Question 1

Correct

1.00 points out of 1.00

Comment

A 10-year-old girl with a history of congenital cerebral aqueductal stenosis treated with placement of a ventriculoperitoneal shunt as an infant is evaluated in the emergency department for a 1-week history of headaches, intermittent emesis, and diplopia. Head computed tomography reveals significant ventriculomegaly and severe obstructive hydrocephalus. The neurosurgeon confirms a diagnosis of ventriculoperitoneal shunt malfunction. The patient is admitted to the pediatric intensive care unit postoperatively after removal of the shunt and placement of an external ventricular drain with a plan for an endoscopic third ventriculostomy procedure after treatment of a presumed shunt infection with intravenous antibiotics. The patient has been in the intensive care unit for 2 days, doing well, with resolution of her symptoms. However, she is now experiencing a return of her symptoms after magnetic resonance imaging (MRI) of the head earlier today. Since the MRI, she has been laying flat on the bed 2 ft below the level of the external ventricular drain. The measured intracranial pressure in this position is 5 mm Hg.

Of the following, the return of the patient's symptoms is MOST likely due to

- A. decreased drainage of cerebrospinal fluid and ventriculomegaly due to the position of the external ventricular drain ✔
- B. increased drainage of cerebrospinal fluid and collapsed ventricles due to the position of the external ventricular drain
- C. factors other than the positioning of the external ventricular drain
- D. laying flat on the bed with a change in cerebrospinal fluid (CSF) flow through the cerebral aqueduct and no downward drainage of CSF into the spinal canal

Your answer is correct.

The placement of an external ventricular drain (EVD) is a lifesaving procedure that is often performed in the intensive care unit. Placement of an EVD allows for drainage of cerebrospinal fluid (CSF), relieving or improving elevated intracranial pressure (ICP), installation of intracranial medications, and intracranial pressure monitoring. The provider must have a clear understanding of the placement procedure, appropriate positioning, and use of the drain. During the procedure, the proximal portion of the drain is surgically inserted, typically into the frontal horn of the right cerebral lateral ventricle to allow for drainage of CSF and to relieve any potential elevated ICP caused by an accumulation of CSF and obstruction to CSF flow. The distal portion of the drain is then attached to a pressure transducer with an adjustable height distal collection system. The amount of CSF drained is determined by the pressure differential between the proximal tip of the EVD in the ventricle and the distal collecting system. Depending on the height of the collecting system, drainage can be a fixed volume per desired time, continuous, or intermittent drainage as needed according to ICP elevations. The height of the pressure transducer should be set at the level of the patient's foramen of Monro, which corresponds to the level of the external auditory meatus of the ear in the supine position. A leveling device should be used to zero the drain at this level and ensure accuracy of placement. The level of the collecting system is adjusted to the desired height level, depending on the clinical scenario and desired amount of drainage. The CSF will drain whenever the interventricular pressure exceeds the pressure set by the height of the collecting system. The flow of CSF through the drain ceases once the pressure equalizes between the CSF compartments in the brain and the distal collecting system.

The collection system and transducer must be adjusted and kept at the level of the external auditory meatus of the ear whenever the patient changes position to avoid inaccurate ICP readings and overdrainage or underdrainage of CSF. If the transducer and collecting system are placed above the foramen of Monro, such as if the bed is lowered, or hung too high above the bed, it will result in falsely low ICP readings and decreased drainage of CSF. If the transducer and collecting system are placed below the level of the foramen of Monro, such as placed on the bed, or falls to the floor, it will result in falsely elevated ICP readings and likely rapid, excessive CSF drainage. The drain must be clamped using a stopcock whenever the patient is being transferred or moving to avoid these fluctuations in drainage and inadvertent rapid decompression or underdrainage.

In the vignette, the measured intracranial pressure of 5 mm Hg should not provide false reassurance to the practitioner because it is likely significantly lower than the actual ICP and unreliable because of the positioning of the EVD. The positioning of the EVD above the patient will result in decreased drainage of CSF, worsening ventriculomegaly, elevated ICP, and the return of symptoms. Increased drainage of CSF and collapsed ventricles would occur if the EVD was placed below the level of the patient which is not consistent with the described vignette. The return of the patient's symptoms is not likely to be secondary to a change in flow through the cerebral aqueduct because the patient has known congenital cerebral aqueductal stenosis, requiring a ventriculoperitoneal shunt, and minimal to no flow via the aqueduct.

PREP Pearl(s)

- Proper alignment of the external ventricular drain transducer and collection of cerebrospinal fluid are essential for intracranial pressure monitoring.
- Placement of an external ventricular drain transducer and collection of cerebrospinal fluid (CSF) above the level of the foramen of Monro will result in decreased CSF drainage and falsely lower intracranial pressure readings.
- Placement of an external ventricular drain transducer and collection of cerebrospinal fluid (CSF) below the level of the foramen of Monro will result in increased CSF drainage and falsely elevated intracranial pressure readings.

Suggested Reading(s)

- Chung DY, Olson DM, et al. Evidence-based management of external ventricular drains. *Curr Neurol Neurosci Rep*. 2019:19:94. doi:10.1007/s11910-019-1009-9
- Dossani RH, Patra DP, et al. Placement of an external ventricular drain. *N Engl J Med*. 2021;384:e3. doi:10.1056/NEJMvcm1805314

Content Domain

- Principles of Monitoring (technique, assessment, complications)

ABP Content Specification(s) / Content Area(s)

- Accurate placement of an external ventricular drain.

The correct answer is: decreased drainage of cerebrospinal fluid and ventriculomegaly due to the position of the external ventricular drain

View Peer Results

Question 2

Correct

1.00 points out of 1.00

Comment

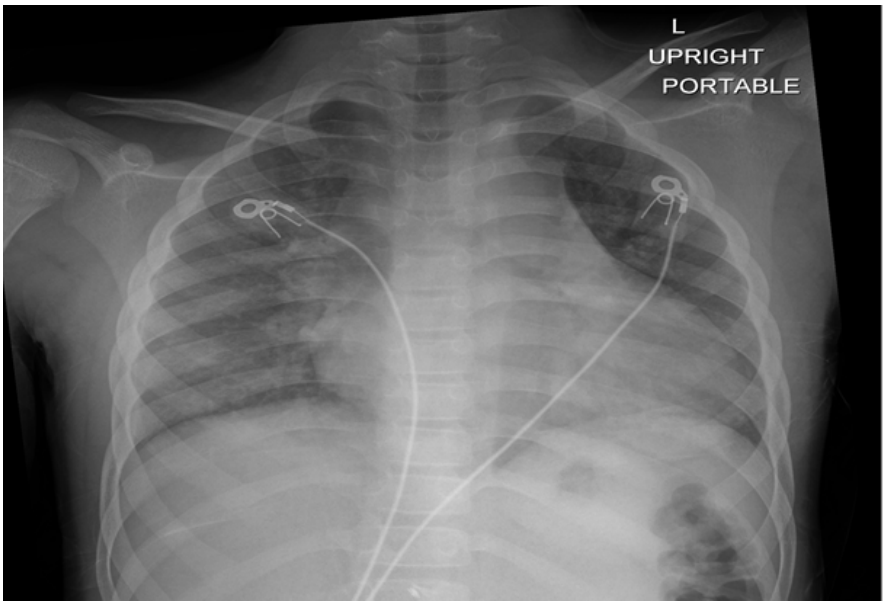
A 9-year-old boy with sickle cell disease is admitted with pain in his left shoulder and upper back.

Laboratory values are shown

Laboratory Test	Results
White blood cell count	14,100/μL (14.1 x 10^9/L)
Neutrophils	70%
Hemoglobin	7.1 g/dL (71 g/L)
Hematocrit	21%
Platelet count	140 x 10^3/μL (140 x 10^9/L)

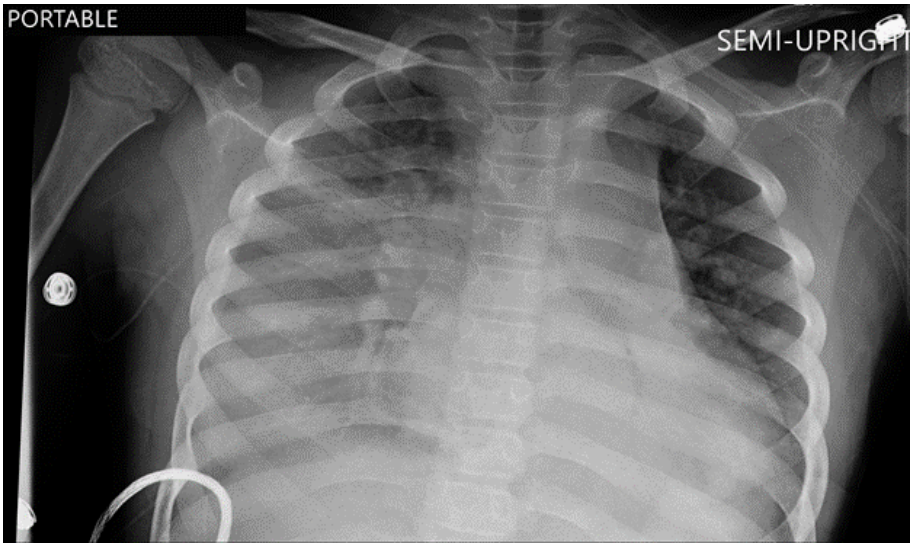
He is started on intravenous hydration at 1.5 times maintenance rate. He is also receiving intravenous acetaminophen and morphine delivered via a patient-controlled analgesia device for pain control. His initial chest radiograph is shown (**Figure 1**). Over the next 12 hours, he becomes febrile to 38.8 °C and develops increased work of breathing. He develops oxygen desaturation eventually requiring high flow nasal cannula at 20 L/min, 80% oxygen in order to maintain pulse oximetry readings above 95%. His breathing has stabilized on a high flow nasal cannula. A repeat chest radiograph (**Figure 2**) is obtained upon transfer to the pediatric intensive care unit. A blood culture is done and intravenous piperacillin/tazobactam and azithromycin are started.

Figure 1. Initial chest radiograph on admission.



Courtesy of M. Mathur

Figure 2. Repeat chest radiograph on transfer to PICU.



Courtesy of M. Mathur

Of the following, the NEXT best step in treating this patient is

- A. endotracheal intubation and mechanical ventilation
- B. increase intravenous fluids to 2 times maintenance rate
- C. red blood cell exchange through apheresis
- D. simple packed red blood cell transfusion ✔

Your answer is correct.

Sickle cell disease is caused by a single nucleotide substitution in the β-globin allele on chromosome 6 resulting in sickle hemoglobin, which polymerizes at low oxygen concentrations. The red blood cells distort into a sickle shape, increasing blood viscosity, and reducing or obstructing blood flow in the microvasculature. In the lungs, this can lead to acute chest syndrome (ACS), characterized by the onset of new respiratory symptoms, fever, hypoxemia, and new opacities on chest radiograph in a patient with sickle cell disease.

Acute chest syndrome commonly develops within the first 3 days of hospitalization for a severe vaso-occlusive crisis. Rib and vertebral infarction can lead to hypoventilation and atelectasis and bone marrow infarction may lead to fat embolism. The underlying pathology in ACS involves a combination of infarction, fat embolism, and infection, and results in significant ventilation perfusion mismatch and hypoxemia. Low oxygen saturation in turn may lead to more sickling precipitating a vicious cycle of worsening lung disease. Infection with an atypical bacteria or virus is present in over half of the patients with ACS.

The patient in this vignette presented in an acute pain crisis, but then developed worsening respiratory distress. He has a fever and desaturation as well as increasing opacities on his chest radiograph consistent with ACS. Transfusion of packed red blood cells reduces the burden of sickle cells and improves microvascular perfusion. Simple transfusion and partial/whole blood volume red cell exchange via apheresis are both effective measures to relieve ACS. However, apheresis requires the placement of a dialysis catheter. This invasive procedure would add the risk of sedating a child in respiratory distress. The patient in this vignette has a hemoglobin of 7.1 g/dL [71 g/L (which is relatively low)], and a simple transfusion [up to a hemoglobin of 10 g/dL (100 g/L)] can lower the percentage of circulating sickle cells with less risk. A red blood

cell exchange may become necessary if he does not improve with a simple transfusion. If the patient started with a hemoglobin higher than 10 g/dL (100 g/L), transfusion would increase viscosity, and red blood cell exchange though apheresis would be considered the preferred option.

Hydration has already been started and any further increase in the intravenous fluid administration rate should be made after assessment of the cardiac function and filling pressures. Since oxygen saturation is above 95% and the work of breathing has improved on high flow oxygen, additional support with mechanical ventilation is not indicated.

PREP Pearl(s)

- Acute chest syndrome has a multifactorial etiology.
- If the patient's hemoglobin is low, a simple red blood cell transfusion may be safer than red cell apheresis.

Suggested Reading(s)

- Rees DC, Robinson S, Howard J. How I manage red cell transfusions in patients with sickle cell disease. Br J Haematol. 2018;180(4):607-617. doi:10.1111/bjh.15115
- Emre U, Miller ST, Gutierez M, Steiner P, Rao SP, Rao M. Effect of transfusion in acute chest syndrome of sickle cell disease. *J Pediatr*. 1995;127(6):901-904. doi:10.1016/s0022-3476(95)70025-0
- Velasquez MP, Mariscalco MM, Goldstein SL, Airewele GE. Erythrocytapheresis in children with sickle cell disease and acute chest syndrome. *Pediatr Blood Cancer*. 2009;53(6):1060-1063. doi:10.1002/pbc.22211

Content Domain

- Hematology/Oncology

ABP Content Specification(s) / Content Area(s)

- Understand the pathophysiology of acute chest syndrome in a patient with sickle cell disease
- Evaluate and treat a patient with acute chest syndrome
- Understand the risks and benefits of a simple packed red cell transfusion and red cell apheresis in the treatment of acute chest syndrome

The correct answer is: simple packed red blood cell transfusion

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Question 3

Correct

1.00 points out of 1.00

Comment

A 6-year-old patient with a complex medical history, including developmental delay and intractable epilepsy is admitted to the intensive care unit for pneumonia and increased seizures. The child is ordered ceftriaxone despite having a documented allergy to cephalosporins. The medication is dispensed by the pharmacist, but the bedside nurse notes the allergy prior to administering the medication. The nurse notifies the providers and the attending discloses the error to the family.

Of the following, the next BEST step in the evaluation of this error is

- A. identify the physician who ordered the antibiotic and mandate additional safety training
- B. perform a failure mode and effects analysis
- C. perform a root cause analysis ✔
- D. report a sentinel event to the Joint Commission

Your answer is correct.

Medical errors are a major public health problem and a leading cause of death in the United States. Taking steps to build a culture of safety, in which events are recognized, reported, learned from, and then prevented can improve patient safety. Errors may be categorized into two major domains: errors of omission, in which an error occurs because of an action that was not taken; and errors of commission, in which an error occurs because of an action that was taken.

A root cause analysis (RCA) identifies causal factors as they relate to problems or errors. Typically, an RCA begins with defining the problem, then establishing a timeline of events leading up to the problem, identifying the root cause from among other potential causal factors, then creating a “causal graph” between the root cause and the problem. Root cause analysis is typically used to evaluate sentinel events that result in injury or death, and are therefore done retrospectively after an event has occurred. A failure mode and effects analysis follows a prospective and preventative approach. Sentinel events (events resulting in unexpected mortality, major permanent harm, or severe temporary harm) must be reported to and are reviewed by the Joint Commission, and health care institutions are required to perform a root cause analysis after any sentinel event. Although not actually resulting in direct patient harm, a near-miss event may still warrant an RCA. Through this process of reporting and investigation, teams work to identify the causative event and factors that resulted in the occurrence of a sentinel event to help prevent future similar errors.

The Joint Commission defines a sentinel event as, “any unexpected occurrence involving death or serious physical or psychological injury, or the risk thereof [...] the phrase 'or the risk thereof' includes any process variation for which a recurrence would carry a significant chance of a serious adverse outcome.” Although the event involved in the vignette was a near miss, it has the potential to be a sentinel event and therefore warrants a root cause analysis.

A failure mode and effects analysis fosters safety and prevention of events through a proactive and iterative process of identifying potential or real sources of error. Failure mode and effects analyses assume that errors will occur even when maximum care is utilized, but then seek to build in redundancies to serve as safety nets; these safety nets are then intended to catch errors before they occur.

Medication errors are among the most common preventable safety events. They can occur at multiple points in the prescribing, dispensing, or administering of medications; therefore, interdisciplinary collaboration to establish safe medication use and distribution is critical for patient safety.

It is also important to note the mental health effects of medical errors on health care professionals, including anxiety, depression, and guilt, and efforts must be taken to debrief events with involved parties to help the team to process the event together.

PREP Pearl(s)

- Medical errors are a major public health problem and a leading cause of death in the United States.
- Medication errors are among the most common medical errors.
- Root cause analyses are used to identify underlying processes or system issues that lead to or contribute to a sentinel event.

Suggested Reading(s)

- D'Errico S, Zanon M, Radaelli D, Padovano M, Santurro A, Scopetti M, et al. Medication errors in pediatrics: proposals to improve the quality and safety of care through clinical risk management. *Front Med*. 2022;14(8):814100. doi:10.3389/fmed.2021.814100
- Kohn LT, Corrigan JM, Donaldson MS; Committee on Quality of Health Care in America. To Err Is Human: *Building a Safer Health System*. National Academy Press; 2000.

Content Domain

- Special Critical Care Issues

ABP Content Specification(s) / Content Area(s)

- Describe processes for evaluating medical error events

The correct answer is: perform a root cause analysis

View Peer Results

Question 4

Correct

1.00 points out of 1.00

Comment

A 23-month-old male infant transferred 4 days ago to the pediatric intensive care unit is treated for acute hypoxemic respiratory failure secondary to viral bronchiolitis. The family reports a 3-day history of fever, fussiness, increased work of breathing, and pallor before admission. He continues to have increased work of breathing without resolution of fever since admission. The child attends daycare but has not been exposed to any known sick contacts.

Vital signs include an axillary temperature of 39.5°C, blood pressure of 90/60 mm Hg, heart rate of 150 beats/min, respiratory rate of 45 breaths/min, and oxygen saturation of 98% on 14 L/min of heated, humidified, high-flow oxygen therapy via nasal cannula.

On physical examination, the child is lying supine with mild subcostal retractions. He is awake, alert, and consolable in the mother's arms. He has non injected sclera with some scleral icterus noted, clear rhinorrhea, no oral lesions, a normal-appearing tongue, no posterior pharyngeal inflammation, and shotty anterior cervical lymphadenopathy. His lungs are clear to auscultation, and he has a normal cardiac impulse and unremarkable cardiac findings to auscultation. His abdomen is soft and nontender, his liver is palpable 2 to 3 fingerbreadths below the right costal margin, and the tip of his spleen is palpable at the left costal margin.

A chest radiograph reveals no focal pneumonia or pleural effusion. The cardiac silhouette appears normal. An abdominal ultrasound confirms hepatosplenomegaly with mild ascites.

Laboratory data are shown:

Laboratory Test	Results
White blood cell count	7,100/μL (7.1×10^9/L)
Hemoglobin	5 g/dL (50 g/L)
Platelet count	81×10^3/μL (81×10^9/L)
Aspartate aminotransferase	400 U/L
Alanine aminotransferase	225 U/L
Lactate	18.9 mg/dL (2.1 mmol/L)
Ferritin	6,800 ng/mL (6,800 μg/L)
Triglycerides	894 mg/dL (10.1 mmol/L)

The child's clinical condition worsens, and he requires intubation and mechanical ventilation.

Of the following, the result MOST likely to be helpful in diagnosing the suspected disease process is

- A. elevated Epstein-Barr virus IgM levels ✔
- B. elevated natural killer cell activity
- C. a fibrinogen level of 400 mg/dL (4.0 g/L)

D. a neutrophil count of 3,100/μL (3.1×10^9/L)

Your answer is correct.

Hemophagocytic lymphohistiocytosis (HLH) is a disorder characterized by pathologic uncontrolled immune activation with extreme inflammation from a cytokine storm. It is often underdiagnosed and is fatal if untreated. The reported frequency of HLH in children is 1.2 cases per million per year. The hallmark of HLH is proliferation of activated macrophages and histiocytes resulting in phagocytosis of red and white blood cells and of platelets. Criteria to diagnose HLH have been established by the Histiocyte Society and require either a molecular diagnosis with a genetic mutation involving *PRF1*, *UNC13D*, *STXBP2*, *Rab27a*, *STX11*, *SH2D1A*, *XIAP* genes, or 5 or more of the following signs and symptoms:

- Fever greater than 38.5 °C
- Splenomegaly
- Cytopenia affecting at least 2 peripheral blood cell lineages
  - Hemoglobin level less than 9 g/dL (90 g/L) in infants younger than 4 weeks of age or less than 10 g/dL (100 g/L)
  - Platelet count less than 100×10^3/μL (100×10^9/L)
  - Neutrophil count less than 1,000/μL (1.0×10^9/L)
  - Hypertriglyceridemia (greater than 265 mg/dL [3 mmol/L]) and/or hypofibrinogenemia (less than 150 mg/dL [4.4 g/L])
  - Hemophagocytosis in a lymphoid organ (bone marrow, spleen, or lymph nodes)
  - Hyperferritinemia (greater than 500 ng/mL [500 μg/L]),
- Low or absence of natural killer cell activity
- Elevated soluble CD25 (soluble interleukin 2 receptor alpha) level greater than 2,400 U/mL

There are 2 classifications of HLH: familial (primary) and acquired (secondary). Primary or familial HLH is inherited in an autosomal recessive fashion and most commonly occurs in infants younger than 1 year. Children with primary HLH may have isolated central nervous system involvement, primary hepatic failure, or isolated fever with cytopenia and splenomegaly. Typically, infectious and malignant causes must be considered and ruled out. Infectious causes may not be identified in many of these cases. Secondary HLH typically has a definable trigger that can include infection, rheumatologic disease, malignancy, or autoimmune disorders.

Children older than 1 year tend to have a secondary infection that triggers HLH and most commonly includes coinfection with Epstein-Barr virus or cytomegalovirus. Virus-associated HLH has been reported with influenza and parainfluenza, herpes simplex, adenovirus, parvovirus B19, respiratory syncytial virus, and enterovirus. Malignancies, particularly lymphoma, are associated with HLH at initial presentation. Children with an immunocompromised state such as those receiving chemotherapy or with a primary immunodeficiency are at increased risk of developing HLH. Additionally, those patients receiving immune-activating therapies such as CAR-T cells or immune checkpoint inhibitors have increased incidence of HLH.

Hemophagocytic lymphohistiocytosis can present with fever of unknown origin or systemic disease involvement including central nervous system dysfunction and hepatitis or acute liver failure. Central nervous system involvement can result in seizures, altered consciousness, irritability, and hypotonia. Results of cerebrospinal fluid analysis are abnormal in more than 50% of patients with HLH and include pleocytosis and elevated protein. Hemophagocytosis may also be observed in the cerebrospinal fluid. The acute nature of cytokine release and progression to multiple organ dysfunction may mislead providers into presuming sepsis to be the primary diagnosis. Hepatosplenomegaly is uncommon in sepsis but often seen in HLH, which may assist in making the appropriate diagnosis. As a result of delayed diagnosis, HLH is associated with significant acute and long-term mortality. Not all HLH diagnostic criteria may be initially present, so a high index of suspicion must be maintained if a child exhibits atypical symptoms after infection or malignancy or with an autoimmune history.

Once a diagnosis of HLH is made, it is imperative to begin aggressive treatment following established protocols including use of intravenous immunoglobulin, cytotoxic agents, immunomodulating drugs, and corticosteroids. Response to therapy can be followed with soluble CD25 levels or serum ferritin levels. Soluble CD25 levels may be a more reliable measure of inflammation in some patients; however, this test may not be readily available in all centers. In the case of refractory treatment, salvage therapy includes monoclonal antibody therapy with alemtuzumab and emapalumab. Patients may require hematopoietic transplantation to prevent recurrence of HLH disease in refractory cases. Without prompt diagnosis and treatment, HLH is uniformly fatal.

Evidence of coinfection with Epstein-Barr virus or cytomegalovirus is most consistent with a diagnosis of HLH in the vignette. The fibrinogen level, neutrophil count, and elevated natural killer cell activity are inconsistent with the Histiocyte Society diagnostic criteria.

PREP Pearl(s)

- Secondary hemophagocytic lymphohistiocytosis is typically triggered by infectious, rheumatologic, immunologic, or malignant comorbidities. Coinfection with Epstein-Barr virus or cytomegalovirus is common.
- The rapid progression of secondary hemophagocytic lymphohistiocytosis to multiorgan system failure and cytokine release may clinically imitate sepsis and delay proper diagnosis and aggressive treatment.
- Hepatosplenomegaly and hyperferritenemia frequently occur in hemophagocytic lymphohistiocytosis and may assist in making the appropriate diagnosis.

Suggested Reading(s)

- Dao D, Xoay, TD, Galeano BK, Phuc PH, Ouellette Y. Etiologies and clinical outcomes of patients with secondary hemophagocytic lymphohistiocytosis at a tertiary PICU. *Pediatr Crit Care Med*. 2019;20:e311-e318. doi:10.1097/PCC.0000000000001980.
- Gregory J, Greenberg J, Basu S. Outcomes analysis of children diagnosed with hemophagocytic lymphohistiocytosis in the PICU. *Pediatr Crit Care Med*. 2019;20:e185-e190. doi:10.1097/PCC.0000000000001827.
- Jordan MB, Allen CE, Greenberg J, Henry M, Hermiston ML, et al. Challenges in the diagnosis of hemophagocytic lymphohistiocytosis: recommendations from the North American Consortium for Histiocytosis (NACHO). *Pediatr Blood Cancer*. 2019:e27929. doi:10.1002/pbc.27929.

Content Domain

- Hematology/Oncology

ABP Content Specification(s) / Content Area(s)

- Recognize primary and secondary hemophagocytic lymphohistiocytosis
- Recognize the complications of hemophagocytic lymphohistiocytosis
- Plan treatment for a patient with primary hemophagocytic lymphohistiocytosis
- Plan the treatment of secondary hemophagocytic lymphohistiocytosis

The correct answer is: elevated Epstein-Barr virus IgM levels

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Question 5

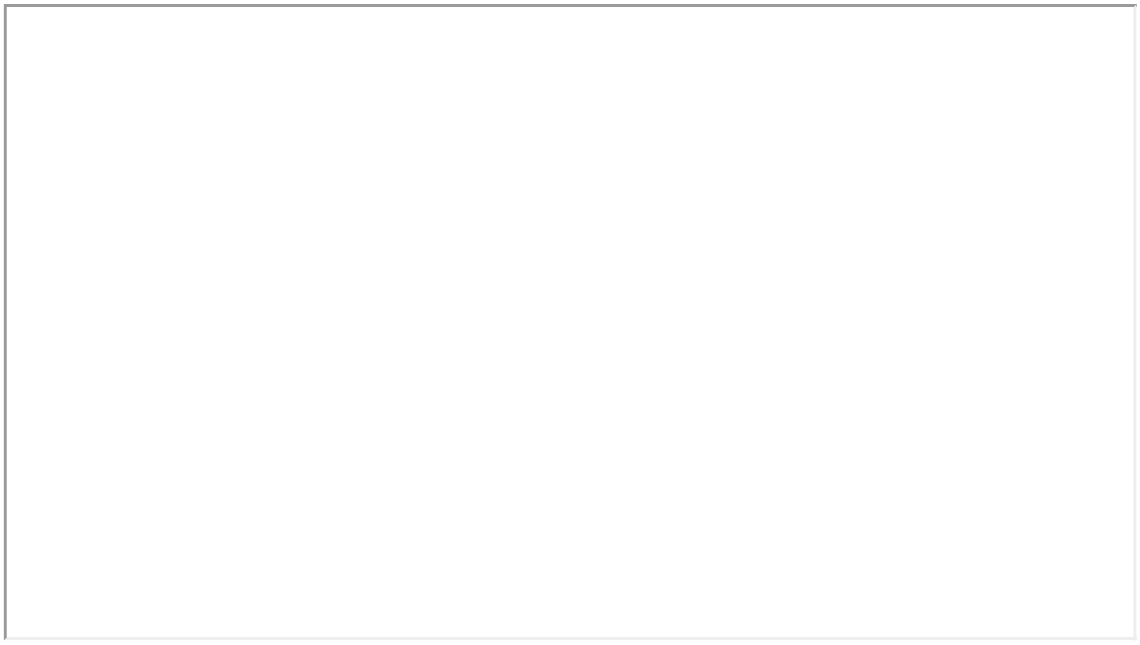
Correct

1.00 points out of 1.00

Comment

A 15-year-old girl living with systemic lupus erythematosus has been admitted to the acute care unit for cough, dyspnea, and fever. She is given ampicillin for suspected community-acquired pneumonia and 3 L/min of supplemental oxygen via nasal cannula to maintain oxygen saturations greater than 92%. Chest radiography on the day of admission shows simple small bilateral pleural effusions. She is eating and drinking poorly, so she is given intravenous 0.9% sodium chloride with 5% dextrose at 1,600 mL/m^2/d. The next day she develops increasing chest pain and is given ibuprofen and prednisone for pleuritis. That evening a rapid response team alert is called due to tachycardia and increased tachypnea. She is awake, alert, calm, and following commands. Her oxygen saturation is 100% on 5 L/min of simple face mask supplemental oxygen. Her blood pressure while supine is 90/68 mm Hg, and her heart rate is 130 beats/min. She is transferred to the pediatric intensive care unit, where echocardiography is performed (Video 1).

Video 1. 4-chamber apical view of the heart.



Courtesy of A. Riley

Of the following, the next BEST step in the treatment of this patient is

- A. endotracheal intubation
- B. furosemide, 1 mg/kg
- C. methylprednisolone, 30 mg/kg
- D. pericardiocentesis ✔

Your answer is correct.

The patient in the vignette has systemic lupus erythematosus (SLE) with evidence of generalized serositis with pleuritis, pericarditis, and effusions related to both. Although her initial symptoms were primarily pulmonary in nature, pericardial disease in patients with SLE can be indolent and mimic other types of pain until an

effusion affects hemodynamic function. This patient had been given both a nonsteroidal anti-inflammatory drug (ibuprofen) and low-dose glucocorticoid for her serositis but has evidence of disease progression. After worsening of her symptoms, the patient has a large pericardial effusion (PCE) with echocardiographic evidence of cardiac tamponade. Video 2 shows the heart surrounded by a large PCE and right atrial collapse, and the Figure shows a significant (47%) decrease in flow velocity across the mitral valve during the respiratory cycle (respiratory variability). Of note, mitral valve inflow variability of greater than 30% is consistent with tamponade.

Video 2. Labeled 4-chamber apical view of the heart

Courtesy of A. Riley and F. Lam

Figure. Mitral valve flow measurements

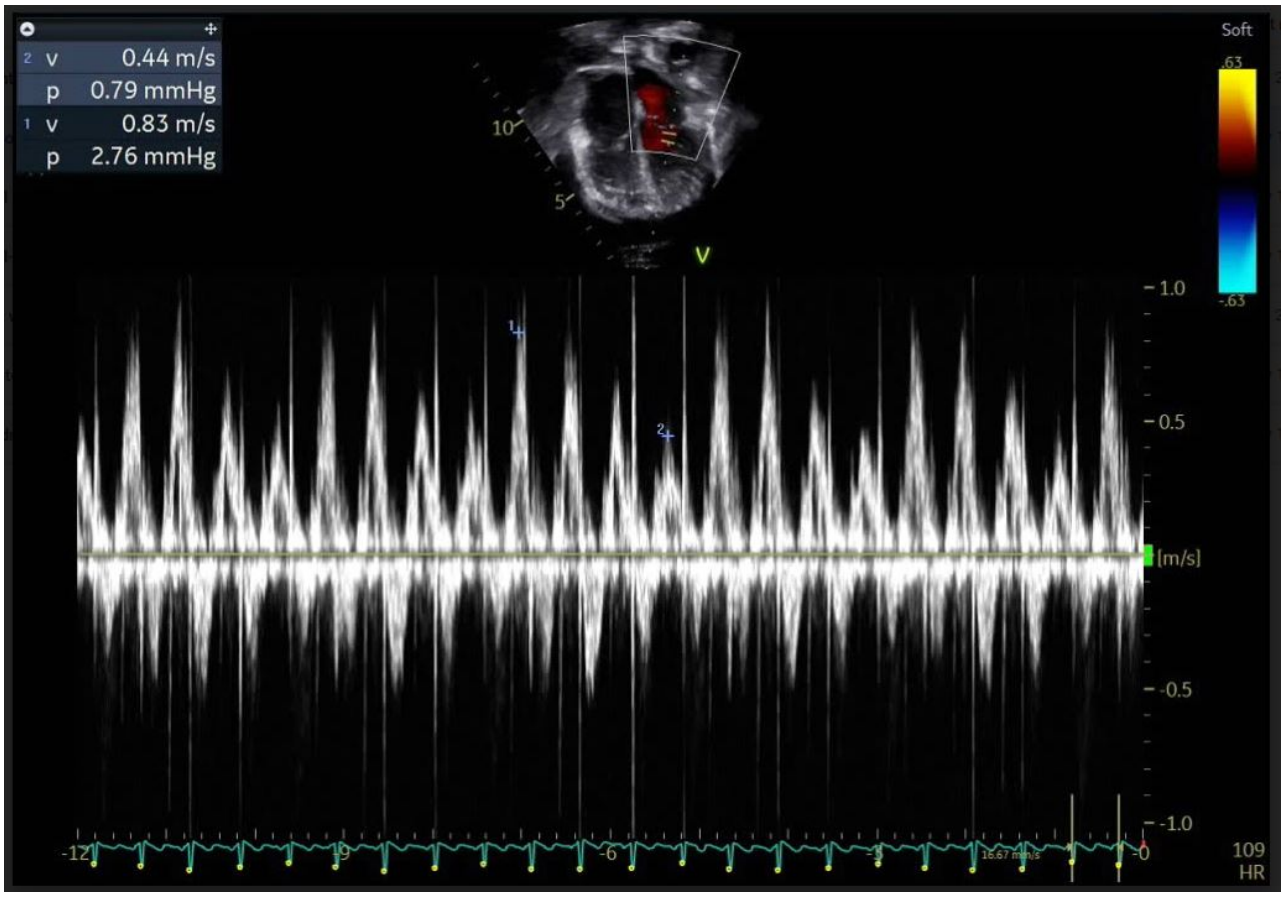
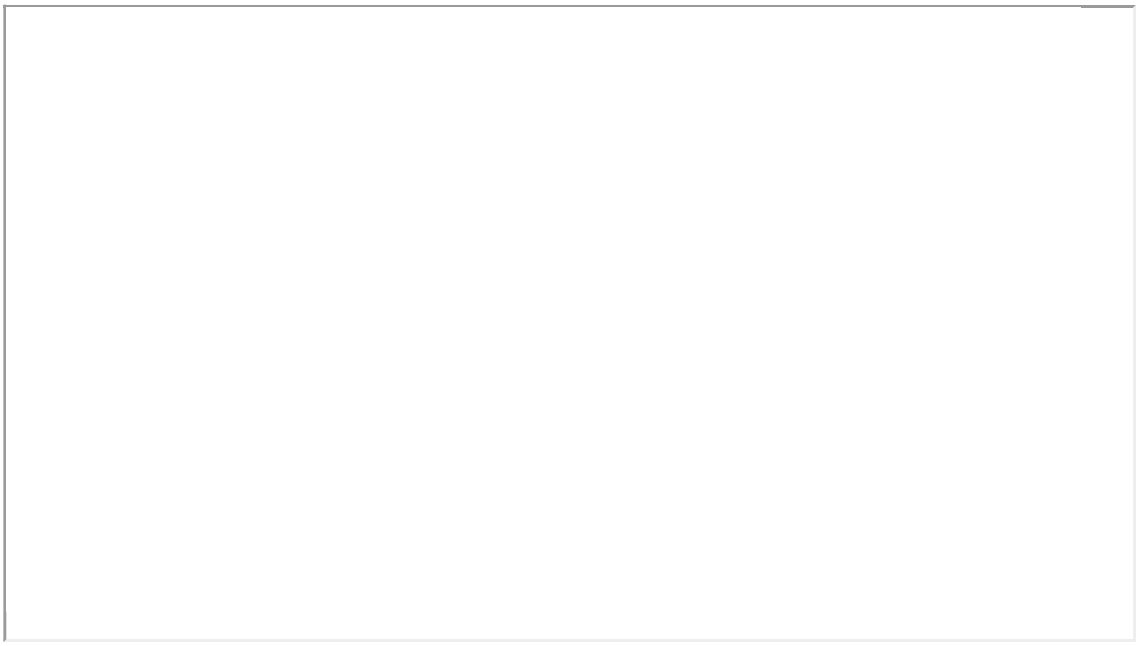


Figure. Mitral valve flow measurements

Courtesy of A. Riley

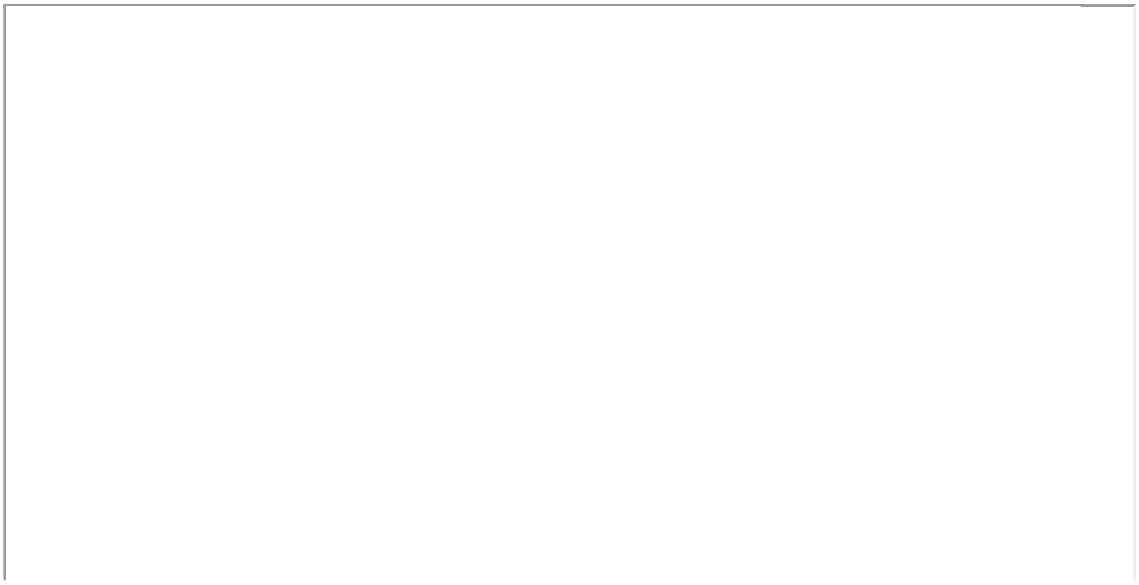
Given her worsening symptoms (tachycardia, hypotension, and tachypnea) and the significant PCE with mitral valve inflow variability greater than 30%, the best next course of action is drainage of the PCE via percutaneous pericardiocentesis or surgical drainage. For effusions without concern for bleeding or purulence in the pericardial sac, percutaneous pericardiocentesis is the preferred approach over a surgical pericardiocentesis (with or without pericardiectomy). Percutaneous pericardiocentesis can be echocardiography or fluoroscopy guided, depending on institutional standards, although echocardiography-guided drainage obviates the need for ionizing radiation and provides more dynamic visualization of the heart and pericardial sac during the procedure compared with fluoroscopy. Video 3 shows the use of agitated saline contrast to visualize the needle in the pericardial sac with Video 4 showing the catheter within the PCE. Finally, Video 5 shows the catheter within the pericardial space with the PCE drained and the heart rate significantly reduced. The reported risks of echocardiography-guided pericardiocentesis are small (<2%) but may include cardiac puncture and/or bleeding within the pericardial space.

Video 3. Echocardiogram of agitated saline contrast within the pericardial sac



Courtesy of A. Riley and F. Lam

Video 4. Echocardiogram of the catheter within the pericardial sac



Video 4. Echocardiogram of the catheter within the pericardial sac

Courtesy of A. Riley and F. Lam

Video 5. Echocardiogram post drainage

Courtesy of A. Riley and F. Lam

Endotracheal intubation for positive pressure ventilation is not recommended for this patient because of the concern for decreasing venous return with the increased intrathoracic pressure. This condition could lead to a worsened decrease in preload to the heart. The patient is calm and alert and would likely tolerate percutaneous pericardiocentesis with local anesthetics and moderate sedation (such as with ketamine). Furosemide is also not recommended because of its effect on decreasing preload and worsening cardiac output. Finally, high-dose methylprednisolone is not the next best option because of the need for urgent drainage of the hemodynamically significant PCE.

PREP Pearl(s)

- Pericardial effusions with evidence of tamponade (mitral valve inflow variation >30%) should be drained immediately.
- Medications or procedures that would decrease cardiac preload should be avoided, if possible.

Suggested Reading(s)

- Adler Y, Charron P, Imazio M, et al. 2015 ESC guidelines for the diagnosis and management of pericardial diseases. *Eur Heart J*. 2015;36(42):2921-2964. doi:10.1093/eurheartj/ehv318
- Herron C, Forbes TJ, and Kobayashi D. Pericardiocentesis in children: 20-year experience at a tertiary children's hospital. Cardiol Young 2022; 32(4): 606-611. PMID: 34304745

Content Domain

- Technical Procedures & Complications

ABP Content Specification(s) / Content Area(s)

- Diagnose and treat pericardial effusion.

The correct answer is: pericardiocentesis

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Question 6

Correct

1.00 points out of 1.00

Comment

A 17-year-old girl presents to the emergency department with 4 days of fever, lethargy, emesis, dysuria, and dark-colored urine. In the emergency department, the patient is hypotensive, tachycardic, and febrile. A urinalysis is positive for leukocyte esterase, white blood cells, and nitrites. After receiving 60 mL/kg of intravenous 0.9% saline, the patient is admitted to the pediatric intensive care unit where she has ongoing hypotension. She is started on a norepinephrine infusion. Over the course of the next few days, her urine output decreases with rising blood urea nitrogen and creatinine levels, and worsening respiratory failure. The clinical team makes the decision to start continuous veno-venous hemodiafiltration. Citrate is chosen as regional anticoagulation.

Of the following, while receiving this therapy, the systemic infusion that a patient MUST also receive is

- A. calcium ✔
- B. magnesium
- C. potassium
- D. sodium bicarbonate

Your answer is correct.

Continuous renal replacement therapy (CRRT) is an important therapy used routinely in both adults and children in intensive care units. Indications for CRRT include fluid overload (causing negative pulmonary and/or cardiac effects that are unresponsive to diuretic therapy) uremic complications, metabolic derangements, and toxin removal. Continuous renal replacement therapy is a blanket term for a variety of therapies, including continuous veno-venous hemodiafiltration, a method which combines ultrafiltration and dialysis (relying on the principles of diffusion and convection) with blood drawn from and returned to a venous access point. Regional anticoagulation (ie, anticoagulation directed only at the extracorporeal circuit) is commonly used in children to prevent circuit clotting. Children are at higher risk of circuit clotting than adults due to lower flow rates through the CRRT circuit.

Citrate regional anticoagulation is the preferred modality for pediatric CRRT. Citrate is infused into the blood after it is removed from the patient and binds with calcium. Most of this calcium-citrate is then removed by CRRT; however, some does return to the patient to be metabolized by the liver. Due to the binding of serum calcium with citrate, calcium must be infused to the patient systemically so the patient does not become hypocalcemic. Serum total calcium and ionized calcium as well as circuit calcium must be monitored closely while a patient is receiving CRRT. Caution must be exercised in patients with liver dysfunction/failure as they cannot clear citrate and may develop citrate accumulation (sometimes termed 'citrate lock') in which total calcium rises but ionized calcium falls as total body calcium is bound to citrate. A ratio of total calcium to ionized calcium over 2.5 is indicative of citrate accumulation. Citrate also binds magnesium and phosphorus; these levels must also be monitored closely. Systemic heparin may also be used as anticoagulation, but incurs a higher risk of bleeding. Although the patient should be monitored for acid-base disturbances with infusion of citrate, sodium bicarbonate is not routinely administered with CRRT.

PREP Pearl(s)

- Anticoagulation is often necessary in pediatric continuous renal replacement therapy.

- Citrate is commonly the anticoagulant of choice in pediatric continuous renal replacement therapy due to its more favorable side-effect profile when compared with heparin.
- Calcium must be administered via continuous infusion while using citrate for anticoagulation due to citrate-calcium binding.

Suggested Reading(s)

- Beltramo F, DiCarlo J, Gruber JB, Taylor T, Totapally BR. Renal replacement therapy modalities in critically ill children. *Pediatr Crit Care Med*. 2019;20(1):e1-e9. doi:10.1097/PCC.0000000000001754
- Davis TK, Neumayr T, Geile K, Doctor A, Hmeil P. Citrate anticoagulation during continuous renal replacement therapy in pediatric critical care. *Pediatr Crit Care Med*. 2014;15(5):471-85. doi:10.1097/PCC.0000000000000148
- de Galasso L, Picca S, Guzzo I. Dialysis modalities for the management of pediatric acute kidney injury. *Pediatr Nephrol*. 2020;35(5):753-765. doi:10.1007/s00467-019-04213-x

Content Domain

- Renal and Electrolytes

ABP Content Specification(s) / Content Area(s)

- Describe options for anticoagulation while on continuous renal replacement therapy
- Identify potential complications of anticoagulation associated with continuous renal replacement therapy
- Identify electrolyte abnormalities potentially associated with continuous renal replacement therapy

The correct answer is: calcium

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