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Question 1

Correct

1.00 points out of 1.00

Comment

A 3-day-old exclusively breastfed infant born at 39 weeks' gestation to a gravida 1 mother was discharged from the hospital 24 hours ago, and is brought to the community emergency department after being found in the bassinet with irregular respirations, lethargy, and emesis on the sheet. On the way to the emergency department, the infant suffers a 5 minute seizure with staring and lip smacking. Emergency medical services administer 0.1 mg/kg of lorazepam and 5 mL/kg of 10% dextrose bolus for a fingerstick blood sugar of 20 mg/dL. On arrival, the infant's rectal temperature is 34.5 °C, heart rate is 130 beats/min, respiratory rate is 10 breaths/min, blood pressure is 61/30 mm Hg, and SpO_2 is 88% on 2 L/min nasal cannula.

Due to altered mental status and hypopnea, the decision is made to intubate and provide mechanical ventilation while further workup ensues. Arrangements are made to transfer the infant to the closest children's hospital. After transport and arrival in the pediatric intensive care unit, the following laboratory values are obtained:

Laboratory Test	Result
White blood cell count	18,000/μL (180×10^9/L)
Hemoglobin	20 mg/dL (200 g/L)
Hematocrit	60%
Platelet count	160×10^3/μL (160×10^9/L)
Sodium	134 mEq/L (134 mmol/L)
Potassium	5.0 mEq/L (5 mmol/L)
Chloride	110 mEq/L (110 mmol/L)
Carbon dioxide	15 mEq/L (15 mmol/L)
Urea nitrogen	30 mg/dL (10.7 mmol/L)
Creatinine	0.8 (70.7 μmol/L)
Glucose	45 mg/dL (2.5 mmol/L)
Ammonia	135 μg/dL (96.39 μmol/L)
Alkaline phosphatase	174 U/L

Aspartate aminotransferase	159 U/L
Alanine aminotransferase	97 U/L
Creatine kinase	601 U/L
Creatine kinase myocardial fraction	62 ng/mL (62 µg/L)
Urinalysis	
Specific gravity	1.015
Ketones	Negative
Blood	Negative
Protein	Negative

The infant is given a 25% dextrose bolus for hypoglycemia and 2 fluid boluses for hypotension. Portable anteroposterior chest radiograph shows the endotracheal tube in good position and bilateral small to moderate pleural effusions.

Of the following, given the infant's likely diagnosis, the study that should be MOST prioritized is

- A. cranial ultrasound
- B. echocardiogram ✓
- C. electroencephalogram
- D. hemodialysis catheter placemen

Your answer is correct.

PREP Pearl(s)

- The hallmark finding for fatty acid oxidation disorders during a metabolic crisis is nonketotic hypoglycemia.
- Prevention of fasting and rapid intervention during mild illness or physiologic stress is important in preventing metabolic crisis in patients with fatty acid oxidation disorders.
- Infants with severe fatty acid oxidation disorders, especially very long-chain–fatty acid oxidation disorders, often present with cardiomyopathy.

Critique

The constellation of this infant's findings of nonketotic hypoglycemia, mild hyperammonemia, and shock, points to a diagnosis of a fatty acid oxidation disorder. The final diagnosis cannot be determined with the information given, but it is possible to narrow the differential diagnosis to a disorder of fatty acid metabolism due to the hallmark finding of nonketotic hypoglycemia. Disorders of fatty acid metabolism are rare and

inherited in an autosomal recessive fashion; they cause acute crises of energy deficiency and production due to an inability to use fatty acids for fuel. Ketone bodies are not produced normally in these disorders, eliminating this key alternative fuel for the body and leading to hypoglycemia with no ketone production. Disorders of fatty acid metabolism include:

- Medium-chain acyl-CoA dehydrogenase (MCAD) deficiency, the most common fatty acid oxidation disorder
- Long-chain–fatty acid oxidation disorders (LC-FAOD)
- Very long-chain–fatty acid oxidation disorders (VLC-FAOD)

Infants with these disorders often present with metabolic crisis in the first few days after birth due to periods of relative fasting before the mother's breast milk supply increases to normal levels. The cause of shock may be multifactorial, with cardiomyopathy contributing to cardiovascular collapse specifically in patients with LC-FAOD and VLC-FAOD. Therefore, assessing cardiac size and function with an echocardiogram is essential to diagnose cardiomyopathy, which can be fatal in neonates and infants with fatty acid oxidation disorders (FAODs).

Patients with FAODs may present with rhabdomyolysis (induced by exercise, fasting, or illness), hepatic dysfunction (including severe hypoglycemia and hyperammonemia), and cardiomyopathy. These clinical manifestations can lead to frequent hospitalizations, failure to thrive, developmental delays, neurologic complications, and premature death.

Medium-chain acyl-CoA dehydrogenase deficiency is the most common FAOD; MCAD is an enzyme that catalyzes the breakdown of fatty acids for energy production when glucose is not available. Children with MCAD deficiency cannot use medium-chain fatty acids for energy; this results in severe hypoglycemia brought on by physiologic stressors (eg, mild illness). Children with MCAD deficiency typically present between ages 3 to 15 months, when nighttime feedings decrease or stop, resulting in hypoglycemia. Hypoglycemic episodes can result in permanent neurologic sequelae before diagnosis; preventive measures which have been shown to reduce mortality and morbidity include:

- Avoiding fasting (often with nighttime continuous feeds or cornstarch as a slowly released carbohydrate source)
- Aggressive intervention during minor illness

Long-chain–FAODs are caused by mutations in nuclear genes encoding mitochondrial enzymes involved in the conversion of dietary long-chain fatty acids into energy. The current standard therapy for LC-FAODs is avoidance of fasting and supplementation of medium-chain triglyceride oil which does not require the typical steps of long-chain fatty acid metabolism. The use of medium-odd-chain fatty acids, such as triheptanoin, is a more recent treatment for LC-FAODs.

Patients with VLC-FAODs have a mutation causing a deficiency in very long-chain acyl-coenzyme A dehydrogenase (VLCAD) enzyme. The phenotype of VLCAD deficiency is heterogeneous, ranging from catastrophic metabolic and cardiac failure in infancy to mild hypoketotic, hypoglycemia, and exertional rhabdomyolysis in adults. Like LC-FAOD, the current standard therapy for VLC-FAODs is avoidance of fasting and supplementation of medium-chain triglyceride oil.

Cardiomyopathy in fatty acid oxidation disorders can be hypertrophic or dilated; it is most common in VLC-FAOD and more rare in MCAD. Pericardial effusion may also be present. While cardiomyopathy correlates with the severity of disease, if detected early, it can be reversed with appropriate therapy for the FAOD.

The use of L-carnitine therapy for management of FAODs remains somewhat controversial; it should be discussed with the patient's metabolic geneticist. The physiologic basis of L-carnitine supplementation is that excess acylcarnitines (generated as a result of enzyme deficiencies) can bind free carnitine; they are then

excreted by the kidneys, leading to secondary carnitine deficiency. Carnitine is involved in the transfer of long-chain fatty acids across the inner mitochondrial membrane for subsequent β-oxidation; thus, a deficiency in carnitine may lead to abnormalities in fatty acid oxidation. Carnitine may not be necessary for MCAD, but it is more commonly used in LC-FAOD and VLC-FAOD/VLCAD.

Newborn screening with tandem mass spectroscopy now identifies many infants with FAODs prior to the development of their first metabolic crisis, allowing for early institution of dietary therapy and monitoring for complications such as cardiomyopathy. Many cases of what was thought to be sudden infant death syndrome have been retrospectively diagnosed as FAODs, either due to subsequent siblings' diagnoses or re-analysis of newborn screening results.

Crucial for the intensivist is to treat metabolic crises in patients with FAODs by rapidly treating acute hypoglycemia and providing continuous dextrose-containing fluids to prevent catabolism. Close monitoring of blood sugar, hepatic function, and ammonia, along with assessment of the patient's neurologic status are required to determine when the patient's normal feeding schedule can resume after a crisis.

Suggested Reading(s)

- McGregor TL, Berry S, Dipple KM, et al. Management principles for acute illness in patients with medium-chain acyl-coenzyme A dehydrogenase deficiency. *Pediatrics.* 2021;147(1):e2020040303. doi:10.1542/peds.2020-040303
- Merritt JL 2nd, MacLeod E, Jurecka A, Hainline B. Clinical manifestations and management of fatty acid oxidation disorders. *Rev Endocr Metab Disord.* 2020;21(4):479-493. doi:10.1007/s11154-020-09568-3
- Vockley J. Long-chain fatty acid oxidation disorders and current management strategies. *Am J Manag Care.* 2020;26(suppl_7):S147-S154. doi:10.37765/ajmc.2020.88480

Content Domain

- Neonatology, Inborn Errors of Metabolism

Learning Objectives

- Understand that cardiomyopathies are common in patients with fatty acid oxidation disorders

The correct answer is: echocardiogram

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Question 2

Correct

1.00 points out of 1.00

Comment

A 9-year-old child has been admitted to the pediatric intensive care unit for septic shock from bacterial pneumonia. He remains intubated and ventilated with a positive end-expiratory pressure of 10 cm H_2O and a FiO_2 of 0.75 to maintain a saturation of > 90%. He requires epinephrine and norepinephrine infusions at 0.03 µg/kg/min and 0.05 µg/kg/min, respectively, to maintain mean arterial pressure > 60 mm Hg. His urine output decreased to 0.3 mL/kg/hr in the last 12 hours. His morning laboratory test results are shown:

Laboratory Test	Result
White blood cell count	4,000/µL (4 x 10^9/L)
Hemoglobin	8.4 g/dL (84 g/L)
Platelet	12 ×10^3/µL (12 x 10^9/L)
Urea nitrogen	42 mg/dL (14.9 mmol/L)
Creatinine	1.6 mg/dL (141.4 µmol/L)
Lactate dehydrogenase	800 U/L
Fibrinogen	300 mg/dL (8.82 g/L)
Factor V	90% activity
ADAMTS-13 activity	Low
Ferritin	683 ng/mL

Of the following, the pathology of the disease process in this child is MOST likely caused by

A. antibody-mediated destruction of platelets

B. disruption of hematopoietic stem cells

C. endothelial dysfunction with microthrombi deposition ✓

D. hyperproliferation of histiocytes and macrophages

Your answer is correct.

PREP Pearl(s)

- Thrombocytopenia-associated multiorgan failure is a distinct clinical phenotype associated with sepsis.
- Children with the thrombocytopenia-associated multiorgan failure phenotypes have higher morbidity and mortality.
- The standard therapy for thrombocytopenia-associated multiorgan failure is therapeutic plasma exchange.

Critique

The child in the vignette has developed thrombocytopenia-associated multiorgan failure (TAMOF), a condition associated with sepsis. A distinct clinical phenotype of sepsis, TAMOF is characterized by the presence of thrombotic microangiopathy. Clinically, it is diagnosed by the presence of thrombocytopenia and multiorgan failure (defined as 2 or more organs affected). Septic children with TAMOF have higher organ dysfunction, severity of illness scores, and mortality.

Thrombocytopenia-associated multiorgan failure exists along the same spectrum of syndromes with disseminated microvascular thrombosis, such as thrombotic thrombocytopenic purpura (TTP), disseminated intravascular coagulopathy (DIC), and hemolytic uremic syndrome (HUS). In both TTP and TAMOF, an activated endothelium releases ultralarge von Willebrand factor (ULVWF) protein clusters which cause spontaneous platelet aggregation. A disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13 (ADAMTS-13, also known as von Willebrand factor cleaving protease), is the metalloprotease that is supposed to cleave ULVWF into smaller and less thrombogenic forms. In sepsis, many inflammatory mediators either inhibit or inactivate ADAMTS-13. The reduced activity of ADAMTS-13 allows for more ULVWF in the circulation, resulting in platelet/VWF-rich thrombi which are deposited in multiple organs. This accounts for the thrombocytopenia and multiorgan failure that manifest clinically. Thrombocytopenia-associated multiorgan failure can occur in the absence of DIC and is less of a consumptive process, which leads to the contrast of TAMOF patients having normal levels of coagulation factors as well as normal fibrinogen levels.

The standard therapy for TAMOF is therapeutic plasma exchange; this removes ULVWF/platelet multimers from the bloodstream and replaces ADAMTS-13, which is contained in fresh plasma. In 2022, the TAMOF network released the results of an observational longitudinal cohort study examining the efficacy of TPE in children with TAMOF. The study reports that the use of TPE was associated with a decrease in organ dysfunction, as well as a lower risk of 28-day all-cause mortality.

In this vignette, the child has sepsis which predisposes him towards developing TAMOF. His laboratory values demonstrate thrombocytopenia with high lactate dehydrogenase (suggesting intravascular hemolysis), but a normal fibrinogen and factor V levels (which distinguish this condition from DIC). He also has multiorgan dysfunction with persistent respiratory and cardiovascular dysfunction, and newly developing renal failure. The low ADAMTS-13 activity level confirms the diagnosis.

Antibody-mediated destruction of platelets is the pathophysiology behind idiopathic thrombocytopenic purpura. Hyperproliferation in macrophages and histiocytes are seen in the hemophagocytic lymphohistiocytosis/macrophage activation syndrome. While some children with sepsis may develop macrophage activation syndrome in their clinical course, this child in the vignette does not have it. Disruption of hematopoietic stem cells happens in bone marrow failure. While sepsis can cause suppression of bone marrow function, this would not be the cause for thrombocytopenia alongside the multiorgan dysfunction exhibited by this patient.

Suggested Reading(s)

- Carcillo JA, Berg RA, Wessel D, et al; Eunice Kennedy Shriver National Institute of Child Health and Human Development Collaborative Pediatric Critical Care Research Network. A multicenter network

assessment of three inflammation phenotypes in pediatric sepsis-induced multiple organ failure. *Pediatr Crit Care Med.* 2019;20(12):1137-1146. doi:10.1097/pcc.0000000000002105

- Fortenberry JD, Nguyen T, Grunwell JR, et al. Therapeutic plasma exchange in children with thrombocytopenia-associated multiple organ failure: the thrombocytopenia-associated multiple organ failure network prospective experience. *Crit Care Med.* 2019;47(3):e173-e181. doi:10.1097/ccm.0000000000003559
- Nguyen TC. Thrombocytopenia-associated multiple organ failure. *Crit Care Clin.* 2020;36(2):379-39. doi:10.1016/j.ccc.2019.12.010

Content Domain

- Critical Care, Multisystem Failure

Learning Objectives

- Recognize thrombocytopenia-associated multiple organ failure
- Discuss diagnosis and treatment of thrombocytopenia-associated multiple organ failure

The correct answer is: endothelial dysfunction with microthrombi deposition

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Question 3

Correct

1.00 points out of 1.00

Comment

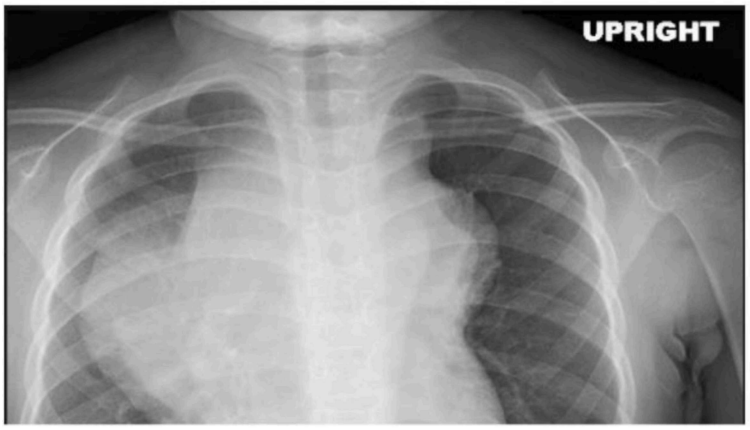
A physician is asked to provide procedural sedation to facilitate chest computed tomography for a 9-year-old female with a history of autism spectrum disorder. She presented to the emergency department with 14 days of progressively worsening cough, sore throat, dyspnea, and right upper quadrant abdominal pain. Her parents report that the girl had a persistent nonproductive cough for the last 3 months. The cough and difficulty breathing are reportedly worse overnight.

A chest radiograph was attempted 2 weeks ago but was cancelled due to patient agitation. The parents state that albuterol improves her work of breathing, although despite its use, the girl had difficulty lying down last night. There is no history of congestion, rhinorrhea, headaches, nausea, vomiting, diarrhea, or rashes.

On examination, while the patient is upright, blood pressure is 114/73 mm Hg, heart rate is 117 beats/min, temperature is 36.2 °C, respiratory rate is 35 breaths/min, and SpO_2 is 94% in room air. When lying down, the patient is noted to have difficulty breathing and hepatosplenomegaly is appreciated. Auscultatory examination reveals normal breath sounds, with mildly decreased air entry at the bilateral bases; no stridor, wheezing, nor rhonchi are appreciated.

Given concerns of the patient's respiratory distress, a chest radiograph is obtained (**Figure**).

Figure. Chest radiograph of the patient in the vignette.



Courtesy of L. Polikoff

Concerning the requested procedural sedation, of the following, the BEST next step is

- A. administer an inhaled β-agonist
- B. administer intramuscular ketamine and place laryngeal mask airway
- C. administer intranasal midazolam for anxiety

D. cancel procedural sedation ✓

Your answer is correct.

PREP Pearl(s)

- Clinicians should be cognizant of clinical examination findings, and symptoms elicited from the patient's history for issues that may compromise respiratory mechanics when considering procedural sedation.
- The American Society of Anesthesiologists has introduced guidelines for sedation and analgesia by non-anesthesiologists that provide specific definitions for procedural sedation.
- Mediastinal masses in pediatric patients confer significant management challenges with sedation.

Critique

Pediatric intensivists are increasingly performing sedation outside of the pediatric intensive care unit. Procedural sedation involves the administration of pharmacological agents to provide both analgesia and anxiolysis while simultaneously ensuring patient immobility to facilitate optimal conditions for a medical procedure. Historically, this practice was known as "conscious sedation," an inaccurate term that should no longer be used because of characterization by vague definitions and diverse interpretations. In 2002, the American Society of Anesthesiologists introduced the practice guidelines for sedation and analgesia by non-anesthesiologists, which provided a more structured and precise framework for procedural sedation. This terminology and guideline adoption marked a pivotal moment in the field, standardizing practices and enhancing the safety and effectiveness of sedation in nonanesthetic contexts. The continuum of sedation for pediatric patients is as follows:

1. Mild sedation (anxiolysis): Intent is anxiolysis with maintenance of consciousness.
2. Moderate sedation: Formerly known as conscious sedation. A controlled state of depressed consciousness during which airway reflexes and airway patency are maintained. Patient responds appropriately to age-appropriate commands (eg, "open your eyes") and light touch.
3. Deep sedation: A controlled state of depressed consciousness during which airway reflexes and airway patency may not be maintained, and the ability to independently maintain ventilatory function may be impaired. The child cannot be easily aroused but responds purposefully following repeated or painful stimulation.
4. General Anesthesia:Loss of consciousness occurs. Patients likely have impaired airway reflexes, airway patency, and ventilatory function. Children are not arousable, even by painful stimulation.

A thorough review of the medical history and a meticulous physical examination are indispensable components in preventing untoward events during pediatric sedation. Clinicians should remain vigilant for signs and symptoms of compromised respiratory mechanics; etiologies may include primary lung parenchymal pathologies, such as viral and bacterial infections, or mechanical obstructions to the bronchial tree that compromise air movement. The constellation of signs and symptoms noted in this vignette should raise concern for airway obstruction due to extraluminal compression, for which a chest radiograph is warranted. Given the patient's history, an anterior mediastinal mass would be high on the differential. Providing sedation might result in loss of the ability to maintain an open airway and result in respiratory arrest. Inhaled β-agonist therapy would offer little help in maintaining airway patency from extrinsic compression.

The presence of a mediastinal mass in a pediatric patient confers significant challenges in airway management, and routinely requires a multidisciplinary approach. As noted in this patient, the trachea or bronchi can be compressed, especially when the patient is supine. Early consultation with pediatric anesthesia and anticipatory preparation for difficult airway management is prudent. If the patient requires endotracheal intubation, techniques such as awake intubation with local anesthetic or the use of a rigid bronchoscope may be considered.

Large mediastinal masses may compress the heart or great vessels, leading to compromised cardiac output or venous return. These patients ideally are best cared for in a center with extracorporeal support. Early consultation with the extracorporeal support team is warranted, given the high risk for cardiac arrest. If a patient experiences cardiac arrest, clinicians should administer chest compressions while the patient is in the prone position, performing compressions in the midline of thoracic spine between T7 and T9, utilizing the same technique as during supine cardiopulmonary resuscitation. Patients with significant tumor burden are also at risk for oncologic emergencies, including electrolyte derangements and renal insufficiency associated with spontaneous tumor lysis syndrome.

Suggested Reading(s)

- Coté C, Wilson S, et al; American Academy of Pediatrics, American Academy of Pediatric Dentistry. Guidelines for monitoring and management of pediatric patients before, during, and after sedation for diagnostic and therapeutic procedures. *Pediatrics*. 2019;143(6):e20191000. doi:10.1542/peds.2019-1000
- Cravero JP, Blike GT, Beach M, et al; Pediatric Sedation Research Consortium. Incidence and nature of adverse events during pediatric sedation/anesthesia for procedures outside the operating room: report from the pediatric sedation research consortium. *Pediatrics* 2006;118(3):1087-1096. doi:org/10.1542/peds.2006-0313
- Verma S, Kalra K, Rastogi S, Sidhu HS. Clinical approach to childhood mediastinal tumors and management. *Mediastinum.* 2020;30;4:21. doi:10.21037/med-19-82

Content Domain

- Medical procedures, Procedural Sedation

Learning Objectives

- Identify contraindications to natural airway sedation

The correct answer is: cancel procedural sedation

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Question 4

Correct

1.00 points out of 1.00

Comment

A 6-year-old child is admitted to the pediatric intensive care unit (PICU) after suffering a cardiac arrest at home. The child was jumping on a trampoline with binoculars while bird-watching. The mother found them unresponsive and laying on the trampoline. She could not detect a pulse. The binocular strap had become twisted and was wrapped around the child's neck. She began cardiopulmonary resuscitation and called emergency medical services (EMS). The child was intubated in the field. The EMS endorses easy visualization of the airway with no resistance in advancing the endotracheal tube and no material in the airway with suctioning. The child's pulse returned after 2 doses of epinephrine.

The child is admitted to the PICU. Assessment at 24 hours shows no purposeful movements or spontaneous eye opening.

Of the following, the MOST likely mechanism of morbidity noted in this patient is

- A. carotid sinus hypersensitivity
- B. cervical spinal cord injury
- C. compression of the jugular vein ✓
- D. obstruction of the airway

Your answer is correct.

PREP Pearl(s)

- Preschool and school-aged children are more prone to accidental neck compression injuries.
- Complete compression of the jugular veins can occur when as little as 5 pounds of force are applied, whereas the carotid arteries and trachea require over 10 and 30 pounds of force, respectively, to completely stop arterial flow or obstruct the airway.
- Obstruction of the bilateral jugular veins increases cerebral venous pressure, and as the carotid arteries are obstructed, the cerebral arterial pressure drops, resulting in hypoxic brain injury.
- Adolescents should be counseled that sexual encounters involving choking or strangulation can have serious consequences.
- Computed tomography angiogram of the carotid arteries should be performed in all strangulation injuries to assess for carotid dissection.

Critique

Young children are at increased risk for strangulation injuries due to the relative laxity of the trachea and lack of neck musculature compared with adolescents and adults. The jugular veins are particularly prone to compression due to their position close to the skin and absence of any structures to prevent their compression when force is applied circumferentially around the neck. Complete compression of the jugular veins can occur when as little as 5 pounds of force are applied, whereas the carotid arteries and trachea require over 10 and 30 pounds of force, respectively, to completely stop arterial flow or obstruct the airway.

Jugular vein compression prevents the outflow of blood from the brain. Obstruction of the bilateral jugular venous system acutely raises intracranial pressure. Initial symptoms may include headache, neck pain, and dyspnea. Complete compression of the jugular veins may cause the child to become unconscious in as little as 10 seconds. As the patient loses consciousness with the ligature in place, neck muscles relax, and the carotid arteries may also become compressed. Cerebral perfusion pressure drops precipitously since it is dependent on the mean arterial pressure minus the central venous pressure. The overall effect is a dramatic decrease in the supply of oxygenated blood to the brain. As a result, the brain becomes hypoxic with neuronal injury and death occurring rapidly if blood flow to the brain is not promptly restored. The rapidity of ischemia and potential brain damage is also a concern in sexual encounters involving choking or strangulation, and adolescent patients should be made aware of the serious consequences of these practices.

As oxygenated blood flow to the brain is compromised, the loss of autonomic control from the brain stem results in profound hypotension and cardiac ischemia. As the ischemia persists, the heart rate slows and eventually asystole occurs. Pressure on the carotid sinus baroreceptors stimulates the parasympathetic nervous system. and is a potential cause of syncope due to pressure on these baroreceptors. The effect of this stimulation is bradycardia, vasodilation and hypotension. However, carotid sinus hypersensitivity is often due to atherosclerosis and calcification of the arterial wall increasing the baroreceptors sensitivity to pressure. Hence, although it can be a potentially fatal result of carotid sinus compression in older adults, it would be quite unlikely in a pediatric patient. Carotid dissection is a rare complication in strangulation injuries. However, a computed tomography angiogram of the carotid arteries is often performed in strangulation injuries to evaluate for signs of this type of injury.

Obstruction of the airway requires significantly more force than compression of the jugular veins; it can occur in hanging injuries. In these cases, the complete obstruction of the airway can result in negative pressure edema and pulmonary dysfunction. Intubation in these cases is often difficult and requires the use of specialized equipment, such as a fiberoptic endoscope to visualize the airway and place an endotracheal tube.

Spinal cord injury is an uncommon finding in pediatric strangulation injuries. Although up to 25% of cases may have a cartilaginous or bony fracture, transection of the spinal cord is only seen when there is significant force applied to the cervical spine with subsequent hyperextension. These are most commonly seen in judicial hangings where the person's body falls from a height of 6 feet or more and then has abrupt deceleration. In these cases the hyperextension causes the pedicles of the C2 vertebrae to fracture with subsequent anterior displacement of the body of the C2 vertebrae relative to C3. As a result, the spinal cord is transected at the level of C2-C3, and there is loss of sympathetic nerve input. Fortunately, this type of injury is rarely seen in pediatric cases.

Suggested Reading(s)

- Davies D, Lang M, Watts R. Paediatric hanging and strangulation injuries: A 10-year retrospective description of clinical factors and outcomes. *Paediatr Child Health.* 2011;16(10):e78-81.
- Hopkins B, Wang A, McKendy K, et al. A retrospective analysis of the clinical use and utility of advanced imaging in the evaluation of near-hanging and strangulation injuries at a Canadian level-1 trauma centre. *Injury.* 2023;54(10):110978. doi:10.1016/j.injury.2023.110978
- Stellpflug SJ, Weber W, Dietrich A, et al. Approach considerations for the management of strangulation in the emergency department. *J Am Coll Emerg Phys Open*. 2022;16;3(2):e12711. doi:10.1002/emp2.12711
- Vilke GM, Chan TC. Evaluation and management for carotid dissection in patients presenting after choking or strangulation. *J Emerg Med.* 2010;40(3):355-358. doi:10.1016/j.jemermed.2010.02.018

Content Domain

- Critical Care, Trauma

Learning Objectives

- Recognize the mechanisms leading to morbidity and mortality in strangulation injuries and the differences in presentation

The correct answer is: compression of the jugular vein

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Question 5

Correct

1.00 points out of 1.00

Comment

A 6-month-old child is intubated and mechanically ventilated for the past 6 days for acute respiratory failure secondary to respiratory syncytial virus bronchiolitis. Over the past 24 hours, the patient has shown signs of improvement and has had satisfactory oxygenation and ventilation without excessive work of breathing, as the level of mechanical ventilation was titrated down to an FiO_2 of 0.35, tidal volume of 6 ml/kg, rate of 14/min, PEEP of 5 cm H_2O, inspiratory time of 0.5 sec, and pressure support of 14 (synchronized intermittent mandatory ventilation pressure-limited volume control mode [SIMV PRVC]).

Of the following, the MOST appropriate next step towards liberation from invasive ventilation is

- A. perform a spontaneous breathing trial ✓
- B. respiratory sprinting with increasing periods of spontaneous supported respiration
- C. wean the ventilatory rate in increments
- D. wean pressure support

Your answer is correct.

PREP Pearl(s)

- Children who are intubated and receiving conventional mechanical ventilation for more than 24 hours should be screened for extubation readiness testing using a protocolized bundle that includes a spontaneous breathing trial.
- A spontaneous breathing trial is defined as a systematic method of reduction of invasive mechanical ventilation support to predetermined settings to assess the likelihood that a patient will be able to independently maintain adequate minute ventilation and gas exchange without excessive respiratory effort, if liberated from invasive mechanical ventilation.
- Extubation readiness testing is defined as a bundle of elements used to assess the patient's eligibility to be liberated from invasive mechanical ventilation.
- There is no evidence to support one ventilator weaning technique over another.

Critique

The patient described in the vignette has stabilized, shown improvement, and they have tolerated a reduction in ventilatory support. This suggests that the patient has transitioned from the acute phase of ventilation into the weaning phase. Weaning is the process by which positive pressure ventilation is decreased and the patient becomes increasingly responsible for generating the energy required for effective gas exchange. Once the patient has reached the weaning phase, liberation from invasive ventilation should be targeted to reduce the duration of mechanical ventilation and complications associated with intubation and ventilation. Weaning may continue until the patient is liberated from respiratory support, or requires noninvasive respiratory support, such as noninvasive ventilation, CPAP, negative pressure ventilation, or high-flow nasal cannula.

There are no accepted criteria that mark the start of the weaning process. Not all patients need to be weaned; some patients can be successfully extubated and liberated from invasive ventilation once they are over the acute phase. There is no evidence to support one weaning technique over another. All the methods described in the vignette have been used to wean patients; these include gradually reducing the number of mandatory breaths in SIMV mode, switching to a spontaneous mode, such as CPAP with PS or volume support, and reducing the amount of positive pressure, or alternating periods of spontaneous ventilation with a time-cycled mode and possibly increasing the amount of time on the spontaneous mode.

Recently, the Pediatric Acute Lung Injury and Sepsis Investigators (PALISI) Network recommended a protocolized screening process for determining if a patient is ready for liberation from invasive ventilation. This includes performing SBT as part of determining extubation readiness testing (ERT). A spontaneous breathing trial (SBT) is defined as a systematic method of reduction of invasive mechanical ventilation support to predetermined settings to assess the likelihood that a patient will be able to independently maintain adequate minute ventilation and gas exchange without excessive respiratory effort if liberated from invasive mechanical ventilation. This differs from weaning where the reduction in ventilator settings occurs in smaller increments over a longer period of time. Extubation readiness testing is defined as a bundle of elements used to assess the patient's eligibility to be liberated from invasive mechanical ventilation. This includes SBT as well as assessment of the level of sedation, adequacy of airway protective reflexes, likelihood of post-extubation upper airway obstruction, assessment of respiratory muscle strength, the magnitude of airway secretions, hemodynamic status, and a plan for post-extubation respiratory support.

In the above vignette, the patient has transitioned into the weaning phase. The most appropriate next step is to perform SBT as part of ERT. If the patient passes SBT and ERT, he or she may be extubated to the planned post-extubation respiratory support. If the patient does not pass the spontaneous breathing trial, he or she should be weaned using any of the techniques described above and screened for ERT daily.

Suggested Reading(s)

- Abu-Sultaneh S, Iyer NP, Fernández A, et al. International clinical practice guidelines for pediatric ventilator liberation, a Pediatric Acute Lung Injury and Sepsis Investigators (PALISI) Network Document. *Am J Respir Crit Care Med.* 2023;207(1):17-28. doi:10.1164/rccm.202204-0795so
- Abu-Sultaneh S, Iyer NP, Fernández A, et al; Pediatric Acute Lung Injury and Sepsis Investigators (PALISI) Network. Operational definitions related to pediatric ventilator liberation. *Chest.* 2023;163(5):1130-1143. doi:10.1016/j.chest.2022.12.010
- van Dijk J, Blokpoel RGT, Abu-Sultaneh S, Newth CJL, Khemani RG, Kneyber MCJ. Clinical challenges in pediatric ventilation liberation: a meta-narrative review. *Pediatr Crit Care Med.* 2022;23(12):999-1008. doi:10.1097/pcc.0000000000003025

Content Domain

- Critical Care, Medical Procedures (advanced)

Learning Objectives

- Manage weaning of ventilatory support and liberation from invasive ventilation

The correct answer is: perform a spontaneous breathing trial

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Question 6

Correct

1.00 points out of 1.00

Comment

A 17-year-old adolescent presents with bradycardia and syncope. In the emergency department, she appears emaciated with weight of 30 kg (50th percentile for a 9-year-old), heart rate of 42 beats/min, and blood pressure 115/70 mm Hg. Bedside cardiac telemetry reveals sinus arrhythmia and bradycardia. Her physical examination is notable for generalized emaciated appearance, with little to no body fat, accentuated rib and facial bones, and low muscle mass throughout. There is significant lanugo of her arms, back, and face. Further history reveals extensive mental health history including prior hospital admissions for anorexia nervosa. She is admitted to the pediatric intensive care unit and started on parenteral fluids containing 10% dextrose at 75 mL/hr. Eighteen hours later, she develops general confusion and weakness with altered mental status.

Of the following, the BEST next step in evaluation and management is

- A. computed tomography scan of the brain
- B. echocardiogram
- C. electroencephalogram
- D. serum electrolyte panel ✓

Your answer is correct.

PREP Pearl(s)

- Patients with starvation or malnutrition are at risk for refeeding syndrome when carbohydrates, including parenteral dextrose, are reintroduced.
- Severe electrolyte derangements including hypokalemia, hypophosphatemia, and hypomagnesemia can cause life-threatening complications in patients with refeeding syndrome; thiamine should be provided to all patients at risk for refeeding syndrome.
- A slow, progressive introduction of carbohydrates is warranted in patients at risk for refeeding syndrome, and frequent electrolyte assessments should be performed with judicious electrolyte replacements.

Critique

The patient in the vignette presents with emaciation/starvation as a result of an underlying eating disorder. Nutrition is initiated with parenteral dextrose, and she then develops neurologic abnormalities. This scenario is suggestive of refeeding syndrome. Refeeding syndrome is a potentially fatal metabolic disorder that occurs when nutrition is reintroduced too rapidly in malnourished patients. It can occur with any type of nutritional replacement (enteral or parenteral), and thus even patients who are not yet enterally fed are at risk for refeeding syndrome when carbohydrates (including parenteral dextrose) are introduced. Refeeding syndrome is associated with a pattern of abnormal laboratory findings, including serum hypophosphatemia, hypokalemia, and hypomagnesemia, which are highly suggestive of this diagnosis; thus, a serum electrolyte panel is the best next step in the evaluation and management of this patient.

Refeeding syndrome is a constellation of findings related to the metabolic response to the introduction of micro and macronutrients in a patient who has been starved or is severely malnourished. Patients who have not had adequate nutrition are in a state of catabolism, and refeeding syndrome is thought to be an exaggerated physiological response to reintroducing glucose (refeeding) after a prolonged phase of starvation. Electrolyte and nutritional derangements that occur with refeeding syndrome include increased serum glucose, hypophosphatemia, hypokalemia, hypomagnesemia, thiamine depletion, and fluid and sodium retention, with resulting impaired end-organ function and cardiac arrhythmias.

In a severe catabolic state, insulin production is decreased, whereas glucagon and catecholamines are stimulated. Glycogen stores are depleted, leading to activation of gluconeogenesis and the production of glucose from endogenous proteins via proteolysis. This results in muscle-wasting and thus functional weakness and weight loss. Vitamin and electrolyte levels are decreased and stores are depleted. Lipolysis increases, causing an increased concentration of serum free fatty acids. This activates ketogenesis, leading to high production of acetoacetate and β-hydroxybutyrate. When carbohydrates are reintroduced, hyperglycemia is seen; consequently, there is an increase in insulin secretion. A transition to anabolic processes occurs, leading to intracellular shift of glucose, water, and electrolytes, and resulting in a potentially severe drop in serum electrolytes and micronutrient concentrations. The resulting electrolyte imbalances, namely hypophosphatemia, hypokalemia, and hypomagnesemia can cause life-threatening arrhythmia, muscle spasms, or tetany. As the intracellular shift of glucose is thiamine dependent, thiamine deficiency can lead to symptoms of beriberi, including heart failure. The more severely compromised the overall nutritional state, the higher the risk of refeeding syndrome.

Patients at risk of developing refeeding syndrome need replenishment of carbohydrates, electrolytes, and vitamins (especially thiamine) as part of their resuscitation from starvation. Carbohydrates should be reintroduced in a slow stepwise fashion and increased slowly to goal caloric intake over the course of multiple days. Excessive (including typically normal daily maintenance requirement) early administration of glucose can lead to complications, including hyperglycemia, and consequently to an osmotic diuresis, dehydration with metabolic acidosis, hyperosmotic coma, ketoacidosis, along with an increased respiratory quotient with excessive production of carbon dioxide, hypercapnia, and respiratory failure. Serum electrolytes should be closely monitored and replaced to achieve normal serum levels of potassium, calcium, magnesium, and phosphate. Because of sodium and water retention, fluid management can be challenging, and it may need to be restricted initially with close monitoring of overall fluid balance, weight change, and serum sodium concentration. Thiamine should be administered in all cases. The full resting energy requirements should be achieved within 5 to 10 days of presentation, depending on the initial risk of refeeding syndrome using a balanced nutritional mix of 40% to 60% carbohydrates, 30% to 40% fats, and 15% to 20% proteins.

Computed tomography of the brain may be required in patients with the development of acute altered mental status, particularly if there are concerns for cerebral edema as in patients with diabetic ketoacidosis. However, in this vignette, evaluation of fluid and electrolyte status in a malnourished patient will likely identify the underlying etiology and allow for provision of therapy to address the cause of her mental status change. Electroencephalogram and echocardiogram are unlikely to reveal the underlying cause of an acute mental status change in a patient with a primarily metabolic condition as their underlying disorder, and thus would not be the preferred response.

Suggested Reading(s)

- Boal AH, Panarelli M, Millar C. Starvation ketoacidosis and refeeding syndrome. *BMJ Case Rep.* 2021;14(12):e245065. doi:10.1136/bcr-2021-245065
- da Silva JSV, Seres DS, Sabino K, et al; Parenteral Nutrition Safety and Clinical Practice Committees, American Society for Parenteral and Enteral Nutrition. ASPEN consensus recommendations for

refeeding syndrome. *Nutr Clin Pract*. 2020;35(2):178-195. doi:10.1002/ncp.10474
- Reber E, Friedli N, Vasiloglou MF, Schuetz P, Stanga Z. Management of refeeding syndrome in medical inpatients. *J Clin Med.* 2019;8(12):2202. doi:10.3390/jcm8122202
- Runde J, Sentongo T. Refeeding syndrome. *Pediatr Ann.* 2019;48(11):e448-e454. doi:10.3928/19382359-20191017-02

Content Domain

- Behavior/Development, Eating Disorders

Learning Objectives

- Identify common electrolyte abnormalities associated with refeeding syndrome
- Plan the management of patients at risk for refeeding syndrome
- Identify clinical complications associated with initiation of nutrition to malnourished patients

The correct answer is: serum electrolyte panel

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