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Duration	2 mins 10 secs
Points	6.00/6.00
Grade	**100.00** out of 100.00
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Question 1

Correct

1.00 points out of 1.00

Comment

A 4-year-old child with a history of developmental delay, seizures, and hearing loss was found to be seizing by their father. Emergency medical services administered intravenous (IV) lorazepam and transported the child to the emergency department.

On arrival, the child has a depressed mental status and continued clinical seizures. The child is given 60 mg/kg IV levetiracetam, followed by 20 mg/kg IV phenobarbital for persistent seizures. They have a temperature of 37.5 °C, heart rate of 154 beats/min, blood pressure 111/69 mm Hg, respiratory rate of 10 breaths/min, and oxygen saturations of 90% on 100% supplemental oxygen by nonrebreather mask. The child is intubated for airway protection and transferred to the pediatric intensive care unit, where an electroencephalogram shows continued status epilepticus. A propofol infusion is initiated and titrated to burst suppression. The seizures stop, and the child is maintained on a propofol infusion at 8 mg/kg/hr (133.3 mcg/kg/min) for 48 hours.

Reassessment of vital signs today shows a temperature 37.0 °C, heart rate of 48 beats/min, blood pressure of 84/40 mm Hg, respiratory rate of 16 breaths/min, and oxygen saturations of 99% on a FiO_2 of 0.4 by mechanical ventilator. On examination, the child's heart has a regular rate and rhythm with normal S1, S2. Lungs are clear to auscultation bilaterally, and the abdomen is soft and nontender with hepatomegaly.

Laboratory data are shown:

Laboratory Test	Result
Venous blood gas	
pH	7.25
PCO_2	42 mm Hg
Bicarbonate	12 mmol/L
Base deficit	-12 mmol/L
White blood cell count	9,800/µL (9.8 x 10^9/L)
Hemoglobin	11.2 g/dL (112.0 g/L)
Hematocrit	33.6%
Platelet	160,000/µL (160 x 10^9/L)
Creatine kinase	2,460 U/L
Triglyceride	660 mg/dL

Of the following, the BEST next step is to

- A. administer an IV sodium bicarbonate bolus
- B. discontinue the propofol infusion ✓
- C. increase the respiratory rate on the mechanical ventilator
- D. start broad-spectrum IV antibiotics

Your answer is correct.

PREP Pearl(s)

- Propofol is approved by the US Food and Drug Administration for induction of anesthesia in children aged 3 years or older, and for maintenance of anesthesia in children at least 2 months of age.
- Propofol-related infusion syndrome consists of refractory bradycardia with hepatomegaly or fatty liver, hyperlipidemia, metabolic acidosis, or rhabdomyolysis or myoglobinuria.
- There is an increased risk of propofol-related infusion syndrome in children who receive propofol infusions for more than 48 hours and at doses higher than 4 mg/kg/hr (66 mcg/kg/min).

Critique

Propofol, 2,6-diisopropylphenol, is an intravenous (IV) hypnotic anesthetic that is valued for its rapid onset and short half-life. It is insoluble in water, and therefore is prepared in soybean oil with an emulsifier (egg lecithin) which gives it its white color. Propofol is used for anesthesia, sedation, and more recently as an anticonvulsant. Its mechanism of action is through binding of γ-aminobutyric acid (GABA)-A receptors which leads to prolonged opening of chloride channels in the postsynaptic neurons that leads to hyperpolarization. Propofol is approved by the US Food and Drug Administration (FDA) for induction of anesthesia in children aged 3 years or older, and for maintenance of anesthesia in children at least 2 months of age. These restrictions are in place due to the risks of propofol-related infusion syndrome and death in children.

Propofol-related infusion syndrome was first described in 1990 in a 2-year-old sedated with propofol for croup. While more commonly discussed in the pediatric population, propofol-related infusion syndrome has been reported in adults. Propofol-related infusion syndrome is comprised of refractory bradycardia in conjunction with hepatomegaly or fatty liver, hyperlipidemia, metabolic acidosis, or rhabdomyolysis or myoglobinuria. Additional clinical features include hypotension, Brugada-like electrocardiogram pattern with convex-curved ST elevation in the precordial leads, arrhythmias, asystole, or death, as well as lactic acidosis, renal failure, hyperkalemia, transaminitis, hypertriglyceridemia, and hyperthermia.

There is an association of propofol-related infusion syndrome in patients who receive propofol infusions for more than 48 hours and at doses higher than 4 mg/kg/hr (66 mcg/kg/min). Additional risk factors include young age, inadequate carbohydrate intake, critical illness, corticosteroid use, vasopressor use, and inborn errors of metabolism. The etiology of development of propofol-related infusion syndrome is unclear, but thought to be related to inhibition of the mitochondrial respiratory chain or impaired mitochondrial fatty acid metabolism. Propofol is contraindicated in patients that have mitochondrial disorders.

Treatment of propofol-related infusion syndrome begins with prompt discontinuation of propofol. Cardiac support, including addition of inotropes, vasopressors, cardiac pacing, or use of extracorporeal membrane oxygenation, have been described. Management of metabolic and lactic acidosis have included sodium bicarbonate, and hemodialysis or hemofiltration, which additionally provide lipid removal. The correct answer in this scenario is discontinuation of propofol infusion. The patient described does not have sepsis, therefore addition of broad-spectrum antibiotics is incorrect. Increasing the respiratory rate on the mechanical ventilator will not correct the metabolic acidosis. Administration of sodium bicarbonate may help correct the metabolic acidosis, but should not precede discontinuation of the propofol infusion.

Suggested Reading(s)

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- Singh A, Anjankar AP. Propofol-related infusion syndrome: a clinical review. *Cureus.* 2022;14(10):e30383. doi:10.7759/cureus.30383

Content Domain

- Critical Care, Sedation and Analgesia

Learning Objectives

- Identify the mechanism of action of propofol
- Identify propofol-related infusion syndrome and its risk factor

The correct answer is: discontinue the propofol infusion

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Question 2

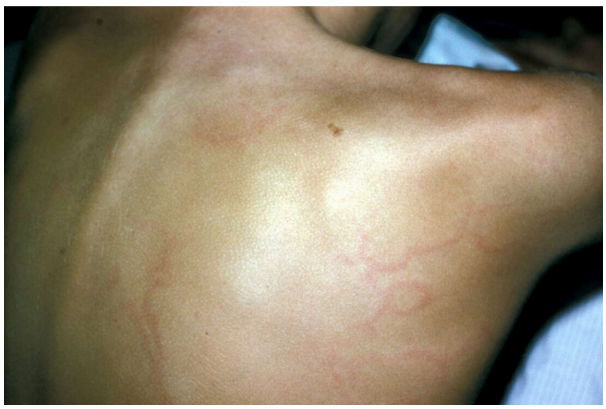
Correct

1.00 points out of 1.00

Comment

A 9-year-old child presents with 3 days of fever, fatigue, and palpitations. She recently immigrated from South America, and her medical history and vaccination status is unknown. On physical examination, her vital signs include a temperature of 39.4 °C, heart rate of 150 beats/min, respiratory rate of 20 breaths/min, blood pressure of 95/65 mm Hg, and oxygen saturation of 99% in room air. Lungs are clear throughout. There is erythema and edema with tenderness of the left knee and right elbow, which she stated also involved the right knee until today. She has a pansystolic murmur at the cardiac apex. Abdominal examination is benign. Her skin is notable for a well-defined pink macular rash with irregular borders and central clearing on her trunk and legs (**Figure**). An echocardiogram reveals mitral regurgitation with moderate left ventricular enlargement. She is admitted to the pediatric intensive care unit for further evaluation and management.

Figure. Rash on trunk.



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Of the following, the BEST medication regimen for initial management is

A. doxycycline

B. parenteral penicillin and corticosteroids ✓

C. parenteral vancomycin and ceftazidime

D. rituximab

Your answer is correct.

PREP Pearl(s)

- Acute rheumatic fever is an inflammatory condition that can occur as a delayed immune response to a group A *Streptococcus* infection.

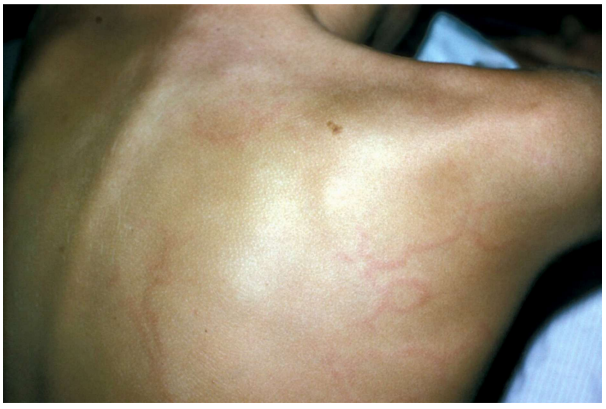
- The diagnosis of acute rheumatic fever requires evidence of a recent streptococcal infection and evidence of some combination of major and minor Jones criteria, including carditis, arthritis, chorea, erythema marginatum, and subcutaneous nodules.
- The mainstay of initial therapy for acute rheumatic fever includes eradication of the inciting group A *Streptococcus* infection using penicillin, and corticosteroids for patients with rheumatic carditis.

Critique

The child in the vignette has an acute febrile illness associated with migratory polyarthritis, carditis with mitral valve regurgitation, and a characteristic rash which raises high suspicion for acute rheumatic fever (ARF). Current recommendation for initial therapy for acute rheumatic fever includes the administration of penicillin and corticosteroids. Doxycycline is first-line antibiotic therapy used for tick-borne illness including lyme disease, which is caused by *Borrelia burgdorferi.* A characteristic rash (ie, erythema migrans) often occurs at the site of the tick bite, and is accompanied by systemic symptoms including malaise, fatigue, headache, arthralgias, myalgias, fever, and regional lymphadenopathy. Erythema migrans is a rash with a bright red border, partial central clearing, and a bull's eye center; it is the presenting manifestation of borreliosis in most patients in the United States. Rituximab is a treatment option for children with systemic juvenile idiopathic arthritis (SJIA) refractory to other therapies. Rituximab is a chimeric monoclonal anti-CD20 B-cell antibody often used to treat certain vasculitis and rheumatologic syndromes. The child in the vignette has arthritis, however, the other features in the vignette make a diagnosis of systemic juvenile idiopathic arthritis less likely; rituximab would not be first-line therapy. Parenteral vancomycin and ceftazidime would be indicated for children with acute bacterial infections (including infectious endocarditis), however, based on the clinical vignette and associated symptoms, a diagnosis of ARF is more likely.

Acute rheumatic fever is an inflammatory condition that can occur as a delayed immune response to a group A *Streptococcus* infection, or often, untreated streptococcal pharyngitis. The classic presentation includes the combination of fever, joint pain (arthritis), and carditis, often accompanied by other manifestations, such as erythema marginatum (**Figure**), subcutaneous nodules, and Sydenham chorea. The diagnosis of rheumatic fever is made using the Jones criteria (**Table**). The Jones criteria were first established in 1944, and since then have undergone multiple modifications, revisions, and updates, most recently in 2015. The major Jones criteria include carditis, arthritis, chorea, erythema marginatum, and subcutaneous nodules. The minor Jones criteria include mono/polyarthralgia, fever, and elevated acute phase reactants (including erythrocyte sedimentation rate, C-reactive protein, and prolonged PR interval). The diagnosis of ARF is made when patients demonstrate 2 major Jones criteria or 1 major and at least 2 minor criteria. In addition, evidence of preceding infection with group A *Streptococcus* must be shown, which is usually done using streptococcal serology.

Figure. Rash on trunk.



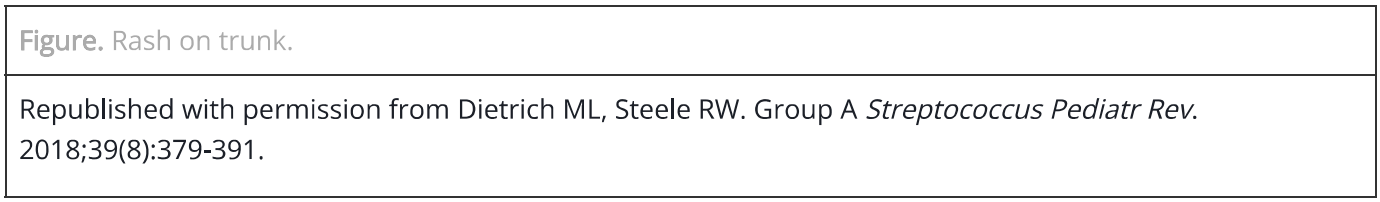


Figure. Rash on trunk.

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The most common clinical manifestations include large joint migratory arthritis and/or arthralgia, usually with fever, and a pansystolic murmur of mitral regurgitation. A recent streptococcal infection is usually endorsed, or in some scenarios, a recent pharyngitis without evaluation by medical practitioners raises concern for an untreated streptococcal pharyngitis. Primary prevention of ARF is through the eradication of group A *Streptococcus* infections, including streptococcal pharyngitis or impetigo. Secondary prevention is used to prevent the recurrence of ARF and the progression of rheumatic heart disease; it consists of prophylactic antibiotics in patients with previous episodes of rheumatic fever or those with prior history of rheumatic heart disease. Typically, an intramuscular injection of benzylpenicillin G every 4 weeks for at least 5 to 10 years (or until patients are in their early 20s) is required.

Suggested Reading(s)

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- Ralph AP, Noonan S, Wade V, Currie BJ. The 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease. *Med J Aust.* 2021;214(5):220-7. doi:10.5694/mja2.50851

Content Domain

- Infectious Diseases, Inflammatory disease

Learning Objectives

- Plan the therapy for acute rheumatic fever
- Identify the signs and symptoms of rheumatic fever

The correct answer is: parenteral penicillin and corticosteroids

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Question 3

Correct

1.00 points out of 1.00

Comment

A 15-year-old adolescent is admitted to the pediatric intensive care unit following presentation to the emergency department for a 1-week history of increasing confusion, extreme fatigue, and weakness. His mother further reports a 1-month history of frequent headaches, blurred vision, and occasional dizziness. She also mentions that his clothes are fitting looser, but she attributes this to a recent attempt to improve his eating habits. He has no significant medical history, does not take any medications, and is not currently being followed by a physician. On examination, the patient arouses briefly to noxious stimuli and mumbles incoherently, with dry mucous membranes, a capillary refill time of 4 seconds, and 1+ pulses throughout. His heart rate is 128 beats/min, blood pressure is 105/60 mm Hg, respiratory rate is 22 breaths/min, oxygen saturation is 100% in room air, and temperature is 36.2 °C. He receives 1 L of isotonic intravenous fluids, with slight improvement in his perfusion. Laboratory data are shown:

Laboratory Test	Result
Sodium	148 mEq/L (148 mmol/L)
Potassium	4.0 mEq/L (4.0 mmol/L)
Chloride	104 mEq/L (104.0 mmol/L)
Bicarbonate	17 mEq/L (17 mmol/L)
Urea nitrogen	40 mg/dL (2.38 mmol/L)
Serum creatinine	1.9 mg/dL (167.9 mmol/L)
Glucose	1,100 mg/dL (61.05 mmol/L)
Venous pH	7.32
β-hydroxybutyrate	1.9 mmol/L
Serum osmolality	380 mOsm/kg
Urinalysis	
Ketones	Trace
Glucose	4+

Of the following, the MOST likely underlying diagnosis contributing to this patient's presentation is

A. acute kidney failure with uremic encephalopathy and hyperglycemia

B. adrenal insufficiency with stress-induced hyperglycemia

C. diabetic ketoacidosis

D. hyperosmolar hyperglycemic state ✓

Your answer is correct.

PREP Pearl(s)

- Hyperosmolar hyperglycemic state is a life-threatening complication of type 2 diabetes mellitus characterized by severe hyperglycemia, dehydration, hyperosmolality, and altered mental status without significant ketoacidosis.
- The key laboratory findings in hyperosmolar hyperglycemic state include extreme hyperglycemia (usually >600 mg/dL), elevated serum osmolality (>320 mOsm/kg), normal pH, absent or minimal ketones, and normal β-hydroxybutyrate.
- Hyperosmolar hyperglycemic state carries a higher mortality risk compared with diabetic ketoacidosis due to delayed diagnosis and the severity of dehydration.
- Patients with hyperosmolar hyperglycemic state require close neurologic observation during volume resuscitation and administration of lower insulin infusion rates to correct metabolic disturbance compared with patients with diabetic ketoacidosis.

Critique

This patient's presentation of extreme hyperglycemia, high serum osmolality, and dehydration without significant metabolic acidosis, nor elevated β-hydroxybutyrate and ketonemia is most consistent with hyperosmolar hyperglycemic state (HHS), often associated with type 2 diabetes. In HHS, there is some residual endogenous insulin function, preventing the significant lipolysis and subsequent ketogenesis that occur in diabetic ketoacidosis (DKA). However, there is an insufficient amount of insulin to prevent the profound hyperglycemia, leading to severe osmotic diuresis and profound dehydration. Subsequent electrolyte derangements associated with this include profound hyperosmolality, causing rapid decrease in intracellular water from brain cells; this gives rise to the associated neurologic symptoms. Moreover, this osmotic diuresis may also give rise to acute kidney injury, due to reduced intravascular volume, and subsequent decreased renal perfusion.

Differentiating HHS from DKA is crucial; although clinically there may be overlap with DKA, the extreme hyperglycemia and the absence of ketones and severe increased-gap metabolic acidosis and normal β-hydroxybutyrate are key features that differentiate DKA from HHS. The initial goal of management of HHS is restoring and maintaining intravascular volume; as such, patients require early and aggressive fluid resuscitation, with eventual administration of low-dose insulin infusions once glucose levels stabilize following fluid resuscitation, compared with the higher insulin infusion rates needed for DKA. The mortality rate in HHS can be as high as 20%, potentially due to under-recognition in pediatric patients. Patients with HHS require close neurologic observation during volume resuscitation and administration of lower insulin infusion rates to correct metabolic disturbance compared with patients with DKA. Patients with HHS are also at severe risk for cerebral edema, and may require hyperosmolar therapy.

While DKA is common in hyperglycemic emergencies, the lack of ketones, normal bicarbonate, and normal pH rule out this diagnosis. Moreover, patients with DKA usually present with an increased anion gap metabolic acidosis and ketonemia, which are absent here. The patient's elevated urea nitrogen and creatinine

are suggestive of acute kidney injury, secondary to the severe dehydration from the osmotic diuresis, rather than primary renal disease. While uremic encephalopathy can cause confusion, it does not explain the severe hyperglycemia and hyperosmolality. Adrenal insufficiency can cause fatigue, confusion, and hypotension, and stress-induced hyperglycemia can occur, but it rarely leads to such extreme levels of hyperglycemia seen in the vignette. Moreover, adrenal insufficiency more commonly presents with hyponatremia, rather than hypernatremia, the latter of which is more consistent with dehydration and hyperosmolality in HHS.

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Content Domain

- Endocrinology, Diabetes Mellitus

Learning Objectives

- Differentiate hyperglycemic hyperosmolar syndrome (HHS) from diabetic ketoacidosis.

The correct answer is: hyperosmolar hyperglycemic state

View Peer Results

Question 4

Correct

1.00 points out of 1.00

Comment

An 8-month-old infant is admitted to the intensive care unit (ICU) for persistent stridor and feeding intolerance. Racemic epinephrine and dexamethasone were administered, and helium/oxygen mixture was started with minimal improvement. This is the infant's second admission to the ICU for stridor. The mother said that while the noisy breathing has persisted for about 3 months, it worsened in the last few weeks. She has attributed it to a recent upper respiratory tract infection, and has intermittently administered a bronchodilator without any improvement in respiratory symptoms. On examination, the infant is anxious but easily consoled, with mildly increased work of breathing and minimal secretions. The infant has good air movement bilaterally despite the stridor, and a viral respiratory panel is negative. Bedside nasopharyngoscopy demonstrates normal supraglottic anatomy and vocal fold function. Chest radiograph is normal.

Of the following, the BEST study to definitively diagnose this infant's condition is

- A. computed tomography angiography of the chest ✓
- B. positron emission tomography scan
- C. pulmonary function test
- D. upper gastrointestinal series

Your answer is correct.

PREP Pearl(s)

- An anatomic abnormality should be suspected in children with repeated admissions for recurrent stridor with no associated infection.
- A computed tomography angiography scan of the chest is a rapid and useful test to diagnose a vascular ring.
- After computed tomography angiography, further testing (genetic evaluation, echocardiogram) should be performed in children with vascular rings, given its association with other congenital conditions.

Critique

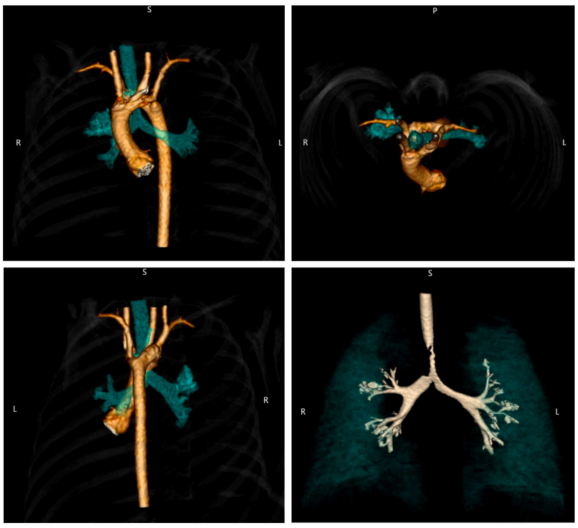
The cause of this infant's current symptoms is a vascular ring. Vascular rings are abnormalities in the position and/or branching of the vessels of the aortic arch. Rings can be complete or incomplete. The clinical manifestations of a vascular ring arise from extrinsic airway compression, resulting in stridor and potentially malacia. In rare cases, tracheal stenosis due to complete cartilaginous rings can be found in association with left pulmonary artery sling.

Presentations of a vascular ring include stridor, apnea, recurrent respiratory infections, barky cough, and respiratory distress, typically in the first year after birth. Some children might be mistakenly diagnosed with asthma and prescribed bronchodilators. Patients with esophageal compression may also have the additional symptoms of dysphagia and failure to thrive.

Some of the common anatomic anomalies resulting in a ring are:

1. Double aortic arch; this is when the ascending aorta divides into two branches, which encircle the trachea and esophagus, and then reunite on the left to form the descending aorta. This variation is considered a complete vascular ring and produces the most severe symptoms. The **Figure** shows imaging of a double aortic arch, with compression of the trachea evident in the last reconstructed image.

Figure. Imaging finding of a double aortic arch.



Courtesy of A. Ruth

2. Left pulmonary artery sling; in this anomaly, the left pulmonary artery arises from the right pulmonary artery and traverses right to left between the trachea and esophagus. Clinical symptoms result from the compression of the right mainstem bronchus and/or trachea.

3. Single aortic arch with aberrant subclavian artery; the level of compression is variable depending on where the aberrant artery crosses, producing different symptoms depending on whether the compression is of the esophagus (dysphagia) or distal airway (stridor). In many cases, this anomaly is asymptomatic.

Radiologic workup may start with a chest radiograph, which can reveal the location of the aortic arch. In a right aortic arch, the compression on the right side of the trachea could potentially be detected in imaging. Historically, contrast esophagrams have been performed to delineate the degree and location of esophageal compression. Currently, the preferred modality for diagnosis is either a computed tomography scan or a magnetic resonance image. A computed tomography angiogram (CTA) can rapidly provide images that can accurately identify the vascular anomaly as well as the degree of compression. While magnetic resonance imaging can also show the anatomy without the use of contrast, it is a much longer study and may require securing the airway of a patient. Bronchoscopy might also be performed for diagnostic evaluation of the vascular ring, but it is not the initial modality of choice, as a CTA is generally easier to obtain. If a bronchoscopy were undertaken prior to diagnosis, it may show narrowing at certain parts of the airway, and should then prompt further imaging. Of note, echocardiography should be performed in children with a diagnosis of vascular ring, as this anomaly is frequently associated with other congenital heart defects. The risk of concomitant congenital heart disease is higher in those with a right aortic arch, with the most common condition being conotruncal anomalies. Aortic arch anomalies and vascular rings are also associated with the 22q11.2 deletion syndrome.

Treatment of vascular rings is surgical repair. Multiple studies examining outcomes show that surgical correction for vascular rings is safe, with very low mortality rates. Reports of long-term outcomes of repair however, are still varied, with some studies showing little residual symptoms and others reporting a significant number of patients continuing to experience post-repair respiratory symptoms and needing reoperation.

The infant in the vignette would benefit from a CTA to visualize their vascular ring. A positron emission tomography scan will not be useful in diagnosing their anatomic abnormality. Similarly, a pulmonary function test will not be diagnostic, and this infant is too young to perform the test.

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Content Domain

- Pulmonology, pulmonary complications from other diseases

Learning Objectives

- Recognize presentation of congenital vascular abnormalities
- Identify preferred imaging modality to diagnose a vascular ring

The correct answer is: computed tomography angiography of the chest

View Peer Results

Question 5

Correct

1.00 points out of 1.00

Comment

A 27-month-old child with an apnea-hypopnea index of 10 is admitted to the pediatric intensive care unit after scheduled tonsillectomy and adenoidectomy. The child weighs 20 kg and had a prolonged operative course due to difficulty placing a peripheral intravenous catheter. The child had initially been placed on 1 L/min of oxygen via nasal cannula when they first returned from the operative suite, but they are now awake and require no respiratory support. The child is now asking for water and keeps climbing out of bed. The mother is requesting that they be discharged home where she would not have to keep placing the child in bed. She states that her older child went home the same day after their tonsillectomy.

Of the following, the BEST recommendation for this child is

- A. continue cardiopulmonary observation in the pediatric intensive care unit ✓
- B. discharge home if Aldrete criteria have been met
- C. discharge home if oxygen saturation remains >95% during nap
- D. transfer patient to pediatric service

Your answer is correct.

PREP Pearl(s)

- Hyperactivity has been associated with sleep apnea in children.
- Obesity can worsen sleep apnea but is difficult to treat in this age group.
- Patients with adenotonsillar hypertrophy are at increased risk for worsening obstructive sleep apnea immediately after tonsillectomy; up to 10% of patients younger than age 3 years develop complications following tonsillectomy.

Critique

Children younger than the age of 3 with obstructive sleep apnea are at increased risk for complications after their adenotonsillectomy. Despite administration of intraoperative steroids, the manipulation of the airway and postanesthetic effects on pharyngeal tone increase the likelihood of airway obstruction and apneic events. Overnight observation is also warranted if the child has any existing comorbidities that would increase the likelihood of sleep apnea or sleep disordered breathing. Some examples of conditions that should be admitted for close observation include Beckwith-Wiedemann syndrome, trisomy 21, Crouzon syndrome, or any other condition that is associated with hypotonia and/or midline facial abnormalities.

The Aldrete score addresses five parameters: respiration, circulation, consciousness, color, and activity with a maximum score of 10. This score is used to determine if a child has sufficiently recovered from anesthesia or procedural sedation before moving them to a lower level of monitoring. However, in this vignette, even if the child had an Aldrete score of 10, they would remain at risk for complications.

In a 2006 study of 2,315 children younger than age 6 years who underwent adenotonsillectomy for obstructive sleep apnea, the complication rate in children 3 years of age and younger was 10%. The most common complication was oxygen desaturation requiring oxygen or airway intervention. Approximately one third of these children required placement of a nasopharyngeal airway, and 8.8% required intubation. Hence, even if the child has an Aldrete score of 10 after adenotonsillectomy, the child should remain where prompt airway intervention can occur.

A decreasing level of care may be possible in hospitals that offer continuous pulse oximetry and cardiopulmonary monitoring on their pediatric service, but these patients have a high potential for decompensation. Patients with sleep-disordered breathing have been noted to have higher rates of hyperactivity. However, this association is controversial, and there is disagreement as to whether hyperactivity is a true disorder or the result of poor sleep hygiene and subsequent difficulty in concentrating on tasks. A meta-analysis found that children identified as hyperactive had an improvement in their symptoms over the next 2 to 13 months following adenotonsillectomy. In some cases, it may be necessary to give medication such as clonidine or dexmedetomidine to keep the patient sufficiently quiet to ensure monitoring equipment (eg, cardiac telemetry, pulse oximetry) is kept in place.

The ability to maintain an appropriate level of oxygenation while asleep is a common discharge criteria for conditions such as bronchiolitis and pneumonia. However, a short nap without desaturation immediately following the surgery would not allow enough time for any delayed airway swelling to occur. Hence, the recommendation is for at least a full 24 hours of observation to ensure any later onset changes to the airway do not increase airway obstruction and subsequent apnea.

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Content Domain

- Pulmonology, Sleep Disordered Breathing

Learning Objectives

- Recognize which children with obstructive sleep apnea require observation after surgery.

The correct answer is: continue cardiopulmonary observation in the pediatric intensive care unit

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Question 6

Correct

1.00 points out of 1.00

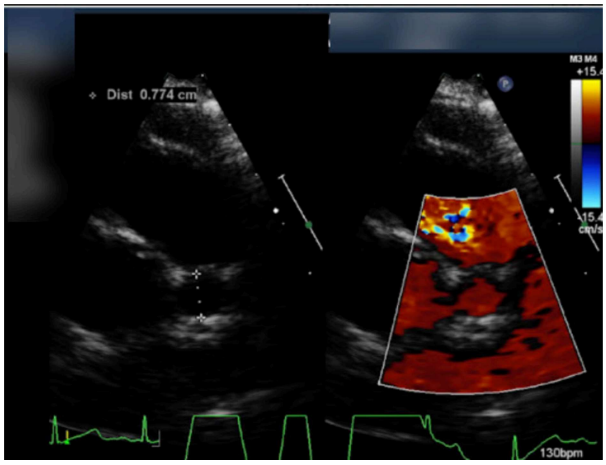
Comment

A 12-year-old child is admitted to the pediatric intensive care unit following presentation to the emergency department with severe chest pain radiating to his left arm and jaw, which began suddenly while he was playing soccer. The pain is associated with nausea and vomiting. His parents report that he has been healthy for most of his life but experienced several unexplained fevers as a toddler, which resolved without a specific diagnosis. However, the family has moved multiple times, and the parents recall that he had a prolonged hospitalization during early childhood for an unspecified "inflammatory condition," and did not receive follow-up care.

On physical examination, the child is diaphoretic and anxious, with labored respirations. His heart rate is 145 beats/min, blood pressure is 90/50 mm Hg, and respiratory rate is 24 breaths/min. His peripheral pulses are weak, and his capillary refill time is 4 seconds. A systolic murmur is appreciated at the left sternal border, and an S3 gallop is noted. There is no peripheral edema, joint swelling, or rash. His lung fields are clear to auscultation, but chest radiograph reveals cardiomegaly with mild pulmonary vascular congestion.

A 12-lead electrocardiogram shows ST-segment elevation in leads II, III, and aVF, and ST-segment depression in leads V1-V4. Laboratory investigations demonstrate a venous blood gas of pH 7.28, $pvCO_2$ 32 mm Hg, pvO_2 80 mm Hg, HCO_3^- 17 mEq/L, lactate 4.5 mmol/L, and high-sensitivity troponin 8.5 ng/mL. A representative echocardiogram of the right heart is shown (**Figure**).

Figure. Echocardiogram of Right Heart.



Courtesy of M. Rowin

Of the following, the MOST likely underlying cause of this patient's presentation is

A. anomalous left coronary artery from the pulmonary artery (ALCAPA)

B. hypertrophic cardiomyopathy with myocardial ischemia

C. myocardial infarction due to unrecognized coronary artery aneurysm ✓

D. viral myocarditis with coronary vasculitis

Your answer is correct.

PREP Pearl(s)

- Coronary artery aneurysms are a known complication of Kawasaki disease, even years after the acute illness, and can lead to myocardial infarction when they thrombose or rupture.
- A detailed history of early childhood inflammatory illnesses, even if vague or poorly documented in a child presenting with a myocardial infarction, should raise suspicion for history of Kawasaki disease, especially if there has been inconsistent follow-up.
- Inferior-wall myocardial infarction is indicated by ST-segment elevation in leads II, III, and aVF, which is typically caused by occlusion of the right coronary artery.

Critique

This patient's presentation is consistent with that of a myocardial infarction (MI), specifically an inferior-wall MI, as indicated by the ST-segment elevation in leads II, III, and aVF on the electrocardiogram (ECG). The finding of an aneurysmal right coronary artery (RCA) on echocardiography strongly points to an underlying cause related to coronary artery aneurysm, which is a known sequela of untreated or inadequately managed Kawasaki disease. The history of unexplained fevers in early childhood, along with the current finding of an RCA aneurysm, suggests that the patient likely had Kawasaki disease as a very young child, which was either not diagnosed or not followed appropriately.

Kawasaki disease can lead to progressive coronary artery aneurysms, which can thrombose or rupture, causing an acute coronary syndrome (ACS) and myocardial infarctions later in life. The patient's previous inflammatory illness and the lack of follow-up further support this diagnosis. Additionally, the systolic murmur and S3 gallop are consistent with heart failure due to ischemic damage from the MI, as reflected in the reduced left ventricular ejection fraction.

Coronary arteries originating from the contralateral (noncoronary) sinus and having an interarterial course, in which they run from the ascending aorta to the pulmonary trunk, is a potentially fatal anomaly. The right coronary artery originating from the left coronary sinus is the most prevalent anomaly of this type, However, it is known that there is a high risk of sudden death among patients with a left coronary artery of anomalous origin from the right sinus. The presence of an RCA aneurysm seen on echocardiogram is also inconsistent with this diagnosis.

Viral myocarditis can mimic symptoms of MI and is associated with elevated troponins, S3 gallop, and reduced ejection fraction. However, patients generally have preceding viral symptoms, and myocarditis typically presents with diffuse ECG changes rather than localized ST-segment elevations. Coronary artery aneurysms are not a common feature of viral myocarditis. Hypertrophic cardiomyopathy (HCM) can lead to myocardial ischemia and sudden cardiac death in young patients, particularly during exercise. However, this patient's ECG findings of inferior-wall MI and the echocardiographic evidence of an RCA aneurysm make HCM unlikely. The clinical findings in the vignette are most consistent with ischemic heart disease due to coronary artery involvement, rather than outflow obstruction or hypertrophy typical of HCM.

Suggested Reading(s)

- Dionne A, Dahdah N, Singh-Grewal D, Burgner DP, Newburger JW, de Ferranti SD. Anti-thrombosis management of patients with Kawasaki disease: results from an international survey. *Int J Cardiol.* 2020;307:154-158. doi:10.1016/j.ijcard.2019.10.045
- Gorelik M, Chung SA, Ardalan K, et al. American College of Rheumatology/Vasculitis Foundation guideline for the management of Kawasaki disease. *Arthritis Rheumatol.* 2022;74(4):586-596.

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Content Domain

- Critical Care, Cardiology Disorders and Heart Failure

Learning Objectives

- Describe the pathophysiology of myocardial infarction in pediatric patients
- Recognize that coronary artery aneurysms from Kawasaki disease or congenital coronary anomalies can lead to myocardial infarction

The correct answer is: myocardial infarction due to unrecognized coronary artery aneurysm

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