manifestations are consistent with type I immediate hypersensitivity, and patients with myelomeningocele are at high risk for latex allergy. In addition to an allergic reaction, the differential would include possible pneumothorax, occlusion of the endotracheal tube with secretions and kinking, or endobronchial intubation. Specific interventions would be to notify the surgeon, stop the volatile agents and provide 100 % oxygen, stop the antibiotic infusion, administer IV epinephrine (10–20 mcg incremental boluses, to effect) along with intravascular volume support if needed, call for help, and exclude pneumothorax and other mechanical causes. Diphenhydramine may be given, but not as the first line of treatment.

5. The loss of 750 mL of blood represents 20 % of the circulating blood volume at a rate of 500 mL/h, and this will decrease the Hct from 32 % to 25 %. If blood loss continues at this rate, it will significantly deplete the patient's oxygen carrying capacity and expose the patient to hypoxemia. 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 1 L 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. Autologous blood donation is probably the most effective preoperative strategy.

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 air flow in the pharynx, and the patient phonates. Your considerations? What therapeutic interventions will you make? Why?

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. As you wake her, she mumbles about the "worst headache she's ever had." Your considerations? Why? How would you evaluate the patient? How would you intervene in this patient, specifically?

Additional Topics

Questions

1. A 9-year-old girl with a history of Angelman syndrome is scheduled for bilateral varus derotational osteotomies, acetabuloplasty, gastrocnemius lengthenings, hip arthrogram, and left hip open reduction and adductor lengthening. Additional medical history includes seizures, hypotonia, and severe global developmental delay. She is on Zarontin (ethosuximide) and valproic acid for seizure control and has been seizure-free for the past 3 years. She has swallowing difficulties. She is known to have difficult intravenous access; does she need an IV for a rapid sequence induction? Would you use succinylcholine or avoid it? How can she have spasticity as well as generalized hypotonia? What would be your choice of regional analgesic technique for postoperative pain management? Would she be better off with a neuraxial or peripheral nerve block technique? What are the advantages and disadvantages of each?

Postoperative Course

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. The patient needs to be checked for increased intracranial pressure due to VP shunt malfunction resulting from kinking or disconnection. The differential diagnoses are opioid-induced headache and hypercarbia, hypotension, cerebral hypoperfusion due to hypotension, low Hct, and intraoperative position-related compression of the brain stem. Increased ICP can result from obstruction of cerebral venous drainage due to intraoperative internal jugular vein compression from the prone position with excessive neck flexion. Interventions would include 100 % oxygen and assisted ventilation via mask. If the patient remains sedated and bradycardic and the headache persists or progresses, then reintubation and hyperventilation should be considered. If the condition does not improve, consider measuring VP shunt reservoir pressure and tapping the shunt. If no improvement follows, then consider emergency MRI to assess cerebral edema and/or compression of the posterior fossa contents.

Additional Topics

Answers

1. Angelman syndrome is a genetic disorder with neurological manifestations of varying degrees of developmental delay, spasticity, seizures, and sleep disturbance characterized by relatively short episodes of sleep. Early in its clinical description, it was characterized as the "happy puppet" syndrome because of characteristically happy-appearing patients who laugh and smile frequently [2]. Their receptive competence is often better than their expressive competence, although still typically quite delayed. With regard specifically to the musculoskeletal system, they are typically ataxic with motor overactivity and are subject to chronic adverse orthopedic effects such as abnormal skeletal stresses resulting in bone, joint, and soft tissue disorders. The swallowing disorder is of some concern; it is important to find out if it is associated with aspiration and whether periods of NPO are satisfactory in minimizing this risk. If that is the case, then a

2. 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?

3. A 12-year-old with Duchenne muscular dystrophy who is wheelchair bound with progressive scoliosis, requires spine fusion. Why? What is the significance of "wheelchair bound at 12-year-old"? 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?

routine NPO period should suffice for a mask induction and subsequent IV placement, which will probably be much more successful. Uncontrolled gastroesophageal reflux disease would warrant a rapid sequence intravenous induction, which could be accomplished with succinylcholine or a priming-dose method with a nondepolarizing muscle relaxant. Because there are GABA-receptor abnormalities associated with the deletions of portions of chromosome 15 that give rise to Angelman syndrome, benzodiazepine premedications should probably be avoided because of the unpredictability of the patients' responses. Regional techniques certainly are helpful in providing perioperative analgesia, assuming that compromises in sensation will not disturb the patient more. Neuraxial techniques such as a lumbar epidural would address all of the surgically treated area, but have the disadvantage of less mobility in the perioperative period and bladder catheterization postoperatively.

2. Dystrophic as well as nondystrophic scoliosis can occur with NF1 [3–5]. 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 be of varying severity. 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 operations or one. 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.

3. Neuromuscular scoliosis as a result of muscular dystrophy is a result of progressive muscle incompetence and resulting weakness, resulting in axial imbalance and unbalanced mechanical forces on the spine [6, 7]. Being wheelchair bound at 12 years old, a relatively common finding in DMD, suggests that his disease 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; 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-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

4. A 13-year-old girl with Charcot-Marie-Tooth disease and progressive pain and difficulty walking is scheduled for multiple foot osteotomies and tendon releases. She has decreased sensation and muscle mass in her lower extremities as a result of her underlying disease and some generalized weakness, but is otherwise doing well in school. What are your anesthetic management concerns in this patient with progressive neuropathy and muscle loss? What if she requests a regional anesthetic? Do you think there is a risk of exacerbation of her neuropathy? If she had reflux, is a rapid sequence induction of general anesthesia with succinylcholine indicated?

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 [8]. 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 [9]. 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.

4. Patients with Charcot-Marie-Tooth disease (CMTD) have a progressive loss of sensation and muscle mass, particularly in distal extremities, and often require surgical reconstruction for extreme pes planus deformities of their feet. Older techniques included multiple joint fusions, but currently, osteotomies and tendon transfers are the preferred repair. The disease has a broad spectrum from mild sensory-motor impairment (which is not necessarily related to the severity of foot deformity and functional impairment) to significant loss of extremity sensation and motor function. In severe cases it may result in impaired respiratory function, aspiration, and vocal cord paresis [10]. Regional anesthetic techniques have been favorably reported notwithstanding the progressive peripheral neuropathy and the theoretical risk of exacerbation. Regional techniques are particularly helpful for perioperative analgesia for the first several days. For patients with comorbidities such as dysphagia, vocal cord paresis, or reflux, rapidly securing the airway should be considered, and succinylcholine has been used without adverse effect in patients who are active. Alternatively, nondepolarizing agents can be used in increased doses or in accordance with the priming principle for more rapid onset.

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Chapter 13
Orthopedics: Scoliosis

Joseph P. Cravero

A 16-year-old female with idiopathic scoliosis is coming for instrumentation and posterior spinal fusion. She has a history of asthma. Weight 60 kg. *P* = 92 bpm, BP = 108/62 mmHg, RR = 20/min, temperature = 36.7 °C.

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Preoperative Evaluation

Questions

1. When does idiopathic scoliosis begin, which sex is more likely affected, and at what age is it generally diagnosed? How does this differ from neuromuscular disease-related scoliosis?

2. Her curve is 75°. Is this cause for concern? How would you assess her pulmonary function? Do you need pulmonary function tests? What type of pulmonary dysfunction is most common with this level of scoliosis? Does it matter if her scoliosis is due to neuromuscular disease or is idiopathic?

3. Do you need more information on her cardiac status? What type of cardiac disease would she be at risk for, and how would you diagnose the extent of her cardiovascular compromise (if any)? How would your considerations differ in a patient with non-idiopathic scoliosis such as that associated with Duchene muscular dystrophy?

Preoperative Evaluation

Answers

1. Scoliosis is a complicated pathological problem that involves lateral curvature of the spine. Idiopathic adolescent scoliosis (AIS) is the most common form of scoliosis and is found in 1–3 % of children/adolescents between 10 and 16 years of age. This accounts for 70 % of all cases of scoliosis. Females are affected 3.6 times as often as males. Neuromuscular disease-related scoliosis can have many causes including neuropathic disease (cerebral palsy, syringomyelia), myopathic disease (muscular dystrophy, amyotonia congenita), neurofibromatosis, mesenchymal disorders (Marfan's syndrome, Morquio disease, Still disease), or trauma. Although variable in nature, neuromuscular scoliosis often begins at an earlier age and progresses faster. It is ultimately more likely to require surgical correction.

2. Yes, there is cause for concern because curvatures of 65° or greater can cause significant restriction of ventilation. The need for pulmonary function testing would depend on her exercise tolerance. If she has excellent exercise tolerance, I would not pursue testing. As scoliosis becomes severe, the rotation of the vertebrae causes the ribs to form a rib hump and restrict the thoracic cage. Vital capacity, forced expiratory volume in 1 s (FEV), and PaO are all decreased. The FEV-to-FVC ratio is largely unchanged. A preoperative vital capacity of less than 35 % is considered a significant predictor of respiratory compromise in the perioperative time frame. Lung function should be optimized prior to surgery. Any lower respiratory tract symptoms or signs (rales, rhonchi, wheezing) should prompt a thorough evaluation and postponement of surgery. In these cases 4–6 weeks should pass before administering general anesthesia for correction.

3. Patients with severe scoliosis are at risk for chronic hypoxia, pulmonary hypertension, and cor pulmonale. Hypoxic pulmonary vasoconstriction is an ongoing risk in patients with chronic hypoxia. Right ventricular hypertrophy and (eventually) cardiomyopathy can result. Any patient with known or suspected cardiac disease should have a preoperative cardiac evaluation. An ECG to evaluate for ischemia or axis deviation is indicated as is an echocardiogram to determine overall function and structural anomalies. Compromised patients will require intraoperative cardiac monitoring with consideration for transesophageal echocardiography during the operative case. A central line for trending of central venous pressures and administration of vasoactive medications would also be indicated. Patients with Duchene muscular dystrophy often have a dilated cardiomyopathy with compromised left ventricular function. In these cases, the cardiac compromise would be expected to be more marked than that with idiopathic scoliosis and accompanying cardiac dysfunction.

4. Her asthma is only a problem when she has a cold. She is maintained on beclomethasone (Qvar®) inhaler bid. She uses an albuterol inhaler when she has active wheezing. Do inhaled steroids require perioperative coverage with "stress steroids?" She was on oral steroids 2 months ago for 2 weeks. Will she need perioperative steroids? How will you prepare her for the operating room?

5. Should she receive erythropoietin preoperatively? What are the relative advantages/disadvantages to autologous donation of blood vs. acute isovolemic hemodilution? What other methods are available to avoid blood transfusion?

6. She is needle phobic; she is crying and keeps pulling her hand away as you try to look for an IV. Should you premedicate – with what medications? Would you use nitrous oxide to help start the IV?

4. Inhaled corticosteroids such as beclomethasone, budesonide, fluticasone, and triamcinolone are the cornerstones of therapy for persistent asthma. While the use of these drugs can result in undeniable improvement in overall symptoms, inhaled corticosteroids are suppressive rather than curative. No clinically important adrenal suppression has been shown to occur with the administration of these medications in their recommended doses. Patients who have been taking systemic corticosteroids for more than 2 weeks in the prior 6 months are considered at risk for adrenal suppression in the setting of major surgery. In this case she would need perioperative systemic steroids. If she were recently on systemic steroids, I would obtain a pulmonary consult to check her current pulmonary function and her responsiveness to bronchodilators. I would prepare her by placing her on oral steroids for 4–5 days preoperatively and continuing for the week after surgery. I would also have her take her bronchodilator therapy during the perioperative period.

5. The use of perioperative erythropoietin therapy has been shown to increase hematocrit levels before and after scoliosis surgery. On the other hand studies have not shown that the administration of the drug decreases the exposure to transfused blood for patients undergoing scoliosis surgery. I would not administer the medication to this patient, but it could be considered in specific cases where preoperative anemia is a major concern. Autologous donation can be performed if the patient is over 50 kg and she is not anemic. The blood can be harvested from the patient and stored for as much as 6 weeks. Donation can occur twice weekly and can be done up to 72 h prior to surgery. There is some evidence that this practice can decrease the exposure to allogenic blood. It is critical to consider the extent of the surgery and the overall likelihood of transfusion. If the correction is going to be large and transfusion is a certainty, then I would offer autologous donation as an option for the patient. Acute normovolemic hemodilution (ANH) is a technique in which blood is removed from the patient around the time of induction of anesthesia. Circulating volume is maintained with colloid or crystalloid. As opposed to autologous transfusion, there is less chance of clerical error and administration of the incorrect blood – since it does not leave the operating room. In addition, the blood that is returned to the patient is fresh and contains all of the factors, 2,3 DPG, and normal electrolyte components of the patient's blood.

6. If the patient is severely anxious, I will offer oral midazolam as a premedication, 0.5–1 mg/kg with a maximum dose of about 15 mg. Yes, if the patient would tolerate a mask on her face, the use of 70 % nitrous oxide in oxygen is an alternative to facilitate intravenous access.

Intraoperative Course

Questions

1. What are the surgical approaches to scoliosis repair and how would they change your anesthetic management?

2. What monitors do you need? Is an arterial line needed? Is CVP monitoring important? Would a TEE be indicated? What are the advantages and disadvantages? Does the underlying cause of the scoliosis change your opinion on this matter?

3. What are somatosensory evoked potentials (SSEPs)? What are motor evoked potentials (MEPs)? What do each they measure? Are both needed? If they are normal, does this assure normal neurological function postoperatively? What does a wake-up test measure? Is it necessary if SSEPs and MEPs are both normal during the course of a correction?

Intraoperative Course

Answers

1. Surgical correction of scoliosis involves the placement of screws and rods on the lamina of the vertebrae to segmentally straighten the spine. After the adjustment and tightening, the vertebral bodies are roughened in preparation for the placement of bone graft with is packed into the intervertebral spaces. The correction can be done exclusively from the posterior approach or from an anterior approach – or an anterior release combined with a posterior fixation. For the anterior approach, the patient is supine, and, depending on the level of the defect, the abdomen or thorax may be entered, generally from the lateral aspect. For the posterior approach the patient must be prone with particular attention to pressure areas since the surgery time is generally prolonged.

2. In addition to standard monitors of ECG, blood pressure, end-tidal carbon dioxide, and percutaneous pulse oximetry, I will use intra-arterial blood pressure monitoring. The arterial line will provide ready access for blood samples during the case, and it provides an accurate measure of blood pressure on a beat-to-beat basis. The arterial pressure tracing can also be used to estimate the intravascular volume by evaluating the difference in systolic blood pressure between inspiration and expiration. Finally the arterial line could be used with a minimally invasive cardiac output monitor to further delineate hemodynamic responses to anesthesia agents and blood loss. I don't expect to use a CVP monitor in most cases. CVP will not offer additional information beyond the monitors mentioned above and is not without potential risks of air embolism, hemopericardium, pneumothorax, and arrhythmias. If the patient has a significant cardiomyopathy, then I would use the CVP to deliver vasoactive medications for support and control of blood pressure. A TEE would be helpful in cases associated with cardiomyopathy (muscular dystrophies) since it can give an excellent estimate of cardiac filling and wall motion abnormalities associated with ischemia. The probe may be difficult to insert and/or maintain in the prone position in a posterior spinal fusion, so significant advanced planning must take place.

3. SSEP monitoring measures electrical activity in ascending sensory pathways. Intraoperative SSEP monitors the integrity of the sensorineural pathways from the site of sensory neuron stimulation (caudad to the surgical site) to a site cephalad to the surgical site such as the brainstem or the prefrontal sensory cortex. The principal goal of intraoperative monitoring is the identification of nervous system impairment at the operative site in the hope that prompt intervention will prevent permanent deficits. The surgeon is alerted to the possible damage, and corrective action is taken to prevent the damage. SSEPs measure evoked electrical wave configuration, peak-to-peak intervals, absolute and inter-peak latencies, and comparative latency delays between the ipsilateral and contralateral pathways. They

4. What agents will you use for induction and maintenance of anesthesia? Are some induction and maintenance agents more compatible with SSEP and MEP monitoring? Does it matter? What muscle relaxant will you choose? How will you place and secure the ETT? What will you choose for maintaining the anesthetic? Will this choice affect the SSEP monitoring? Do you need a baseline reading of SSEP or MEP before starting surgery? How will you assure amnesia? Is a BIS monitor helpful? What is the utility of antithrombolytics in spinal fusion surgery?

measure the proximal neural electrical responses (brachial plexus, spinal cord, brainstem, or cerebral cortex) to peripherally applied standard sensory stimuli (such as median or posterior tibial nerves), thereby testing the integrity sensory pathways' function. No, because of the potential for false-positive and false-negative results. No, because wake-up test evaluates the gross motor function. Yes, motor evoked potentials (MEPs). MEPs are performed by electrical stimulation of the motor neurons directly or indirectly by trans-osseous stimulation. MEPs are more specific test of descending motor pathways but are not more specific or sensitive than SSEPs in detecting spinal cord injury during surgery. The use of both MEP and SSEP monitors during spinal surgery provides the optimal sensitivity and specificity in detecting neurologic impairment of spinal cord during surgery. The wake-up test is a specific test for gross motor function of the spinal cord but has its own limitations. The wake-up test is warranted and a reliable test if MEPs are absent or SSEPs are abnormal. If simultaneous recordings of MEPs and SSEPs are normal during spine surgery, they are of sufficient sensitivity and specificity to negate the need for a wake-up test.

4. Planning for anesthesia must start with the appreciation that many anesthetics can interfere with the signals used to test SSEPs and MEPs. All of the inhaled anesthetics have significant effects on the amplitude timing of these potentials. Barbiturates, benzodiazepines, and propofol have less profound inhibition of these signals. Etomidate, ketamine, and opioids are not associated with significant diminution of the potentials. With this in mind, for induction of anesthesia, I would choose propofol and fentanyl. If MEP monitoring were planned, I would avoid muscle relaxants (as they obliterate this signal) and provide local anesthesia for the airway. If absolutely needed, I would use a relatively small dose of an intermediate-acting muscle relaxant such as rocuronium or cisatracurium with the idea that they would diminish in effect during the preparatory phase of the procedure and be gone by the time the procedure was actually starting so MEPs could be used. Induction with propofol has a slightly greater effect on SSEP readings than etomidate or ketamine, but this is usually not a reason to avoid the drug for induction as its effect would be diminished by the time testing of nerve conduction was started. Placement of an endotracheal tube via the nasal route will ensure that the tube can be more firmly secured against accidental dislodgement and less likely to kink than via the oral route in patients positioned prone during surgery. For maintenance, a low-dose, balanced technique including N_2O 50 %, inhalation agent at less than 0.25–0.5 MAC, and a combination of continuous infusion of propofol and infusion of a short-acting opioid such as remifentanil or fentanyl can be used. I would choose these because these agents in the above-described doses have minimal effect on suppressing both motor and sensory evoked potentials. It is critically important to obtain a reliable baseline recording after the induction of induction and prior to surgical manipulation. The BIS monitor uses a proprietary algorithm to interpret a limited number of EEG leads and derive a measure of depth of anesthesia. The monitor reading correlates with other measures of anesthesia depth but has not been proven to prevent awareness. It may be very useful

5. After labetalol is given to control hypotension, you notice that the peak inspiratory pressure, which had been 24 cm H_2O, has risen to 52 cm H_2O. Tidal volume (exhaled) is decreased to 200 mL from 450 cc. Oxygen saturation falls to 92 %. How do you determine and correct the problem?

6. You note copious bleeding. A hematocrit sample comes back 28 %. Does the patient need to be transfused? How do you determine when a transfusion would be appropriate? You are using a cell saver. What efficiency of blood salvage can you expect to obtain with this device? What are you giving back to the patient when you transfuse "cell saver blood?" What is the hematocrit on "cell saver blood?" When is DDAVP indicated and how does it work?

Postoperative Course

Questions

1. What are your criteria for extubation?

in avoiding "overshooting" the depth of anesthesia. While many anesthesiologists utilize the BIS for scoliosis repair surgery, use is not universal. The BIS monitor may be most helpful in cases where muscle relaxant is being used (since patient movement cannot be used to detect very light anesthesia) or patients who have physiology that prevents the use of significant amounts of anesthesia during the case. In most cases, attention to patient movement and vital signs can alert the anesthesiologist to light anesthesia levels that are associated with awareness. Antifibrinolytics such as tranexamic acid (TXA) and epsilon-aminocaproic acid (EACA) have been shown to decrease blood loss in posterior spinal fusion surgery. Their use is recommended during these surgeries.

5. The beta-blocker effect of labetalol may have caused bronchospasm. Other causes may include pneumothorax (asymmetry of breath sounds and shift of precordial apical pulse), blood transfusion reaction (hives, rashes, fever, darkening of urine, and direct and indirect Coombs test), endobronchial intubation (asymmetric breath sounds), obstruction of the endotracheal tube by secretions, or kinking of the endotracheal tube. If the head position correction fails to rectify the kinking of the endotracheal tube, the tube must be replaced.

6. I will transfuse when the Hct is between 20 and 25 %. The exact point of transfusion will depend on the rapidity of blood loss and the stability of the patient (cardiovascular parameters) as well as her blood gas (the presence or absence of acidosis). The amount of blood that can be salvaged with the use of the cell saver device ranges from 50 to 70 % of the shed blood. I would transfuse packed red blood cells unless there is autologous whole blood available. The hematocrit of the re-transfused shed blood ranges from 50 to 70 %. DDAVP is desmopressin, a synthetic analog of natural arginine-vasopressin. The difference in chemical structure enhances the pressor to antidiuretic potency (2,000–4,000:1), and prolongs the duration of action of the compound due to its resistance to enzymatic cleavage. As with arginine-vasopressin, desmopressin stimulates the endothelial cells to release factor VIII, prostaglandin I2, and tissue plasminogen activator but is more potent than arginine-vasopressin. It may also increase platelet adhesiveness.

Postoperative Course

Answers

1. The decision to extubate patient's trachea will depend on the following criteria: the patient is awake, is spontaneously breathing, is comfortable, and follows command, full recovery of muscle twitch response to nerve stimulation in the train-of-four and sustained tetanus at 50 Hz for 5 s or sustained double burst stimulation prior to administration of cholinesterase antagonists, ETCO <50 mmHg, head lift for >5 s, and the ability to generate maximum inspiratory pressure of greater than −25 mmHg.

2. How would you determine the postoperative disposition for this patient?

3. What would be your plan for postoperative analgesia?

Additional Topics

Questions

1. A patient with cerebral palsy (CP) needs a posterior spinal fusion. What are the anesthetic considerations for a patient with cerebral palsy and spastic quadriplegia? Can you use succinylcholine for rapid sequence induction?

2. A patient with Duchene muscular dystrophy needs an anterior/posterior (AP) spinal fusion. What is Duchene muscular dystrophy and how will it affect your anesthetic plans? Is this patient at risk for malignant hyperthermia?

2. This patient does not need the ICU if she maintains adequate spontaneous gas exchange, has stable hemodynamics, and produces adequate urine output. Moreover, she has other risk factors. Major concerns about postoperative care include moderate restrictive chest wall disorder due to a greater than 60° curvature, respiratory depression from large amounts of postoperative opioid requirements, the potential for flare-up of asthma, and low postoperative Hct – which can compromise tissue oxygenation

3. I would plan on multimodal analgesia for the postoperative period. This should include acetaminophen, gabapentin, and opioids (delivered by patient-controlled analgesia pump). If the surgeon agreed to allow NSAID use, I would add ketorolac. Muscle relaxation with benzodiazepines such as Valium is useful in treating the discomfort from muscle spasm.

Additional Topics

Answers

1. CP is a term that is applied to patients with static encephalopathy that is present at birth. Causes range from perinatal asphyxia to intrauterine infection or kernicterus. The signs and symptoms of the disorder become more obvious as the patient ages. The findings are varied and can range from minor spasticity (in one or two limbs) with normal intelligence to patients with severe spastic quadriplegia and limited intellectual development. There are many anesthetic implications of this disorder. Because of their spasticity, positioning patients with CP can be difficult. Simple supine or prone positioning may require extensive planning and padding. Gastroesophageal reflux disease (GERD) is a common problem and must be considered when planning induction. Seizures are a common problem. Many of these patients will be on multiple anticonvulsants as well as the antispasmodic baclofen. Spasticity in these patients can be severe and often leads to extensive scoliosis. This fact, together with the small size of many of these patients, leads to the need for careful attention to blood conservation and replacement as well as dilutional coagulation abnormalities. Vascular access can be very challenging. Postoperative pain control will depend on attention to behaviors that (sometimes) only close family members recognize as indicative of pain. Close communication and appreciation of these difficulties are critical for perioperative care. CP is not a denervation problem; thus these patients tolerate succinylcholine without difficulty.

2. Duchene muscular dystrophy is a hereditary muscle disease associated with skeletal muscle weakness and abnormal function of smooth muscles including cardiac muscles. It is due to defects in the dystrophin gene, on the X chromosome.

The dystrophin defect is inherited as X-linked recessive diseases. Therefore, only males are affected and female relatives of affected males may be carriers. The anesthetic plan must take into account that the defective skeletal muscles are at risk for breakdown (rhabdomyolysis) from succinylcholine and inhalation agents. Furthermore the defective heart muscles result in dilated and occasionally restrictive cardiomyopathy. Some patients may have RBBB and other rhythm defects. Later in adolescence, degeneration of the tongue and replacement with fatty infiltrate may pose difficult to direct laryngoscopy and glottis visualization. Also in adolescence, muscle weakness is moderate to severe, and patients usually require postoperative ventilatory control in the ICU. Scoliosis tends to be very severe in this patient population. Because of the extent of posterior spinal fusion surgery, these patients tend to lose a substantial amount of blood – ranging from 50 to 200 % of the circulating blood volume. While the patients are at risk for rhabdomyolysis with succinylcholine or inhaled agent exposure, DMD patients are not specifically at risk for MHS at a rate that significantly exceeds that of the general population.

Suggested Readings

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Chapter 14
Otolaryngology

Robert S. Holzman

A 10 kg, 2.5-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? 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?

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?

Preoperative Evaluation

Answers

1. In contrast to the popular association of hypoventilation and airway obstruction with the Pickwickian 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. For that reason, 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 reside 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.

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 judgement of the anesthesiologist with regard to the effects of immediate preoperative stress on the cardiopulmonary system, and the 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.

Intraoperative Management

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? Mom insists on being present for induction – is this a good idea? As the patient enters stage II, he develops "seesaw" 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 s it goes to 88 %. What is 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 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?

Intraoperative Management

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 $ETCO_2$ 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 preexisting IV. That is not very likely, as this child is probably coming in from home. That said, 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, particularly with the diagnosis of airway obstruction. 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, not really "sleep."

   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 as well. 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. 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 wake-ups are less problematic. Do you agree? Would you choose that strategy? Why?

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

Perioperative Care

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?

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.

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.

Perioperative Care

Answers

1. Impaired oxygenation in the immediate post-op period can be for 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 nonrebreathing 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.

Additional Questions

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?

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?

Additional Questions

Answers

1. The CHARGE syndrome is an acronym that stands for colobomas of the eye, heart disease, atresia of the choanae, retarded growth, genital anomalies, and ear 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.

   Surgical correction at this point has progressed to transnasal endoscopically guided membranous and bony resection in the majority of case, although transpalatal approaches are still utilized. The surgical goal is to create choanal patency, preserve mucosal integrity, and minimize the chance of restenosis. If stenting is utilized, then 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. 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 fiber-optic intubation? How is this situation different than a Ludwig's angina patient?

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 in. beyond her mouth, and the distal 3 in. 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 preoxygenate? What are your postoperative considerations?

3. This is a very typical presentation in an older pediatric patient with a sore throat, difficulty swallowing, often sick for a few days, and occasionally, voice changes 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 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 fiber-optic 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 fiber-optic 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. Differentiate epiglottitis, croup, and bacterial tracheitis. What difference does it make to your anesthetic management to consider these as different entities?

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?

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 appearance 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 laying 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–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 need to be supported with endotracheal intubation and perioperative intensive care. It is important to differentiate the disorders because airway support is often necessary for epiglottitis and bacterial tracheitis, while airway instrumentation and endotracheal intubation should usually be avoided for croup.

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 endotracheal intubation. 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. 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.

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 15
Head and Neck

Robert S. Holzman

An 8-year-old girl with Klippel-Feil syndrome, scoliosis, a solitary kidney, neurogenic bladder, sacral agenesis, and tethered cord was scheduled to undergo cervical spine fusion following 1 month in halo traction. She was on no medication and developmentally was apparently doing well. She had a known difficult airway from multiple prior surgeries.

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Preoperative Evaluation

Questions

1. Will this patient have a "difficult airway?" What are the contributing reasons that you would be concerned about? Why has she been in traction? Is it likely that she has to remain in traction, or can this be released for induction? Should the halo device be removed, or should it remain? Are you surprised by the patient's comorbidities? What developmental explanation can you offer?

2. Does this patient require further evaluation or consultation, and if so, with whom? Any other studies you would like?

Preoperative Evaluation

Answers

1. This patient is highly likely to have a difficult airway for several reasons. First of all, she has Klippel-Feil syndrome as a result of fusion of several cervical vertebrae. Klippel-Feil syndrome severity is typically classified as type I, when patients have a single-level fusion; type II, when patients have multiple, noncontiguous fused elements; and type III, with multiple, contiguous fused segments. It may also be associated with branchial arch anomalies like Goldenhar syndrome, fetal alcohol syndrome, and anomalies of the extremities. Preoperative traction has been known to relieve sensorimotor impairment and improve the quality of the fusion repair. Depending on the type of halo fixation device, it may very well be released or portions released in order to get to the face and the patient's head held carefully in traction by the spine surgeon. The halo in all likelihood will be replaced by Gardner-Wells tongs or a similar device for the procedure. The comorbidities of renal anomalies often occur with Klippel-Feil syndrome; hearing impairment is not uncommon as well. Sacral agenesis and a tethered cord are not typically associated, even though they represent neural crest migration defects.

2. It would be advisable to consult with the ORL service in case their help is needed for securing the airway or for emergency tracheotomy. Soft tissue imaging studies, such as a soft tissue x-ray of the neck, would help define the anatomic relationships, especially the relationship of the posterior pharyngeal wall, which is likely to have moved anteriorly, thus narrowing the pharyngeal cross-sectional diameter and the larynx. This is also an important consideration for the patient's airway status postoperatively.

Intraoperative Course

Questions

1. What monitors will you choose? Why? Does this patient need an arterial line? Why? Does this procedure require any other special monitors? Would a precordial Doppler be a reasonable choice? Why or why not?

2. What are your considerations for anesthetic induction? Your colleague stops by and suggests an awake intubation? What do you think? You select an intramuscular preinduction technique in the pre-op holding area with ketamine because of the extreme separation anxiety, and the patient obstructs within 30 s in the mother's arms and begins to turn blue. What do you do next? Will an oral airway help? Is this patient a difficult intubation? Should you continue with the case?

Intraoperative Course

Answers

1. Routine noninvasive monitoring plus an arterial line to follow mean arterial pressure. In addition, there is a chance of air embolism because as bone is decorticated, the potential point of air entry is superior to the venous system and the right atrium of the heart. The Doppler should be placed in the typical position on the anterior chest wall, but should not be allowed to compress the skin with undue pressure.

2. Depending on the airway assessment and the patient, anesthetic induction might precede intubation of the trachea or follow intubation. An "awake" (actually, sedated, with topical and/ or local anesthetic) can be accomplished in almost any age patient with adequate planning and time. Topical lidocaine can be nebulized or gargled, specific nerve blocks for the glossopharyngeal and superior laryngeal nerves can be administered, a transtracheal injection of lidocaine can be delivered, and intravenous sedation can supplement the entire procedure. At that point, either a direct laryngoscopy, a video laryngoscopy, or a fiber-optic intubation (transnasal or transoral) can be accomplished. The patient can also undergo an inhalation induction with preservation of spontaneous breathing. Neuromuscular blockade can also be utilized once the ability to ventilate by mask is ensured. All of these possibilities are ultimately determined by the anticipated difficulty of the airway as well as the maturity, cooperation, and willingness of the patient and family.

   Sedation or preinduction strategies in the pre-op area have to be judiciously weighed against the risks of proceeding to the operating room, where more familiar emergency surroundings and equipment are available in case of an emergency. The first intervention with acute airway obstruction should be mask positive-pressure ventilation. It may be possible to insert a laryngeal mask airway, but this may also be difficult if the approximation of the laryngeal inlet and the posterior pharyngeal wall is very close, as a result of the traction. This may also make the insertion of an oral airway difficult, although a nasopharyngeal airway might be easier and more effective. Desperate situations almost always evoke some measure of desperate interventions, but if mask ventilation can be provided, then the patient should be moved to the operating room directly where the full range of definitive treatment can be provided.

   The patient is likely a difficult airway because of the underlying anatomic problem and also because of the halo device bringing the posterior pharyngeal wall into approximation with the laryngeal inlet. In addition, exposure of the larynx with a standard laryngoscope may prove challenging in the patient in a halo.

3. What will be your primary anesthetic technique? Why? Will you use nitrous oxide? Why/why not? What is your choice of muscle relaxant, if any? Why? What would you consider optimal fluid management for the case? Choice of fluid? Volume?

4. During surgery, the patient suddenly develops a drop in blood pressure, bradycardia, and a drop in end-tidal CO_2. What is your differential diagnosis? How can you go about narrowing the possibilities? What would you do? Why? Blood pressure is 60/40 with a heart rate of 60; how would you manage his depth of anesthesia? Why?

If adequate ventilation is ensured and the patient's airway is secured, then the team should discuss whether to proceed or not. Reasons not to proceed would include, for example, undue manipulation of the head and neck and the desire to wake the patient up and reevaluate the patient's neurological status.

3. The main anesthetic consideration intraoperatively is choosing a technique that will interfere least with measurement of sensory and motor evoked potentials, so the inhalation agent use has to be minimal, neuromuscular blocking drugs should be avoided, and a hypnotic and narcotic technique would probably be optimal. That combination might typically include propofol and fentanyl, sufentanil, or remifentanil by infusion. While the intraoperative goal would be minimal interference with evoked potential monitoring, emergence and perioperative goals would be directed to comfort as well as ease of assessment of neurological status, which is another reason that narcotic techniques are favored. Minimizing intravenous fluid administration would decrease the amount of tissue edema in dissected areas, which is particularly concerning neural tissue. The choice of fluid should be normal saline rather than lactated Ringer's solution because of the hypo-osmolality of Ringer's, and the volume administered should be conservative.

4. Several causes can be speculated. Sudden volume loss from surgical bleeding is a possibility, and because of gravity, blood loss will not necessarily be visible and obvious. A venous air embolism can produce an identical clinical scenario, as can reflex-mediated neural output from the brainstem while dissecting in the upper cervical spine. While the precordial Doppler is very sensitive for detecting air, it is not infallible, so the surgeon should be informed, the field flooded, and other causes ruled in or out. In that context, the surgeon should be immediately informed of the patient's instability and flood the field. Release of traction will relieve extrinsic pressure from the brainstem if this is a contributing factor. In any event, the patient's depth of anesthesia should be decreased in an effort to ultimately increase regional blood flow and perfusion pressure.

Postoperative Course

Question

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? Is it likely that it will be a "difficult extubation?" What would you look for at the end of surgery to help you predict the success of your extubation strategy? How could you use a tube exchanger to facilitate extubation? At the end of the procedure, following extubation, desaturation recurs while the patient is struggling, bearing down, and breath holding. Your management? Why? Is it likely that the patient's airway will be worse after surgery than before?

Additional Questions

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?

Postoperative Course

Answer

1. Postoperative intubation may be a very reasonable choice if the airway was significantly altered by the surgical procedure, as it often is, because of a shortened anteroposterior pharyngeal diameter.

   Edema may be a further consideration following the prone position and significant fluid exchange. But leaving the trachea intubated has relatively little to do with waking the patient up enough to obtain a neurological exam, which should be easily achievable in order to evaluate the neurological status. Another possibility is extubation over a tube exchanger, in order to facilitate reintubation, and this should be considered as well. The mental status does help in assessing fitness (and motivation) for extubation.

   Tube exchangers are a means of providing a stent with oxygen flow to a patient, as well as facilitating reintubation. They are fitted with a locking 15 mm OD circuit connector at the end and can be inserted easily.

   The patient can be struggling, bearing down, and breath holding because he is light and was intubated prematurely or because of secretions or airway irritability.

   The patient's airway sill likely be worse – more narrowed – immediately after surgery, but will remodel itself as an adaptive measure.

Additional Questions

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 leads 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) 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.

2. What is Goldenhar's 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. A 5-year-old was hit by a car while riding his bike and sustained an over-the-handlebar fall into the pavement, suffering a Le Forte I fracture, mandibular fracture, a Colle fracture, and various bumps and bruises. He comes to the operating room in a soft collar in preparation for oral surgery to repair his facial trauma. What is the difference between the various Le Forte fractures, and what is the significance for your anesthetic plan? Your considerations for anesthetic induction? Maintenance? What are your considerations for anesthetic emergence? What are your considerations for how you will wake him up? Would it be safer to leave him intubated? Why or why not?

Considerations for anesthetic induction include the patient and family's emotional state – the patient is fearful because of multiple visits to the clinic as well as "stranger anxiety" typical for this age, and the parents are fearful as well because they know this is a big operation with significant morbidity. There is also the larger context of the uncertainties of the family with a chronically ill, syndromic child. While there may be advantages to a parent present during induction in the operating room such as decreased crying and decreased aerophagia, thereby lowering the risk of intragastric air and regurgitation, these advantages may be outweighed by the relatively minimal chance that this infant will be consoled by a parent's presence, the likely difficulty with the mask fit because of midfacial hypoplasia, and the presence of impaired secretion elimination because of the sinus abnormalities with the strong possibility of airway irritability, laryngospasm, or bronchospasm on induction.

A balanced technique with fentanyl/air/oxygen and isoflurane would be my anesthetic technique of choice. Nitrous oxide should be avoided because of the risk of venous air embolism and also because body cavities will be opened and subsequently closed. Muscle relaxation should be utilized to ensure lack of movement.

2. Goldenhar's 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. You have to be concerned as much about what you don't see as what you do see; the fact that he has multiple facial fractures and therefore the potential for airway trauma or difficult laryngoscopy and intubation may be the tip of the iceberg; he may also have chest and cardiac contusions, blunt abdominal trauma, closed head injury, and a period of unconsciousness and altered sensorium with or without concussion. All of these are less obvious in physical exam and therefore must be suspected and inquired about during the history, with any additional appropriate laboratory tests. Assuming that these other issues have been ruled out, then the amount of bleeding, soft tissue injury with swelling, bony injury, and trismus will affect the ease of induction, mask fit, and intubation. A 5-year-old will not

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

5. A 9-month-old is scheduled for exploration of the posterior triangle of the neck for a large right-sided “cystic hygroma” that is half the size of his face. What is a cystic hygroma? From where does it arise? Why does the surgeon want to do a posterior triangle exploration? What are your anesthetic considerations? Are these patients difficult to intubate? Under what circumstances? He asks you to leave the right arm free of IVs in case he has to do an axillary exploration. Why would he ask that?

likely tolerate an awake laryngoscopy and intubation, so a decision will need to be made about the amount of trismus present that is likely to be relieved with the induction of anesthesia (and therefore facilitate intubation) versus the amount of soft tissue swelling and/or bony injury that may make laryngoscopy and exposure difficult. Radiological evaluation may be very helpful in this regard, especially if stridor is present. It is important to discuss the extent of the repair, as it is likely that a nasal intubation will be necessary if internal maxillary fixation is to be applied. Emergence considerations are important as well; coughing, bucking, and Valsalva maneuvers should be avoided, and in general, a smooth wake-up is best. It may be well advised to keep the patient intubated for several days until soft tissue swelling decreases and an air leak is reestablished. Extubation may be accomplished in the intensive care unit or in the operating room.

4. 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's syndrome) [3]. 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 endpoint 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.

5. "Cystic hygromas," more properly known as cystic lymphangiomas or macrocystic lymphatic malformations, typically present in this area of the head and neck as lobular, multiloculated collections of cysts and lymphatic vessels filled with clear or serous fluid. There are a variety of often bewildering and challenging presentations because, while most arise in the posterior triangle of the neck in continuity with the fascia of the longus colli muscle, they may extend into the axilla, the anterior triangle of the neck, and intraorally. In addition, bleeding may occur into the cysts, forming clot and therefore hard, extrinsically compressive masses to the oral cavity and therefore the airway. The airway may therefore be difficult to visualize in these patients, and each should be looked at individually for the potential for difficult mask fit, visualization, and tracheal intubation.

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Chapter 16
Ophthalmology

Robert S. Holzman

A 2-year-old, 8.5 kg girl is scheduled for bilateral 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.

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Preoperative Evaluation

Questions

1. Of what significance is the early history of prematurity? Why? Do you still anticipate problems? Should this patient have a retinal exam preoperatively for retinopathy of prematurity (ROP)? Why/why not? Is the concentration of inspired oxygen a concern in your anesthetic plan? Should this patient receive narcotics?

2. Is this patient at greater risk for the development of malignant hyperthermia? Why/why not? How can you further determine this? Should the patient have a muscle biopsy? Would you use a clean technique? Why/why not?

Intraoperative Course

Questions

1. What are your considerations for anesthetic induction? Why? Your colleague stops by and suggests pretreating with an anticholinergic? What do you think? Should this patient have been premedicated with an anticholinergic an hour earlier? Is intravenous administration during induction more or less effective for prevention of the oculocardiac reflex? Why/why not?

Preoperative Evaluation

Answers

1. She is quite small for her age, so the first thing to be concerned about is failure to thrive; an average 2-year-old should weigh about 12 kg. If she is simply small for her age and has been consistently at the lower end of the growth curve, then reassurance from the pediatrician along with an adequate nutritional history would probably suffice, but it is very possible that with poor intake, she may be anemic and immunodeficient and have other issues that require further medical consultation. 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 F_iO_2 of 0.3–0.4 following 100 % oxygen prior to intubation. Likewise, I would not be concerned about a significant risk of respiratory depression with judicious amounts of opioids, with this history of 2 years of age in an otherwise asymptomatic ex-34 weeker.

2. 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.

Intraoperative Course

Answers

1. My principal consideration for this child, who is certainly small for her age but in reasonable health according to her vital signs and hematocrit, is her separation/preoperative anxiety. This can be dealt with by anxiolytic premedication with or without a parent-present induction. The premedication is more likely to be anxiolytic than the presence of the parent, although there is much variation. Moreover,

2. Is a muscle relaxant necessary for this case? Why? What is your choice of muscle relaxant? Why? Will you use nitrous oxide? Why/why not? Should this patient routinely receive antiemetics? What's your choice? Why? What are the disadvantages of metoclopramide in a child compared with an adult? Is total intravenous anesthesia with propofol an advantage? Would ondansetron be of any greater benefit? Is dexmedetomidine part of your anesthetic plan? Advantages? Disadvantages?

many parents wish to be present for a variety of reasons beyond "reassurance," such as curiosity and education, and these may be very reasonable because ultimately satisfying these needs will inform future decisions and may be helpful for siblings as well. However, parents should not feel compelled to go into the operating room or participate in the parent-present induction if they do not wish to do so. Many in the past have felt that an anticholinergic premedication in the setting of strabismus surgery is required to decrease the incidence of the oculocardiac reflex. When working with experienced surgeons, the bradycardia of the oculocardiac reflex is most rapidly treated by asking the surgeon to stop the traction on the extraocular muscle. If an anticholinergic is needed to treat the bradycardia, it can be administered intravenously at that time. Most patients, however, do not experience the bradycardia of the oculocardiac reflex in the hands of a skilled surgeon, so their routine exposure to an anticholinergic is not necessary.

2. A muscle relaxant is not necessary for this case, as long as the patient is well anesthetized and motionless. Movement under the microscope will be magnified, and there is the possibility of significant compromise of the surgical procedure and injury to the patient. Surgeons often have a preference for operating with or without the administration of a neuromuscular blocking agent and will often make that part of their preoperative discussion with the anesthesiologist. The most important thing, however, is consistency of anesthetic technique with regard to the surgeons' caliper measurements for muscle resection and recession. The choice of muscle relaxant will depend on the anticipated length of the surgery (i.e., the number of muscles). Pancuronium, with its associated tachycardia, may be a very reasonable choice for this particular procedure. Neuromuscular blockade should be monitored carefully because of the echothiophate treatment. I would use nitrous oxide as part of the anesthetic technique, with only a minor concern about its possible contribution to perioperative nausea and vomiting (PONV), which is likely to occur in any event with this patient because of the association of PONV with strabismus surgery. Prophylactic antiemetic administration is very reasonable given the high association of strabismus surgery with PONV. For years, droperidol was routinely administered to strabismus surgery patients, but now there is concern about its effect on cardiac conduction and its possible contribution to Torsades, an extremely rare event which nevertheless resulted in a "Black Box" warning from the FDA which effectively stopped the use of droperidol as an antiemetic. Other antiemetics include ondansetron, a selective 5-HT3-receptor antagonist which blocks serotonin, both peripherally on vagus nerve terminals and centrally in the chemoreceptor trigger zone. Metoclopramide can also be used, but is associated with a higher incidence of tardive dyskinesia in children than in adults, although these are typically in larger dose ranges than those administered by anesthesiologists. Propofol-based TIVA may have some advantages in that it has an antiemetic effect and would also allow avoidance of the use of potent inhalation anesthetic agents. Its antiemetic effect, however, may be of questionable duration into the postoperative period, so a more sustainable strategy might involve the use of ondansetron, dexamethasone, and possibly metoclopramide.

3. During surgery, the patient suddenly develops a drop in heart rate from 110 to 54 beats per minute. What is your differential diagnosis? How can you go about treating? What would you do? Why? Why does this happen? What is the specific pathway of this reflex? Does greater anesthetic depth block this reflex? What is a traction test? What is a duction test?

Postoperative Course

Questions

1. The anesthesia resident would like to do a deep extubation. What are the advantages of a deep extubation? Would you choose this method? Why/why not? What are the risks of a Valsalva maneuver in this patient? What if she had just undergone an open globe procedure?

3. The most likely cause of this bradycardia is the oculocardiac reflex, mediated in its afferent limb by the long and short ciliary nerves, synapsing in the ciliary ganglion and traveling through the ophthalmic division of the trigeminal nerve. The efferent limb travels from the motor nucleus of the vagus nerve through the peripheral portion of the vagus nerve and terminates in the heart. Other causes include hypoxia, hypertension, elevated intracranial pressure, and other noxious stimuli mediated through the vagus. Direct myocardial depression from the inhalation anesthetic, specifically through its antichronotropic effects, or cardiac effects of other drugs, such as opioids, should also be included in the differential diagnosis. The first maneuver would be to make the surgeon aware of the heart rate and request that he or she discontinue their procedure; the patient may then be observed for a relatively rapid return to a normal heart rate. If this does not occur, then an anticholinergic should be promptly administered IV. An increase in anesthetic depth may block this bradycardia, but it may also contribute, because of the antichronotropic effect, to a worsening of the bradycardia. When traction is applied to the extraocular muscles, it will eventually produce a bradycardic response followed by a recovery tachycardia upon release of the traction. Fatigue of the oculocardiac reflex typically occurs with subsequent manipulation; this is generally referred to as a traction test. A forced duction test is a maneuver used to evaluate mechanical restriction to ocular movement. The sclera is grasped with a forceps and the eye is moved into each field of gaze. This allows the surgeon to differentiate between a paretic muscle and a muscle with restricted movement. Many surgeons prefer that neuromuscular blockade be used during a forced duction test to eliminate the variability in muscle tone with the changing depth of anesthesia. This test is most useful for those undergoing repeat strabismus surgery or in those with paralysis or prior trauma causing a mechanical restriction.

Postoperative Course

Answers

1. A deep extubation, when well conducted, will allow for a motionless emergence without bearing down or "bucking" on the endotracheal tube with resultant strain and an increase in mean arterial and central venous pressure as well as head and neck venous pressure. There is, in addition, some evidence that suggests higher oxygen saturations through emergence following deep extubation in children [1]. The Valsalva maneuver would increase venous pressure in the head and neck and influence the accumulation of edema in the surgically resected tissue; however, this is less of a problem in extraocular surgery than with intraocular surgery, such as an open globe. Most pediatric patients, however, will cry after these procedures, testing the closure of the globe anyway.

2. You are called urgently to the PACU 15 min post-op because the patient has been screaming, banging her head against the side rails, and not making any sense and is inconsolable. She is scratching at her eyes. What is your differential diagnosis? Should you sedate her? Get a blood gas? Give her physostigmine?

3. As part of the plan, the surgeon would like to adjust the correction in the PACU via the adjustable sutures:
   (a) What will the surgeon need from you in order to obtain the best result?
   (b) What medication will you use?
   (c) Are there any premedication administration criteria that you use to determine the patient's fitness for this portion of the procedure?
   (d) What monitoring will you choose?

Additional Topics

Questions

1. A 5-year-old just fell from the Jungle Gym at MacDonald'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, hematocrit 36 %. What anesthetic plan and counseling will you give to the parents?

2. Emergence delirium can occur for a variety of reasons; the most common in this situation would be the delirium associated with emerging from a sevoflurane anesthetic (the etiology of which remains unclear) and the central nervous system effects of anticholinergic syndrome, resulting from the administration of anticholinergics during surgery or as part of the reversal of neuromuscular blockade, particularly if atropine was used (tertiary ammonium molecule which can cross the blood-brain barrier). There is a lower chance of this syndrome with the use of glycopyrrolate, a quaternary ammonium molecule which is much less likely to cause CNS effects because it is a polar molecule. Hypoxia, respiratory distress, and pain should be considered in the differential diagnosis as well. Sedation may work if the patient is having a difficult time adjusting to the emergence and is at risk for self-injury, although ideally she should simply be protected from hurting herself until the delirium (if that is what it is to be determined) passes. A blood gas is unlikely to help in further delineating hypoxia beyond the reading of a pulse oximeter probe. The administration of antilirium (physostigmine), a centrally acting anticholinesterase, may be useful diagnostically, but its synergy with echothiophate should prompt concern.

3. 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 [2]. 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.

Additional Topics

Answers

1. 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

2. What happens to intraocular pressure with a vigorous Valsalva maneuver? (Describe the pathway, specifically, and how it affects the aqueous humor.) Is it rational to use a muscle relaxant if a patient performs a Valsalva maneuver? Are there any relaxants to be avoided? Why? What about propofol or any other hypnotic?

3. What are the systemic effects of the following antiglaucoma drugs that are of significance to the anesthesiologist?

   Timolol
   Brinzolamide
   Pilocarpine

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 barbiturates 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 2–3 min for a partial depletion of acetylcholine quanta at the neuromuscular junction, and then intravenous induction (with thiopental or propofol) followed by an intubating dose of the nondepolarizing neuromuscular blocking agent. The onset of adequate relaxation for tracheal intubation usually follows within 90 s, 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 [3].

2. The effect of the Valsalva maneuver is as follows: it raises the central venous pressure, it impedes drainage of the aqueous humor through the canal of Schlemm into the episcleral venous system, it increases intraocular blood volume, and therefore it increases intraocular pressure. The administration of a nondepolarizing muscle relaxant may be necessary to immobilize a patient if a Valsalva maneuver occurs during an alteration of the depth of anesthesia. If succinylcholine is chosen, it may actually elevate intraocular pressure as a result of its tonic effect on extraocular muscles. It may be more efficacious to administer thiopental or propofol intravenously in order to acutely lower the intraocular pressure.

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 s 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 adjunctives 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.

Alpha-adrenergic agonists decrease aqueous production and may increase uveoscleral outflow. Brimonidine 0.15 and 0.2 % crosses the blood-brain barrier. They are relatively contraindicated in children, due to side effects of somnolence and fatigue, presumably due to central nervous system depression. It was recently reported that two infants, both younger than 2 months, experienced apnea, lethargy, hypotension, hypothermia, and hypotonia, after the administration of one drop in each eye of topical brimonidine. Both infants recovered without sequelae after brimonidine was discontinued. There have also been reports of syncopal episodes in two children, both 10 years of age [44]. A case of intermittent coma in a neonate treated with brimonidine was remarkable for findings of plasma brimonidine concentrations 12–24 times higher than the mean plasma concentration observed in adults. Symptoms were reversed temporarily with naloxone and ceased after brimonidine was discontinued. The other agent in this class, apraclonidine 0.5/1.0 %, is much less selective for alpha-2 adrenoreceptors than is brimonidine, and it is also more useful for short-term rather than long-term therapy.

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

4. An ex-premie is scheduled for an examination under anesthesia and cryotherapy for retinal detachment. What exactly is cryotherapy? What is the difference between that and a scleral buckle procedure? How will you anesthetize him? Where will you measure his oxygen saturation? Should you anticipate any other problems specific to his ex-premature state?

their ability to relax nocturnal ciliary muscle tone. Dosing more often than once daily may mediate inflammatory effects, causing an increase in intraocular pressure.

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.

4. Cryotherapy is used when retinopathy of prematurity has led to cicatrix formation in the retina and impending retinal detachment and loss of vision. Cryotherapy is used to produce a chorioretinal scar, sealing a retinal break as a result of the accumulation of subretinal fluid with subsequent detachment. Cryotherapy may be used as part of a sequence including a scleral buckling procedure for reattachment of the retina to the pigment epithelium. In scleral buckling, the sclera is physically indented in order to bring the retinal tear closer to the pigment epithelium. External drainage of the subretinal fluid may be necessary as well. The anesthetic required is a general endotracheal anesthetic. Special consideration has to be given to the use of nitrous oxide; if chosen as part of the anesthetic technique, the air in the posterior chamber may actually increase in volume if nitrous oxide is continued once the eye is closed; therefore, nitrous oxide should be discontinued approximately 20 min prior to sealing the chamber. Alternatively, nitrous oxide can be avoided as part of the anesthetic technique. Oxygen saturation can be measured in any extremity or the earlobe and should provide an accurate reflection of retinal saturation. While there may be theoretical concerns about oxygen treatment in the setting of established retinopathy of prematurity, the patient already has very significant injury from multifactorial causes, and the entire patient has to be considered. Patients with coexisting conditions such as cardiac lesions may require additional considerations for oxygen monitoring.

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Chapter 17
Respiratory System

Robert S. Holzman

A 3-year-old male, 15 kg, is scheduled for functional endoscopic sinus surgery, bilateral myringotomy and tubes, and nasal ciliary biopsy. Vital signs are BP 90/60 mmHg; P 125 bpm, as palpated by his PMI in the right precordium; and T 37.2 °C. His hemoglobin is 13.0 gm/dL. 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.

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Preoperative Evaluation

Questions

1. Why is this patient wheezing? How would you like to further evaluate the wheezing? Why? How will it affect anesthetic management?

2. What are the implications of dextrocardia for this patient?

3. Is this patient adequately treated with his current medication regimen? What would you add? Why? Any specific implications for anesthetic management? Are there significant neuroendocrine effects to be expected from the use of beclomethasone through the inhalation route?

Preoperative Evaluation

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 breath, the less efficient their breathing becomes. They therefore have to work harder and utilize more calories to breathe, worsening their failure to thrive.

   The evaluation of wheezing is initially clinical, with attention to the respiratory rate, utilization of accessory muscles of respiration such as intercostal muscles and neck muscles, and gross movement of the entire rib cage. In addition, intercostal retractions may be seen. Tachypnea is the first sign of respiratory distress unless the patient is already exhausted. The wheezing may be inspiratory, expiratory, or biphasic, and this will lead to a further refined diagnosis of supraglottic, infraglottic, or mixed airway obstruction. A careful history and the patient's responsiveness to bronchodilators will provide further information as to the etiology of the wheezing. There may be cardiovascular consequences as well, including pulmonary hypertension and right heart failure. Anesthetic management should be directed to optimal preinduction medical management through sympathomimetics, anticholinergics, and leukotriene modifiers, with additional steroids as needed. Theophylline is used less often.

2. 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.

3. 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. An albuterol inhaler and ipratropium may be appropriate additional medications for achieving short-term control. Leukotriene modifiers (montelukast, zafirlukast) will become effective more slowly. The beclomethasone can have systemic effects, although these may not be clinically significant.

4. Should this patient be premedicated? With what? Why? What about a parent-present induction?

Intraoperative Course

Questions

1. How will you set up the ECG monitor? Why? Would an arterial line be a good idea Why/why not? Would you place it preinduction? Is a sidestream capnometer more accurate than a mainstream capnometer for this case? What about transcutaneous CO_2 analysis?

2. Would an anticholinergic be helpful prior to induction? Would you administer? Why/why not? Is this patient at risk for inspissation of secretions as a result? How can you minimize airway reactivity during the induction of anesthesia? How will you know when he is "deep" enough? Should any specific intravenous agents be avoided? Why/why not? Is ketamine better than propofol for induction?

4. Premedication may be helpful for several reasons; it will lessen anxiety (for the parent as well as the child) and will also decrease the secretion of endogenous catecholamines and lower the risk of induction laryngospasm. In addition, lower anxiety will decrease crying and aerophagia, which may increase the chance of aspiration. A benzodiazepine alone should not shift the CO_2 response curve and cause significant respiratory depression. A more complete "preinduction" premed such as a benzodiazepine + ketamine + an anticholinergic orally will serve several purposes: amnesia, anxiolysis, bronchodilation, and a decrease in secretion volume.

Intraoperative Course

Answers

1. The ECG monitor should be configured for dextrocardia with a mirror image of the usual lead placement. An arterial line is probably not necessary for this type of surgery, which should be relatively short, without a large blood loss. On the other hand, if the patient's respiratory status deteriorates as the case proceeds, one should have a low threshold for arterial line placement in this patient with known severely reactive airways. Sidestream and mainstream capnography differ because the sidestream capnograph aspirates a bulk sample of respiratory gases continuously, while the mainstream capnograph detects respiratory gases by an infrared beam transmitted through the gases in the endotracheal tube without bulk gas sampling. The main disadvantage of the mainstream capnometer in children is that the detector is bulky relative to the diameter of the endotracheal tube. Under the drapes, in a warm patient, there is a tendency for the weight of the mainstream capnometer to kink the endotracheal tube. The aspiration side channel in the sidestream capnometer is much lighter and does not kink the tube. Transcutaneous CO_2 analysis can be helpful when skin blood flow can be relied upon; in cold patients and low cardiac output states, it becomes less reliable. General anesthetics can also alter skin blood flow, but children preserve their cutaneous blood flow better than adults, and therefore, in the warm child, transcutaneous CO_2 analysis is relatively reliable.

2. It is worthwhile administering an anticholinergic prior to induction; however, it is important to be aware of the dose administered. The most logical choice is a reasonable dose of glycopyrrolate intravenously (if an IV is present) or inhalation administration of ipratropium bromide. Inspissation, or drying, of secretions is not a significant concern, because anticholinergics change the volume of elaborated secretions but not their water content [1–3]. In general, patients should be deeply

3. What is your choice for anesthetic maintenance? Why? How would the use of sevoflurane for maintenance compare to isoflurane? Would desflurane be a good choice after induction and during maintenance? Is MAC different in this age group? In what way? A colleague suggests that he would avoid nitrous oxide in this patient? Do you agree? 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?

4. What muscle relaxant would you choose? Why? What are the varying histamine-release properties of the relaxants you typically use? Which releases the least amount of histamine?

5. Are there advantages to allowing this patient to breath spontaneously? Would you do it? Why/ Why not? What breathing circuit would you choose? Why?

anesthetized as well as adequately treated with sympathomimetics, anticholinergics, and steroids, in order to additionally attenuate their bronchoreactivity. The usual clinical signs should be followed for depth, such as heat rate, blood pressure, and abdominal muscle tone. The eye signs, particularly pupillary dilatation, may be less reliable because of the administration of the sympathomimetics and anticholinergics both causing mydriasis. Intravenous induction agents do not need to be avoided as long as you remember that they are primarily hypnotics and anesthetic depth is more influenced by the volatile agents and narcotics. Ketamine may also act synergistically for depth of anesthesia and sympathomimetic effects. The choice of intravenous induction agent makes relatively less difference as long as an adequate depth of general anesthesia is achieved by one of the above strategies.

3. A balanced anesthetic with a significant component of deep inhalation anesthesia with a volatile agent would probably be best. The opioids are excellent for providing analgesia and rapid attenuation of surgical stress, while the potent volatile agents are excellent bronchodilators because of their effect on airway smooth muscle. At equi-MAC concentrations, the volatile agents are almost all equally efficacious at bronchodilatation. Desflurane, because of its pungency, has been associated with laryngospasm during induction of anesthesia in children, but has certainly been employed successfully as an intraoperative maintenance agent following a sevoflurane induction, with the added benefit that as an insoluble anesthetic, the emergence is more rapid. Ketamine may be a reasonable choice because of the reasons outlined above. Avoiding nitrous oxide allows you to use a higher F_iO_2 and may also be important if the pulmonary vascular resistance is elevated and pulmonary hypertension is present. Cocaine is an excellent topical vasoconstrictor, but it is unnecessary to use a 4 % concentration in this age patient; 1–2 % would be enough for topical analgesia and vasoconstriction, up to 3 mg/kg. This is a relatively uncommon practice in pediatrics, however, where oxymetazoline has been found equally effective.

4. A variety of muscle relaxants can be chosen as long as their histamine release and potential for allergic reaction are minimal. Atracurium (mixed cis and trans enantiomers) and rocuronium are associated with histamine release, but vecuronium and cisatracurium are minimal histamine releasers.

5. There may be an advantage to spontaneous breathing due to less turbulent airflow distal to an area of obstruction, according to the Hagen-Poiseuille equation. For the majority of patients, muscle relaxation with positive pressure ventilation will provide optimal operating conditions and maximize patient safety because of less chance for movement. For severely obstructed patients, the spontaneous breathing strategy may become necessary. I would choose a circle absorption system for the economy of fresh gas flow. An alternative would be a variation of the Mapleson circuits, either a D (Jackson Rees modification of Ayre's T) or an A (Magill system, optimal for spontaneous breathing). However, both of these Mapleson systems require a much higher fresh gas flow, with its attendant loss of temperature and wastefulness of volatile agent, than a circle absorption system, which can have fresh gas flows lowered to within very low flow or closed circuit range.

6. Shortly following the application of 4 % cocaine, the patient's blood pressure is noted to be 180/110, with a heart rate of 220 bpm. As the surgeon begins, it increases to 210/120 with a heart rate of 230. Why? What else is in your differential? What would you do? Should you attempt to slow the heart rate? Why? What will you do to slow the heart rate? Any potential problems with that?

7. A sudden increase in peak airway pressure and desaturation (bronchospasm): the patient desaturates to 87 % over 30 s, and diffuse biphasic wheezing develops with a rise in peak inspiratory pressure to 55 cm H_2O. What would you do first? and next? Why? Where could the problem be? If this is truly bronchospasm, what could you do to fix it? Pharmacologically? Mechanically?

Postoperative Course

Questions

1. When would you extubate? Why? Would you extubate deep? Is this patient at risk for postextubation croup? More so than anyone else? For postextubation laryngospasm? More so than anyone else? Tracheomalacia?

2. What are the risks to this patient of reversal of neuromuscular blockade? Why? What are the alternatives? Would you avoid a reversal agent? Is neostigmine better/worse than pyridostigmine or edrophonium?

6. The direct effect of absorbed cocaine is the etiology of these findings. There are two options – if the cocaine is packed in the nose and is continuing to be delivered, the packing should be removed and suction applied to remove any trace quantities pooled at the mucosal level. Medications that act directly as antihypertensives can be administered if these expectant measures are ineffective, such as sodium nitroprusside (Nipride) in doses ranging from 0.5 to 10 mcg/kg/min and esmolol or propranolol, also in small divided doses and titrated to effect.

7. There should be a logical and rapid progression through a differential diagnosis of mechanical obstruction at the level of the breathing circuit and endotracheal tube and then to the level of the patient's trachea and major conducting airways. Such obstruction can be due to a kinked tube or a tube obstructed with secretions or a mucus plug or the airway obstructed with a mucus plug. If there is no obvious kink in the endotracheal tube, then a suction catheter can be passed through the endotracheal tube until you are convinced that the tube is clear. If mechanical obstruction is ruled out, then diffuse inspiratory and expiratory wheezing can be accounted for by bronchospasm. Albuterol can be delivered via the endotracheal tube, but it may be more effective if the bronchospasm is severe to deliver dilute epinephrine (i.e., 0.1 mcg/kg) intravenously to see if the bronchospasm will break. Anesthetically, the patient should probably be deepened with the volatile agent; ketamine and intravenous glycopyrrolate may also be effective. In order to promote exhalation due to the impaired expiratory flow rate, the expiratory phase of the I: E ratio should be lengthened; otherwise there may be breath stacking.

Postoperative Course

Answers

1. Arguments can be made for deep and awake extubation. I would extubate awake because there is a better assessment of oxygenation, ventilation, and other airway needs, even though some studies have shown a higher saturation with deep extubation [4]. Postextubation croup is probably more related to subglottic irritation from the intubation as well as a loose subglottic basement membrane. Postextubation laryngospasm is a much more likely possibility in any child with irritable airways.

2. Neuromuscular blockade and controlled ventilation often result in less efficient respiratory mechanics because of the cephalad migration of the diaphragm, decrease in FRC, and decreased efficiency of alveolar airflow because of positive pressure ventilation. The reversal of neuromuscular blockade with an anticholinesterase and anticholinergic may result in bronchospasm and increased salivation due to the anticholinesterase.

Additional Questions

Questions

1. What are the anesthetic induction considerations in the patient with pulmonary sequestration? What immediate interventions can be performed therapeutically for deterioration during induction?

2. Is infantile lobar emphysema a surgical emergency? Why/why not? Should the patient be bronchoscoped first? Why/why not? What muscle relaxant would you choose? Why? A colleague of yours suggests the avoidance of nitrous oxide; do you agree?

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?

Additional Questions

Answers

1. Pulmonary sequestration is the result of early isolation of pulmonary tissue from the developing lung bud. Sequestration may occur as intralobar or extralobar, depending on whether the abnormal tissue is located within the pleura or outside of it. The abnormal tissue may have cystic and solid areas with mixtures of air, rudimentary air sacs, and bronchi and chronically inflamed areas. Deterioration during the induction of anesthesia may be a result of communication of the sequestration with the respiratory or gastrointestinal tract, resulting in respiratory distress or GI symptoms. Spontaneous breathing may be more optimal in this regard until the communication can be identified, isolated, and ligated along with the blood supply to the sequestration. Extralobar sequestration, although extrapleural, generally presents at an earlier age with recurrent infections. Arteriovenous malformations are also commonly associated.

2. It may or may not be, depending on the timing and severity of presentation. A deficiency of bronchial cartilage, bronchial stenosis, or extrinsic vascular compression from the pulmonary artery may result in congenital lobar emphysema, characterized by overinflation and air trapping in the affected lobe with compression atelectasis of adjacent parenchyma and possible mediastinal displacement. Males are affected more often than females. There is progressive respiratory distress in the newborn period or in early infancy. Rapid deterioration requires urgent surgery, and when accompanied by a mediastinal shift, increased intrathoracic pressure, impaired venous return, and decreased cardiac output may result. The diagnosis is made by chest X-ray when bronchovascular marking is present in a hyperlucent area of the lung. Bronchoscopy should be considered in the older infant if there is a possibility of intraluminal obstruction causing lobar emphysema. Surgical excision in the neonate, however, may be an emergent procedure if the lobe is expanding rapidly. Vigorous positive pressure should be avoided during anesthetic induction, and spontaneous ventilation should be preserved. Nitrous oxide should be avoided, as should muscle relaxants.

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 cava 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

4. 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		
FEV_1 = 18 % of predicted		
FEV_1/FVC = 96 % of predicted		
TLC = 64 %		
RV = 194 %		
RV/TLC = 78 %		
ABG: (0.5 L/min O_2) pH = 7.27, pCO_2	= 95 mmHg, pO_2	= 183 mmHg

Her ECG is unremarkable apart from a sinus tachycardia. Her echocardiogram 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 gastroenterologist's feel should take about 0.5 h to 45 min.

What are your plans for intraoperative anesthetic technique and postanesthetic care? What are the likely complications and pitfalls?

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.

4. 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 [8]. 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, 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 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 or most importantly during controlled ventilation [9]. 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 [10].

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Chapter 18
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?

Preoperative Evaluation

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

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?

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 and hip flexion

Intraoperative Course

Answers

1. Phenylephrine, infusions of dopamine, epinephrine, milrinone, and tranexamic acid.

2. ECG (5 lead including V_5), pulse oximeter X 2 (upper and lower), BP X 2 (upper and lower, arterial line and BP cuff), and CVP. ECG, pulse oximeter, and BP cuff prior to induction. The right arm may not be a reliable source of central BP measurement given the presence of a right Blalock- Taussig shunt (BTS).

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?

7. What are your transport considerations from the OR to the intensive care unit?

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.

   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 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.

Postoperative Course

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?

Postoperative Course

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 the 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 an 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.

   Neither cardioversion nor adenosine is effective. Treatment of JET is atrial pacing at a rate slightly faster than the junctional rate reinitiating AV 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 AV synchrony is offset by the reduction in diastolic filling time present at these rates.

Additional Questions

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?

Additional Questions

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 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 non-beating, unvented heart.

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 vascular remodelling with a gradual decrease in left to right shunting.

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Chapter 19
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 S_pO_2 is 75–80 %.

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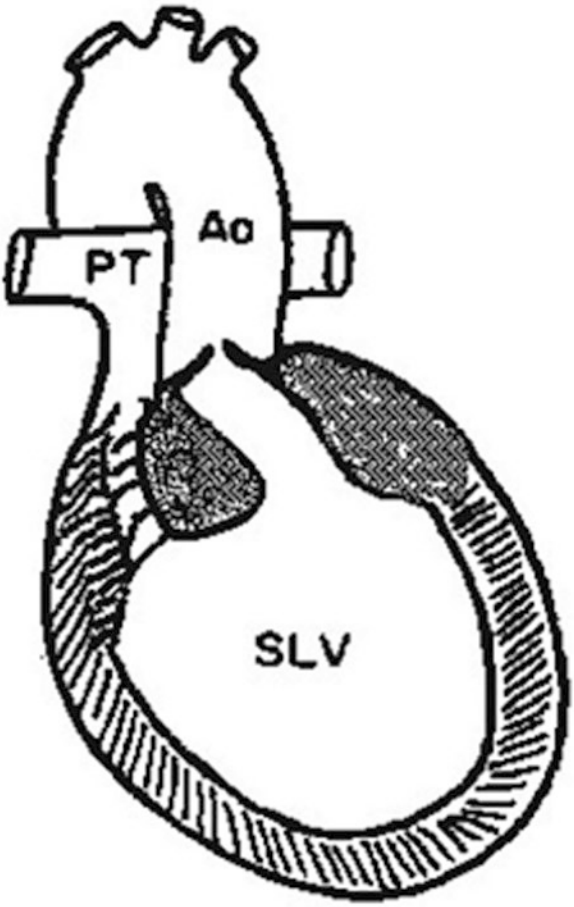


Preoperative Evaluation

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?

Preoperative Evaluation

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. The right "ventricle" is composed of only an outlet portion and communicates with the left ventricle via a bulboventricular foramen (BVF). Transposition of the great vessels (TGV)

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?

refers specifically to the anatomic circumstance 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 is 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 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 derived from the SVC to RPA and LPA anastomosis. 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. Following a 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 Glenn pathway. Cardiac catheterization would be necessary to rule out decompressing venous collaterals from the Glenn pathway to the IVC or directly to 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 ketamine 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?

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.

The hormonal factor present in hepatic venous blood that promotes this process has not been identified. 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.

Intraoperative Course

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 PAOP. However, placement of such a catheter would be highly impractical as it would require fluoroscopy and a trip to 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 and avoidance of dynamic obstruction to pulmonary blood flow across the BVF. In the case of a bidirectional Glenn, dynamic pulmonary outflow obstruction would be less relevant. 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

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 s 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 effect venous return? Particular concerns in this patient?

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 is 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 1:2–1:3, 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_D/V_T). 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.

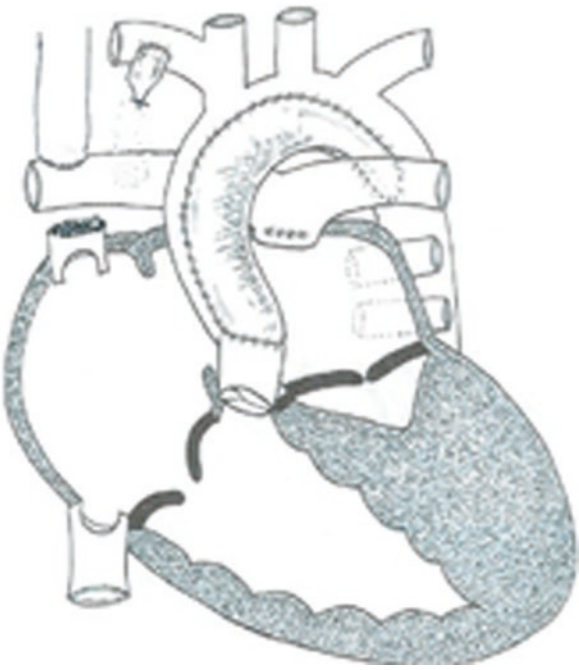
Postoperative Management

Answers

1. V_d/V_t will be increased in a patient with a Glenn shunt due to an increase in Zone 1 ($P_{airway} > P_{alveolar} > P_{pulmonary\ venous}$) 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 F_iO_2 will have no direct effect on the oxygen carrying capacity of this portion of the cardiac output. Increasing F_iO_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 than 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.

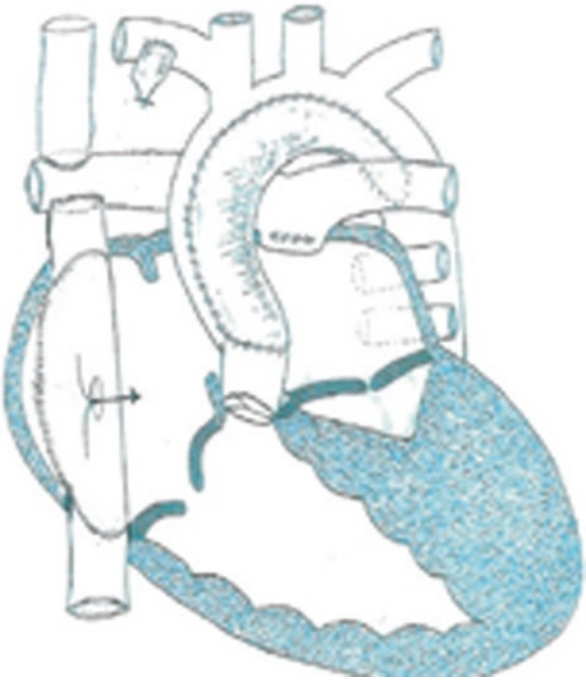


Bidirectional Glenn

Additional Questions

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

Additional Questions

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 you going to tell the parents? What are you going to tell the surgeon? 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 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 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.

3. The patient needs to be either chemically or electrically cardioverted. This is an unstable rhythm that will likely deteriorate to ventricular fibrillation (VF).

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Chapter 20
Cardiac III

James A. DiNardo

A 2-month-old with hypoplastic left heart syndrome (HLHS), s/p Stage 1 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 is 75–80 %, weight 3.8 kg, and Hct 42 %.

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Preoperative Evaluation

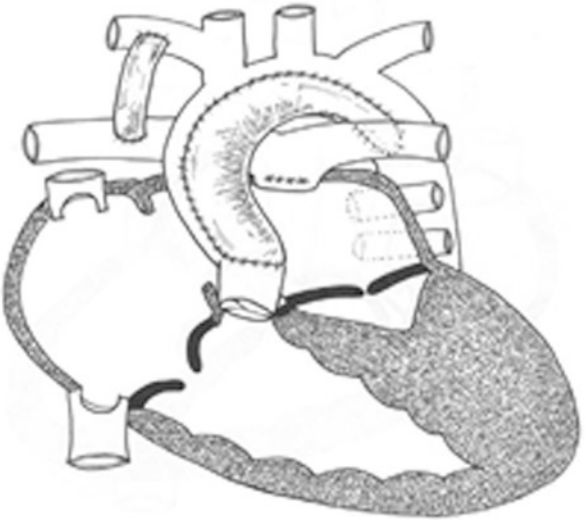
Questions

1. What is a Norwood procedure?

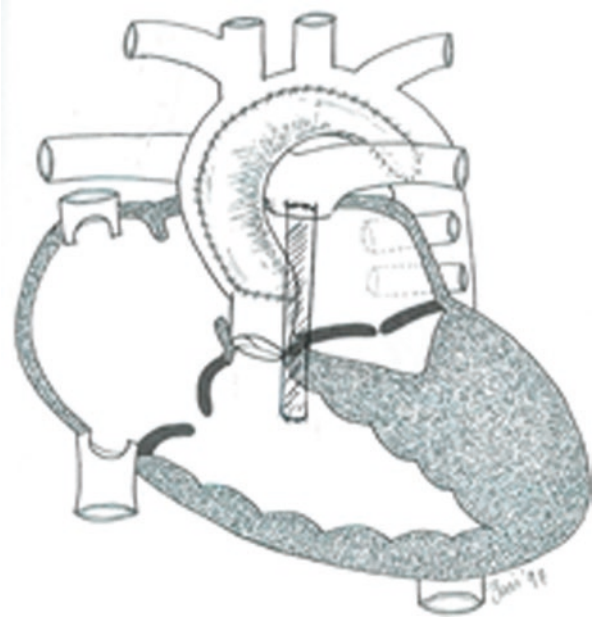
Preoperative Evaluation

Answers

1. Norwood with BT shunt



Norwood with RV-PA conduit (Sano)



2. What does the term “parallel circulation” mean?

3. What anatomic problems may arise after the Norwood procedure? How can you assess these clinically? What investigations are indicated?

Intraoperative Course

Questions

1. How would you induce anesthesia for this patient? Maintenance technique?

The Norwood procedure (Stage 1 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; autologous pericardium or graft material is used; 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 one ventricle then distributes output to both the systemic and pulmonary beds (parallel circulation). As a result of this physiology, the ventricular output is the sum of pulmonary blood flow (Q_p) and systemic blood flow (Q_s); 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 such as truncus arteriosus and 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.

Intraoperative Course

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 should not exceed 4 %, while maintaining spontaneous ventilation can be considered to facilitate 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? Why?

4. How would you treat hypotension (systolic BP <60 mmHg) and a reduction in SpO_2 to 70 % occurring 30 min 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 SpO_2 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 Course

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?

Postoperative Course

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 (SaO_2) 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 SaO_2 are:

   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 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 Questions

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?

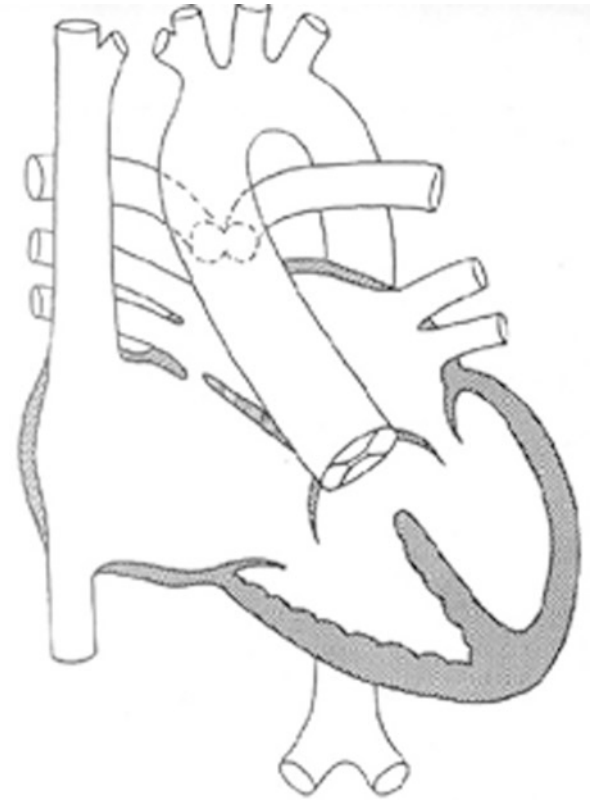
2. A 5-year-old male, S/P truncus arteriosus repair as a newborn, regularly followed by his cardiologist, but full details not available because he recently moved to the area, presents to the ER with an acute abdomen. The general surgeon suspects acute appendicitis and wants to proceed with surgery as 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?

Additional Questions

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 is 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.

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?

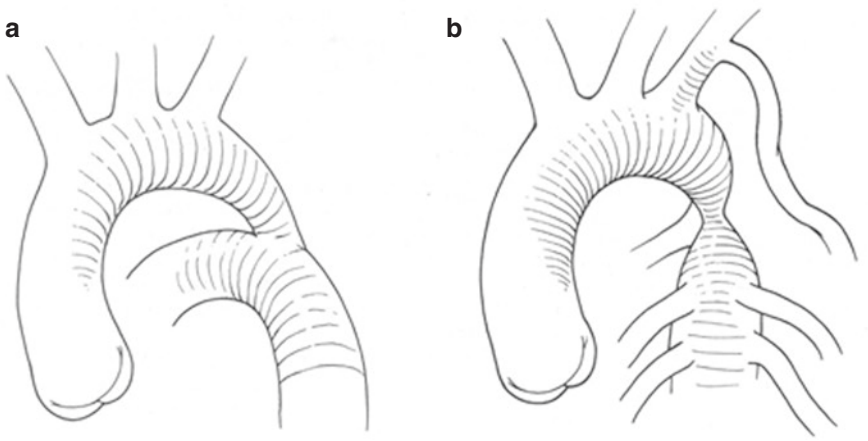
4. A 10-year-old with coarctation of the aorta is scheduled for repair by thoracotomy. Describe your clinical assessment of this patient. What preoperative investigations are important? How will they affect surgical management? Would you perform one lung anesthesia? What is the smallest double-lumen 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 from 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 valved 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 should focus on work of breathing and the ability of the baby to maintain systemic oxygen delivery and peripheral perfusion given limited cardiac reserve. Signs of systemic venous congestion such as an enlarged liver and jugular venous distention would be consistent with RV failure secondary to pulmonary hypertension. A CXR will likely reveal interstitial pulmonary edema, large central pulmonary arteries, bi-atrial enlargement, and a relatively normal sized LV as these children have small LVs as a characteristic of Shone's complex. An inhalation induction would be problematic in this patient. Hypotension and tachycardia during an inhalation induction in a baby with pulmonary hypertension and systemic or near systemic RV pressures will likely cause RV ischemia and cardiovascular collapse due to an inability of the RV to deliver an adequate volume of blood to the left heart. In addition, airway obstruction will elevate PVR compromising the ability of the RV to deliver an adequate volume of blood to the left heart. A functional examination could be performed with use of a ketamine delivered as an intermittent infusion or repeated small boluses. This would provide hemodynamic stability and dissociative anesthesia in conjunction with maintenance of spontaneous ventilation.

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, intercostals, 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 would be a 5FArndt 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.

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Chapter 21
Thoracic Surgery

Robert S. Holzman

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.

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Preoperative Evaluation

Questions

1. Will this patient need any diagnostic procedures prior to coming to the OR?

2. Would you obtain any imaging? Why? What studies? What if they require an anesthetic? Does this patient need any specific laboratory evaluation?

3. Should the patient be transferred to the ICU for stabilization, intubation, and airway securement and sedation?

4. Would you anticipate any comorbidity?

Preoperative Evaluation

Answers

1. 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. Typical signs and symptoms include tachypnea, grunting, retractions, use of accessory muscles, cyanosis, and failure to thrive. Impaired clearance of sections due to impaired air flow can result in infectious processes such as pneumonia or lung abscesses. It sounds like this situation is turning into an acute emergency after 2 days, contraindicating diagnostic procedures beyond those already done. The patient needs to come directly to the operating room and have the worsening respiratory status treated before any further compromise. Preoperative evaluation may require an echocardiogram to rule out congenital heart disease and may also be useful for identifying patients with (1) decreased cardiac output due to increased intrathoracic pressure from a large lung lesion and (2) pulmonary sequestrations with a large blood supply, in which high-output cardiac failure develops.

2. 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. If time permits, more thorough preoperative evaluation is necessary to determine the patient's current pulmonary status, the impact of the anticipated surgery, and the ability to tolerate OLV if needed.

3. The patient should come directly to the operating room for definitive surgical treatment.

4. Comorbidities may be congenital or due to acutely compromised physiology. CCAM is a lung lesion that develops from adenomatous overgrowth of terminal bronchioles. These lesions can be cystic, solid, or mixed intrapulmonary masses that lack bronchial cartilage and glands. Communication with the tracheobronchial tree is usually maintained and vascular supply and venous drainage are derived from the pulmonary circulation. Approximately 20 % of patients with CCAM have an associated congenital anomaly, with renal agenesis and cardiac defects being the most common. When the lesions are large, pulmonary growth may be compromised leading to pulmonary hypoplasia in normal contiguous lung areas.

5. Is this an unusual problem? Common? Relatively common for neonatal surgical emergencies? For neonatal thoracic surgical emergencies?

6. Would your considerations be any different if this was a congenital lobar emphysema or a pulmonary sequestration with regard to planning the anesthetic?

Intraoperative Course

Questions

1. What are your considerations for anesthetic induction? Your colleague stops by and suggests an awake intubation. What do you think? You select a spontaneous breathing inhalation induction. Would nitrous oxide enhance the uptake of your sevoflurane? What if the patient rapidly develops depressed ventilatory drive and positive pressure ventilation is necessary – what are your specific considerations in this circumstance? Should neuromuscular blockade be used to facilitate anesthetic induction following the administration of intravenous propofol?

5. CCAMs comprise approximately 25 % of all congenital lung malformations, with an estimated incidence of 1:25,000–1:35,000.

6. Each congenital lung lesion 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.

Intraoperative Course

Answers

1. In neonates with significant cardiopulmonary compromise, urgent/emergent preoperative intubation and ventilation may be required. For lesions that can become overdistended with positive pressure ventilation (PPV) (CLE in particular, but rarely CCAM as well) due to a ball-valve effect, spontaneous ventilation should be maintained. Because most CCAMs are solid, patients generally tolerate PPV without cardiopulmonary compromise. Pulmonary sequestrations have no bronchial connection, so there is no risk of overexpansion with PPV. An awake intubation is reasonable as long as it goes smoothly. It has the advantage of preserving airway reflexes with regard to aspiration as well as spontaneous breathing. The disadvantage is that the stressed neonate may hold his breath, and the stimulation from awake laryngoscopy might result in stress and elevated intracranial pressure and might be the cause of regurgitation (rather than conferring protection). It will ultimately depend on the strategy used (i.e., topical anesthetic of the airway and skillful technique, in experienced hands).

   Inhalation or IV induction may be performed with general endotracheal anesthesia. In cases of significant intrapulmonary shunting, the rate of rise of inhaled anesthetics may be slowed, but in most circumstances, this is more theoretical than practical in this age group. In patients with CCAM or CLE, an inhalation induction is preferred because spontaneous ventilation should be maintained until either the chest is opened or one-lung ventilation (OLV) of the contralateral lung is achieved. Air trapping may cause compression of the normal lung, mediastinal shift, and a reduction in cardiac output. Because oxygen consumption is higher in the infant compared to the adult (8–10 mL/kg/min vs. 3.5 mL/kg/min in an adult), this coupled with a small FRC predisposes the infant to hypoxemia.

   In cases where overinflation of the lung is not a concern, muscle relaxation may be given to facilitate intubation. Positive pressure ventilation may be utilized at

2. What are your monitoring considerations? Is an arterial line needed? Why? How about a central line? What if a PICC line was scheduled in addition to the surgical procedure; would this be sufficient for monitoring blood gases and overall state of well-being intraoperatively?

3. Is there any way you can offer one-lung ventilation in order to facilitate the surgeon's exposure for thoracoscopic surgery? 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?

low peak inspiratory pressures/low volumes; hand ventilation may actually be better during many portions of the procedure. Unlike adults, infants with unilateral lung disease do not improve their oxygenation when the healthy lung is dependent and the diseased lung is nondependent. In infants, oxygenation is optimized when the healthy lung is nondependent. This difference is due to the more compliant chest wall of the infant, which cannot completely support the dependent lung. The FRC is consequently closer to the residual volume and airway closure becomes more likely in the dependent lung.

2. Monitoring should include standard noninvasive monitoring, an arterial line for blood pressure and arterial blood gas monitoring, and two peripheral IVs. A central line is probably not necessary unless needed because of difficult IV access. The accuracy of venous blood gases obtained from the central line accurately reflecting oxygenation and ventilation may be called into question with an open chest; arterial blood gas analysis is really needed here.

3. Thorascopic procedures may require OLV to improve visualization and reduce the risk of injury to the lung and adjacent structures Techniques for OLV in infants include the use of selective mainstem intubation or a balloon-tipped bronchial blocker. Although the mainstem bifurcation angle at the carina is close to equal (approximately 55°) in neonates, there remains a tendency for most "blind" mainstem intubations to go to the right; therefore, fiberoptic bronchoscopy guidance facilitates accurate endobronchial intubation. Other specialized tubes such as the Univent or double-lumen tube are too large for infants. In neonates, it may be necessary to place the blocker alongside the ETT because it will occupy a significant portion of the cross-sectional area of the ETT and consequently impede ventilation. With the use of a closed-tip bronchial blocker, the operative lung cannot be suctioned or have continuous positive airway pressure (CPAP) delivered to it. Particularly in neonates and small infants, there is a risk of dislodgment of the bronchial blocker with positioning of the patient and during the surgical procedure itself. The tracheal lumen may be occluded by the balloon tip. The position of the blocker should be continuously checked by monitoring airway pressures, auscultating the chest, and ultimately confirming position with direct visualization using a fiberoptic bronchoscope. A fiberoptic bronchoscope is needed for accurate placement of a bronchial blocker. The bronchoscope must be readily available throughout the surgery to confirm and check the position of the blocker in order to reposition it as necessary. If OLV is not established in patients with CLE, either spontaneous ventilation or gentle, cautious PPV with minimal PIP should be employed.

   Hypoxia or hypercarbia may occur during OLV or with surgical retraction of the lung. Reexpansion of both lungs periodically may be required. Some degree of hypercarbia will probably occur and may be permissible.

4. Following induction, the baby is turned laterally for prepping. There is progressive difficulty with mechanical ventilation, increasing peak inspiratory pressures, and slowly progressive desaturation. Concerns? Differential? Your next steps? The arterial line trace is dampened, the pulse pressure is 5 mm Hg, and the blood pressure cuff is continuously recycling without giving you a blood pressure. How do these findings affect your assessment? What would you like to do next? Anything you would like your surgeons to do?

5. Any plans for regional anesthesia for perioperative analgesia? At what level would you place an epidural catheter? Would ultrasound imaging help? Why? How specific do you have to be about tip placement of an epidural in a 2-day-old? Is it any different than for an adult? For a 2-year-old? Will it be worth it for this thoracoscopic procedure in a 2-day-old?

Postoperative Course

Question

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?

4. Infants with unilateral lung disease do not improve their oxygenation when the healthy lung is dependent and the diseased lung is nondependent. In infants, oxygenation is optimized when the healthy lung is nondependent. This difference is due to the more compliant chest wall of the infant, which cannot completely support the dependent lung. The FRC is consequently closer to the residual volume and airway closure becomes more likely in the dependent lung. In addition, shifting of the mediastinum, compromised venous return, and decreased cardiac output may all contribute to impaired gas exchange, hypoxemia, and ischemia. The patient should be turned back to the supine position, and the adequacy of cardiac output should be reevaluated. The dampening of the arterial trace probably reflects this decrease in cardiac output, as would a decrease in $ETCO_2$. In addition to repositioning, a volume bolus would be indicated in an effort to enhance the blood volume of the pulmonary circulation.

5. Postoperative pain can cause significant splinting and an effective regional anesthetic technique may smooth the perioperative course. A thoracic epidural catheter can be placed from thoracic, lumbar, or caudal approaches. The position of the tip of the catheter can be confirmed radiographically or by ultrasound imaging (in the case of wire-reinforced catheters) when it is placed after induction. Other regional techniques may be employed, with the paravertebral catheter as the most common emerging technique.

Postoperative Course

Answer

1. After most thoracic procedures, the patient may be extubated at the end of the procedure. Extubation is desirable because an air leak can develop at the bronchial suture (or staple) line with PPV. Adequate oxygenation and ventilation must be confirmed before extubation. In patients with limited cardiopulmonary reserve, continued postoperative intubation and ventilation may be required. There are numerous reasons why hypoxemia may occur following extubation. Breath-holding, bearing down, an increase in vascular resistance, exacerbating the splinting, and atelectasis that is likely to be present account for pulmonary shunt. In addition, intracardiac and extra-cardiac shunts may be present, with venous admixture during reversals of normal intrathoracic pressure relationships. A certain amount of patience is necessary to see if the infant will “get through” this emergence phase; otherwise, the endotracheal tube may need to be replaced.

Additional Questions

Questions

1. You have a full schedule of esophageal dilations in seven infants who are post various TE fistula repairs. What common medical problems do you expect to encounter in these patients? Which rare problems would you expect? What will the most vexing part of the anesthetic likely be? What metabolic consequences would you expect to take care of as a result of all of these patients being "veterans" of the operating room?

2. Under what circumstances would you insist on a preoperative cardiology evaluation and echocardiogram prior to a TE fistula repair?

3. What is the controversy about surgical repair of a TE fistula with or without a gastrostomy?

Additional Questions

Answers

1. The most common problem is overt or silent aspiration. The majority (in some estimates as high as 85 %) of these children will have methacholine-provocable reactive airways, and "asthma" is the most common medical diagnosis in these children. Several causes may coexist – silent aspiration, alterations of gastric emptying, changes in the normal position or pressure relationships of the lower esophageal sphincter, dysmotility of the distal third of the esophagus, and the presence of a laryngeal cleft as another manifestation of a foregut anomalous communication. Bronchospasm and airway reactivity intraoperatively is not uncommon and rarely surprising, with the presence of an increased amount of airway secretions and often accompanied by compromise of the tracheobronchial connective tissue manifested as tracheomalacia and bronchomalacia.

   Oxygen consumption is normally higher in the infant compared to the adult (8–10 mL/kg/min vs. 3.5 mL/kg/min). This coupled with a small FRC predisposes every infant to hypoxemia. For infants who are veterans of the operating room due to multiple surgical repairs, such as TEF repair, they are in a state of "tissue repair" for months to years later, with a consequent increase in oxygen consumption and CO_2 production, requiring much greater minute volumes in the operating room to eliminate their increased CO_2 load.

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.

3. The routine placement of a gastrostomy as part of the primary repair of TEF has been controversial for decades. While many feel it is simply unnecessary, some surgeons feel it is detrimental to the development of reactive airway disease and compromises the potential future use of the greater curvature if a neo-esophagus needs to be created in reconstruction.

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Chapter 22
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.

Preoperative Evaluation

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.

Intraoperative Course

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

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 conjoined 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?

administration. 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) cross-matched blood should be available. The blood bank should be at least 1–2 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.

Postoperative Course

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.

Additional Question

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 L3–4. 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.

Suggested Readings

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Chapter 23
Gastrointestinal Disease

Joseph P. Cravero

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 room air and appears comfortable. There is an oral tube/drain in place that is helping to drain oral secretions. Vital signs are BP = 85/50 mmHg, P = 120 bpm, R = 22/min, and T = 37 °C. Her hemoglobin is 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?

Preoperative Evaluation

Answers

1. TEF/EA has an incidence of 1:2,500–3,000 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 T-E 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?

Intraoperative Course

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 H_2O 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 SpO_2 suddenly decreases to 82 % from 99 % and the inspiratory pressure increases from 25 to 45 cm H_2O. 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?

Postoperative Course

Questions

1. At the end of the surgery, during planned "awake" extubation, the bronchospasm recurs. Your management? 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 FiO_2. 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 FiO_2 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

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

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 re-intubate? Why/why not?

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?

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 well 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

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 arousal, 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

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?

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?

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 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.

Chapter 24
Bariatric Surgery

Robert S. Holzman

A severely obese 17-year-old female is scheduled for laparoscopic sleeve gastrectomy. She is 5′9″ and weighs 310 lb (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 SpO_2 = 94 %. Her HbA1c = 9.2, which is part of a screening study for the surgery. She takes no other medication.

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Preoperative Evaluation

Question

1. Will this patient need any additional diagnostic procedures prior to coming to the OR? Would you obtain any imaging? Of what? Why? What studies? Does this patient need any specific additional laboratory evaluation? Would pulmonary function testing help? What would you expect it to show? Are there any additional comorbidities you should expect? 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?

Preoperative Evaluation

Answer

1. First, a thorough evaluation and physical exam is needed, which may lead to additional studies. Morbidly obese patients, including adolescents, are at risk for significant comorbidities such as cardiovascular (hypertension, dyslipidemia), endocrine (type 2 diabetes – T2DM), and, in girls, hyperandrogenism and polycystic ovary syndrome (PCOS), pseudotumor cerebri, nonalcoholic fatty liver disease, obstructive sleep apnea, asthma, slipped capital femoral epiphysis (SCFE), tibial torsion with varus deformity (Blount's disease), and psychological issues such as depression and low self-esteem.

   Imaging studies would be guided by the above considerations. Ultrasound is an easy screen for concerns about fatty infiltration of the liver, although the physiological significance of that finding would have to be determined by liver function studies including an evaluation of synthetic function.

   Pulmonary function testing is unlikely to be helpful unless there are reversible components to reactive airway disease that are undertreated. However, 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.

   While there can be other comorbidities, the above are the most common.

   Most of the patients are highly motivated and therefore do well. Even those whom you would expect might need postoperative mechanical ventilation are very motivated for extubation and mobilization in the immediate perioperative course. Efforts should be made to make them as comfortable as they need to be to promote such mobilization, including regional anesthesia techniques that will decrease systemic opioid requirements but allow pulmonary toilet and early mobility. It would be ideal to place the epidural with the patient awake but sedated, in order to self-report paresthesias or other adverse events. That said, if the patient is very anxious, then the amount of sedation in order to accomplish this effort might also blunt ventilatory drive significantly as well as efficiency of oxygenation. Another advantage is that the patient can self-report placement in or off the midline, which can be a substantial problem in morbidly obese patients when asleep.

Intraoperative Course

Questions

1. What are your considerations for monitoring? Your colleague stops by and suggests awake placement of an arterial line – what do you think? Why? Is it likely that an arterial line will make a difference in quality of care/perioperative outcome?

   You select awake/sedated placement of an epidural catheter for perioperative analgesia. What landmarks will you use? What imaging would be helpful, if any?

   Does this patient need central access?

   If a PICC line was planned anyway, would you forego the arterial line?

2. Does this patient need a rapid sequence induction? In general, do bariatric patients require a RSI? How about a sedated fiber-optic or glidescope intubation; 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?

3. 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?

Intraoperative Course

Answers

1. Monitoring can probably be accomplished noninvasively unless there are concerns about perioperative ventilation, accuracy of blood pressure monitoring by noninvasive methods, or frequency of metabolic monitoring. Placement of the line prior to induction is probably not necessary. The need for an arterial line will be dictated by the presence of the abovementioned comorbidities more than the procedure itself.

   Bony landmarks will be difficult to palpate for epidural placement, so patient self-report may be crucial. A sitting position is the preferred position because it will be easier to judge the midline according to a line imagined between C7 and the gluteal cleft.

   A central line will not be needed to manage the patient during surgery, but if venous access is difficult, then a central line or a PICC line may be a good idea for several days following surgery.

   If an arterial line were needed based on the comorbidities or concerns identified above, it could not be replaced by a PICC line (does not help with blood pressure monitoring, arterial blood gases, etc.), so a chronically ill patient would probably require both.

2. Signs and symptoms of reflux should be evaluated carefully; the majority of obese patients are NOT at increased risk for pulmonary aspiration. Nevertheless, induction of anesthesia with a 30° "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 video laryngoscopy without the need for fiber-optic laryngoscopy or "awake" intubation. They will, however, have a tendency to desaturate more rapidly.

3. 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.

Dosing of succinylcholine, if chosen?

Choice of inhalation agents – desflurane, sevoflurane, nitrous oxide?

Choice of opioids – fentanyl, sufentanil, remifentanil?

Choice of analgesic adjuncts – acetaminophen, ketorolac, dexmedetomidine, clonidine?

4. 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 100 mL/kg/min. Are you surprised? The surgeons begin to insufflate; the peak inspiratory pressure increases to 48 cm H_2 O and the rate is 24, for an $ETCO_2$ of 46. Is this optimal? Is it the best it can be? What can you do to make things better? Does it matter? If so, why?

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.

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.

The same considerations apply to the opioids. Remifentanil has the additional advantage of biotransformation via nonspecific plasma esterases. Fentanyl clearance is higher in obese patients, although comorbidities such as obstructive sleep apnea may affect dosing.

All of these drugs may reduce the amount of opioid required, which would be particularly beneficial for this patient population; this should include the use of local/regional anesthesia.

4. 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. Airway manipulation should begin with preoxygenation in the head-up position for several minutes, whether a rapid sequence induction is chosen or not [1]. 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 8 mL/kg (ideal body mass, around 70 kg would be my estimate) 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, insufflation of the abdomen, and the various positional changes required for the surgery, including steep Trendelenburg positioning.

   Not really surprised, this finding is actually encouraging. With significant ventilatory restriction, the breathing circuit in this circumstance may be ventilated far in excess of the patient. Secondly, there may be an increase in VO_2 as well as the VO_2/VCO_2 relationship. I would anticipate an increase in oxygen consumption and CO_2 production. There may be some further metabolic changes with regard to the respiratory quotient in morbidly obese patients, and their

5. What is your strategy for providing segmental analgesic blockade? Which agents would you choose? Would hydromorphone or fentanyl be an optimal choice? What effects on the CO_2 response curve would you expect these epidurally administered narcotics to have? Will it be worth it for this laparoscopic procedure?

minute CO_2 production may be increased at a baseline [2]. Normal minute volume for this age patient would be about 9–10 L/min; 100 mL/kg/min = 14 L/min.

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 plethysmographic 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).

5. The catheter tip should ideally be located at that spinal segment that innervates the dermatome in the middle of the surgical field. In this case, the patient will have a laparoscopic approach with surgical stimulation throughout the peritoneum and the diaphragm (therefore the shoulder, C3, C4, and C5.) A low thoracic epidural block should be sufficient to provide segmental analgesia from L1 to T6. Pain relief for the referred shoulder pain will be more difficult to provide via a regional technique because the shoulder pain is referred from the central portion of the diaphragm, embryologically innervated by mesenchyme contributed from C3, C4, and C5. Most effective regional techniques such as an interscalene or cervical epidural block are not worth the risk/benefit ratio because the pain will start getting better on the first postoperative day and is relatively easy to treat with systemic narcotics.

   A weak solution of bupivacaine (0.1–0.125 %) along with fentanyl or hydromorphone, when well administered, will both be effective for perioperative analgesia. You can make a theoretical case for fentanyl based on its greater lipophilicity; you can make a stronger case for hydromorphone based on its hydrophilicity and better "spread" in the epidural space. The CO_2 response curve has been shown to be blunted with neuraxially administered narcotics, and a patient like this, who will probably do very well, should nevertheless be monitored with an apnea monitor during the time of the infusion.

   "Worthiness" is really a matter of "best fit." Many patients will do perfectly well in the perioperative period with patient-controlled analgesia (PCA) because they are typically highly motivated, do not sit around feeling sorry for themselves, and mobilize early. For those patients who have particular issues with severe

Postoperative Course

Question

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?

underlying respiratory disease or for whom acute postoperative pain will likely affect their immediate post-op recovery, an epidural may very well be helpful with gas exchange, restoration of the FRC, and pulmonary toilet. Other techniques short of an epidural would include local infiltration at the end of surgery or transversus abdominis (TAP) or rectus sheath blocks as a single shot or with a catheter technique. Even with ultrasound guidance, placement may be challenging and results variable.

Postoperative Course

Answer

1. Unless there is recognized difficulty with ventilation and gas exchange during surgery, it is not likely that the patient will 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 usual criteria of a vital capacity maneuver of 10 mL/kg (lean body mass) and negative inspiratory force of greater than -20 cm H_2O.

   No special equipment should be required for extubation if intubation was routine; the same equipment (e.g., video laryngoscope) 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 positive pressure can be utilized.

   If the patient is in pain, that pain should be treated. This is best assessed if the patient is responsive and conversational. A rapid determination should be made as to whether the patient's airway can be manipulated (e.g., jaw lift, oral or nasal airway) that further emergence will be facilitated with this level of interventional support. If not, then a quick decision for re-intubation would be appropriate.

Additional Questions

Questions

1. What is the mechanism of insulin resistance in childhood obesity? Are 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?

2. Should morbidly obese patients with obstructive sleep apnea undergo tonsillectomy/adenoidectomy prior to bariatric surgery?

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?

Additional Questions

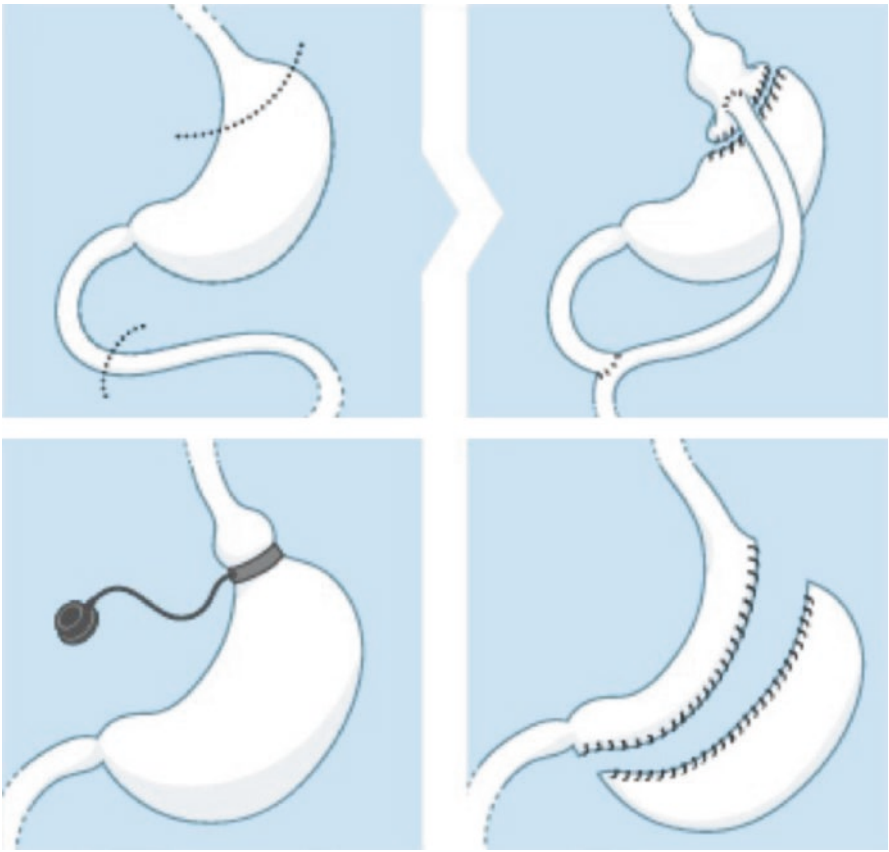
Answers

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 [3]. More than 50 % of obese adolescents in the United States exhibit insulin resistance [4].

2. There is no a priori reason to suppose that airway surgery prior to bariatric surgery decreases the risk of perioperative airway obstruction, despite the fact that OSA is six times more likely in the morbidly obese patient. One could look at it from the opposite perspective – the bariatric surgery may decrease the risk of OSA, without operating on the airway itself. What is important is to evaluate the patient for OSA prior to the bariatric surgery and design the perioperative care around those findings [5, 6]

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. RYBG is the most commonly performed bariatric procedure. A small proximal gastric pouch is created with an anastomosis to a Roux limb (food collection pouch) of jejunum that bypasses 75–150 cm of small bowel. This arrangement restricts intake, as well as limits digestion and absorption of ingested nutrients. In addition, it limits gastrointestinal hormone secretion. Adjustable gastric band is a purely restrictive procedure that involves placing a tight, adjustable prosthetic silicone band around the gastric inlet. Sleeve gastrectomy is a technically easier restrictive procedure that involves a partial gastrectomy and creation of a tubular stomach from a portion of the greater curvature.

4. What specific concerns do you have about positioning and how would you manage them?



From Chatterjee [7]

4. The supine and Trendelenburg positions should be avoided or minimized because of the drastic reduction in lung volumes from cephalad displacement of the diaphragm. These positions can also result in increased venous return, cardiac output, pulmonary blood flow and volume, and arterial blood pressure. Elevating the upper body 25–30° in a semi-recumbent or reverse Trendelenburg's position improves pulmonary mechanics. The prone position is usually well tolerated by obese patients provided the abdomen is not compressed although an extremely large pannus, even if freely hanging (e.g., on a spine fusion or Jackson table), may place undue traction on the mesenteric circulation. The lateral position is typically well tolerated.

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Chapter 25
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? 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?

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?

Preoperative Evaluation

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. 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 acidemia. 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 dilute 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 2-3DPG. 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 $\text{GFR mL/min/1.73M}^2 = 0.55 \times \text{Length(cm)/plasma creatinine}$.

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 seconds, and impaired skin turgor.

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 hydronephro-

Intraoperative

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, and 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?

sis and cause kidney damage because chronic inflammation of the glomeruli and tubules results in scarring. If infection is ongoing during the procedure, the infection may spread into the bloodstream causing bacteremia and septic shock under general anesthesia or in the immediate postoperative period.

Intraoperative

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 preexisting elevation of serum potassium and metabolic acidosis can be aggravated by hypoventilation, respiratory acidosis, and laryngospasm that could accompany inhaled induction. 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 as 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. Caution is advised, and measures to mitigate the increased potassium by assuring adequate ventilation and preparing to give calcium, insulin, and glucose should be made. 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?

Emergence

Question

1. The anesthetic vapor has been off for 30 min, 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 h 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–T9. 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 Transverse Abdominis Plane (TAP) block with continuous local anesthetic 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.

Emergence

Answer

1. 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, brain stem ischemia/anoxia, or drug-induced effects, e.g., anticholinergic 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, brain stem 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?

Additional Questions

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; 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, and impaired renal function, 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 26
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- = 40, 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. 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?

5. Why does K^+ depletion cause alkalosis?

Preoperative Evaluation

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 mm Hg 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.

4. The most common cause of alkalosis is acid loss through vomiting and/or NG suction. H_2 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.

5. 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.

Intraoperative

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 pre-existing 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?

Intraoperative

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 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 chloroprocaine would be considered a better choice in a patient with compromised cardiac performance. Local anesthetics are weak bases. 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 $pH = pKa + \log([\text{base}]/[\text{cation}])$. Since it is the unionized base form of local anesthetics that determines the onset of the drug, the more acidic the patient is, the slower the onset will be.

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?

Postoperative

Quesitons

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. 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 hypo-osmolar (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

Answers

1. Acidosis can have multiple effects. With pH <7.22, myocardial and smooth muscle depression can result in decrease in 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-Hgb dissociation curve) and add to acidosis. The response to vasopressors and inotropes is impaired in this setting. Pulmonary 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-Hgb 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. Your patient is not waking up after completion of the surgery. What possible effects of acidosis could be a cause of delayed emergence?

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?

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, monoquaternary 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

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.

Suggested Readings

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Chapter 27
Genitourinary Disorders

Joseph P. Cravero

A baby in the first day of life presents for closure of an exstrophy of the bladder. The product of 36 weeks gestation, he is 2.3 kg, with a preoperative hematocrit of 56 %, BP 65/35 mmHg, HR 130 bpm, and RR 24/min.

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Preoperative Evaluation

Questions

1. Is this baby premature? 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?

Preoperative Evaluation

Answers

1. Yes, because the infant was born before 37 weeks gestation. Infants born between 36 and 37 weeks gestational age are categorized as borderline premature. Those born between 31 and 36 weeks GA are considered moderately premature and those born between 24 and 30 weeks gestation are considered severely premature. Small gestational age babies weigh less than 2.5 at birth. The more premature the infants are, the greater the risk for perioperative complications. Premature 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 linearly between 36 and 28°C, therefore increasing oxygen consumption, which can lead to hypoxemia, acidosis, apnea, and respiratory depression. Premature 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 premature 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. Premature 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 successful epidural analgesia is provided and minimal opiates are 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 work-up) should be evaluated to ensure the anatomy is normal.

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 next? If the case will take 8 h, 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?

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?

2. Yes; 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 h) 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].

Intraoperative Course

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 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 deleterious acidosis and impaired cardiovascular performance.

2. No, 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

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 CO_2 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. 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?

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.

Postoperative Course

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), 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), is breathing regularly, and has an empty stomach. 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. 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

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?

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?

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, CT, or MRI of the spine reveals no cord tethering [4].

Additional Questions

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-1, 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. 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 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 28
Bone and Connective Tissue Disorders

Joseph P. Cravero

An 11-year-old boy with achondroplasia presents for Ilizarov leg lengthening. His admission VS are HR 92 bpm, BP 110/80 mmHg, and RR 16/min. Hct 34. No prior history of surgery.

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Preoperative Evaluation

Questions

1. The patient has a large head. Is this normal? Why/why not? What might it signify? How could you evaluate the presence or absence of hydrocephalus? Is this important? Why/why not? How would it alter your anesthetic management if he had normal pressure hydrocephalus? What if this patient had progressive difficulty walking; what would this suggest? How would you evaluate it? How could this be distinguished from his short-limb difficulties?

2. What are the anesthetic implications of his bony abnormalities? Is this more likely to occur cranially or caudally? Is the spinal defect only a mesodermal defect, or does it involve neuroectoderm as well? The parents want to find out about a spinal; what would you tell them? Is this likely to be easy in this patient (aside from psychological issues)? What level would you choose to perform the block?

3. 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?

Preoperative Evaluation

Answers

1. Yes, a large head is normal in achondroplasia patients. While macrocephaly is typical of the syndrome, there is a higher incidence of hydrocephalus due to foramen magnum stenosis. Various imaging modalities will show dilated ventricles. If found, foramen magnum stenosis predisposes to brainstem compression and respiratory arrest. The absence of hydrocephalus or normal pressure hydrocephalus does not rule out the presence of foramen magnum stenosis, and the risk of brainstem compression remains unchanged. This may be due to spinal cord compression from spinal canal stenosis. Imaging of the thoracic and lumbar spine would help with a more specific diagnosis. Difficulty walking owing to short limbs is not associated with neurological symptoms. However, spinal cord compression is associated with lower extremity changes in sensation and motor function. Bladder and rectal sphincter dysfunction may also occur with thoracolumbar stenosis.

2. 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. Lumbar spinal anesthesia has been performed safely in patients with spinal stenosis and vertebral malalignment but is associated with a potentially higher incidence of direct neural injury by the block needle or as a result of epidural bleeding. Therefore, it is advisable to perform spinal anesthesia if it offers distinct advantages over general anesthesia. In this particular patient, there is no obvious benefit of spinal anesthesia over general anesthesia. It is likely to be difficult due to the lordotic deformity and vertebral malalignment. I would choose a site that is not involved with lordosis, kyphosis, stenosis, or vertebral malalignment after reviewing the imaging studies and documenting any preprocedural neurological deficit. L5–S1 may be safe because the deformities are usually located above this level. Nevertheless, the risk of cord compression is possible with the use of a large volume of local anesthetic and epidural venous bleeding.

3. This patient does not need soft tissue films of the neck. Achondroplasia is a disease of underdevelopment of bone. Midface hypoplasia is expected with this disorder. MRI of the airways can be helpful to determine the extent of airway involvement. As a disorder of bone underdevelopment, achondroplasia can be associated with dysgenesis of the odontoid process, resulting in atlanto-axial instability. The presence of atlanto-axial 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, now readily available, 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 midface hypoplasia, micrognathia, or macrognathia and prepare the patient and the equipment accordingly.

Intraoperative Course

Questions

1. What kind of monitoring (aside from routine) would you choose for this case? Why/why not? Does this patient need an arterial line? Why/why not? A CVP? A precordial Doppler? Why/why not?

2. Your colleague walks by the room, takes a look in as you are placing monitors, and suggests that you consider a rapid sequence induction because the patient is a preteen, and they all have full stomachs. What do you think? How would you decide which approach to use? Would you use succinylcholine for a rapid sequence induction? Why/why not? Any association with malignant hyperthermia? The patient can extend his neck through an arc of 15–20°. Does this influence your choice of anesthetic induction techniques? How will you do an awake intubation? Would a light wand be just as good as a fiber-optic bronchoscope? Would an anterior commissure scope work just as well for this problem? What are the advantages and disadvantages of each in patients with odontoid hypoplasia and decreased neck extension?

Intraoperative Course

Answers

1. I would monitor the patient with standard noninvasive monitoring. I would not use an arterial line or a CVP catheter for this low-risk procedure. I would, however, use a precordial Doppler because of the chance of significant air or fat embolism with this procedure. I would not approach this as a rapid sequence induction. Achondroplasia is associated with a difficult tracheal intubation due to midface hypoplasia and micrognathia. Minimal manipulation of the cervical spine is advisable because of the potential of atlanto-axial joint instability. Rigid videolaryngoscopy or flexible fiber-optic laryngoscopy may be necessary to secure the airway with minimal cervical spine manipulation. Careful evaluation of the airway during the preoperative visit is crucial. In the operating room, airway access could be assessed after providing sedation in a spontaneously breathing patient.

2. An electively prepared patient should not ordinarily need a rapid sequence induction, and I would not use succinylcholine. Succinylcholine causes fasciculations that may produce atlanto-axial instability and spinal cord compression. Achondroplasia is also associated with hypotonia, and the use of succinylcholine may produce clinically significant hyperkalemia. Even if the patient demonstrated good range of motion of the neck, it would not necessarily be reassuring with regard to routine laryngoscopy and intubation.

   Intubation of trachea in a conscious child is performed after effective intravenous sedation, with incremental doses of a short-acting opioid such as fentanyl 0.5 mcg/kg IV repeated at 5 min intervals to effect supplemental IV non-opioid sedation with midazolam in 25 mcg/kg increments, and should achieve a state of anxiolysis, antegrade amnesia, and sedation while maintaining the patient's ability to respond to verbal commands. The airway mucosa of the nares, oropharynx, and supraglottic area can be anesthetized with topical local anesthetic and/or glossopharyngeal and superior laryngeal nerve blocks to markedly reduce the discomfort associated with instrumentation and reduce the requirement of intravenous sedation. Lidocaine 1 % and cocaine 4 % spray are effective topical anesthetics for the mucosa. The advantage of cocaine over lidocaine is that it produces vasoconstriction and minimizes bleeding with turbinate instrumentation. The oropharynx is anesthetized with an intraoral glossopharyngeal nerve block with submucosal infiltration into each palatoglossal fold. The supraglottic mucosa, including the epiglottis, is anesthetized with bilateral superior laryngeal nerve blocks. This is accomplished by percutaneous injection of 2–3 mL of lidocaine 1 % at the junction of greater cornu of the hyoid bone and thyroid cartilage. Alternatively, the supraglottic mucosal anesthesia is achieved by placing lidocaine-soaked gauze pad into the pyriform fossae.

3. After the first 3 h of the case, six transfemoral Ilizarov struts have been placed; the pulse oximeter reads 94 % on 50 % nitrous and oxygen. What do you make of this? Why/why not?

   There are at least three more hours of distraction to go. What will you do? Is PEEP effective for fat embolism syndrome? How would you choose the most effective PEEP level? What are the problems with PEEP? Any particular problems with this patient? What are they? Is this different than any other patient?

4. After the first 4 h of the case, you note that the current nasopharyngeal temperature is 39.7°; the heart rate, which started at 100/bpm, is now 140/bpm. What do you think is going on? What else could it be? How will you evaluate? Is there any way to test? Would you place an arterial line at this point? Give dantrolene prophylactically? What if the end-tidal CO_2 was 43? 57? What if you increased the minute ventilation by 100 %, and the end-tidal CO_2 decreased from 53 to 50? What could this be? Why?

A light wand can be used for blind oral intubation while the neck is stabilized. An anterior commissure scope may not be useful because of restricted neck mobility during stabilization. Often, excessive neck extension is required to visualize the glottis with an anterior commissure scope. The advantage of the light wand is it allows blind oral intubation without the need for excessive neck extension and flexion movement. The disadvantage is that advancing of the endotracheal tube may require flexion of the neck that can be a substantial risk in the presence of atlanto-axial instability. The advantage of an anterior commissure scope is that it facilitates visualization of an anteriorly located larynx. The disadvantage is that it requires hyperextension of the neck.

3. The potential for fat embolism should be considered. Optimize ventilation. Increase the inspired oxygen to maintain oxygen saturation above 95 %. Notify the surgeon. Place an arterial cannula to monitor arterial blood gasses. PEEP is an effective strategy for fat embolism syndrome. Optimal application of PEEP involves a stepwise increase of PEEP to optimal arterial saturation without compromising hemodynamics. It may compromise right ventricular function by increasing intrapulmonary pressure and left ventricular function by interventricular septal shifting. The hemodynamic effect of PEEP could be exaggerated in this patient due to scoliosis and restricted intrathoracic capacity.

4. With these findings, malignant hyperthermia must be included in the differential diagnosis list. Passive hyperthermia from active patient heating and the use of insulating drapes are also likely. Other less likely causes are mismatched blood transfusion reaction, drug-induced pyrexia, a cerebral bleed or thermogenesis from fat embolism, and an allergic reaction. Monitoring closely for a progressive rise of end-tidal carbon dioxide ($ETCO_2$) concentration is crucial, as is obtaining a venous or arterial blood gas. Active cooling of the patient should be started. Depending on the clinical context and monitoring results, I might place an arterial line at this point, particularly if the tachycardia and elevated $ETCO_2$ were out of proportion to efforts to deepen the anesthetic and increase minute ventilation or were accompanied by muscle contracture. I would administer dantrolene if the arterial blood gas analysis reflects uncompensated metabolic and respiratory acidosis and a wide A-a oxygen gradient despite increased minute ventilation. The values presented are not suggestive of malignant hyperthermia. These low values could be due to increased dead space ventilation. However, an inability to reduce $ETCO_2$ despite doubling the minute volume is suggestive of an inability to compensate for excessive production of carbon dioxide. This may suggest MHS. If the hypercarbia is due to increased dead space, it would decrease with hyperventilation.

Postoperative Course

Questions

1. 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? Why? Do you think this patient would meet spirometric or inspiratory force criteria for extubation? Why/why not? Is there any reason for him to have weak muscles? Should he be extubated in the operating room?

2. Is this patient at any higher risk for PCA methods of pain management? Would a preoperative blood gas have been likely to reveal this problem? How would you write the orders for his PCA?

Additional Questions

Questions

1. A 1.5-year-old with osteogenesis imperfecta is on the add-on list for open reduction internal fixation of a femur fracture. His preoperative temperature is 39.4°, and he has no signs of a URI. How will you evaluate this? Could this be a urinary tract infection? Is this temperature normal? Why? 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 management? Hematologic support? Does he need platelets? What if he is 6 kg; how will you manage his fluid therapy? Would you use succinylcholine to gain rapid control of the airway?

Postoperative Course

Answers

1. 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. If the intraoperative course was uneventful (i.e., if the above events did not occur), then careful extubation after meeting standard criteria would be reasonable. 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 will use the following criteria for extubation and 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.

2. Yes. Chronic respiratory insufficiency with carbon dioxide retention may accompany patients with severe scoliosis and/or kyphosis as well as hypotonia. A preoperative arterial blood gas may reveal hypercarbia with a relatively normal pH as a result of a compensatory metabolic alkalosis, although the primary process with be the respiratory acidosis. A more conservative PCA dosing regimen may be called for, as well as nurse-controlled analgesia if the patient is cognitively impaired.

Additional Questions

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. 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

What are the drawbacks? One of your partners suggests that this patient needs an arterial line instead of a blood pressure cuff. Do you agree? Why or why not?

2. 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

3. 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?

bleeding. Fluid replacement requires careful management because some patients with OI may experience hyperhidrosis and may need addition fluid replacement of insensible losses. I would not use succinylcholine because succinylcholine-induced contractures increase the risk of fracture of the brittle bones. 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.

2. Radial aplasia and Hallermann-Streiff syndrome: Narrow upper airways, obstructed nares, micrognathia, tracheomalacia, obstructive sleep apnea and cor pulmonale, and structural heart disease. Birdlike 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: It is 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.

   Thrombocytopenia-absent radii syndrome (TAR syndrome) similar to Fanconi anemia 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), and 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).

3. Patients with rickets usually have hypocalcemia that may cause arrhythmias and potentiate non-depolarizing muscle relaxants. Patients with untreated rickets may have soft bones, which predispose to fracture with mild pressure. Poor

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?

4. With what degree of curvature would you counsel a family for postoperative mechanical ventilation? Why? How will this influence your choice of monitors? At what degree of curvature would you expect to have to use a pulmonary artery catheter? Why?

5. A 16-year-old is scheduled for a sagittal split osteotomy and mandibular advancement; she has severe juvenile rheumatoid arthritis. What are your airway considerations? Why do these patients have airway problems? Are these mainly at the level of the mandible, the temporomandibular joint, or the larynx? Is there any cervical spine problem?

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 non-depolarizing 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 (vitamins D3, D2, 25(OH)D3, 1,25(OH)2D3, 24,25(OH)2D3).

4. A curve of 65° or greater increases the chances for postoperative mechanical ventilation because of a significant decline in ventilation-perfusion matching in addition to an increased dead space to tidal volume ratio (>0.6). If the scoliosis curvature progresses beyond 65°, adequacy of ventilation should be monitored with serial arterial blood gas analysis. A pulmonary artery catheter or transesophageal echo monitoring may be required in the presence of a curve of 90° or greater, clinical evidence of right ventricular compromise, or the presence of preoperative hypoxemia and carbon dioxide retention. The use of TEE in the postoperative period can be very useful to monitor right ventricular function during positive pressure ventilation and to guide fluid and inotropic therapy.

5. In advanced or severe rheumatoid arthritis, the airway can be compromised. The earliest manifestation is micrognathia due to ankylosis of the temporomandibular joints. Other manifestations are ankylosis of the cricoarytenoid joint and a small glottic aperture, cervical spine ankylosis, flexion deformity, and subluxation of the atlanto-axial joint. A small mouth opening, limited neck range of motion, severely restricted neck extension, and small glottis due to restricted movement of cricoarytenoid joint may be present in varying combinations and severity. Airway difficulties should be expected at all levels of the upper airway passages. Flexion deformity of the cervical spine and subluxation of the vertebral bodies and atlanto-axial joint should be anticipated.

Chapter 29
Skin Disorders

Robert S. Holzman

A 4-year-old is scheduled for a circumcision for recurrent balanitis as well as dental restorations. He has junctional epidermolysis bullosa, Herlitz type 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.

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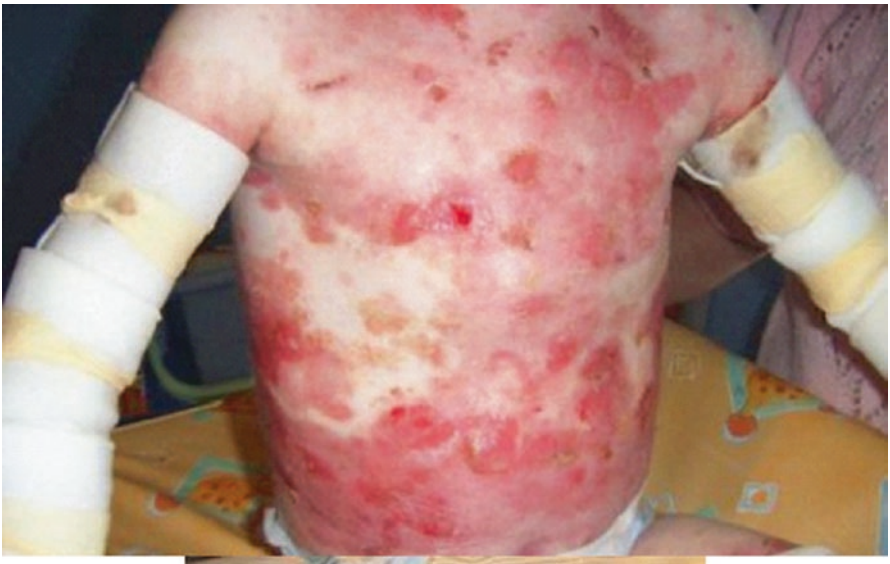

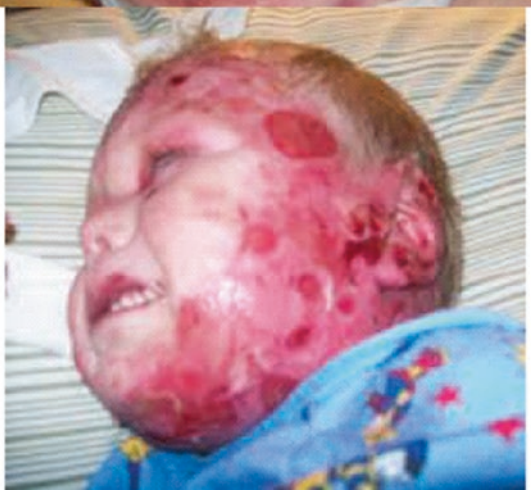

Preoperative Evaluation

Questions

1. What is the basic histological lesion? Are there different forms of epidermolysis bullosa? What are the broad categories? Of what medical, clinical, and anesthetic significance are the associated comorbidities?

Preoperative Evaluation

Answers

1. There are four basic types of epidermolysis bullosa (EB) that differ phenotypically, genotypically, and histologically [1]. While milder forms of EB simplex will commonly present in late childhood or even during adult years, pediatric anesthesiologists will encounter earlier onset and more severe forms such as junction and dystrophic EB and Kindler syndrome, with multiple skin levels of

2. With specific reference to the planned surgery, what unique areas are essential to your preoperative discussion with your surgical colleagues? Nursing colleagues? PACU nurses?

blister formation. Mechanically fragile skin with easy blistering or erosions is characteristic. Nails may be dystrophic or absent, and granulation tissue may be exuberant especially periorally. In general, the more severe and widely distributed the blistering on the skin, the more likely it is for extracutaneous sites (trache, esophagus, ocular, corneal) to be involved. Dental enamel hypoplasia is seen in all subtypes of junctional EB. There is an increased risk (44 % by age 55) of developing squamous or basal cell carcinomas and even melanomas [1]. The Herlitz type of junctional EB, which is what this patient has, is present from birth and typically involves the upper airway as well as all skin surfaces. The severity of intraoral blistering (and their inevitable healing pattern which ultimately results in chronic scarring) may eventually narrow the mouth and the laryngeal inlet and also result in ankyloglossia, similar to the glossoptosis in Pierre-Robin syndrome. The cumulative risk of laryngeal stenosis or stricture is 40 % by 6 years of age. Fifty percent of patients die within the first 2 years of life [1]. Children are also often anemic because of the constant injury and healing process and the skin's metabolic utilization of iron.

Essential planning for the case [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.
> Preoperative assessment should include the likelihood of anemia, dehydration, electrolyte abnormalities, and poor nutrition.

2. 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 sterile synthetic nonadhesive hydrocolloid dressings (e.g., Duoderm®) is a must.

   For an intraoral procedure like the dental rehabilitation, whether endotracheal intubation, laryngeal mask, or natural airway techniques are chosen, the airway will be shared by the anesthesiologist and dentist/surgeon for almost the entire case, and it is likely that if the tube is not secured with tape, the anesthesiologist will be holding it and moving it from side to side. Extra care must be taken around carious teeth because of their poor enamel. A well-lubricated, undersized endotracheal tube should be chosen if tracheal intubation is contemplated. If using an LMA, movement should be gently applied with as little manipulation as possible because the inflated cuff of the LMA is constantly in contact with oropharyngeal soft tissue.

3. What is the significance of his treatment with anabolic steroids?

4. How will you evaluate the airway? What are the risks of airway instrumentation/manipulation?

Intraoperative Course

Questions

1. Does this patient need an IV in order to facilitate a "quiet" induction? How will you accomplish this?

2. The medical student suggests an IM preinduction 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?

3. 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. Obviously, transdermal delivery systems are contraindicated; parenterally administered anabolic steroids are acceptable. Orally administered anabolic steroids may have associated hepatotoxicity. The risks of anabolic steroids include personality (i.e., aggressiveness) alterations, hypertension, hypercholesterolemia (paradoxical), feminization as a result of suppression of natural testosterone levels, focal segmental glomerulosclerosis, and an increased oxygen consumption (therefore an increased carbon dioxide production and minute ventilation requirement under anesthesia) in the setting of an already increased metabolism from skin turnover.

4. Airway evaluation begins with physical diagnosis; anticipated problems would include perioral blisters, scarring, microstomia, intraoral blisters and scarring, restricted head and neck motion, esophageal strictures/stenosis, and laryngotracheal stenosis. Any of these can be made worse with airway manipulation following the procedure [3].

Intraoperative Course

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. With skin blistering, scar formation, and possibly pseudosyndactyly in certain forms of EB, the parent will be the best guide for IV placement. 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, it is a perfectly acceptable idea as long as the perianal area is carefully examined with regard to blistering and skin irritation [4]. The usual induction dose (methohexital) or 25–30 mg/kg (10 % solution) may have to be increased if the patient is on concurrent drug therapy such as anabolic steroids.

4. You proceed with a 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?

7. How will you monitor this patient? For each "routine" monitor, how are your choices influenced by the underlying disease?

8. What are your positioning considerations?

9. If a "natural" airway is chosen, how will you maintain anesthesia?

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. Liberal lubrication should be generously applied between the cuff of the mask and the skin of the face. "Shearing" types of movements to reseat the mask must be avoided.

5. As a result of the EB, the small mouth and any intraoral lesions will restrict access of any visualizing device except a fiber-optic 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 video laryngoscope like a Glidescope. A flexible fiber-optic scope would be needed for a more limited mouth opening, as well as for any use of a LMA. Of course, limited mouth excursion would limit the access of the dentists as well.

6. Yes, although not without careful discussion with the surgical team. A spontaneous breathing technique, with ready availability of suction, can be chosen, and a pharyngeal screen can be placed for debris [4]. The decision is subject to individual assessment, based on the severity of the EB with its cutaneous and extracutaneous manifestations and a candid discussion about risk/benefit options with the surgical team and the parents. Although progressive scarring of the esophagus and trachea is a well-known association of the Herlitz form of EB, it does not seem to be related to airway manipulation directly but rather to the duration and severity of the disease.

7. 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 around a finger after application of a transparent dressing/plastic wrap or the adhesive cutoff. 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].

8. 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.

9. Any spontaneous breathing technique is fine; all have their advantages and disadvantages. Potent inhalation agents can be utilized, but will not be able to be scavenged adequately during the dental procedure. A total intravenous technique can be utilized with various combinations of propofol, propofol-remifentanil, dexmedetomidine, or ketamine.

10. The surgeons would like to do a penile block for the circumcision. What are your recommendations?

11. The dentists would like to inject local anesthetic with a vasoconstrictor to control bleeding and provide for analgesia post-op. What are your recommendations?

Postoperative Course

Questions

1. When would you extubate? Deep? "Awake?" Narcotics or dexmedetomidine?

2. You extubate, and the patient is stridorous in the OR. What will you do? How will you evaluate? Should you do a rigid bronchoscopy?

3. What concerns do you have for the prophylaxis management of emergence delirium?

10. 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).

11. Same considerations apply – as long as the injection is not superficial (i.e., minimal chance of raising a wheal), then it should be fine.

Postoperative Course

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 avoid struggling. This could be accomplished with narcotics and/or dexmedetomidine.

2. Depending on the degree of stridor, management will likely be medical, with standard nebulized racemic epinephrine. Preexisting tracheal stenosis might predispose to post-intubation croup, but unless the patient was in extremis, one should be loathe to re-instrument the airway in this circumstance. 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.

3. As before, it is crucial to manage pain and anxiety as well as any emergence delirium effectively in order to minimize skin trauma as a result of patient movement or the need for restraint. Family interaction as well as adequate pharmacology will be key, although clearly the patient is familiar with how to behave himself in order to avoid skin trauma.

Additional Topics

Questions

1. Anesthesiologists usually encounter the skin as the organ 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 its functions in the context of the following:

   Protection
   Immunology
   Fluid and protein balance
   Thermoregulation
   Sensation
   Metabolism
   Social interaction
   Storage

2. An otherwise healthy 5-year-old boy with vesicoureteral reflux was started on Bactrim two 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-gauge IV.

   How will you evaluate and manage his airway?
   What fluid/electrolyte problems can you anticipate?
   Nutritional problems?
   What is the nature of this problem? How long will it take to resolve? Can it recur? Is this problem different than erythema multiforme?

Additional Topics

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 and 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. 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) [5]. 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.

   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 avoid a general anesthetic if possible and consider a TIVA technique for PICC line placement, perhaps in the radiology suite.

   Weeping of proteinaceous fluid from affected sites and transdermal loss of free water and blood may result in hypoproteinemia and volume contraction as well as anemia for acute (blood loss) as well as chronic (bone marrow depression) reasons. Isotonic intravenous therapy and/or blood, fresh frozen plasma, platelet, or other volume expanders such as albumin need to be considered. Calorie stress during massive tissue injury and repair as well as the inability to eat will be the likely problem here and will require nutritional support during the acute phase of

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?

illness until the acute phase is over. This may require enteral (gastrostomy) or parenteral hyperalimentation.

It is an immune-complex binding disorder that results in the acute onset of painful skin lesions, fever, sore throat, and conjunctivitis. It is most commonly associated with sulfonamides, allopurinol, and anticonvulsants. Systemic steroids are the typical pharmacological treatment that usually work acutely but are associated with higher mortality. It is typically a first-exposure medication reaction. There may be cross-reactivity with structurally similar drugs, and the reaction can occur with reexposure. Erythema multiforme is more typically associated with herpes virus, and it also can reoccur [6].

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 more rare [7].

In addition to the physical exam, 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 to subluxation. The tendency toward easy bruising and bleeding is more a matter of vascular fragility in the absence of coagulopathy. The vascular type of EDS is more associated with cardiac and vascular abnormalities like valvulopathies and aneurysms. Generalized hypotonia is a consideration and should be assessed carefully when planning the need for muscle relaxants or respiratory support perioperatively [3].

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 predispose to 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. [7]. There is a more significant risk of such events when intravascular monitoring is indicated so 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, and early mobilization is key to preserving muscle strength in the perioperative period [3].

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Chapter 30
Inborn Errors of Metabolism

Robert S. Holzman

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, and RR 36.

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Preoperative Evaluation

Questions

1. What problems would you expect? Why didn't this show up earlier? Is this typical? Atypical?
2. What phy.sical characteristics will you see on exam that are of concern to you as an anesthesiologist?

3. What physiological derangements should you anticipate?

4. Any particular considerations for intravenous fluid management?

5. Does the hepatomegaly occur together with splenomegaly and are there any specific consequences you would have to worry about?

Preoperative Evaluation

Answers

1. and 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 respiratory 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 may also have impaired function of platelets and neutrophils and so may be subject to bacterial infection or have a prolonged bleeding time [1].

3. Hypoglycemia, lactic acidosis, and elevation of free fatty acids. Children should not be fasted for more than 3 h 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.

4. 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 given and glucose checked frequently.

5. The spleen is not enlarged. Nevertheless, the increase in abdominal girth is a consideration for mask induction, adequacy of respiration especially spontaneous respiration, and the influence the enlarged liver has on gastric emptying.

Intraoperative Course

Questions

1. Induction method?

2. Choice of monitoring?

3. How would you constitute intravenous fluids? Why?

Perioperative Management

Questions

1. How should glucose be managed postoperatively and how will you monitor the effectiveness of your strategy?

2. The patient is having difficulty with control of recurrent epistaxis after you placed an NG tube following induction. They are waiting for discharge as a day surgery patient. Should you delay discharge? Is this normal? Can you reassure the parents that the epistaxis will resolve? Should the nose be packed?

Intraoperative Course

Answers

1. Because of the protuberant abdomen, strong consideration should be given to a rapid sequence induction with endotracheal intubation even for a "simple" case like a liver biopsy. For any case lasting longer than an hour, a second IV large enough to withdraw blood samples or an arterial line should be placed for frequent glucose monitoring. Alternatively, point of care testing methods may be used when appropriate for minor cases or outpatient surgery. The key is the frequency of glucose monitoring and the plan for administration of complex carbohydrates as well as simple glucose for rescue [2].

2. Invasive monitoring is not needed for this case, but an extra line to draw blood from would be a convenience. For glucose monitoring, finger-stick point of care testing may be used as well, but I would reserve this for minimally invasive cases of short duration.

3. A 10 % dextrose infusion should be administered at 1.5–2X maintenance, with electrolytes as needed. D10 NS would be a good choice for most patients; nephropathy associated with GSD Type I will influence this choice as well. Lactated Ringer's should be avoided because of the lactate. The glucose infusion should be titrated to a blood glucose level greater than 70 mg%.

Perioperative Management

Answers

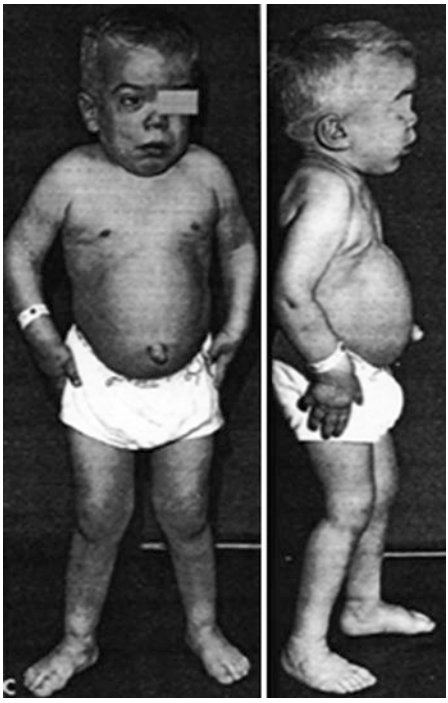
1. Glucose-containing IV fluid should be continued until the patient is taking POs and is checked for an acceptable glucose level. The progression of weaning from IV to PO glucose-containing fluid should be slow, and the glucose should probably be rechecked, making sure that the parents know that they should be checking the glucose at home as well.

2. Prolonged bleeding is not uncommon in these patients; the reasons are unclear. Normoglycemia will actually help the bleeding tendency, but nasal trauma should be ruled out by a nasal exam and the patient closely followed in the PACU until the bleeding stops.

Additional Questions

Questions

1. A 5-year-old with Hurler syndrome needs a recurrent umbilical hernia repaired. He cannot come in for evaluation prior to surgery, but will be coming in early on the day of surgery for his pre-op work-up. He is not yet in school and is 24 kg.



   (a) What is the basic defect? Are there any significant comorbidities?
   (b) How do they affect your anesthetic plan?
   (c) What is the significance of the protuberant abdomen and enlarged liver and spleen?
   (d) How does this disease affect the cardiovascular system?
   (e) Do you think this patient has restrictive or obstructive lung disease? Should PFT's or arterial blood gases be obtained preoperatively?

2. 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?

Additional Questions

Answers

1. Lysosomal Storage Disease: Hurler Syndrome
   (a) 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.
   (b) 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 [3].
   (c) The liver and spleen 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.
   (d) 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.
   (e) 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. 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 if untreated may lose as

3. An 8-year-old with a history of familial hypercholesterolemia type II B is scheduled for a portacaval shunt as a last-ditch treatment effort to lower his cholesterol; he is treated with cholestyramine and clofibrate. Dietary treatment has been of no benefit. His preoperative ECG shows Q waves in leads II, III, and avF and leads V4–V6. What are your thoughts about management? Do you think he is homozygous or heterozygous for this disorder? What difference does it make? What if his brother was heterozygous?

4. A community pediatric dentist calls to ask your advice about a 4-year-old who has been 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?

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 has 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 pro-convulsant 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 B_{12} deficient.

3. Familial hypercholesterolemia is inherited as a dominant disorder. Patients who are heterozygous often do not develop coronary atherosclerosis until the third or fourth decade of life. By age 60, 85 % of these individuals have suffered an MI. The rare (1:1,000,000) patient who is homozygous for the abnormality in lipoprotein metabolism has a much more severe form of the illness. These patients have plasma cholesterol levels of >600 mg%. Dietary management is of little or no help in lowering the cholesterol. Cholesterol-lowering medications also offer little benefit. Coronary disease is evident by 10 years of age, and most patients die by 30 years of age. Therapies such as plasmapheresis, ileal bypass surgery, and portacaval shunt placement have had some success. Liver transplantation has helped several patients. There is a possible genetic treatment involving transfecting the patient's own hepatocytes with a gene for the missing receptor.

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) [4, 5]. 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.

5. An 8-year-old 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. 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? How will you "design" your anesthetic technique?

5. 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. Similar to many other metabolic disorders, prolonged fasting will tend to make MELAS syndrome worse, so the NPO time should be minimized or supplemental glucose provided. Lactated Ringer's may not be 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, the most common being total intravenous anesthesia although the full range of anesthetic medications have been utilized. Perioperative hyponatremia and hyperkalemia have been noted [6, 7].

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Chapter 31
Allergy/Immunology

Robert S. Holzman

An 8-year-old 25 kg boy is scheduled for functional endoscopic sinus surgery. He has a history of common variable immunodeficiency (CVID) with hypogammaglobulinemia, was diagnosed at 4 months of age with recurrent sinus and respiratory tract infections, and was seen in the emergency department 2 weeks ago requiring an epinephrine injection for moderate respiratory distress. He has a productive cough constantly and bronchiectasis by x-ray. His medications include Singulair, Atrovent, and an infusion of intravenous immune globulin G every 3 weeks. Vital signs are BP 90/60 mmHg, P 100 bpm, R 18/min, and T 37 °C, and his Hb is 13 gm/dL.

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Preoperative Evaluation

1. Is this a B-cell or T-cell deficiency, or both? Of what significance is the comparison? What is the immune globulin that is deficient?

2. What is bronchiectasis? Of what importance is it to anesthetic management? Of what significance is the recent visit to the E.D? Do you desire additional information regarding his chronic lung disease? Specifically? Why? Is a chest x-ray necessary? Why/why not? Do you need to identify which inhaler he is using? Why? Relevance?

3. What effect does his lung disease have on his cardiovascular system? How will you evaluate this preoperatively? Why? What are the anesthetic implications? Are there tertiary effects of such cardiac disease on other organ systems?

Preoperative Evaluation

1. B cells are functionally deficient rather than decreased in number. Progression through the normal stages of B-cell development into memory B cells appears to be delayed or blocked. In addition, non-B cells may have abnormalities as well, for example, low numbers of CD4+ cells and high numbers of CD8+ T cells. Natural killer (NK) cells are typically lower than normal. CVID is a pan-hypoglobulinemia; there are decreased levels of IgG and IgA. IgM antibodies are decreased in about 50 % of affected patients as well. Recurrent infections are the typical presentation, with *H. influenza*, *S. pneumoniae*, and *S. aureus* the frequent causes. Respiratory tract infections are most common, along with sinuses, eyes, skin, and gastrointestinal manifestations. Bronchiectases are the anatomic result of severe recurrent pulmonary infections.

2. Bronchiectasis is characterized by weakness and loss of cartilaginous integrity of the bronchial wall as a result of chronic inflammation, infection, and abscess. It is usually the result of chronic focal or diffuse lung infections. Anesthetic management is influenced by the presence of chronic secretions, mucous plugging of the airways, impaired gas exchange, and air trapping affecting shunt as well as dead space. Patients often have chronic pulmonary disease such as cystic fibrosis or impaired immune defense mechanisms such as hypogammaglobulinemia. The recent visit to the ED suggests that the patient's compensation may be marginal. It would be helpful to know whether he is on bronchodilators and what those are, whether supplemental oxygen is required at home, whether he has been treated with steroids recently, and whether his respiratory difficulties are associated with any other medical abnormalities. A chest x-ray is helpful if the history points to an acute exacerbation above his usual baseline. It is important to identify the inhaler he uses because you might wish to employ additional inhalers or intravenous medications that may work additively or synergistically with his home medication.

3. Chronic pulmonary disease with hypoxemia may result in gradual elevation of pulmonary artery pressure and pulmonary vascular resistance, with eventual right heart strain. Ultimately, right heart failure will affect not only pulmonary circuit volume but also systemic cardiac output and contribute to left ventricular failure and impaired perfusion. Because general anesthesia typically involves the use of positive pressure ventilation, which can adversely affect cardiac filling as well as cardiac performance and also involves the use of volatile anesthetics, myocardial performance can be severely affected. Tertiary effects of impaired cardiac disease include hypoperfusion to the splanchnic circulation and protein-losing enteropathies, hepatic and splenic congestion, and renal failure.

4. The child is quite anxious and the mother tells you the anxiety can initiate asthma symptoms. Would you order premedication? If not, why not? If so, what? Rationale. Should this patient receive preoperative antibiotics? Why/why not?

Intraoperative Course

1. What monitors will you select? Why? How will your chosen monitors assist in evaluation of his pulmonary function? Will $P_{ET}CO_2$ accurately represent his $PaCO_2$? Explain. Would you consider an arterial catheter for ABG assessment? Why/why not? How will you place lines? What is the risk of infection? Is this patient colonized? Why?

2. This child is terrified at prospect of an IV and will not cooperate. How will you induce anesthesia? Rationale. Colleague suggests IM ketamine. You respond? How will you prepare the skin? Why? Should antibiotics be administered before oral intubation? What is your choice? Why?

4. Mothers usually know their children best, and premedication may not be a bad idea for this patient. On the other hand, he is 8 years old and conversation may do well at this age; children younger than school age will need premedication more often. Oral midazolam would probably work well, but should be used in an effective dose; 15–20 mg seems reasonable to achieve a good anxiolytic effect. Other alternatives might include intramuscular ketamine, midazolam, and glycopyrrolate. Preoperative antibiotics, if given well in advance (e.g., 2–3 days), may aid in decreasing various sites of infection within ectatic segments of the patient's lungs. In addition, these patients may have other foci of infection, such as the sinuses, which may become important as a source of infection in the perioperative period.

Intraoperative Course

1. At this point, standard noninvasive monitors should be sufficient for the planned procedure. SpO_2 should be sufficient to judge the efficiency of oxygenation and changes in shunting throughout the case, and the $ETCO_2$ should be sufficient to judge the adequacy of ventilation and any changes in dead space. $PETCO_2$ should be a reasonable reflection of $PaCO_2$ because carbon dioxide is so much more diffusible than air; however, much depends on the extent of the bronchiectasis. While one lobe may not change the dead space to tidal volume ratio significantly, multi-lobe bronchiectasis could. If the bronchiectasis was extensive or perioperative ventilatory failure was anticipated, then an arterial line would be useful for following blood gases frequently. In this case, the line should be placed with good sterile technique because the risk of infection from opportunistic organisms as well as the patient's own colonized flora could be significant.

2. I don't think it would be bad to start off with a mask induction, but if he is terrified of the IV, he may be terrified of the mask. Alternatives include an oral or intramuscular premed (but what about the needle stick with the IM shot?) or the use of eutectic mixture of local anesthetics (EMLA) cream applied about an hour in advance. Antibiotics may not be a bad idea, and a broad-spectrum antibiotic such as ampicillin, clindamycin, or Zosyn would probably be reasonable choices to cover a wide range of oral and respiratory flora.

3. What agent(s) would you use to maintain anesthesia? Why? Would you avoid N_2O? Benefits? What is the role of nitrous oxide use with regard to immunosuppression? Methionine synthetase regulation? Would you favor spontaneous ventilation or would you control ventilation? Why? Would you use a muscle relaxant? Why/why not?

4. Toward the end of surgery, the inspiratory pressure during controlled ventilation rises from a previous high of 24 cm H_2O to 60 cm H_2O, along with a drop in oxygen saturation to 85 % and a drop in blood pressure to 72/40. Discuss your considerations? Diagnostic steps? Therapy?

3. Anesthetic maintenance should ideally be carried out with a balanced technique for a few different reasons. First of all, the volatile agent will act to attenuate bronchoreactivity and will also allow a higher F_iO_2 than would a high inspired concentration of nitrous oxide. The use of an opioid, on the other hand, would decrease the amount of postoperative coughing that would undoubtedly follow the anesthetic and would also allow for more rapid emergence because of the decreased requirement for the volatile agent. If the patient remains well oxygenated, then nitrous oxide need not be avoided, although nitrous oxide has been shown to worsen elevated pulmonary artery pressure in patients with known elevated pulmonary vascular resistance due to heart disease. Nitrous oxide has also been shown to interfere with methionine synthetase when used in inspired concentrations greater than 50 %. There may be a role for spontaneous ventilation in decreasing turbulence in already turbulent airways and therefore enhancing gas exchange, but if the patient's airflow is that turbulent, one might be better off delaying the surgery anyway. The use of a muscle relaxant may allow a lighter plane of anesthesia and therefore faster emergence, but the lighter plane of anesthesia may worsen bronchoreactivity and bronchospasm. There is also a relationship between bronchospasm, airway reactivity, and laryngospasm, so it may nevertheless be worthwhile considering the use of muscle relaxants as an adjunct, but they are not intrinsically necessary to the procedure.

4. The most likely consideration is bronchoreactivity and bronchospasm with an acute elevation in airway pressure and significant impairment of systemic cardiac output because of the mechanical effect of the increased intrathoracic pressure on left ventricular output (due to shifting of the interventricular septum) and impaired venous return because of the increase in intrathoracic pressure. This picture would account for the acute elevation of airway pressure, the drop in oxygen saturation, and the drop in blood pressure. The differential would also include a pneumothorax and pneumohemothorax, an accidental mainstem intubation with resulting bronchospasm, and pulmonary edema, all of which should be carefully considered. A judgment should be made about any changes in the depth of the endotracheal tube, the chest auscultated, the length of the tube examined, and the beneficial effect of withdrawing the tube if deemed necessary. Pneumothorax should initially be evaluated by auscultation and a chest x-ray if needed; placement of a chest tube may be required and nitrous oxide should be discontinued. Pulmonary edema will likely be obvious because of the frothy material in the endotracheal tube and the copious secretions during suctioning.

Postoperative Course

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? What if you had given ketamine? Why?

Additional Topics

1. The embryologic derivation of the thymus and the parathyroids has certain specific anesthetic implications besides an associated T-cell dysfunction. Explain. What are the immunological implications? Why? What are the metabolic abnormalities you would anticipate? Why?

Postoperative Course

1. Here the data is controversial. Although many would prefer to extubate deep, I would tend 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.

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 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, but in the end, the patient often ends up where he started off, and one still needs mild (personal) restraint and careful protection of the patient to get through the emergence stage.

Additional Topics

1. The thymus and parathyroids are derived from the third and fourth pharyngeal pouches. Although anatomic abnormalities may result from congenital malformations, such as abnormalities of the tracheal or branchial cleft sinuses, these are rare in the third and fourth pouches. Concurrent abnormalities in other organ systems may include cardiac abnormalities, micrognathia, and low-set ears (from a concurrent failure of the second branchial pouch). However, failure of normal formation of the thymus and parathyroid results in the DiGeorge syndrome, with low parathormone levels, hypocalcemia, and immunological problems. If the thymic hypoplasia is minimal, T-cell function may be almost normal; there may be variable degrees of severity. The decreased serum ionized calcium may result in tetany, myocardial depression, and seizures.

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?

3. 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. Severe combined immunodeficiency syndrome is a primary T-cell abnormality that also results in 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 a month-old blood should be considered.

3. 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 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 more risky because of its in vivo nature. Epinephrine in doses of 0.1 mcg/kg intravenously can be used to treat anaphylactic reactions.

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Chapter 32
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 carinii infection.

VS: HR 150/min; RR 50/min with retractions; BP = 76/55 mmHg; T = 38.5° C. Lab Hct = 25 %, WBC 3,500, 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?

Preoperative Evaluation

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 S_pO_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.

Intraoperative Course

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 health-care personnel after accidental percutaneous exposure to blood is 0.3 %. After a parenteral exposure to a patient with HIV, the health-care 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

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?

three-drug regimen), the timing is not [7]. 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

Questions

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.

Postoperative Course

Answers

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.

Additional Topics

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 vasculitis of meningococcemia will lower blood pressure generally, and the inflamed intima of the cannulated artery will make the likelihood of thrombosis or embo-

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?

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

lism 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. 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, it is safer to provide mechanical ventilation until recovery has occurred. Administration of calcium is not recommended since the antagonism to neuro-

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?

muscular 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 thorascopic drainage of empyema (VATS) or bronchoscopy [8]. *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 is 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 [9].

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Chapter 33
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 bpm, RR = 28/min, 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? 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?

Preoperative Evaluation

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 is 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 hand. Pulmonary impairment can be defined by vital capacity less than 40 mL/kg, impaired expiratory effort (maximum static expiratory volume), and flow-volume loops showing decreased flow on expiration in the supine and sitting position.

   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 somewhat 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 actual 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 particular patient because she is scheduled for a sternotomy. Ideally, the patient should not take her pyridostigmine preoperatively. Omitting the morning dose of pyridostigmine will weaken the patient and obviate the need for intraoperative muscle relaxants in most cases. Added weakness may provoke her anxiety so the issue should be carefully discussed prior to

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 pulmonary artery catheter would help?

the day of surgery. Yes, if pulmonary function is impaired despite optimal medical therapy, the morning 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 s (FEV_1) are decreased due to weakness. If FVC is normal and the FEV_1 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. Yes, 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 particular 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.

Intraoperative Course

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 pulmonary artery catheter is indicated if cardiac dysfunction is present and monitoring of filling pressures, cardiac output, and systemic vascular resistance was needed.

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 breath spontaneously? Explain your choice of inhalation agent.

3. Assume that you have administered a dose of propofol and started sevoflurane; the patient is now in laryngospasm and her saturation is 85 %. There is no $ETCO_2$ 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 agent? How much should be given? Which nondepolarizer would you choose?

4. The patient was 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?

5. During the thymus dissection, the patient begins wheezing, and you note a prolonged and slow 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. No, because volatile agents produce muscle relaxant effect and spontaneous ventilation is almost certain to be inadequate under anesthesia. I would choose sevoflurane. Sevoflurane has the least respiratory depressant effect compared to other currently used volatile agents, isoflurane and desflurane.

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. If a large dose is given, they are at risk for phase II block with prolonged muscle relaxant effect. Yes, 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 ED95, which is a much smaller dose than typically required for non-myasthenic patients.

   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 T1/T4 ratio before the administration of a nondepolarizing agent and titrate the dose of the selected agent to maintain a visible T1 response. There are many surgical cases where muscle relaxation is not necessary, and this issue should be discussed with the surgeon prior to surgery.

5. 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.

Postoperative Care

Questions

1. Should you reverse the neuromuscular blockade? Why? What is a cholinergic crisis? What are the symptoms and signs of cholinergic crisis? How is it treated?

2. 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?

3. How should pain relief be managed? With a thoracic epidural or a morphine PCA? Why? What are the advantages and disadvantages of each?

Postoperative Care

Answers

1. I would not reverse the neuromuscular blockade. It is advisable not to administer anticholinesterase drugs to reverse the neuromuscular blockade as this may precipitate a cholinergic crisis. It is fatal if not treated in a timely fashion. 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.

2. 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. No, I would not extubate until reflexes were present.

3. The 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.

Additional Questions

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. 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?

Additional Questions

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 acetylcholinesterase enzyme (the esteric site), blocking its activity and causing an overabundance of acetylcholine. I would offer cardiorespiratory support and symptomatic treatment.

   If organophosphate toxicity is suspected, decontamination is initiated. The patient is stripped 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 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, 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 h 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 min 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 is 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 fiber-optic 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

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 this 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?

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. 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 release of calcium in to 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, adult onset CCD may experience transient worsening with intense activity. However, unlike other myopathies, exercise improves muscle strength. In general, 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

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 h ago. Should he be intubated awake? Would you perform?

nuclei in the brain stem. 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 respiration 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. In addition, these patients are intolerant to sedatives, 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. 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.

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-Barre 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-Barre disease because there is an active demyelination process that likely predisposes to serious hyperkalemia.

Suggested Readings

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Chapter 34
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?

Preoperative Evaluation

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?

Intraoperative Course

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_3^- 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:

$$(Na + \mathrm{K}) \times 2 + \text{glucose}/18 + \text{BUN}/3$$

If we assume the contribution of BUN to osmolality is 10, then the equation reduces to

$$S_{osm} = (132.5 \cdot 2) + \text{glucose}/18.\ (132.5 \cdot 2) + 10 + 990/18 = 330.$$

The hyponatremia may only be apparent, not real, if the water content of the plasma is reduced by the presence of excess lipids.

Postoperative Care

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 specific gravity determination and will also cause an osmotic diuresis. While a fluid bolus is indicated, care must be taken with the speed of administration given the propensity of these patients to develop cerebral edema [3]. A CVP catheter would be a good guide to fluid administration and help minimize 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?

3. What is the relationship of U_{osm} to P_{osm} 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?

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.

Additional Topics

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 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, $U_{osm} > P_{osm}$ 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

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?

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 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 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 pulmo-

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?

nary 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 is indicated [14, 15].

Annotated References

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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.

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Management of the cardiovascular changes expected during surgical removal is also reviewed.

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Chapter 35
Transplantation

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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?

Preoperative Evaluation

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 contributes 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. S_vO_2 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? Is the hepatorenal syndrome present?

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 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 includes 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].

Intraoperative Course

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 posttransplantation 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.

Additional Questions

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 cells; increasing cAMP levels with resulting increased contractility. The sys-

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?

temic 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 cath 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 preop. 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 coadministration 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. Posttransplantation 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 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 (gamma aminobutyric acid) 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 36
Minimally Invasive Surgery

Robert S. Holzman

A 16-year-old girl is scheduled for right-sided VATS resection of an anterior mediastinal mass (thymus). She has intermittent bronchitis since childhood, most recent episode several months ago. Labs unremarkable; VS unremarkable; 50 kg.

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Preoperative Evaluation

Questions

1. What else would you like to know about her history?
   (a) Respiratory effects of an anterior mediastinal mass?
   (b) Other symptoms?
   (c) Is her perioperative plan likely to be influenced by the above?

2. Her labs are unremarkable.
   (a) What other studies would you like to know the results of or obtain?
   (b) How will the results influence your planning? Of what consequence will that be for the patient and family?

Intraoperative Course

Questions

1. Routine noninvasive monitors are already planned for this case; does it require anything else?
   (a) Do you need an arterial line? Why/why not?
   (b) 1 IV or 2?
   (c) What other nonroutine monitors might be required?

Preoperative Evaluation

Answers

1. While thymomas are rare, they nevertheless are the most common tumor of the anterior superior mediastinum. Paraneoplastic syndromes are relatively common, with myasthenia gravis symptoms in about 50 %. In addition, mechanical effects on the anterior mediastinum include cough, dyspnea, dysphagia, and signs of superior vena cava syndrome. There may also be comorbidities associated with immune deficiencies because of abnormalities of T-cell function. Depending on the range and severity of associated abnormalities, the site of her recovery as well as perioperative management such as postoperative mechanical ventilation may be influenced.

2. Additional studies might include pulmonary function testing with a specific emphasis on a dynamic airway assessment such as a vital capacity maneuver. Imaging studies should have included chest x-ray and CT scan to evaluate the encroachment of the mass on the trachea and main conducting airways. There is an association of airway collapse with tracheal encroachment of more than 50 % (age and gender adjusted). For patients with significant airway compromise from an anterior mediastinal mass, induction in an upright position with spontaneous breathing may be the safest technique. A sedated "awake" intubation with nerve blocks is another technique. Neuromuscular blockade in patients with high-grade tracheal obstruction is fraught with danger as far as airway collapse, and to the extent that the mass encroaches on venous return or the right ventricular outflow tract, cardiovascular function may be compromised as well [1, 2].

Intraoperative Course

Answers

1. The need for an arterial line is influenced by the extent of the tumor and its local effects as well as how much anticipated dissection there will be according to the surgeons. In addition, if there is a component of myasthenia gravis, regardless of whether the patient will be mechanically ventilated postoperatively, in all likelihood the patient will go to the ICU, who will undoubtedly appreciate the presence of an arterial line for frequent checking of arterial blood gases. Because of the proximity to vital structures during the dissection, an arterial line is a very reasonable choice. Adequate vascular access is crucial, and this patient should have (at least) two large-bore IVs. Routine noninvasive monitoring is indicated, plus an arterial line. 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

2. The surgeons request single-lung ventilation.
   (a) How can you accomplish this in a 50 kg adolescent? What are the available choices? What effective lumen will you be dealing with, based on your choices? Why can't you find a right-sided double-lumen tube in the anesthesia workroom?
   (b) What ventilatory adjustments will you need to make as you change over from double-lung to single-lung ventilation? Does this changeover affect minute ventilation?

3. Assume that the patient did have a myasthenia picture preoperatively; what are your considerations for neuromuscular blockade? Does this patient require it?

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 F_iO_2 of 1.0.
   (a) Why is this happening? Are there equipment-related causes? Mechanical causes? Pulmonary causes? Cardiac causes?

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 [3].

2. There are several devices that are useful in small adults. Double-lumen tubes for a 50 kg adolescent girl 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 CO_2 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. There are several compelling reasons to ensure a good level of neuromuscular blockade. First of all, motionlessness is required for safe initial puncture and subsequent manipulation via the thoracoscope. Secondly, this patient will likely have an increased level of end-tidal CO_2 from the CO_2 insufflation as a stimulus to breathing. Diaphragmatic movement is at best disturbing to the continuous view through the thoracoscope and at worst may cause visceral organ damage while working with these small instruments. Therefore, I would carefully assess the level of relaxation in order to ensure reversibility at the end of surgery. On the other hand, effective relaxation may be achievable simply through the use of a volatile agent in a myasthenic, so I would begin with assessment of neuromuscular blockade following intubation of the trachea without the use of relaxants, then start with 1/10th the intubating dose of a nondepolarizing relaxant, and assess again with a twitch monitor. It is important to incorporate a clinical assessment of the adequacy of neuromuscular blockade since the general anesthetic state itself will enhance muscle relaxation and may in fact be adequate clinically. This assessment will also guide the postoperative plan for extubation versus mechanical ventilatory support until the recovery of adequate neuromuscular function.

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

(b) What will likely correct the situation? How would each of these interventions work?
(c) No matter what you do, you are not able to get the saturation above 93 %; what maneuvers are left?

5. The SpO_2 is now 88 % on 100 % O_2 and the blood pressure 60/40 mmHg; how does this influence your analysis? What would you do?

(a) What do you think is going on?
(b) Can altering your ventilation affect this situation?
(c) Would you ask the surgeons to do anything differently?

Postoperative Course

Questions

1. Assume that the patient did have some signs and symptoms of myasthenia gravis preoperatively but was not taking any medication; after waking up uneventfully she is now complaining of dyspnea in the PACU 1.5 h later. Why would this have been delayed? What do you think is the underlying physiology? What would your sequence of interventions be? Is it likely that anticholinesterases will help? Should she be reintubated right from the beginning? Would you administer anticholinesterases in the ICU after intubation in an effort to extubate early?

2. Pain management?

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 with fiber-optic bronchoscopy, optimal ventilatory strategies (including "best tidal volume" in relation to lung expansion, oxygen saturation, and capnograph morphology) for the ventilated lung, passive oxygenation of the operative side 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 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.

Postoperative Course

Answers

1. The stress response to surgery often results in an immediate perioperative improvement in weakness, due in part to elevations in endogenous catecholamines and cortisol. Depending on the patient's preoperative statement, this honeymoon period will be of variable duration. Whether quantitating the effects subjectively ("are you getting enough air to breathe" or objectively with vital capacity maneuvers with bedside spirometry), it is important to be able to judge the adequacy of interventions. Titrating doses of anticholinesterases (e.g. pyridostigmine) may be administered during this assessment in an effort to avoid mechanical ventilation, however, if the context does not seem to suggest that this will be successful, the trachea should be reintubated and the patient supported mechanically. The administration of anticholinesterases in an effort to extubate early is probably unwise.

2. The puncture sites for thoracoscopy lend themselves to local infiltration, and the mediastinal dissection may not be very painful in the postoperative period. Incremental doses of opioids and nonsteroidals should be sufficient. Regional anesthetic techniques might include a thoraacic paravertebral catheter for the affected side. A thoracic epidural catheter could be appropriate depending on the patient's preoperative level of neuromuscular competence and use of accessory muscles.

Additional Topics

Questions

1. A 15-year-old female 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 SpO_2 of 89 % is in the ICU, getting ready to come to the OR for an evacuation and thoracoscopy. The surgeon wants you to use one-lung ventilation to the left lung while he insufflates the pleura in order to visualize the empyema better. How can you do this?

3. An 18-year-old with pansinusitis and cystic fibrosis is scheduled for functional endoscopic sinus surgery (FESS). His preoperative pulmonary function tests indicate a FVC of 80 % of predicted prebronchodilator and 87 % postbronchodilator

Additional Topics

Answers

1. Cholecystectomy in a sickle cell disease teenager 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, it 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 V/Q 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 various end-organ effects of sickle cell disease and the kind of anesthetic technique that would minimize such effects. In order to minimize 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. An 8-year-old, 27 kg child is too small for a 28 French double-lumen endotracheal tube, so there are several options left to isolate the lungs. A left mainstem intubation can be planned with fiber-optic guidance, allowing the right lung to decompress and not be ventilated. A bronchial blocker could be placed along the side of the endotracheal tube in order to further isolate the lung. I would, however, favor placement of an Arndt coaxial endobronchial blocker through an in situ endotracheal tube under fiber-optic guidance. The difficulty with this is the inability to passively supply additional oxygen through the endobronchial blocker in order to take advantage of passive oxygenation in the nonventilated lung, and therefore, one has to be open to the possibility that the patient may require bilateral lung ventilation and the surgeon may have to compress the lung if oxygenation is difficult.

3. These pulmonary function tests are not bad for an 18-year-old and should be judged against a history and physical examination, but his level of function is probably pretty good. He does not show dramatic improvement pre- and postbronchodilator

and FEV1/FVC of 82 and 85 % pre- and postbronchodilator, respectively. He is not wheezing. Any special considerations? He wants to avoid a general anesthetic and wonders if there is a way to do this with sedation and blocks? Any blocks you can suggest?

4. A 2.5-year-old, 13 kg female is scheduled for a 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? Is their recovery likely to be different? You note that the patient's temperature has not decreased during the course of surgery but has rather increased such that by the first 2 h of surgery the temperature is 37.2 °C. Over the next hour, it has increased to 38.3 °C. What do you think may be going on? How can you figure it out?

   What are your considerations for management of intraoperative mechanical ventilation?

PFTs so his medical management is probably close to optimal and will therefore likely tolerate a general anesthetic well. He is curious about the possibility of not going completely to sleep and that is understandable, but should be explored further not only with the patient but also with the surgeon. There is substantial danger to the patient if he moves during the FESS procedure, and many surgeons are also concerned about an awake patient and the possibility of uncontrolled bleeding or ocular damage during the FESS. Theoretically, it should be possible to sedate the patient and provide analgesia through a sphenopalatine block, anesthetizing the ipsilateral palate and maxillary sinus. It may be carried out transorally with injection of local anesthetic through the greater palatine foramen or transnasally at the level of the middle turbinate (which it often is anyway because of the application of local anesthetic by the surgeon prior to instrumentation). However, the intraoperative discomfort likely extends beyond the area that is immediately affected by the sphenopalatine ganglion, and because of these concerns as well as bleeding and comfort and control by the surgeon, I would opt for recommending against a local/deep sedation approach.

4. If the surgical goal is accomplishable by either route, then the decision-making will be most influenced by the overall medical condition of the patient. Perioperative recovery will be faster and less painful with the laparoscopic route, and it should be anticipated that adequacy of postoperative ventilation will be improved with a minimally invasive approach. Extraperitoneal approaches will affect respiration less than intraperitoneal approaches, and retroperitoneal approaches will probably be most favorable for recovery, particularly in the prone compared to the flank position. Intraperitoneal gas insufflation will affect respiratory mechanics and cardiovascular performance as well as impose an extrinsic load of the particular insufflating gas [5–8]. The creation of the pneumoperitoneum will increase peak airway pressure and plateau airway pressures. Cardiac output and splanchnic blood flow decrease, and cardiac output decreases, while systemic vascular resistance increases, leading to recommendations of peak intra-abdominal pressures of 8–12 mmHg. Positioning of the patient in Trendelenburg position and various lateral tilting maneuvers impose further influences on the cardiopulmonary system. While several avenues of gas insufflation have been pursued (air, nitrous oxide, and carbon dioxide), carbon dioxide is most favored because of its increased solubility and lack of support of combustion [9]. The elevation in temperature is not surprising for several reasons. First of all, this is closed cavity surgery, and although cold anhydrous gas is being insufflated, there is no open abdomen to radiate heat. Although the mechanism is unclear, the increase in temperature is common in small children and may be a result of brown fact activation either through the mechanical stimulation of the increased abdominal or retroperitoneal pressure or as a response to the increase in CO_2 load. Even an increase to 38.3 °C is not disturbing as long as CO_2 production (plus CO_2 load from exogenously administered CO_2) is manageable by a proportional increase in minute ventilation. If it is unmanageable by an increase in minute ventilation, then the differential diagnosis needs to be broadened to include malignant hyperthermia.

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Chapter 37
Trauma I

Robert S. Holzman

A 5-year-old, 22 kg boy, previously healthy, is 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.

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Preoperative Evaluation

Questions

1. What systems do these various signs and symptoms point to? Why? What is the most important monitor in the ER? Why? What are you most worried about? Should the patient receive supplemental oxygen en route to the OR? Does the patient need a head CT scan as part of his work-up? A body CT scan? A C-spine series? Chest x-ray? EKG? Fundoscopic examination?

2. If he lost consciousness for a period of time, would this influence your work-up and plan? Why?

3. What is the significance of bleeding from the ear? What if he had a clear discharge from his nose as well as bleeding from the ear?

Preoperative Evaluation

Answers

1. 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 [1]. 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 [2]. 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 [3]. 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.

2. A loss of consciousness should prompt more aggressive investigation for closed head injury and the C-spine.

3. Bleeding from the ear is highly suggestive of moderately severe blast injury, and gas-containing body systems (ear, sinuses, lungs, bowel) should be carefully examined. 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.

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? What about a central line? The patient's neck veins are distended and don't appear to be varying with respirations; what is the significance? What is the differential diagnosis; most likely diagnosis? 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.

2. Assume no pneumothorax; how would you proceed with induction? Would you leave the saphenous vein IV in place, or insert one above the diaphragm prior to induction; just after induction? Why?

3. Does this patient need a rapid sequence induction? With what? Why not inhalation induction with sevoflurane? What about use of nitrous oxide? Why/why not?

4. Exploratory lap reveals a large hepatic subcapsular hematoma and a fractured spleen. Unfortunately, the vascular clamp has come off the splenic pedicle, and the capsule has ruptured; there is a unit of blood in the belly; the BP is 53/20 and the HR 200. What to do next? Get an ABG? Give bicarb? Can the surgeons help you at all? How? Is the saphenous vein IV of any use? How much?

Intraoperative Care

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 preinduction (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). 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 [4], 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 to anesthetic induction, it may be more practical with conscious children to induce anesthesia first and then be prepared to place a chest tube immediately following induction. Subcutaneous emphysema would suggest disruption of the tracheobronchial tree with air leak, and it would be unwise to proceed to positive pressure ventilation until the site of the air leak was identified and could be bypassed or repaired.

2. If there were no pneumothorax (or subcutaneous emphysema), then, 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.

3. The patient probably needs a rapid sequence induction because of the risk of recent ingestion along with the stress of the trauma and possible abdominal injury resulting in an ileus. Because the intravascular volume status and myocardial status are unclear, it may be safest to proceed with ketamine or etomidate as the hypnotic agent and succinylcholine for muscle relaxation. The circumstance that would mitigate against this strategy is an uncertain airway or a worsening pneumothorax with subcutaneous emphysema. Nitrous oxide should be avoided because it does not permit an F_iO_2 of 100 % and it may accumulate in gas-filled spaces.

4. Vascular access and volume replacement are critical. As an immediate measure, the surgeons should try to control ongoing blood loss in the field with the use of direct pressure and control of the aorta with a cross-clamp until additional access is established. This can be most readily accomplished by the anesthesiologist via the internal jugular or subclavian veins; the surgeons can also insert a catheter in

5. Following vigorous resuscitation, the patient is now 31.2 °C. How did this happen? Why did it happen? What mechanisms of heat loss are most significant? Is it worthwhile putting in a heat and moisture exchanger in the breathing circuit at this time? Earlier? What advantage over heated humidifier? What are some of the problems with both? How can you warm the patient? Is this desirable? Should your goal be normothermia? You begin to see a widening of the QRS complex; clinical significance?

6. You note ST segment elevation on your lead II monitor; differential? Significance? Options available? Insert PA line? Add modified V5 monitor? Start nitroglycerin? What if hypotensive as well? What if SpO_2 is now 91 % on 100 % FiO_2? Would physical exam help? TEE? What would you expect to find? What if pink froth is coming out of the endotracheal tube; etiology?

Postoperative

Questions

1. Should this patient remain intubated? Why? What are your criteria for extubation in this setting?

the inferior vena cava, and once access is established in another site, close the venotomy with a purse-string suture. The saphenous vein access will not be of much use in this setting until the bleeding site in the abdomen is repaired.

5. With a temperature of 31.2 °C, the risk of arrhythmias is increased, and the patient must be warmed aggressively. Chances are the mechanism of the heat loss was primarily radiation, but conduction and convection also come into play here. A heat and moisture exchanger will do very little good at this point. It would be more useful to warm the patient by lowering the fresh gas flow rate to minimal or closed circuit flows, deliver heat calories in the form of a Bair hugger and a warm water blanket, use radiant heat lamps, and consider core warming with warmed saline and peritoneal lavage. The widening of the QRS is a worrisome sign that conduction system delay is occurring; this can worsen cardiac conduction, progressing to heart block, ventricular irritability with fibrillation, and cardiac arrest.

6. 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. A V5 monitor would aid in looking at the lateral wall, but would probably be of relatively low yield in this age group, with this particular history, which seems more referable to the anterolateral precordium. If the patient was hypotensive, it could be due to volume depletion or primary cardiac dysfunction as a result of tamponade. 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 low SpO_2 may reflect pulmonary contusion with resultant shunting or myocardial injury with a drop in cardiac output and increased venous admixture with shunting. Physical exam might help with regard to pinning down a cardiac cause. A transesophageal (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. Oxygen desaturation may be accounted for by worsening venous admixture, primary myocardial failure, and pulmonary contusion. Pink froth coming out of the endotracheal tube suggests congestive heart failure (pump failure), volume overload, or pulmonary hypertension.

Postoperative

Answers

1. 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. Mitigating against this would be the need to see the patient awaken in order to assess mental status. Both, however, can be accomplished. The usual criteria for extubation include adequacy of tidal volume, minute ventilation, and bellows

2. Pain management for post-op multiple trauma patient – would you utilize an epidural if you had to cover exploratory lap pain as well as chest tube pain? Should the patient be on PCA because the bowel was not prepped? What if a bowel resection was required?

Additional Questions

Questions

1. A 6-year-old boy was brought into the local ER in Omaha after playing in the corn fields while a crop duster flew by. He is short of breath and bradycardic, pupils are miotic, and he is complaining of intense abdominal cramps. His SpO_2 is 88 %. What are your thoughts?

2. A 16-year-old went to the store to get milk for his grandmother when he was shot by "some dude" with an automatic weapon. You see an entry wound at the lower left abdomen and no obvious exit wound, and the patient complains of shoulder pain. What is the first thing you should do when he arrives, mostly dressed including his leather jacket, in the OR? Once you disarm him, what should you do next? What are your anesthetic considerations? What is a dumdum bullet, and what is its clinical significance?

3. You are preparing a patient with a chest tube for MedEvac transport via non-pressurized 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?

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.

2. If it is anticipated that the patient will remain intubated for several days, then continuous infusion opioids would be a reasonable perioperative pain management plan. Early extubation would probably favor a segmental neuraxial block, although that would have to be balanced against the risk of perioperative infection and coagulopathy if the patient had been massively transfused. Traumatic bowel injury, therefore, might be a relative contraindication to neuraxial blockade.

Additional Questions

Answers

1. The shortness of breath, bradycardia, miosis, increased gastrointestinal motility, and history are all strongly suggestive of organophosphate poisoning, with signs and symptoms of anticholinesterase poisoning [4]. The hypoxia is probably due to a proliferation of airway secretions and worsening of shunt. The patient may also have neuromuscular weakness. Medical treatment with atropine and pralidoxime chloride (2-PAM) is indicated if there is time for a medical intervention; otherwise, he should be intubated, have his airway protected, and receive ventilatory support as well as medical intervention.

2. The first thing to do if the victim of violence arrives fully dressed is to disrobe and disarm him and fully assess for entry and exit wounds. 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 (dumdum 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. Anesthetic considerations include stability of the cardiovascular system, volume loss, injury to the lungs with pneumo- or hemothorax, and therefore the need to avoid positive pressure ventilation, recent food intake, and history of intoxication/drug use/and drug intolerance.

3. 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.

4. 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?

5. What is the difference between a high-velocity missile injury and a low-velocity missile injury? A fragmentation bullet vs. a non-fragmentation bullet?

4. 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.

5. High-velocity missile injury is caused by missiles (bullets or fragments) traveling at a rate >750 m/s [5, 6]. 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 are engineered to break apart after a certain traveling distance following penetration. The bullet fragments penetrate radially from the bullet path for a variable distance, usually of several centimeters. The degree of bullet fragmentation decreases with increasing shooting distance.

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Chapter 38
Trauma II

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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; RA SpO_2 96 %

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Preoperative Evaluation

Questions

1. How will you evaluate him for other injuries?

Preoperative Evaluation

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.

   Activity Best Response Score (changes pediatric patients)

   Eye opening

   Spontaneous 4
   To verbal command 3 (young child: To shout) To pain 2
   None 1

   Verbal

   Oriented 5 (words, phrases, smiles/coos based on age)
   Confused 4
   Inappropriate words 3
   Nonspecific sounds 2
   None 1

   Motor

   Follows commands 6 (young child: spontaneous)
   Localizes pain 5
   Withdraws from pain 4
   Flexion to pain 3
   Extension to pain 2
   None 1

   In addition to the neurologic exam, the child should be evaluated for injuries to the thorax and abdomen. 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

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?

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 [4].

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?

Intraoperative Care

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 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 fiber-optic 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) [5]. Cerebral perfusion pressure is equal to mean arterial pressure (MAP) minus ICP or CVP, whichever is higher.

$$CPP = MAP - ICP\left(\text{or } CVP \text{ if } CVP > ICP\right)$$

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) [6–8]. If associated with VAE, the bradycardia is accompanied by hypotension, the "mill-wheel" murmur from the precordial Doppler, and lowered $ETCO_2$ 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 N_2O, 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 $ETCO_2$ 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 $PaCO_2$ 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 [9, 10]. Decompressive craniectomy may be needed if these maneuvers are not successful in reversing the vital sign abnormalities [11, 12].

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

Questions

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 affects 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.

Postoperative

Answers

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 post-intubation edema in the subglottic area, leading to both hoarseness and inspiratory stridor. It is possible that, during the fiber-optic 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 (C2–C4), and the respiratory insufficiency is leading to hoarseness. Spinal cord injury without radiographic abnormality (SCIWRA) 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 [13]. In addition to hoarseness, neurogenic pulmonary edema may occur.

Additional Topics

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 fiber-optic 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.

   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.

4. Describe the advantages/disadvantages of various locations for vascular access.

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.

Annotated References

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Chapter 39
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.

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Case 4

You are asked to provide anesthesia for an 18-year-old male who has been working on a commercial fishing boat. He has a large fishhook deeply implanted in the plantar surface of his right foot. There are multiple lacerations to repair as well. The hook needs to be removed, but it will take a significant amount of dissection over a wide area. Due to the nature of the injury, local anesthesia alone is likely not going to be sufficient. The patient has a family history of malignant hyperthermia and does not want sedation or anesthesia.

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?

Case 1

Answers

1. Depending on the exact 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 h while a case like this is performed – so this is really not a viable option. Psychologically, operations of the perineum are difficult to perform on an awake adolescent. The choice would be to accomplish the case with regional anesthesia plus monitored anesthesia care/deep sedation 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. A dorsal penile nerve block 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 midline bilaterally. This could be done with ultrasound guidance or by landmarks alone. This block does not provide as complete nerve blockade (in general) as an epidural or spinal block. For a case of this duration, I would choose to simply provide GA with an LMA in addition to the nerve blockade. I would choose epidural anesthesia to allow for the duration of the surgery and to provide the option of using the block for postoperative pain control. If the child were only 1-year-old, I would choose a caudal block with a catheter insertion. 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.

2. On awakening the patient has bilateral foot drop. He is otherwise neurologically intact. What are the possible causes and what is your management?

during the block placement, several large observational studies of central and peripheral nerve blocks in pediatric age patients have not shown a significant incidence of injury from blocks performed while patients are anesthetized. I would place an epidural catheter at L4–5, and I would choose 0.25 % bupivacaine or ropivacaine – 0.2 % for this block. To maintain the block, an infusion of bupivacaine could be continued at 0.4 mg/kg during the case.

2. Bilateral foot drop 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 the placement of epidural catheter is atraumatic. The presence of arteriovenous malformation of the epidural vessels poses a risk. Needle or catheter trauma of the epidural vein can result in excessive bleeding and development of hematoma that may compress the spinal cord and cause 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 h 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?

Case 2

Answers

1. The block is performed with the patient supine and the neck slightly extended. The C6 transverse process tubercle is identified with the index and middle fingers placed at the level of cricoid ring between trachea and 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 a contact with the C6 or C7 transverse process. 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 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 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 phrenic nerve and pneumothorax. Shortness of breath due to recurrent laryngeal 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 is 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 C6–7. 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 anesthesia slowly and in a fractionated manner 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

What would be the best option for this case?

Case 3

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 analphylaxis and allow effective antagonism of systemically released antigens.

Case 4

Answers

I will perform an ankle block and anesthetize specifically the posterior tibial nerve that innervates the plantar aspect of the foot. The posterior tibial nerve runs behind the medial malleolus, under the flexor retinaculum, and lies posterior to the pulsation of the posterior tibial artery. With the patient supine in bed, the knee is flexed and the medial malleolus area is prepped. An ultrasound probe, if available, could be used to identify the posterior tibial artery, accompanying veins, and posterior tibial nerve in the position posterior to the medial maliolus. (The nerve will be most posterior.) A skin wheal is made with a 25-guage needle over the area of the nerve. Under direct ultrasound visualization, the needle is advanced posterior to the vascular structures, near the nerve, but not touching the nerve. After negative aspiration for blood, a total of 2 mL of the local anesthetic solution is injected provided there is no resistance to injection. The injection should be painless.

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?

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?

Case 5

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 the 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 h. 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 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 pre-existing 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 post-anesthesia/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.

Suggested Readings

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Chapter 40
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.

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Preoperative Evaluation

Questions

1. What is complex regional pain syndrome (CRPS)?

2. What classes of drugs do gabapentin 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?

Preoperative Evaluation

Answers

1. Complex regional pain syndrome is a painful syndrome of unclear etiology and pathophysiology. 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 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.

2. Gabapentin is an antiepileptic and pain medication that was synthesized to mimic the structure of gamma-aminobutyric acid (GABA); however, it is 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. The drug also interacts with NMDA receptors, protein kinase C, and cytokines, but the exact nature of its 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. Concerning anesthesia effects, its use has been associated with a lower requirement for intraoperative and postoperative opiates. A small percentage of patients receiving gabapentin have been found to exhibit a prolongation of the QT interval when also taking other medications that affect repolarization. Amitriptyline is a tricyclic drug that inhibits norepinephrine and serotonin reuptake at the second-order neuron synaptic transmission. This drug has been 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, AV 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, local anesthetics for nerve blocks, and intravenous ketamine (for extreme cases). Other therapies include application of heat and cold, topical analgesics, physical therapy

3. Are there any techniques for this anesthetic that can reduce her chance of further neuropathic pain?

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?

(perhaps most important), transcutaneous electrical nerve stimulation (TENS), biofeedback, and spinal cord stimulation.

3. 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 or epidural anesthesia with local anesthetics in these patients could block the nociceptive impulses generated at the surgical site from reaching the dorsal horn neurons and hence minimize further excitation of the dorsal horn neuron and reduce the potential for flare-up of RSD pain postoperatively. For the same reasons, it would be reasonable to plan an anesthetic that included a multimodal pain management strategy, with acetaminophen, nonsteroidal anti-inflammatory drugs, gabapentin, and opioids as needed. In this case, it would be appropriate to administer a regional anesthetic prior to starting surgery.

Intraoperative Course

Answers

1. 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 and expression interaction is 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 should offer good coverage for the lower extremity surgery described in this case. This could be done with adequate sedation in the preoperative time frame or after induction of general anesthesia. Alternatively, analgesia could be provided by placing a femoral nerve 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. These could also be done under significant sedation or while the patient was 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 h in a case where the patient has a CRPS history. This case 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. How would you maintain anesthesia in this patient?

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?

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. Chronic use of amitriptyline can deplete noradrenergic catecholamines. 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.

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 as 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.

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?

Postoperative Course

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. 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 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.

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?

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 how should it be treated?

Additional Questions

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. 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 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 is all that is 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. 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?

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 min. 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 41
Burns

Joseph P. Cravero

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 and tachypneic, with a blood pressure of 130/90, a heart rate of 160, and a temperature of 39.4 °C. He has a headache and 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?

Initial Evaluation

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.

   Clinically, a facial burn, difficult breathing, and tachypnea are indicators of probable thermal inhalation injury. 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 PaO_2. 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 h in room air to 90 min with the administration of 100 % inspired oxygen. Prophylactic antibiotics can be helpful

3. What are your initial considerations in volume resuscitation? 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?

to treat opportunistic infection but should not be given indiscriminately. They are not part of the primary treatment for this problem.

3. Large volume resuscitation is necessary initially and can be guided by the Parkland formula [crystalloid 4mL/kg × percent burn × wt(kg)] or the Brooke formula [crystalloid 0.5 mL/kg + colloid 1.5mL/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. 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 of 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.

Initial Critical Care Management

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.

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 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. 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?

Anesthesia Consultation

Questions

1. When called to intubate this patient, an anesthesia resident asks you if he/she should do a rapid sequence induction – what is your answer? What about using rocuronium? Any special considerations for burn patients with regard to non-depolarizing neuromuscular blocking agents? What if this is the first day post burn and the patient is going for escharotomy?

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. 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 s, 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 are 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 unrefrigerated whole blood usually does not contain adequate coagulation factors and platelets to replace the high requirement during DIC.

Anesthesia Consultation

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. Although succinylcholine is generally safe to use within the first 24 h after a burn injury, even a small acute rise of serum potassium in

2. How can you keep a burn patient warm in the OR? Any special considerations? Is this different from a non-burn patient?

Additional Questions

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?

2. A 14-year-old boy was standing in a cornfield when he was hit by lightning. He suffered a cardiac arrest, but fortunately he was resuscitated 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?

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 non-depolarizers, it is well recognized, for reasons not well understood, that burn victims are resistant to non-depolarizing muscle relaxants, and hence larger doses must be used to achieve adequate intubation conditions.From 24 h 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 °C, intravenous solutions should be heated, and the inhaled anesthetic gases should be humidified and heated.

Additional Questions

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 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. Lighting 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 2,000–2 billion volts of an extremely short duration of 0.1–1 ms.

3. A chemistry major 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?

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 the 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 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.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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Chapter 42
Anesthesia Outside the Operating Room

Robert S. Holzman

An 18-month-old, 18 kg boy is scheduled for an MRI of the brain to evaluate new-onset grand mal seizures, photophobia, and increasing irritability. He has been well previously. He is currently on Keppra (levetiracetam) 350 mg po q 12 h.

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Preprocedural Evaluation

Questions

1. What would you like to know about his current anticonvulsant regimen? Is it important to know levels? Why? Would you insist on this information prior to anesthetizing him? Why or why not? If he is poorly controlled on this multimodal regimen, would you cancel the case in favor of adding another medication to hopefully improve his seizure control? What if he is not likely to improve and will require the diagnostic procedure anyway?

2. Is he at any increased risk for perioperative morbidity from seizures? Why?

3. Would you insist on an IV prior to induction or is it okay 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?"

Preprocedural Evaluation

Answers

1. The efficacy of his anticonvulsant therapy has to be questioned. This may relate to the frequency and duration of his seizures. Grand mal seizures of the tonic-clonic variety are usually the easiest to control, particularly if they originate in a single seizure focus, but generalized tonic-clonic seizures associated with a progressive metabolic disease or complex partial seizures are more difficult to control. Multimodal therapy is often required in these circumstances. The history is important here; he may be optimally (although poorly) controlled with this regimen and yet require a diagnostic study to determine whether there is a specific focus that needs mapping, particularly if surgical resection is anticipated. Blood levels are important to obtain for a baseline, but often, pediatric patients may be optimally controlled with levels either below or above recommended ranges. In this regard, communication with the neurologist is critical because they can add to the historical perspective of the child's treatment.

2. Seizure thresholds may be affected by the administration of or the withdrawal from a general anesthetic, in the first case raising the threshold and in the second case lowering it. Therefore, during emergence, the patient maybe at increased risk for a seizure. All of the general inhalation anesthetics as well as the majority of intravenous hypnotic agents are anticonvulsant. Some inhalation agents produce myoclonus or promote seizure activity in association with hyperventilation (enflurane, sevoflurane) but can also act as anticonvulsants. Methohexital, particularly in association with hyperventilation, can act as a pro-convulsant medication. Propofol is anticonvulsant. Given the multiple attempts at intravenous access, the increased stress of the patient and the likelihood that much air has been swallowed during the attempts, a parent-present inhalation induction may produce a calmer child. In a sitting to partially upright position, with the application of gentle cricoid pressure, a completely asleep child will be a much better candidate for IV placement and the completion of this induction intravenously with the use of a muscle relaxant if placement of an endotracheal tube is considered. Rather than dealing with insistence one way or the other with the parents, it is probably wiser to educate them about the various options. I would be inclined to proceed with a mask induction as outlined above, but if there were other considerations supervening, then another individual could attempt intravenous access and continue with a rapid sequence induction if, for example, active reflux was a consideration.

3. I would not insist on an IV prior to induction. While there is always some risk to a mask induction in this age group, it usually revolves around fear of separation and fear of the unknown. The separation can be dealt with by a parent-present induction and the fear of the unknown with a preinduction medication strategy of

4. What are the relevant equipment issues related to the MRI environment?

Intraoperative Course

Questions

1. The patient is crying, screaming, and fussing with the placement of routine non-invasive monitors; is it all right to go ahead without them? Does he need any additional monitors?

2. What anesthetic technique will you choose? The nurses have already tried X3 to place an IV; the patient is now crying, sweaty, and terrified. The parents insist on a mask induction; would you go ahead or cancel the case?

a rectally administered barbiturate such as thiopental (not methohexital because of its seizure-lowering potential). If crying and struggling are minimized, then the patient's risk of preinduction seizure is lowered because of less stress and the risk of aspiration lowered because of less aerophagia.

4. Ferrous-containing elements of the anesthetic equipment have to be eliminated for the sake of patient safety as well as the test results. Iron-containing materials become missiles in the MRI scanner, depending on iron content and mass, but magnetic attraction obeys the inverse square law such that the closer it gets to the bore of the magnetic, the greater the attraction becomes and therefore, with greater mass, can easily become a projectile. Oxygen tanks, tables, and anesthesia machines have been "sucked into" the bore of the magnet and attest to the risks of not considering these issues with regard to patient safety. For the provider, anything in pockets that contains iron can become projectile as well, such as a stethoscope, scissors, etc. In addition, personal identification cards such as hospital ID cards and credit cards can have their information rendered useless; beepers and telephones will have their radiofrequency chips scrambled to the point of uselessness.

Intraoperative Course

Answers

1. This becomes a matter of risk/benefit ratio while dealing with a fearful toddler. What often happens is despite being surrounded by several adults, the toddler will succeed in pulling off the monitors at a rate faster than that which the adults can place them, all the while being terrified and swallowing more and more air. I would opt for placement of a pulse oximeter to begin with and then rapidly progressing to either an intravenous or inhalation induction in a sitting or semi-sitting position and application of cricoid pressure after the loss of consciousness. Others can place additional monitors when the patient will no longer resist their placement.

2. As outlined above, the smoothest anesthetic technique would probably be the best for the patient. Depending on my assessment of the potential ease of intravenous access, it might lead me to choose IV placement first followed by propofol as an induction medication and then continuous propofol infusion with a natural airway or, alternatively, a mask induction with sevoflurane, oxygen, and nitrous oxide and then continuation either with an LMA and sevoflurane or

3. Every time the patient is positioned in the head holder, he begins to cough and obstruct his airway; what is your next move? Would you deepen the anesthetic (whether infusion or inhalation)? Should an LMA be placed? Endotracheal tube?

Postoperative Course

Questions

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? If not, how will you counsel the parents?

continuous infusion with propofol. A rectally administered preinduction agent such as methohexital would have the advantage of a complete and gentle induction in the presence of the parents, but the disadvantage of using an agent that is recognized for its pro as well as anticonvulsant properties. This is particularly true for difficult to control seizures or complex partial seizures.

3. Because the study requires a motionless patient, at some point, the anesthesiologist will have to produce this condition, no matter what the technique. It would be worthwhile trying to deepen the patient with incrementally larger doses of propofol according to the usual depth assessment criteria, but if it turns out that the patient is intolerant of this approach for any reason (breath holding, coughing, impaired pharyngeal competence, or a partially obstructed airway because of positioning considerations), then an airway device would be indicated. The LMA could be used if the patient could be deepened sufficiently to tolerate the LMA, breathe spontaneously, and remain motionless otherwise. If this approach fails, then the patient should probably be relaxed and intubated so the study can be accomplished.

Postoperative Course

Answers

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 to home, distance to closest hospital, etc.). I would probably keep the patient for 4–6 seizure-free hours in the PACU, with the resumption of his previous p.o. medications, consultation with his neurologist, and postoperative blood levels, which should be easy to obtain.

2. When would you restart his Keppra? Should he receive a supplemental dose intravenously in the PACU? Why or why not?

Additional Questions

Questions

1. A 6-year-old status post a CVA 1 month ago is scheduled to undergo cerebral angiography to rule out Moyamoya disease. What is Moyamoya disease? Why does he need cerebral angiography? Would it be sufficient to do magnetic resonance angiography? Any difference in the anesthetic considerations? What are the anesthetic considerations? Anything you should especially consider with regard to volume status? Blood pressure? Depth of anesthesia? Mechanical ventilation? Control of CO_2 tension?

2. A 7-year-old boy is scheduled to undergo esophagogastroduodenoscopy (EGD) for symptomatic reflux disease. He would like a mask induction because of his fear of needles – what do you think? 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 eructating – what is your next move? Is a deep extubation preferable?

3. A 3-year-old is receiving weekly intrathecal methotrexate for a brain tumor. His oncologist requires motionlessness, and his parents request lack of recall and

2. I would restart his anticonvulsants following the scan. The half-lives are long enough, and the onset is long enough that his level will neither decline nor be increased dramatically if he resumes his usual p.o. schedule a few hours later. It will probably be unnecessary to give him any intravenous equivalent doses.

Additional Questions

Answers

1. Cerebral angiography requires motionlessness and exquisite control of ventilation. Additional considerations specific to Moyamoya disease include strict control of blood pressure, depth of anesthesia, and CO_2 tension in order to control cerebral blood flow [1]. The same considerations apply to Moyamoya disease as those for cerebral vascular insufficiency, such as a patient with high-grade carotid disease, because the impairment of blood flow is due to a congenital absence of the middle cerebral artery and in its place, exceedingly small vessels giving the appearance on angiography of a "puff of smoke," which is what Moyamoya means in Japanese. Patients should be adequately volume replete, compensating for NPO deficits, have a mean arterial pressure within 10 % of baseline (shouldn't be too low because of inadequate perfusion and shouldn't be too high because of the chance of intracranial bleed), and have their CO_2 tension controlled from normal to slight hypercarbia (e.g., end-tidal CO_2 concentration from 38 to 45 mmHg). An arterial line may facilitate close monitoring for unstable patients, but is not ordinarily required. Adequate depth of anesthesia will help with control of blood pressure.

2. 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. With the amount of insufflation typically necessary for EGD, I would have the patient wake up completely rather than consider a deep extubation.

3. 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

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?

4. 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?

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) plus a small dose of fentanyl and 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 will often acquire various opportunistic infections or even URIs from other patients 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 h (but hardly longer because of the context).

4. Approximately 5 % of radiological exams with radiocontrast media (RCM) are complicated by adverse reactions, with one third 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 allergy, the risk of a reaction is increased from 1.5- to tenfold. Reactions may be mild, subjective sensations of restlessness, nausea and vomiting, and 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, 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 reexposure. Pretreatment of these high-risk patients with prednisone and diphenhydramine 1 h before RCM administration reduces the risk of reactions to 9 %; the addition of ephedrine 1 h before RCM administration further reduces the rate to 3.1 % [2].

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Chapter 43
Vascular Anomalies

Robert S. Holzman

A 15-month-old is scheduled to undergo sclerotherapy for a vascular lymphatic malformation of the thigh and buttock that has been enlarging over the last 5 months.

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Preprocedural Evaluation

Questions

1. Is it important to differentiate whether the vascular malformation is arteriovenous, venous, or lymphatic? Are there comorbidities to expect?
2. How does this diagnosis influence the anesthetic technique? What if it is characterized as a "slow-flow" lesion?

3. What are the anesthetic issues if alcohol embolization is planned? Is there a difference between the use of alcohol and Sotradecol (sodium tetradecyl sulfate) as a sclerosing agent? What are the risks of using platinum coils? How does it affect your anesthetic management?

Preprocedural Evaluation

Answers

1. and 2. The various kinds of vascular malformations arise from different vascular tissues and therefore have different characteristics that are important for the anesthetic technique chosen and the success of the radiologist's intervention. The broad categories from a blood flow perspective are either fast-flow or slow-flow lesions. Arteriovenous malformations are fast flow, and venous, lymphatic, capillary, and mixed lesions are typically slow flow. The flow characteristics are particularly important because they affect how volume, ventilation, and hemodynamics are managed and may contribute to patient comorbidity, e.g., high-output cardiac failure, which is also relevant to the anesthetic technique chosen. Specifically for lymphatic malformations, accumulated, static lymph in the malformation typically results in bleeding and infection as well as enlargement, disfigurement, and distortion of adjacent structures [1].

   As a result of increased volume and pressure within the malformed lymphatics, leakage and fluid accumulation can occur in contiguous areas or areas of upstream impaired drainage. Ascites and pleural and pericardial effusions can be of significance. Peripheral edema and hypoproteinemia are of anesthetic significance because they affect volume of distribution and drug binding. Infection commonly accompanies areas of skin breakdown with oozing of proteinaceous fluid. Pulmonary consequences such as restrictive and obstructive lung disease can occur with large lesions.

   Very often, high-flow lesions are not autoregulated, and therefore hyperventilation, while acting to vasoconstrict blood vessels in general, may actually enhance imaging of non-autoregulated vessels. Adequate intravascular volume and depth of anesthesia as well as preservation of cardiac output is necessary in order to provide sufficient perfusion pressure and transit time to the intended area of embolization. Lymphatic malformations are typically slow-flow lesions, and therefore the anesthetic technique influences the transit time less.

3. The embolization material is important as well. Absolute alcohol has the locally desirable effects of sclerosis, but the undesirable effects locally of skin blistering and nerve injury. In addition, constitutional effects of nausea and intoxication are significant considerations in younger age groups. Sodium tetradecyl sulfate, or Sotradecol, has the advantage of less severe tissue necrosis, hence less severe postoperative swelling and pain and much less nausea than ethanol. Because of tissue destruction, it is still a significant concern for hemoglobinuria and skin blistering. There is no intoxication because it is a detergent. Ethanol may also cause pulmonary arterial hypertension [2–4]. While various types of coils and

4. How painful will this procedure be in the perioperative period, and how will it influence your postoperative management in the PACU?

Intraoperative Course

Questions

1. What anesthetic technique will you choose? Does this patient need invasive monitoring or are standard noninvasive monitors acceptable?

2. What ventilation management strategy will you choose? Will ventilation affect the radiologist's treatment? What if this was a venous malformation? An arteriovenous malformation with high flow?

3. How will you plan for IV fluid management with regard to the quality of imaging? Effects of contrast/sclerosing agents on the kidneys? What will you do if the urine, which previously had been clear and yellow, is now rose color? Anything you can do diagnostically? Therapeutically? Recommendations for perioperative monitoring?

glues [5] have been used (which may result in paradoxical or misplaced embolization), alcohol or Sotradecol is now more commonly utilized.

4. The goal of sclerotherapy is to produce endothelial necrosis; therefore, pain typically accompanies successful treatment in the perioperative period. The pain is more marked with ethanol than with Sotradecol. This may be superimposed on the chronic pain that these patients often have [1]. Ethanol causes more tissue swelling and edema than Sotradecol. Therefore, patients may require increased doses of opioids for adequate analgesia. Regional techniques are often relatively contraindicated because of the risk of vascular proliferation in treatment areas.

Intraoperative Course

Answers

1. The choice of anesthetic technique is influenced by comorbidities, duration of case, age of patient, and the specific requirements of the procedure, such as breath holding. All of these issues should be understood and when appropriate discussed with the radiologist and specialty consultants such as cardiologists or hematologists well before the scheduled procedure. Invasive monitoring may be appropriate for specific intra-procedural considerations (not so much in interventional radiology but when patients with vascular anomalies are scheduled for the operating room, in anticipation of massive blood loss) or perioperative care such as planned intubation for airway protection until post-procedural edema resolves, which is sometimes a matter of several days.

2. Lymphatic malformations are less affected by positive pressure ventilation than venous malformations or arteriovenous malformations. The pattern of ventilation will not affect the lymphatic malformation significantly nor will control of CO_2, as it would with a high-flow lesion like an arteriovenous malformation. A venous malformation, as a slow-flow lesion, will also be less affected by the control of CO_2.

3. Optimal imaging, as a general rule, depends on a relatively full intravascular volume status and a relatively slow transit time. This is most important with high-flow lesions and relatively less of a concern with slow-flow lesions, particularly with lymphatic malformations. However, since hemoglobinuria is a risk of both ethanol and Sotradecol administrations as well as the use of ionic contrast media, brisk urine flow with a generous intravascular volume status is a

4. What perioperative concerns do you have about the method(s) chosen to embolize this patient (alcohol, Sotradecol, foreign bodies, e.g., platinum coils; cyanoacrylate glue)?

5. Should neuromuscular blockade be incorporated into your plan? Why/why not?

6. Effects on the pulmonary circulation?

Postoperative Course

Question

1. Should this patient go to the ICU after the procedure? What if this was a cervicofacial lymphatic malformation? What if he had a preexisting tracheostomy?

reasonable strategy for the majority of vascular anomaly patients in interventional radiology. Alkalinization may be considered in addition. Diagnostically, hemoglobinuria should be distinguished from myoglobinuria through laboratory testing. Subsequent monitoring for acute renal failure is important.

4. Additional considerations to those mentioned previously include the dose of ethanol administered with regard to systemic effects, e.g., anticipated emergence delays especially in smaller children [6]. The use of cyanoacrylate glue is more common in high-flow AVMs where the risks include local adhesion to the vascular endothelium and pulmonary embolization [5].

5. Motionlessness is key to the success of the procedure, whether achieved by deep anesthesia or neuromuscular blockade or both. The advantage of neuromuscular blockade is that it enables a lighter anesthetic for a potentially very long procedure, which would allow a faster wake-up time because of less accumulated agent in the tissues.

6. Ethanol injection even in the absence of vascular occlusion techniques is directly associated with an elevation in pulmonary artery pressure, especially during recovery [2, 4].

Postoperative Course

Answer

1. Recovery and observation in the ICU will probably not be necessary even for long procedures unless respiratory care, pain management, or other complications occur. Pain management is often the principal consideration because of ischemic pain in the treated areas. ICU observation/prolonged intubation and airway protection would likely be needed for a cervicofacial lymphatic malformation because of the postinjection swelling and the potential for airway compromise, even if the patient had a preexisting tracheostomy. Alcohol intoxication may also affect emergence. In addition, alcohol is nephrotoxic, so enhanced urine output is particularly important. Alcohol may produce a coagulum of blood and endothelial necrosis, leading to emboli distant from the intended area of intervention.

Additional Questions

Questions

1. A 4-month-old with stridor is scheduled for a diagnostic laryngoscopy and bronchoscopy. She has three 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?

2. A patient with consumptive coagulopathy related to a slow-flowing (involuting) giant capillary hemangioma has received a recommendation from the consulting hematologist for both support with fresh frozen plasma and platelets and anticoagulation. How can this be? What is the cause of this coagulation conundrum? Is it common in vascular malformations? Is it serious? How serious? Is it the same as Kasabach-Merritt syndrome?

3. 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?

4. At what point in the procedure should you be concerned about airway swelling following sclerotherapy near the airway? For how long?

5. A patient with Sturge-Weber syndrome is scheduled for a head MRI. Why? What are they looking for?

6. A 2-week-old with a vein of Galen aneurysm is scheduled for embolization. What accompanying conditions would you anticipate? How would you plan for effective anesthetic management? What are the consequences of success of the procedure? What are the procedurally related risks?

Additional Questions

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 [7, 8]. Cervicofacial hemangiomas should be investigated for subglottic lesions when infants have stridor accompanied by cutaneous hemangiomas.

2. Lesions with stagnant blood flow due to ectatic vessels can cause a consumptive coagulopathy and elevated risk of thromboembolism. This is different than Kasabach-Merritt syndrome, which is thrombocytopenia and factor consumption in association with an inflammatory hemangioma of infancy [9].

3. 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 [10, 11].

4. Swelling after sclerotherapy does not peak until several hours after the procedure. Patients with lesions near the airway may need to remain intubated postprocedure for several days, with extubation in the operating room following an airway exam. Tube exchangers are useful adjuncts, and close collaboration with the ORL service may be needed.

5. Six to 10 % of patients with capillary malformations in the facial V1 dermatome have Sturge-Weber syndrome, which includes the facial capillary malformation, 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.

6. Very high-flow arteriovenous malformations of the brain such as vein of Galen malformations and some arteriovenous fistulae can present in utero or in early infancy with high-output cardiac failure and loss of brain tissue (“melting brain”) and are associated with a poor outcome. Effective anesthetic management consists of a thorough preoperative evaluation to determine the extent of cardiac failure and impaired contractility as well as pulmonary hypertension as a comorbidity. The procedure itself is fraught with high morbidity and mortality because it is a high-flow lesion of the brain [12–14].

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Chapter 44
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 could be considered autism spectrum disorder. He is difficult to direct, likes to do as he likes, and does not like to be touched; he is nonverbal. He is on no medications but his mother gives him gingko 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 is "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?

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 antibiotics prophylaxis for this procedure?

Preoperative Evaluation

Answers

1. Behavior and psychological disorders are overrepresented in the dental population undergoing anesthesia. Oppositional defiant disorder (ODD) describes an ongoing pattern of uncooperative, defiant, and hostile behavior toward authority figures that 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. Most are unable to communicate their concerns. 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 a specific video or musical recording is helpful, and this information 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 mg/kg) can be effective but may be associated with paradoxical agitation in approximately 10 % of patients. For patients who are completely uncooperative, IM ketamine 4 mg/kg is used produce a dissociated state that will allow induction to proceed. The "dissociated state" refers to the 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 and a quiet state 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 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

Intraoperative Course

Questions

1. How would you induce anesthesia? How would you secure the airway? What are the options for airway management? How would you maintain anesthesia in this patient?

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_2 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. 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.

Intraoperative Course

Answers

1. As stated above, an IV or inhaled induction is acceptable. If the patient was sedated, 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 dental rehabilitation, a nasotracheal tube is preferred 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 is generally helpful. 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 also helpful in determining the appropriate side for tube placement and 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 via a fiberoptic scope. Anesthesia may be maintained with an inhaled agent but in this case I would opt for a total

2. During the case there is a sudden decrease in the oxygen saturation. Is this likely due to the residual effect of the VSD?

Postoperative Course

Questions

1. On emergence, the child is crying and thrashing about for 30 min. 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) is the cause of the problem?

intravenous anesthesia technique utilizing propofol and fentanyl with the goal to minimize the emergence delirium. Emergence phenomena are less problematic after a TIVA (propofol based). A dose of dexmedetomidine 1 mcg/kg 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. It is more likely that the decrease in saturation would be due to problems with the ET tube (main stem intubation, plugging, or kinking) or lung pathology such as bronchospasm or lobar collapse.

Postoperative Course

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. 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, and throat procedures are accompanied by more of this type of behavior than peripheral orthopedic procedures. Delirium and agitation generally do not last longer than 30 min in the post-anesthesia setting. 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

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?

collagen as 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 eightfold increase in the concentration of VWF and is effective for most mild/moderate episodes. Gingko biloba is thought to produce neuroprotective effects as an antioxidant, a free radical scavenger, a 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 conjunction with warfarin, aspirin, or other antiplatelet agents. Gingko should be stopped 36 h to 14 days prior to surgery. Depending on the dose given and the presence of other medications, gingko 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.

Additional Topics

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 %. Numerous 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. It is therefore important to identify individuals who are pregnant. 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 (third molars) are wisdom teeth that do not fully erupt 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, then general anesthesia is appropriate. For less involved cases, deep sedation with copious local anesthesia can suffice. There is no indication

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?

(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 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. 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 epirubicinare. 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 Q-T interval) unrelated to the cumulative dose. These problems can evolve hours or days after administration.

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Chapter 45
Ambulatory Surgery Procedures

Joseph P. Cravero

A 6-month-old female is scheduled for inguinal hernia repair. 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.

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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?

Preoperative Evaluation

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 inci-

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?

dence of adverse respiratory events such as bronchospasm, laryngospasm, and coughing. On the other hand, anesthesia in children with these illnesses has not 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].

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?

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?

Intraoperative Course

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 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

3. What would you use for maintenance of anesthesia? Is total intravenous anesthesia (TIVA) appropriate?

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?

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

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 doses of 0.2–0.5 mg/kg during surgeries that are not at risk for significant blood loss – such as inguinal hernia. 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

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?

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

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 (decadron). 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 46
Equipment and Monitoring

Robert S. Holzman

A 13-year-old who developed abdominal pain and claudication was diagnosed with severe mid-aortic syndrome. She is very hypertensive, 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 or 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 exercise. Meds: amlodipine, metoprolol, minoxidil, and clonidine. VS: 142/92, 86, 16. T 37^0C. SpO_2=99 %

She is scheduled for left thoracotomy, partial left heart bypass, proximal anastomosis, exploratory laparotomy, aortobiiliac bypass with bifurcated graft, and reperfusion and reimplantation of the celiac, superior mesenteric, and left and right renal arteries.

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Preoperative Evaluation/Preparation

Questions

1. How will you evaluate the severity of her ischemic disease?

   (a) *Cardiac*: Standard Bruce protocol stress test? Dobutamine stress scan? MRA? Cardiac angiogram? Persantine scan? Echo? Can you do a CT angiography and accomplish the same findings in one study? What are you most worried about?

   (b) *Peripheral vascular*:

   (c) *Renal circulation*:

Intraoperative Course

Questions

1. Does this patient need an arterial line? Why/why not?

   (a) Can you get the same information from a pulse oximeter + end-tidal CO_2 analysis? What is the difference?

   (b) How does the pulse oximeter work?

Preoperative Evaluation/Preparation

Answers

(a) *Cardiac*: She will not be able to do a standard treadmill stress test because of her claudication. The results of pharmacologically induced stress testing evaluation by echocardiogram or dipyridamole scanning are acceptable in this circumstance, although exercise stress testing is more accurate. 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.
(b) *Peripheral vascular*: 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.
(c) *Renal circulation*: This 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.

Intraoperative Course

Answers

1. The patient definitely needs an arterial line for careful monitoring of blood pressure, blood gases, and additional laboratory data.

   (a) 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.
   (b) 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 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 the skin, bone, and connective tissue, would also be part of the final saturation.

(c) How does a transducer work?

(d) Is $ETCO_2$ equivalent to $PaCO_2$?

(e) Would a TEE add any additional information not otherwise obtainable with the monitors you have in place?

(f) 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?

(c) Transducers, in general, 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 output and resulting 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. The transducer system must also have an appropriate damping coefficient because the addition of tubing, stopcocks, and air 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.

(d) 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 because of the use of uncuffed endotracheal tubes) 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.

(e) 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, as well as 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.

(f) 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

2. She has had a series of cross clamp times that add up to about 5 h in various splanchnic circulations. What consequences would you expect? How would this likely be reflected in your available monitoring devices? Anything else available that you would monitor with? Treat with?

3. Is this patient at risk for hypothermia?

   (a) 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?

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. Multi-site oximetry can monitor multiple organs, often the brain plus another organ system such as the kidney, liver, intestine, or muscle and may detect hypoperfusion in regional circulations during shock.

2. 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 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 preexisting lactic acidosis, is an important alternative since it doesn't result in the production of a CO_2 load.

3. Yes, she is at risk for hypothermia.

   (a) 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 ° centrally. PTT increases below 34 °C and coagulation factor activity and platelet function decline below 33 °C. The room should be sufficiently warm (approx. 21 °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

(b) What are your concerns about choosing a temperature maintenance system? What about her ischemic extremities?

(c) Are there risks of hyperthermia?

4. About 3 h into the case, the serum lactate has increased from 2.4 to 5.2 and the temperature is 34.5°C.

(a) What do you think is going on?

(b) What would you need to confirm the diagnosis?

(c) What will your strategy be? Let's say the pH is 7.22? 7.15? Is bicarbonate the best choice?

(d) Would you hyperventilate? Use THAM (tris(hydroxymethyl)aminomethane)? Wait awhile and get another ABG?

5. The patient is now 33.8 °C.

(a) How did this happen?
(b) Where did it happen?
(c) What mechanisms of heat loss are most significant?

address conduction and heat loss when applied under the patient and will address convection and radiation heat loss (the majority of heat loss) when applied over the patient.

(b) 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.

(c) Hyperthermia is not a likely possibility because cross clamping and massive tissue exposure is 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.

4. 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.

(a) 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 delivery 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.

(b) 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 multi-site NIRS measurements that were consistent with regionally impaired perfusion in major vascular beds like the bowel, liver, or kidneys.

(c) 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.

(d) 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.

5. 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 levels reflecting this.

(a, b, and c) Warming should be undertaken cautiously, lest it obligates the patient to metabolic activity she is incapable of. Heat loss should address all three mechanisms: convective heat loss should be

(d) Is monitoring of temperature a standard of practice?

(e) Is it worthwhile putting in a heat and moisture exchanger in the breathing circuit at this time?
(f) Earlier?

6. You decide to transfuse, and the nurse brings you cold blood from the refrigerator, which she checks with you to your satisfaction.

(a) Now what? How to warm? Why?
(b) What different methods are available?

(c) What about filters? How large? What particles? How important?

Postoperative Course

Questions

1. The patient remains intubated after 18 h of surgery and is brought directly to the ICU. The respiratory therapist inquires: "How do you want the vent set up, doc?" Your answer?

(a) What is the difference between volume, pressure, and pressure support ventilation?

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/min for an adult (at normothermia; 7 % less for every degree C decrease).

(d) Yes; temperature "shall be continually evaluated" according to ASA standards.

(e and f) 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.

6. There are a variety of fluid/blood warming systems.

(a and b) Routine and rapid infusers are the broad breakdown. Routine fluid warmers will deliver approximately 150 mL/min compared with 750–1,000 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.

(c) Particulate filters (170–260 μ) 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.

Postoperative Course

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 $ETCO_2$ approximately 34 mmHg, if the dead space to tidal volume ratio is normal). For adults, this is typically around 80 mL/kg/min. 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).

   (a) *Volume control* ventilation relies on delivering a preset tidal volume. The inspiratory flow and volume are constant, but the peak inspiratory pressure can vary. *Pressure control* ventilation delivers an accelerating inspiratory

(b) What are the similarities and differences between “ICU” ventilators and “anesthesia” ventilators?

(c) What determines your settings for pressure support as a weaning strategy?

Additional Questions

Questions

1. The PACU nurse attaches an automated oscillometric blood pressure cuff to a baby but has difficulty obtaining consistent readings.
   (a) Why is this so?
   (b) What can you do to make it better?
   (c) How do you select the appropriate-sized cuff?

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.

(b) 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.

(c) 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.

Additional Questions

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 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

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?

5. How can you safely perform surgery in a patient with an implanted cardioverter defibrillator (ICD)?

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 04–0.5 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 accuracy of reading with different levels of patient activity. This works by changing the averaging time; a 5–7 s averaging time is typical for an inactive patient, while a 2–3 s averaging time is useful for sleep studies but is more affected by patient motion. Lengthening the averaging time up 10–15 s 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 can be applied, a digital nerve block, or vasodilating cream. 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. 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 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 s) intervals at the lowest effective cutting or coagulation settings possible [1].

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Chapter 47
Thermoregulation: Hypothermia and Hyperthermia

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You are taking care of a 4 kg, 5-month-old infant scheduled for a hernia repair.

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Preoperative Considerations

Questions

1. What are the mechanisms of heat loss in humans? What are the mechanisms of heat generation? Is heat generation different for an infant vs. an older child or adult? What is "thermoneutrality"?

2. What effect does general anesthesia have on thermoregulation? Contrast this with the effect of regional anesthesia on thermoregulation.

Answers

1. Heat loss can occur through several mechanisms:
   (a) Radiation – heat lost between objects that are not in contact. This could occur between the baby and the cool walls (or any other object) in the OR.
   (b) Convection – heat lost from a body to moving molecules such as air or liquid. The amount of heat lost depends on the temperature of the air and the speed of the air moving around the patient.
   (c) Evaporation – heat that is lost through the latent heat of vaporization of water or any other liquid evaporating on the patient. This can occur from the skin, a surgical incision, or from mucosal surfaces (which can be 30 % or more of heat loss if dry air is used for ventilation in an infant).
   (d) Conduction – the heat loss that occurs between objects that are in direct contact. The extent of this loss depends on temperature differences and the area of contact. Heat generation can occur through voluntary muscle activity, non-shivering thermogenesis, shivering (non-voluntary muscle activity), and dietary thermogenesis. Non-shivering thermogenesis occurs from metabolism of brown fat in infants and toddlers (perhaps up to 2 years of age). Brown fat contains an increased number of mitochondria and is therefore very effective at generating metabolic energy/heat. The effect is significantly decreased in children under anesthesia, whether this is with inhaled agents or those that have received fentanyl/propofol. Shivering is of minimal importance in maintaining body temperature in newborns and infants as the musculoskeletal system is immature and the muscle mass is limited. Young infants will shiver between 35 and 35.3 °C, but the effect is generally negligible in maintaining core temperature. Dietary thermogenesis is stimulated by nutrients (proteins and amino acids). An infusion of a small amount of amino acids under anesthesia may increase heat generation by up to fivefold in adult models. Infants have a larger surface area to body mass ratio and a higher thermal conductance (lose heat faster due to less fat and greater surface area.). The combination of faster heat loss and reduced ability to generate heat markedly predisposes the infant to hypothermia.

      Thermoneutrality or thermoneutral environment is the ambient temperature at which the oxygen demand is minimal and temperature regulation is accomplished by non-evaporative physical processes alone. For the adult, the neutral temperature is 28 °C, while for neonates and young infants, it is approximately 32 °C.

2. General anesthesia interferes with thermoregulation. This is due to many factors but includes redistribution of core heat to the periphery, a 30 % reduction in heat generation, inhibition of central thermoregulation, and increased exposure (depending on the procedure).

3. How do you define hypothermia? Where would you measure the temperature? Describe the advantages and disadvantages of each site.

Intraoperative

Questions

1. What would be your strategies for preventing heat loss during this case?

Vasoconstriction and non-shivering thermogenesis are the only thermoregulatory responses that are active under anesthesia. Maximal vasoconstriction is similar in the awake and anesthetized patient, although the threshold for vasoconstriction is reduced under anesthesia. Non-shivering thermogenesis is profoundly inhibited under general anesthesia within 10–15 min of induction. Under regional anesthesia in older children and adults, central temperature regulation is preserved, but areas that are anesthetized cannot sense temperature and therefore have inappropriate redistribution of blood flow by vasodilation. In addition, there is no vasoconstriction in the blocked areas. With major neuraxial blockade, vasoconstriction may be lost in a large portion of dermatomes with the result being a large amount of heat redistribution, which can be as marked (or worse) than general anesthesia. Contrary to this finding is the fact that caudal block in infants does not affect the temperature for vasoconstriction.

3. Hypothermia can be mild (33.9–36 °C), moderate (32.2–33.8 °C), or severe (below 32.2 °C). Temperature may be measured from the tympanic membrane (by placing a probe in the auditory canal and sealing it to the external environment), nasopharynx (convenient but can be associated with nosebleeds or inaccuracy if an uncuffed tube is in place), esophagus (convenient but can be confounded by transmission of respiratory gas temperature if not in the distal third of the esophagus), axillary (very convenient but can be very inaccurate if not placed carefully), rectal (can cause trauma and may be inaccurate if embedded in stool or during laparotomy in an infant), bladder (accurate if urine output is copious, not widely available), and skin (wildly inaccurate depending on body area and vasoconstriction).

Answers

1. I would prevent heat loss by:
   (a) Using a radiant heater during induction
   (b) Covering the skin as much as possible – particularly the head which comprises as much as 20 % of the surface area in an infant. A plastic head wrap in an intubated patient is particularly effective.
   (c) I would use a convective forced-air warmer, which should prevent heat loss and rewarm the patient as needed.
   (d) I would avoid a warming mattress as these are minimally effective after the newborn period.
   (e) I would be cautious with fluid administration and I would rewarm fluid – administering through a short length of tubing.
   (f) I would use a heat/humidity filter to attempt to minimize evaporative losses and maximize ciliary function in the respiratory tract. In addition, I would use low fresh gas flows to assist in maintaining heat and humidification.
   (g) I would warm the operating room prior to bringing this patient into the OR.

2. After 1 h of surgery, the temperature is 34.5 °C. Outline the positive and negative effects of hypothermia on an infant.

3. You institute a series of interventions to warm the patient and 90 min later the temperature is 39 °C. What is your differential diagnosis? What do you do for this situation?

Additional Questions

Questions

Case 1 A 16-year-old Asian male is brought to your ICU after a car accident where he experienced multiple traumas and a head injury. He was intubated in the field with fentanyl and succinylcholine. As he is rolled into your unit, he appears to be rigid in appearance. His temperature is 42 °C. His ECG has peaked T waves.

Diagnosis:

1. What is malignant hyperthermia (MH)? What is happening on a cellular level when this occurs? What is the incidence in the pediatric population? What is the inheritance of MH? Which myopathies are associated with MH?

2. Hypothermia can increase the surgical site infection rate threefold and impair the coagulation cascade. This occurs because of changes in perfusion as well as a decrement in immune function including decreased activity of natural killer cells. Platelet function is significantly inhibited, adding to the coagulopathy. Hypothermia decreases drug metabolism – by as much as twofold for a 10 °C reduction in temperature.

3. Excessive temperature can increase oxygen consumption and CO_2 production. The same anatomical and physiological factors that make heat loss a problem in this age group also make rewarming easier than it is in older children or adults. Iatrogenic warming is the most common problem causing hyperpyrexia. Generally this situation is easily reversed by removing the heat generating/preserving strategies outlined above. The convective blanket may be placed on "ambient." Other causes of hyperpyrexia could include viral or bacterial infection. Some diseases such as arthrogryposis and osteogenesis imperfecta as well as those with autonomic dysfunction (Riley-Day syndrome) are associated with increased temperature under anesthesia. Other possible causes could include thyroid storm, pheochromocytoma, neuroleptic malignant syndrome, and meperidine administration.

Answers

Case 1

Diagnosis

1. MH is an inherited disorder associated with a potentially fatal hypermetabolic response to certain pharmacological agents – notably succinylcholine and inhaled anesthesia agents. On a cellular level, exposure to these agents triggers the release of excessive myoplasmic Ca^{++} that in turn leads to diffuse and sustained muscle contraction associated with a hypermetabolic response. The incidence of MH has been reported extensively and varies greatly with the population studied and the definition of the problem that is used. Widely quoted rates report an incidence of approximately 0.2:10:000 in adults and 1–2:10:000 in children. There are reports of specific populations that set the rate at 0.67:10,000 in adults with a quadrupling of that rate when succinylcholine is combined with inhaled agents. The rate of severe, fulminant MH, which includes evidence of a rapid increase in temperature accompanied by critical changes in metabolism, dysrhythmias, and increases in creative kinase (CK), has been reported to be between 0.04 and 0.05:10,000. MH is inherited as an autosomal dominant disorder with variable penetrance. It is often difficult to follow exact inheritance patterns and

2. What is the typical clinical presentation for MH? What is the relationship between masseter spasm and MH? How would you make the diagnosis of MH? What other problems could present in this same manner?

Treatment:

1. What would be your first interventions for this patient? How would this differ if he were presenting in the OR with the same set of symptoms/signs? How does dantrolene work? What are the advantages of using Ryanodex® rather than standard dantrolene?

not everyone who is susceptible will have an episode on first exposure to triggering agents. As a general rule, anyone with first- or second-degree relatives with a history of MH should be considered at risk. Myopathies associated with MH include central core disease and King-Denborough syndrome, which have a high concordance with MH and have been shown to have similar abnormalities in the RYR1 gene that is associated with MH. Other myopathies such as Duchenne muscular dystrophy and Becker muscular dystrophy have been associated with rhabdomyolysis when exposed to succinylcholine or (less commonly) inhaled agents but have not been convincingly associated with the full MH pathology. It is best to avoid exposure to the "triggering" agents, but these entities are not (strictly speaking) associated with MH.

2. MH presents with hypermetabolism. The earliest clinical sign is that of an elevated end-tidal CO_2 which is not controlled by increasing the minute ventilation. Other nonspecific signs include an increase in heart rate and blood pressure. Sinus tachycardia is the most consistent change associated with this problem. Masseter spasm is considered a highly suspicious sign but not pathognomonic of possible MH. Significant (severe) masseter spasm is associated with tetany of the masseter muscle that prevents mouth opening to the degree that would allow the insertion of a laryngoscope blade regardless of the force applied. Muscle biopsy testing of those with masseter rigidity reveals that 20–50 % are susceptible for MH. Generalized muscle rigidity develops as Ca^{++} accumulates in muscle. This can be the case even in the presence of non-depolarizing muscle blockade. The rigidity is very marked, making a person "boardlike" where lifting the feet results in a fulcrum at the cranium. Hyperthermia is a late sign and will be extreme if not treated. Temperatures up to 44 °C have been reported. Acidosis results from accumulation of CO_2 as well as lactate. Initial findings will appear as a pure respiratory acidosis – with the metabolic component more obvious as time goes on. Oxygen consumption will be high enough to result in hypoxia in spite of supplemental O_2 therapy. Skin mottling will occur accompanied by rhabdomyolysis and severe hyperkalemia. Death usually occurs due to ventricular dysrhythmias, pulmonary edema, cerebral hypoxia/edema, disseminated intravascular coagulation, and renal failure.

   Other pathological entities that present in a somewhat similar manner include hyperthyroidism, fulminant sepsis, pheochromocytoma, metastatic carcinoid, cocaine intoxication, neuroleptic malignant syndrome, serotonergic toxicity, and rhabdomyolysis associated with myopathies such as Duchene muscular dystrophy.

Treatment

1. To treat MH, it is critical that all possible triggering agents be stopped. The patient should be ventilated at a maximal rate with 100 % O_2. A call for help should be made immediately, and the MH cart should be brought to the bedside where dantrolene should be drawn up for immediate administration. Approximately 2.5 mg/kg are needed for initial treatment. The drug is prepared

2. What are the late complications of MH – how would you manage this patient in the hours/days after the episode?

with mannitol – so the two are given simultaneously. Mannitol will force a diuresis of the myoglobin-containing serum, which can significantly damage renal function. Improvement should be extremely rapid. Clinical endpoints would include resolution of hypercapnia and tachycardia, resolution of muscle rigidity, return of consciousness, correction of acidosis, and electrolyte abnormalities. Doses should be repeated until symptoms and signs of MH resolve. It should be appreciated that large doses of dantrolene may cause muscle weakness that requires prolonged ventilation. Cooling measures should be initiated if the body temperature has reached dangerous levels. The administration of cooled IV fluids or the administration of a cool "bath" of fluid externally can help to bring hyperthermia under control. Avoid direct contact with ice on the skin since this can cause vasoconstriction and impair heat dissipation. A urinary catheter should be inserted to allow collection of copious urine that should result from the infusion of diuretics along with dantrolene. An arterial catheter should be inserted for serial blood gas determination as well as CK and electrolyte analysis. Metabolic acidosis can be treated with 1–2 mEq/kg of bicarbonate. Hyperkalemia should be aggressively treated with glucose and insulin infusions as well as exogenous calcium. Release of potassium from cells combined with acidosis makes hyperkalemia one of the most life-threatening aspects of this syndrome. If this scenario were to occur in the OR, it would be critically important to discontinue the anesthetics that could trigger MH and communicate with the surgeon concerning the diagnosis. Efforts to end the surgery and position the patient in a manner that will allow maximal therapy must be accomplished quickly. Dantrolene acts by binding to the ryanodine receptor and thus decreasing the concentration of free calcium in the intracellular milieu. Ryanodex® is a lyophilized powder form of dantrolene sodium. It comes supplied as 250 mg and can be reconstituted in 5 cc of sterile water. Compared to older preparations of dantrolene, this form can be prepared in approximately 1 min and requires only one vial for a patient. Older formulations required multiple vials to be prepared and required 15–20 vials depending on the dose required for weight. In addition, Ryanodex® is much more concentrated and a full dose can be administered from one vial.

2. Late complications of MH could include injury to any of the critical organ systems (neurological, cardiac, hepatic, renal) depending on the nature and duration of the MH episode. The child should remain in the ICU for close monitoring for whatever duration is required to assure recovery in terms of mental status, urine output, muscle tone, temperature regulation, and laboratory findings (including muscle enzymes, electrolytes, and liver functions). A major late complication is recurrence of the symptoms/signs of MH that may take place hours after initial exposure to the triggering agent. For this reason, repeated doses of dantrolene should be available and administered every 6 h until all physical and laboratory signs indicate the MH episode is completed. Weakness from repeated administration of dantrolene should be anticipated and provisions for respiratory support should be made.

Posttreatment:

Questions

1. What kind of testing would you suggest for members of the family? How would you compare muscle contracture testing to genetic testing?

Case 2 A newborn child has experienced an episode of severe hypoxic ischemic injury. Is it appropriate to institute cooling to protect neurological outcome? If so how would you do this? What would be your concerns?

Answers

1. The most sensitive and specific testing for MH is the in vitro contracture test that is performed on a live preparation of muscle prepared in a physiological medium. The muscle (obtained from the vastus lateralis) is attached to a strain gauge and is electrically stimulated at baseline and then after exposure to halothane, caffeine, or both. There are a couple of protocols for this testing, but both are between 97 % and 99 % sensitive for MH susceptibility with slightly lower specificity. The testing is performed at a limited number of centers in the USA. Gene testing for a limited number of RYR1 gene mutations is also available. This testing can be done on blood collected at any location and sent to one of two centers in the USA. This testing can be helpful when positive but does not rule out MH susceptibility when negative. It is most helpful when the gene mutation in a family is known and is one that has been described and is part of the testing procedure. If this is the case, such genetic testing could be used instead of muscle biopsy.

Case 2 I would agree to pursue cooling as an effort to limit the chance of death or severe injury as long as the child had residual brain function. Hypothermia attenuates blood-brain barrier damage, release of excitatory neurotransmitters, and free radical production. Anti-inflammatory cytokines are increased. Hypothermia decreases cerebral metabolic rate for glucose and oxygen and reduces the loss of high-energy phosphates during ischemia. Finally hypothermia appears to decrease apoptosis. In animal models, mild hypothermia reduces damage in the cortex, thalamus, and hippocampus. Cooling may be accomplished by a "cooling cap" which is a cap of coiled tubing filled with cooled fluid wrapped around the head. Cooling can also be accomplished by passively lowering the whole body temperature to 33.5 °C. Hypothermia is associated with physiological changes including decreases in heart rate and blood pressure. The QT interval of the electrocardiogram increases with cooling, and arrhythmia has been observed in adults with these temperatures although studies in infants have failed to indicate there is a significant effect. Hypothermia can alter clotting, however, in the trials that have been undertaken to date; no significant biochemical changes have been noted in the patients that were cooled in this manner.

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Chapter 48
Adolescence

Robert S. Holzman

Three teens arrive in the emergency room after being transported by ambulance from the same party.

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. SpO_2 = 97 %. Height 6 ft; weight 85 kg.
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 J-collar. He is also actively hallucinating when he is not in pain. VS: HR = 123, BP = 180/110, RR = 26. SpO_2 = 97 %. Height 6 ft 3 in.; 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 cannabinoids and amphetamine. His blood alcohol level is 0.15.
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 ft 4 in.; weight 45 kg. She is in a soft cervical collar. She admits to taking an unknown amount of methylenedioxyamphetamine (ecstasy) 2 h earlier.

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Preoperative Evaluation/Preparation

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 "sedation?" Any specific implications for 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?

Preoperative Evaluation/Preparation

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 and 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 GlideScope 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

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 1.5 h after arrival at the ED for a closed reduction of the Colles fracture, an ORIF of the ankle fracture, and a 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 SpO_2, which was 97 %, is now 94 %. Your evaluation? After bronchodilator administration, his SpO_2 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?

combative and should be treated as a full stomach, 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.

Patient 3. This patient has a full stomach as well, 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 [1]. 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.

Intraoperative Course

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. 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

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 GlideScope. The surgeon would like go directly to the OR for an exploratory lap and 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? 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°; 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°? 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?

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 he be "safer" remaining intubated?

operating room. Care is typically supportive with regard to mechanical ventilatory support and monitoring of improvements in shunt fraction [2]. For severe fat embolism, high-dose steroid therapy (methylprednisolone) has a role in decreasing the inflammatory response.

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 a one blood volume reserve with additional FFP and platelet support. Hopefully there would have been time to have a FAST (focused assessment 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. If muscle relaxants were not administered, there may be evidence for evolving malignant hyperthermia, particularly if the muscle exam reveals contracture or cogwheel rigidity. If there is evidence of an increased end-tidal CO_2 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 in this setting are severalfold 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 [3, 4].

Postoperative Course

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

Patient 2. His liver laceration was repaired with good hemostasis and excellent surgical technique, and you administered three units of packed red blood cells without any additional products. He has four 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 h 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?

Additional Questions

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?

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 H_2O, 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 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, 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 reevaluated.

Additional Questions

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 frequently "perfectionistic" patients, typically high-achieving female adolescent athletes such as gymnasts or dancers. These children often live 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

2. What are your considerations for a 17-year-old, 135 kg boy scheduled for sleeve gastrectomy? Is any special monitoring required? What preoperative comorbidities concern you? Would you delay the surgery to normalize the HbA_1c? Does this patient need an arterial line? Does this patient need the ICU post-op? Mechanical ventilation support? What are the consequences of poorly controlled hyperglycemia for perioperative care?

3. 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? Is 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?

tempting to not order pre-op lab tests in athletic (albeit "thin") adolescent patients who seem otherwise well. In a tip-off to a deeper underlying problem, these patients are often angry and defensive when you probe more deeply and may refuse laboratory testing because they are well aware that abnormalities may accompany their underlying disorder.

2. Childhood obesity is on an epidemic increase by more than 50 % in the last 10 years. The risks of most significant concern to the anesthesiologist are perioperative adverse respiratory events such as a higher incidence of a difficult mask fit and ventilation, airway obstruction, major oxygen desaturation, and overall critical respiratory adverse events. Comorbidities include asthma, hypertension, sleep apnea, and type 2 diabetes. HbA_1c may be elevated, reflecting chronicity of disease and heralding difficulty with perioperative glycemic control [7].

3. 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." The efficacy of these medications is remarkable; about two-thirds to three-fourths 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 for patients treated with methylphenidate [8, 9].

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Chapter 49
Postanesthesia Care Unit (PACU)

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Pain Management

Questions

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, crying, and he and his parents are wild.

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?

3. You test it, and it doesn't appear to be working. The parents say that their other child received "shots" at another hospital and didn't have any of these problems. The patient doesn't have any relief after 20 min. What other treatment is appropriate?

Pain Management

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. It may not be possible to quickly accomplish this at the bedside, however, and the assumption should always be that the child is indeed experiencing surgical pain until proven otherwise. The pain can be treated with IV opioids or the administration of an appropriate dose of a rapid-onset 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 narcotic given during the surgical procedure. If the choice is to administer IV opioids, it makes sense not to expose the child to another narcotic. Dosing must be done carefully, however, in view of the possibility of synergy between the neuraxial opioid and intravenous opioid leading to excessive depression of respiratory drive. Once the child is comfortable, the utility of the epidural catheter should be assessed as described below. This will be complicated by the age of the child and the fact that the child now has adequate analgesia.

2. If the child's discomfort is such that the few minutes needed for rapid-onset epidural local anesthetics to provide comfort would not cause excessive distress, the catheter position can be tested. Epidural analgesia is a safe and effective analgesic regimen for children [1–3]. Administration of lidocaine will provide a solid sensory block in a relatively short period of time. When administering lidocaine, it is important to consider the dose of local anesthetic that has already been given via the epidural catheter. Local anesthetic toxicity is additive so that a maximum dose of lidocaine given to a child who has been receiving an infusion of local anesthetic will result in that child receiving a total dose above the safe limit. An alternative to assess the epidural catheter is to use 3-chloroprocaine, an aminoester local anesthetic with an even quicker onset and also a very short duration and better safety profile. In addition, the blockade achieved will be both sensory and motor, perhaps allowing a more accurate assessment of the effectiveness of the epidural catheter.

3. If it appears that the child is receiving no analgesia from the drugs given into the epidural space, that technique must be abandoned and another modality begun as soon as possible. If, after administration of appropriate doses of IV opioid, the child still is uncomfortable, additional doses should be given, titrated to

4. He pulled out his IV in the meantime; should he receive IM morphine before you attempt to reinsert the IV?

Postoperative Nausea and Vomiting

Questions

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.

1. 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?

respiratory rate. In addition, adjuncts should be considered. These include IV, oral, or rectal acetaminophen or parenteral ketorolac, an effective nonsteroidal analgesic. Although there is some evidence that ketorolac may affect new bone formation, an inflammatory process, it seems to be much more of a problem following spinal fusions and also in cases where many doses are used. Many orthopedic surgeons agree with the use of ketorolac for 24–36 h in the treatment of pain associated with long bone fractures. However, it is important to discuss this therapy with the operative orthopedist prior to administration of ketorolac. In addition to other analgesics, antiemetics and/or anxiolytics should be considered for this child. If benzodiazepines are used, the clinician must be aware of the synergistic 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 EMLA, Synera or, alternatively, the use of other topical analgesics prior to placement of an IV. With the 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. IM medication should be used only as a last resort. Opioid administration to children via PCA/NCA is a good option [4, 5].

Postoperative Nausea and Vomiting

Answers

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 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].

1. Since prophylaxis with a serotonin antagonist has failed, further treatment with a drug in the same class is not likely to yield good results. Another class of medication, such as promethazine or droperidol, can be given. Promethazine (Phenergan®) is a phenothiazine derivative with antiemetic, antihistaminic, anticholinergic, and sedative effects. Although emptying the stomach might have helped decrease this girl's PONV, the trauma of NG tube placement puts that particular intervention lower on the list. Inadequate fluid resuscitation can

2. Should droperidol or haloperidol be added at this point?

3. On which limb of the emetogenic response does each of the typically utilized antiemetics work?

4. Are some anesthetic techniques better than others for perioperative emesis control?

5. Are some operations more emetogenic than others?

contribute to PONV, and if there is any indication that she has not received sufficient IV fluid, this should be corrected. Dexamethasone is another treatment 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 at or below the recommended doses. 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 h following administration of droperidol is recommended. This report is based on ten cases collected over many years. 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. Vomiting is mediated via the vomiting center and the chemoreceptor trigger zone (CTZ) located in the brain stem. The vomiting center is in the reticular formation, and the CTZ is located in the floor of the fourth ventricle. The vomiting center 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 that cause vomiting. Stimuli associated with nausea include pain and memories as well as labyrinthine stimulation.

4. Various alterations in anesthetic technique have been shown to reduce the incidence of PONV. Patients who receive 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, its use 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 [12]. Dexamethasone, given at the start of a surgical procedure, has some 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, laparotomy, ENT procedures, breast surgery, and strabismus surgery [8, 10, 13].

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?

8. What are the optimal times to administer antiemetics?

Cardiovascular System

Questions

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 pasty on arrival, BP = 50/20 mmHg and a heart rate of 76 bpm.

1. What do you think is going on? Why?

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 [12]. 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, 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.

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 h IV hydration and administration of another antiemetic of a 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 no consensus regarding prophylactic versus rescue treatment of PONV [14]. While it is true that many patients must be given prophylactic treatment in order to prevent one case of clinically significant PONV, the administration of a serotonin receptor antagonist is very safe. Prophylactic treatment for patients with moderate to high risk for PONV seems a sensible course [15].

Cardiovascular System

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 3,500 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 2,000 mL crystalloid. If we assume a one to one replacement of blood lost with PRBC and cell saver, those products have replaced approximately 1,300 mL of her 3,500 mL blood loss. She also was given approximately 2,200–2,500 mL of crystalloid as replacement for the approximately 2 L of EBL not replenished by

2. Why did these findings become apparent now and not earlier?

3. Why do you think she is not tachycardic? Could this be anything else? What?

4. She seems sleepy when you try to awaken her? What are you concerned about? What can you do?

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 6,000 mL, not the 2,000 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. 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 high-dose 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, 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.

Respiratory Failure

Questions

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 %.

1. What do you think is going on? Is this simply hypoventilation? Could it be anything else?

2. What effect would his underlying disease have on the pulmonary circulation that might be contributing to this picture?

3. What if he had laryngospasm at the end of the case, would this change the differential diagnosis? Why?

Respiratory Failure

Answers

1. This is an urgent situation. As the delivered FiO_2 is increased, the child must be quickly evaluated and the cause determined. Hypoxemia in the PACU can be, and often is, the result of hypoventilation. If the child had been receiving supplemental oxygen and still developed hypoxemia, he/she must have had significant hypoventilation to result in an SpO_2 in the mid-80s.

   Alternatively, there may be another cause for the hypoxemia. 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. 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. If the child is breath holding, it is possible that the Valsalva maneuver that accompanies breath holding has "arterialized" the blood the oximeter light is passing through and 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 %.

2. In addition to hypoventilation, other possible causes of this situation include shunt or 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).

3. 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.

4. You obtain an ABG and his pH = 7.24, pCO_2 = 48 mmHg, and pO_2 = 56 mmHg. What now? What do you think is wrong? Should he be reintubated?

Postintubation Croup

Questions

A 3-month-old just underwent direct laryngoscopy/bronchoscopy for stridor evaluation, has subglottic stenosis as a result of intubation as a neonate, and received some dexamethasone in the OR. In the PACU, the baby is crying, is working very hard to breathe, and is retracting intensely, and the nurses are unable to obtain a sat reading, but appears pink although in distress.

1. What might be going on?

4. 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 min which is as important as the laboratory evaluation.

Postintubation Croup

Answers

1. The baby likely has postintubation stridor. Postoperatively, postintubation stridor occurs in approximately 1 % of children, particularly in children < 4 years of age [22, 23]. The syndrome is the result of edema of the tracheal wall below the 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:

$$P = \frac{8Q\mu L}{\pi R^4}$$

Q=volumetric flow rate
μ=viscosity, lb·sec/in2

2. What is a croup score?

3. How does dexamethasone work to prevent postintubation edema? Is it effective if administered post hoc? What will you do?

4. Will sedation help? Does cool mist help? In what way?

5. Would the administration of nebulized racemic epinephrine help the child? How much would you give? How would you administer it?

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 h, and the presence of a URI may predispose the child to the development of postintubation croup [22, 24, 25]. 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. Downes and Raphaely developed the croup score as an aid to the clinical evaluation of children with this syndrome, either due to infection or postintubation [26]. The score does not include the SPO_2 measurement and uses clinical parameters to assign a score that reflects the severity of respiratory embarrassment. A score of 0 to 3 is given for each of five aspects of respiration: stridor, retractions, air entry, color (normal of cyanotic), and level of consciousness and a score derived (below). The higher the score, the more severe the respiratory compromise. While the determination of a croup score does help the clinician apply a more rigorous assessment to the child with stridor, the variables assessed are not independent. As the condition worsens, all variables will worsen together.

	0	1	2
Inspiratory breath sounds	Normal	Harsh with rhonchi	Delayed
Stridor	None	Inspiratory	Inspiratory and expiratory
Cough	None	Hoarse cry	Bark
Retractions and flaring	None	Flaring and suprasternal retractions	Flaring and suprasternal retractions plus subcostal and intercostal retractions
Cyanosis	None	In air	In 40 percent oxygen

3. 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 [27, 28].

4. 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 h or less.

5. 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

6. Should this baby be reintubated? Why? What size tube would you choose?

Hyperthermia

Questions

You are called to the bedside of a 10-month-old who has just undergone a cleft palate repair. He has a temperature of 41.2oC.

1. What might have happened that his temperature got to this point?

2. Could this represent malignant hyperthermia? Why/why not?

mask is held near the child's face with 100 % oxygen used to nebulized the solution.

6. 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 cannot 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.

Hyperthermia

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 case 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 was 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 should be done prior to instituting any therapy [28, 29].

Annotated References

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The authors, leaders in the field of pediatric pain treatment, review all central neuraxial blocks in children. Relavant anatomy and physiology is discussed first followed by review of the technical aspects and medications used in caudals, epidurals and spinals in that order

Yaster M, Kost-Byerly S and Maxwell, LG. Opioid agonists and antagonists. Chapter 12. In pain in infants children and adolescents 2nd Lippincott Williams & Wilkins Philadelphia 2002:181–224

In this chapter, the authors undertake a thorough review of opioid receptor physiology, structure and functional differences and similarities among the commonly used opioids. In addition, dosing regimens, schedules and various routes for administration are discussed. Tolerance, addiction and physical dependence are defined and contrasted.

Gan TJ, Meyer TA, Apfel CC, et al. Society for ambulatory anesthesia guidelines for the management of postoperative nausea and vomiting. Anesth Analg. 2007b;105:1615–28.

The present guidelines were compiled by a multidisciplinary international panel of individuals with interest and expertise in postoperative nausea and vomiting (PONV). These guidelines 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

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Chapter 50
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?

Case 1 Management

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 two 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 MSO_4 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.

Additional Questions

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 antegrade 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 S_pO_2. 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 O_2 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].

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Chapter 51
Critical Care

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A 4-year-old with gram-negative sepsis develops increasing respiratory distress concomitant with antibiotic treatment and her admission to the ICU.

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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?

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?

Case # 1: Acute Respiratory Distress Syndrome (ARDS)

Answers

1. This child may be developing acute respiratory distress syndrome (ARDS) [1, 2, 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 not reversed the pathophysiology nor decreased mortality in ARDS [7, 8].

4. In ARDS many of the pathophysiologic abnormalities are due to diminished activity of surfactant. Functional residual capacity (FRC) is reduced and lung

5. Will PEEP help? Why? What are the adverse effects of high ranges of PEEP? How can these effects be minimized?

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 (Q_s/Q_t). 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 Q_s/Q_t. 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. 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. Under what circumstances would you consider extracorporeal membrane oxygenation (ECMO)? What about extracorporeal CO_2 removal ($ECCO_2R$)?

7. What effects do ARDS have on shunt (venous admixture)? Are there any effects on Vd/Vt? What are they? At what phase of the disease process? What, if anything, can you do about it?

Case # 2: Shock/Multi-organ 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?

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 Q_s/Q_t. 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. Extracorporeal removal of CO_2 ($ECCO_2R$) has not been shown to benefit adults with ARDS and has not been used much at all in the care of children with ARDS.

Case # 2: Shock/Multi-organ System Failure

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 preexisting 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

$$\begin{aligned} P_AO_2 &= \left(1.0 \times \left(713\right)\right) - \left(32 / 0.8\right) \\ &= 713 - 40 \\ &= 673 \\ A-a \text{ gradient} &= P_AO_2 - P_aO_2 \\ &= 673 - 97 \\ &= 576 \end{aligned}$$

For a well person breathing room air, the calculation is

$$\begin{aligned} P_AO_2 &= \left(0.21 \times \left(713\right)\right) - \left(40 / 0.8\right) \\ &= 149 - 5.70 \\ &= 99 \end{aligned}$$

$$\begin{aligned} A-a \text{ gradient} &= P_AO_2 - P_aO_2 \left(\text{in a well person with } SpO_2 = 100\%, \text{assuming } PaO_2\right. \\ &= 110 \\ &= 110 - 99 \\ &= 11 \end{aligned}$$

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 function of many enzymatic systems, including those responsible for myocardial performance. In cases of severe acidosis, drugs such as 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

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 only goes to the OR for evidence of 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?

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?

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 $NaHCO_3 = 0.3 \times (\text{weight in kg}) \times (\text{base deficit})$.

Case # 3: Neurologic Intensive Care

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. 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 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

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?

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 $PaCO_2$ 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 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. He begins to develop “raccoon eyes” and discoloration of the eyelids. Now what do you think?

8. He becomes increasingly difficult to ventilate, with O_2 saturations that are slowly but progressively decreasing and coarse sounding breath sounds, reminiscent of rales. Next step? Diagnostic or therapeutic?

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?

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 P_aO_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

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. 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?

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?

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 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. 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?

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.

Case # 5: Hemolytic Uremic Syndrome

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 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,

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?

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.

Annotated References

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Chapter 52
Resuscitation

Joseph P. Cravero

A 4-year-old white 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?

Management of Prolonged Submersion

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. 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 – and not wait for ventilation equipment, or other advanced resuscitation equipment. prior to starting

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?

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?

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. 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

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?

cause adverse effects and should be reserved for cases such as beta-blocker overdose. If there were no response 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 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 arrest) failed to show improvement in neurological outcome, or survival overall in a hypothermic (33 °C) group. 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 [2, 3].

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?

Additional Questions

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 and 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 [4].

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 [5]. 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 CO_2. 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 $ETCO_2$ [6]. 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 [7].

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