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**Status** Finished

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**Grade** 100.00 out of 100.00

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**Question 1**

Correct

1.00 points out of 1.00

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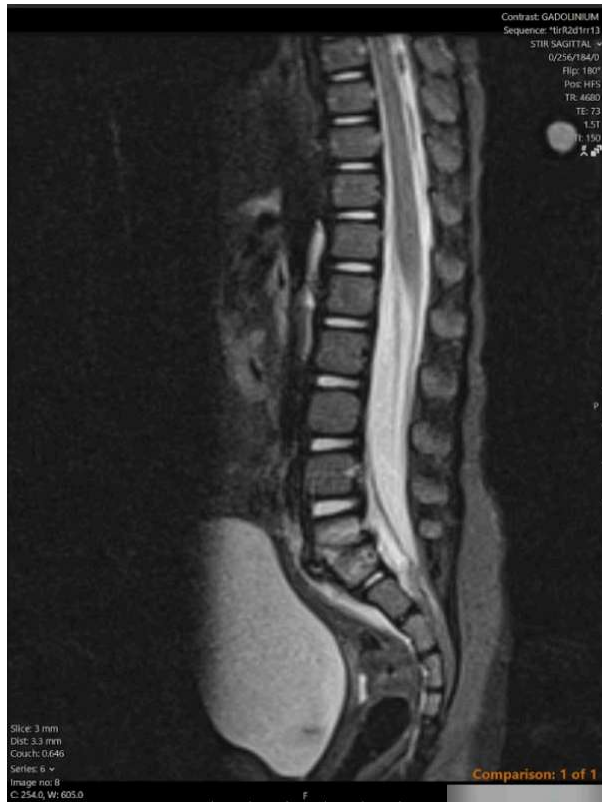
A 15-year-old high school cheerleader presented to the pediatric intensive care unit with hypotension, fever, and tachycardia. The patient also reports severe lower back pain. The patient denies any recent trauma, although she states that cheerleading practice has been intense lately. She was seen by her primary care physician for back pain 3 days ago; she was diagnosed with a muscle strain and prescribed rest and meloxicam. However, the pain became so severe that she did not want to get out of bed, and her parents became concerned when she developed a fever. On further questioning, she has had decreased urine output all day in addition to the fever. Her vital signs are a temperature of 39 °C, heart rate of 120 beats/min, blood pressure of 75/40 mm Hg, respiratory rate of 30 breaths/min, and oxygen saturation of 98% in room air. Laboratory results are shown:

Laboratory Test	Result
White blood cell count	18,000/ $\mu$ L ( $18 \times 10^9$ /L)
Hemoglobin	15 g/dL (150 g/L)
Hematocrit	45%
Platelets	$150 \times 10^3$ / $\mu$ L ( $150 \times 10^9$ /L)
Sodium	145 mEq/L (145 mmol/L)
Potassium	4.5 mEq/L (4.5 mmol/L)
Chloride	108 mEq/L (108 mmol/L)
Carbon dioxide	18 mEq/L (18 mmol/L)
Urea nitrogen	30 mg/dL (10.7 mmol/L)
Creatinine	1.3 mg/dL
Glucose	100 mg/dL (5.5 mmol/L)
C-reactive protein	98 mg/dL (980 mg/L)
Erythrocyte sedimentation rate	80 mm/hour

On physical examination, she has pain to palpation of her lumbar spine, but there is no discoloration or rash on the back. With lower-extremity strength testing, she has severe pain when lifting her leg off the bed.

She is given 2 boluses of lactated Ringer solution and started on ceftriaxone for presumed sepsis. Blood and urine cultures are obtained. Her blood pressure improves to 90/50 mm Hg with fluid boluses, and she makes some urine. However, overnight she has a precipitous drop in her urine output, and a bladder scan reveals a bladder volume of 800 mL. She continues to be febrile, and her back pain becomes more severe, requiring multiple doses of fentanyl to provide relief. A urinary catheter is placed with appropriate drainage of large amounts of urine. However a few hours later she calls out to the nurse because she is feeling as if she cannot move her legs normally. Magnetic resonance imaging with gadolinium contrast is obtained (**Figure**).

**Figure.** Magnetic resonance image of the spine of the patient in the vignette.



Courtesy of C. Guimaraes

Of the following, the MOST likely diagnosis is

- A. acute flaccid myelitis
- B. Guillain-Barré syndrome
- C. spinal cord tumor
- D. spinal epidural abscess

Your answer is correct.

### PREP Pearl(s)

- The diagnosis of spinal epidural abscess requires a high index of suspicion, and patients may not receive a correct diagnosis until neurologic deficits develop.

- Spinal epidural abscess should be suspected in patients with fever, back pain, and spinal tenderness with or without neurologic deficits and/or urinary retention.

## Critique

The patient in this vignette has a spinal epidural abscess (SEA), an infection of the epidural space along the spinal cord. Spinal epidural abscess should be considered in any patient with fever, back pain, and spinal tenderness with or without neurologic deficits and/or urinary retention. If left unrecognized, SEA may progress to irreversible neurologic deficits and/or sepsis. Progression can be extremely rapid, from weakness to paralysis in less than 24 hours. The literature (largely adult) suggests that paralysis lasting more than 48 hours in cases of SEA is likely to be permanent. Therefore, prompt recognition followed by antibiotic therapy and often surgical management are necessary. Spinal epidural abscess is a high-risk low-prevalence condition, and thus requires a high index of suspicion for diagnosis.

The epidural space is the area between the dura mater and the vertebral wall; it is larger in the lumbar and sacral region versus the cervical spine, and therefore, SEA is more common in the thoracolumbar area. Hematologic spread is hypothesized as the mechanism of infection, as the epidural space is very vascular with arteries and a venous plexus. Most epidural abscesses are located posterior to the thecal sac.

Magnetic resonance imaging (MRI) is the diagnostic modality of choice for SEA. The MRI of the patient in the vignette reveals discitis/osteomyelitis at L5-S1 and a resultant SEA. This represents a spectrum of vertebral infections which are most often caused by *Staphylococcus aureus* (>50% of cases). Infections with both methicillin-sensitive and methicillin-resistant *S aureus* have been reported. Other organisms reported to cause SEA in children are group A *Streptococcus* and nontuberculous mycobacterium. Spinal epidural abscess may be associated with spinal instrumentation. In immunocompetent nonpostoperative patients, the source is often a skin or soft tissue infection with hematologic spread of the infection into the spinal epidural space.

Because of the rare nature of this condition, guidelines for management are not available. Case series report prolonged antibiotic treatment (average 6 weeks, with a range of 2-14 weeks), and most patients had operative drainage of their abscess with laminectomy.

The differential diagnosis for SEA includes: spinal cord trauma, spinal cord tumor, discitis, transverse myelitis, and acute flaccid myelitis.

Acute flaccid myelitis (AFM) is most commonly associated with enterovirus D68 infection, and it is characterized by limb weakness and variable cranial nerve involvement following a febrile or upper respiratory tract illness. Other viruses have also been implicated. This patient has weakness, but the MRI findings are not characteristic of AFM. The MRI for AFM reveals T2 hyperintense lesions restricted to the gray matter or predominantly involving the gray matter and some white matter. In addition, most cases of AFM span multiple lesions of the spine and often the entire spine.

Guillain-Barré syndrome (GBS) presents with paresthesias of the distal extremities and ascending paralysis. This patient did not demonstrate this pattern, although bladder dysfunction and autonomic dysfunction may be seen in GBS. Spinal MRI in GBS frequently shows enhancement of the spinal cord roots and cauda equina.

Spinal cord tumors present with back pain and/or weakness, but are not characterized by fever or a sepsis picture. Patients with spinal cord tumors may also have an abnormal gait. Symptoms are largely dependent on the tumor location and extension.

## Suggested Reading(s)

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- Spennato P, Renedo D, Cascone D, Mirone G, Imperato A, Di Martino G, Cinalli G. Spinal epidural abscess in children: a case-based review. *Childs Nerv Syst.* 2020;36(7):1385-1392. doi:[10.1007/s00381-020-04609-3](https://doi.org/10.1007/s00381-020-04609-3)

## Content Domain

- Infectious Diseases, Central Nervous system infections

## Learning Objectives

- Recognize the diagnosis of spinal epidural abscess
- Recognize risk factors and pre-existing conditions associated with spinal epidural abscess
- Plan initial treatment of spinal epidural abscess

The correct answer is: spinal epidural abscess

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**Question 2**

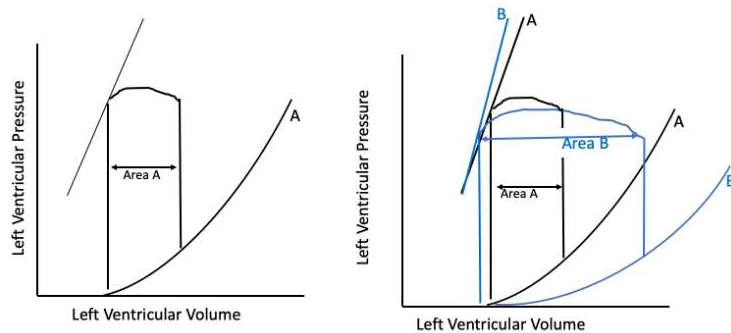
Correct

1.00 points out of 1.00

[Comment](#)

Left ventricular pressure-volume loop measurements are performed on a patient recovering from cardiac surgery. The data are shown (Figure 1):

Figure 1. LV pressure volume loop of cardiac cycle with baseline loop A and changes seen in loop B.



Courtesy of J. Kane

A baseline loop is generated at time A (black lines). The patient then receives a therapy and a new pressure-volume loop is generated at time B (blue lines).

Of the following, the therapy administered resulting in the change observed in loop B (area B-blue lines) is MOST likely

- A. epinephrine infusion
- B. milrinone infusion
- C. nicardipine infusion
- D. preload bolus infusion

Your answer is correct.

**PREP Pearl(s)**

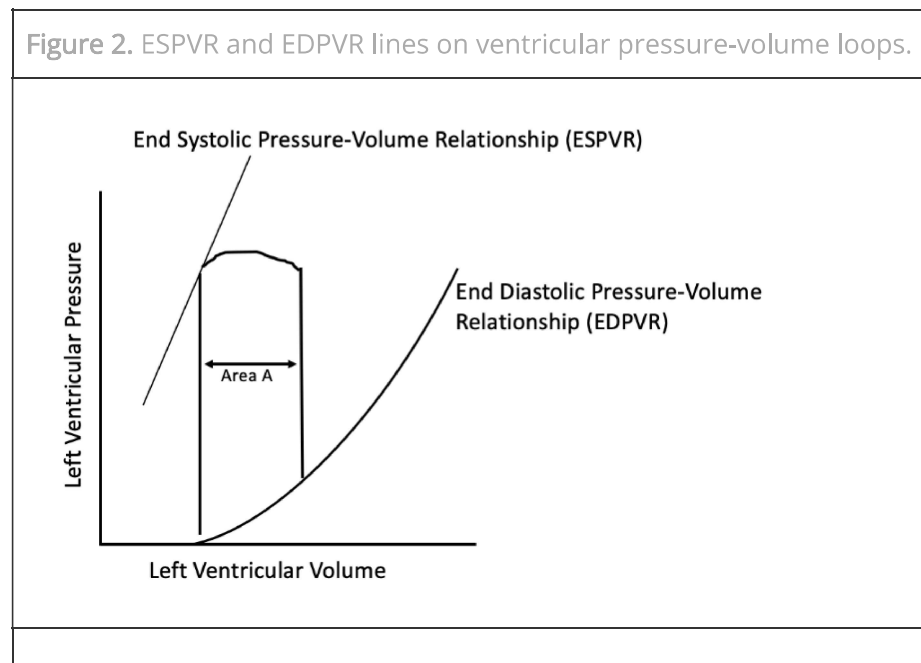
- In pressure-volume loops representing the cardiac cycle, changes in ventricular preload are represented by different points on the same end-diastolic pressure-volume relationship line.
- In pressure-volume loops representing the cardiac cycle, changes in ventricular contractility are represented by different slopes of different end-systolic pressure-volume relationship lines.
- Milrinone increases inotropy and lusitropy and reduces afterload, resulting in a different end-diastolic and end-systolic pressure-volume relationship and a larger stroke volume.

**Critique**

A mastery of the left ventricular (LV) pressure-volume loop is key to true comprehension of the dynamics of cardiac function, especially when considering changes in the loop diagrams as a result of medical interventions. During diastolic filling, the LV pressure is initially lower than the left atrial pressure, allowing blood to flow through the mitral valve. Volume increases along the end-diastolic pressure-volume relationship line with a gradual increase in pressure when the volume of blood in the left ventricle is at its highest immediately before contraction. Next, filling stops due to closure of the mitral valve, as left ventricular pressure surpasses left atrial pressure. The ventricular pressure continues to increase (isovolumetric contraction) until the aortic valve opens, causing decrease in LV volume throughout the duration of contraction. Left ventricular volume decreases while LV pressure is sustained during contraction. Subsequently, as the LV relaxes and aortic pressure exceeds LV pressure, the aortic valve closes in diastole, causing a rapid decrease in LV pressure, resulting in mitral valve opening and returning to the beginning of the cycle with initiation of diastolic filling of the LV.

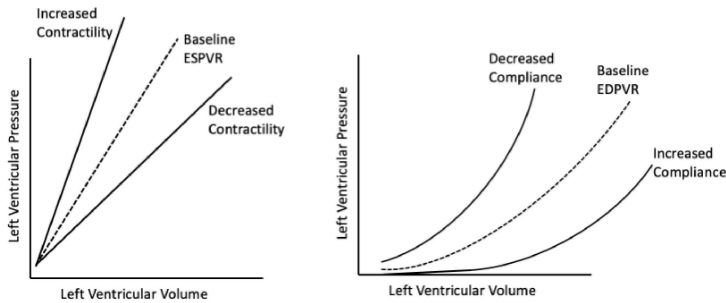
In the vignette, curve B shows a different end-diastolic pressure-volume relationship compared to curve A, with higher ventricular compliance, resulting in more ventricular filling before the start of systole and isovolumetric contraction. Aortic valve opening occurs at a lower pressure in curve B compared to curve A, suggesting lower afterload. There is also a higher slope-to-end systolic pressure-volume relationship line in curve B, indicating higher contractility. Finally, the total stroke volume for curve B is greater than curve A. Of the medications listed, the use of milrinone would result in changes observed in curve B (lusitropy causing increased ventricular compliance, inotropy increasing contractility, and lower afterload resulting in larger stroke volume).

Physiologic changes in preload, afterload, and contractility are easily represented on the ventricular pressure-volume loops (**Figure 2**). Changes in preload affect the end-diastolic pressure-volume relationship. The end-systolic pressure-volume relationship represents the maximal pressure developed by the LV at any given volume and is a measure of cardiac contractility.



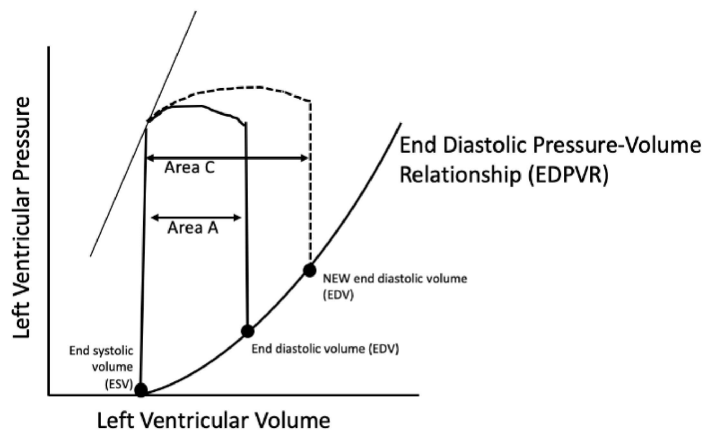
Left ventricular compliance is the reciprocal of the end-diastolic pressure-volume relationship slope. Therefore, decreased compliance (stiffer ventricle) is indicated by an upwards shift of the end-diastolic pressure-volume relationship, and increased compliance (increased lusitropy) is represented by a downward rightward shift of the end-diastolic pressure-volume relationship. The slope of end-systolic pressure-volume relationship, referred to as  $E_{es}$ , is an index of end-systolic elastance, and a change in  $E_{es}$  can be viewed as a change in contractility (**Figure 3**).

**Figure 3.** Contractility changes represented by different slopes in the ESPVR; compliance changes represented by EDPVR lines.



A fluid bolus would increase LV filling, however, it would not affect LV compliance. Thus, a pressure-volume curve demonstrating increased preload (as would be seen with a fluid bolus) would have the same end-diastolic pressure-volume relationship line, and result in higher stroke volume. Each pressure-volume loop with increasing preload has its own unique end-systolic and end-diastolic point but along the same end-diastolic and end-systolic pressure-volume relationship lines (**Figure 4**).

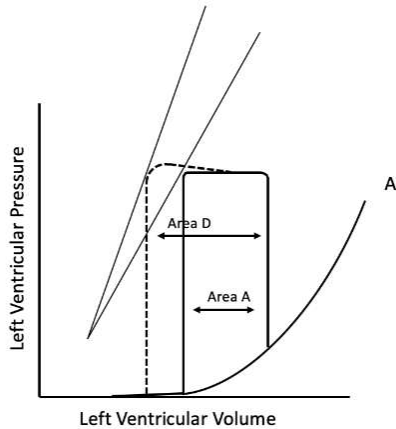
**Figure 4.** Pressure volume loop changes due to increased preload (dotted line area C): larger stroke volume, same EDPVR and ESPVR relationships.



Pressure volume loop changes due to increased preload (dotted line Area C): Larger stroke volume, same EDPVR and ESPVR relationships. Stroke Volume= EDV-ESV

An epinephrine infusion would result in increased cardiac contractility. There would be no expected change in end-diastolic pressure-volume relationship, and the slope of the end-systolic pressure-volume relationship line would be shifted to the left (**Figure 5**).

**Figure 5.** Pressure volume loop changes due to increased contractility: larger stroke volume, increase in ESPVR slope.



Pressure volume loop changes due to increased contractility: Larger stroke volume, increase in ESPVR slope

All images Courtesy of J. Kane

Nicardipine would affect afterload without affecting preload or contractility. There would be no expected change in the end-diastolic nor the end-systolic pressure-volume relationship; however, LV ejection would occur at a lower pressure along the isovolumetric contraction line, and stroke volume would be expected to be larger.

## Suggested Reading(s)

- Brenner MI, Masoumi A, Ng VG, et al. Invasive right ventricular pressure-volume analysis: basic principles, clinical applications, and practical recommendations. *Circ: Heart Fail.* 2022;15(1):e009101. doi:[10.1161/circheartfailure.121.009101](https://doi.org/10.1161/circheartfailure.121.009101)
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- Hiremath G, Batlivala S, Callahan R, et al. Clinical applications of pressure-volume assessment in congenital heart disease. *J Soc Cardio Ang Interv.* 2023;2(3):100599. doi:[10.1016/j.jscai.2023.100599](https://doi.org/10.1016/j.jscai.2023.100599)

## Content Domain

- Physiology, Cardiovascular

## Learning Objectives

- Describe the characteristic changes in pressure-volume loops due to common interventions.
- Identify key characteristics of pressure volume loops in relation to the cardiac cycle.

The correct answer is: milrinone infusion

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**Question 3**

Correct

1.00 points out of 1.00

[Comment](#)

A 17-year-old is a lap-belt restrained passenger in a motor vehicle collision where the driver dies at the scene. The patient is extricated from the vehicle by emergency services; they are crying and able to speak, open both eyes to voice, and are able to move all extremities. Vital signs are a heart rate of 154 beats/min, blood pressure of 135/70 mm Hg, respiratory rate of 24 breaths/min, and oxygen saturation of 92% in room air. On examination, they are found to have slightly diminished breath sounds on the left side, with paradoxical motion of the ribs on the left side, such that they move inward on inspiration and outward on exhalation. Their abdomen is soft and nontender to palpation. Bedside ultrasonography demonstrates left-sided lung sliding

Of the following, the BEST next step in management is

- A. chest tube placement
- B. intubation
- C. needle decompression
- D. supplemental oxygen with a nonrebreather mask ✓

Your answer is correct.

**PREP Pearl(s)**

- Flail chest occurs when 3 or more ribs are fractured in 2 or more places. Paradoxical motion of the flail segment occurs, such that it moves inward on inspiration and outward on exhalation.
- Flail chest is commonly accompanied by pulmonary contusion.
- In children, there is no preference for nonoperative versus surgical stabilization of the rib fractures.

**Critique**

The patient in this scenario has a flail chest, which occurs when 3 or more ribs are fractured in 2 or more places. Flail chest is typically seen in older adults, often following high impact chest trauma. The thoracic cage in children is more compliant than adults, making it harder for children to fracture ribs. Therefore, flail chest is seen very infrequently in children, with reports of a 2% to 4% occurrence following blunt chest trauma. Flail chest can be difficult to diagnose initially, as spasms of intercostal muscles may mask the degree of paradoxical movement of the flail segment.

Due to the large mechanical forces required to cause flail chest, flail chest is frequently associated with concomitant intrathoracic or abdominal injuries. Most commonly, flail chest is accompanied by pulmonary contusion. It is thought that the presence of pulmonary contusion contributes significantly to the mortality rate of 10% to 15% seen in adults with flail chest, as pulmonary contusions are associated with evolving edema, inflammation, and worsening pulmonary function. Hemothorax and pneumothorax may also be present in this population.

Treatment initially should focus on the airway, breathing, and circulation. Patients should be given supplemental oxygen, and in cases of respiratory failure, mechanical ventilation may be required. Patients with flail chest have significant pain associated with rib fractures. This makes them susceptible to atelectasis from splinting and difficulty with secretion clearance due to poor cough. There may also be reduction in diaphragmatic movement and the paradoxical movement of the chest wall may decrease effective air entry, thus resulting in decreased minute ventilation.

Stabilization of the flail segment can be performed with rolled towels or tape. Nonoperative management relies on adequate pain management (opiates, lidocaine patches, intercostal nerve blocks, or epidural catheters) and aggressive pulmonary toilet. Surgical stabilization of rib fractures has gained interest in adults, with studies showing shorter mechanical ventilation and hospital stays. However, there is no agreement on whether nonoperative or operative strategies are preferred in children.

In this vignette, the best next step is supplemental oxygen with a nonrebreather face mask. The patient does not have respiratory failure, therefore intubation is incorrect. Pneumothorax and hemothorax can be associated with flail chest; however, the patient in this scenario does not have those associated symptoms or ultrasonographic evidence of pneumo- or hemothorax. Therefore, needle decompression and chest tube placement are not the best next step.

### Suggested Reading(s)

- Majercik S, Pieracci FM. Chest wall trauma. *Thorac Surg Clin.* 2017;27(2):113-121. doi:[10.1016/j.thorsurg.2017.01.004](https://doi.org/10.1016/j.thorsurg.2017.01.004)
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### Content Domain

- Critical Care, Trauma

### Learning Objectives

- Identify the presence of a flail chest.
- Describe the treatment strategies for flail chest

The correct answer is: supplemental oxygen with a nonrebreather mask

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**Question 4**

Correct

1.00 points out of 1.00

[Comment](#)

A 10-year-old child was admitted to the pediatric intensive care unit with severe traumatic brain injury after a motor vehicle accident. He sustained severe hypoxic ischemic injury, and despite aggressive care for the past 7 days the patient remains unresponsive, and the medical team believes he has no chance of meaningful neurologic recovery. The family wishes to proceed with withdrawal of life-sustaining therapies, and they would like him to be an organ donor. He is determined to be a candidate for donation after circulatory death.

After preparations are made with the organ procurement organization, the team proceeds with extubation as well as discontinuation of vasoactive medications. The operating room team is informed and remains on standby. Ninety minutes later, the patient has a heart rate of 35 beats/min, a blood pressure of 40/15 mm Hg, respiratory rate of 5 breaths/min, and an oxygen saturation of 55% in room air.

Of the following, the BEST next step is

- A. continue with the current plan to see if he would go into asystole at the 2-hour mark
- B. inform the operating team that he is no longer a donor candidate, as circulatory death has not occurred
- C. resume life-sustaining therapies
- D. start oxygen via nasal cannula to increase saturations and oxygen delivery to organs

Your answer is correct.

**PREP Pearl(s)**

- Organ donation after circulatory death is an accepted practice in pediatric critical care and has been endorsed by the American Academy of Pediatrics.
- Time to death after withdrawal of life-sustaining treatments is highly impactful to organ viability, and accepted warm ischemia time ranges from 30 to 90 minutes.
- Outcomes of donor organs from donation after circulatory–death donors are equivalent to those organs retrieved from donors following neurologic death.

**Critique**

Organ donation after circulatory death (DCD) has become an increasingly accepted practice in critical care. Pediatric acceptance of this practice has been slower, with continued national and regional variations and also fewer overall organ donors compared to adults. Donation after circulatory death is defined as the retrieval of organs for the purpose of transplantation from patients whose death is diagnosed and confirmed via cardiorespiratory criteria. The number of pediatric DCD donors has increased annually, and children account for 4% to 5% of all DCD donors in the United States. The proportion of organs transplanted into pediatric recipients from DCD donors has also increased annually, and DCD as an important source of organs

for children and adult transplant recipients. Nevertheless, DCD donors are fewer in numbers compared to donation after death determination by neurologic criteria (DNC), and there remains unfamiliarity surrounding the practice.

In general, DCDs are categorized by the Maastricht criteria, as follows:

**Category 1:** The donor is declared dead before arriving at the hospital.

**Category 2:** The donor dies after unsuccessful resuscitation attempts.

**Category 3:** The donor's death is declared after circulatory arrest occurs following withdrawal of life sustaining treatment (WLST).

**Category 4:** The donor experiences cardiac arrest while brain dead.

Categories 1, 2, and 4 are considered uncontrolled DCD. Category 3 is a controlled DCD and is primarily practiced in the United States. Controlled DCD typically occurs in the intensive care unit or the operating room.

There are currently no internationally accepted standards for pediatric controlled DCD. However, national guidelines do exist, such as the 2017 Canadian guidelines, which include practice statements and GRADE recommendations for the ethical and practical aspects of DCDs.

A challenge of DCDs is the unpredictability of determining when circulatory arrest will occur after WLST. Time to death after WLST is key to organ donation, as warm ischemia time may reduce the viability of organs to be transplanted. The maximum warm ischemia time varies by organs, as well as local transplantation centers. The most common time-range for circulatory arrest to occur is between 30 and 90 minutes; if circulatory arrest occurs within this time period, there is a minimum of 2 minutes, up to 5 minutes, of observation time for sustained mechanical asystole to ensure there is no return of spontaneous circulation. If there is no return of spontaneous circulation during this observation (or "hands off" period), the patient is declared dead, and recovery of organs occurs. Importantly, these patients receive the same treatment to alleviate pain and discomfort as any other patient undergoing end-of-life care.

Outcomes for transplanted organs procured via the DCD pathway have significantly improved with standardized processes to determine death and recover organs. In 2023, *The New England Journal of Medicine* published the results of a randomized noninferiority study comparing transplantation outcomes. The study found that the 6-month risk-adjusted survival of recipients receiving an organ from a circulatory death donor were not inferior to those who received a heart from a brain-death donor. Multiple reports and studies support that outcomes have improved for DCD liver, kidney, and lung transplantation with graft survival rates comparable to organs recovered from brain dead donors. The use of evolving technology that includes normothermic regional perfusion is a method of maintaining potential transplantable organs in situ by instituting a modified veno-arterial extracorporeal membrane oxygenation (VA-ECMO) technique to allow the transplant teams more time to recover organs after warm ischemia time. This method was used mainly for abdominal organs, however, thoracoabdominal regional perfusion is emerging as a strategy to maintain donor hearts as an alternative to the organ care system that is a machine perfusion device. External machine perfusion devices allow for organ recovery and support once organs are removed from the body.

There are many ethical considerations surrounding DCD. Ethical safeguards are thus essential. Transparency in communications and clarity in roles holds critical importance; any health care system that participates in DCDs should have established processes. The decision to pursue WLST must be kept as separate from the decision to pursue organ donation. To avoid real or perceived conflicts of interest, the organ-procuring organization, organ recovery, and transplant team must not be involved in the decision to pursue WLST or the determination of death. Discussions for organ donation should not be mentioned by any treatment team member prior to WLST decisions, unless the family initiates this discussion. Offering DCD as part of pediatric end-of-life care is universally supported by multiple professional medical societies internationally, as well as

the American Academy of Pediatrics. However, some health care professionals may have differing views of the ethical acceptability of this practice depending on societal, personal, cultural, or religious beliefs. These health care professionals should not be asked to participate in this type of donation. Institutions should work to make accommodations for these professionals, as well as honor the wishes of substitute decision-makers and families who wish to participate in the DCD process.

## Suggested Reading(s)

- Antommaria AHM, Fallat ME, Katz AL, et al; American Academy of Pediatrics Committee on Bioethics. Ethical controversies in organ donation after circulatory death. *Pediatrics*. 2013;131(5):1021–1026. doi:[10.1542/peds.2013-0672](https://doi.org/10.1542/peds.2013-0672)
- Kwon J, Blanding W, Shorbaji K, Scalea J, Gibney B, Baliga P, Kilic A. Waitlist and transplant outcomes in organ donation after circulatory death. *Ann Surg*. 2023;278(4):609-620. doi:[10.1097/sla.0000000000005947](https://doi.org/10.1097/sla.0000000000005947)
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## Content Domain

- Critical Care, End of Life

## Learning Objectives

- Recognize eligibility criteria for donation after circulatory death
- Identify ethical considerations around donation after circulatory death

The correct answer is: inform the operating team that he is no longer a donor candidate, as circulatory death has not occurred

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**Question 5**

Correct

1.00 points out of 1.00

[Comment](#)

A 10-year-old child presents with a 2-day history of confusion and drowsiness. These symptoms were preceded by 2 weeks of fatigue, poor appetite, and nose bleeds. On examination, he is pale, confused, and drowsy but arousable to physical stimuli. A cranial nerve examination is normal, showing good tone and strength in all extremities with no signs of meningeal irritation. He is afebrile and mildly tachypneic, with oxygen saturations of 92% in ambient air, and has clear lungs on auscultation. He is well-perfused, has normal heart sounds without a murmur, rub, or gallop.

Laboratory studies show a hemoglobin of 8 g/dL (80 g/L), white blood cell count of 202,000/ $\mu\text{L}$  ( $202 \times 10^9/\text{L}$ ) with 30% myeloblasts, and a platelet count of  $58 \times 10^3/\mu\text{L}$  ( $58 \times 10^9/\text{L}$ ). Chest radiograph and noncontrast computed tomography scan of the brain are normal. Electrolyte profile shows hypocalcemia, mild hyperuricemia, mild hyperkalemia.

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

- A. ceftriaxone
- B. hyperhydration with intravenous fluids ✓
- C. packed red blood cell transfusion
- D. platelet transfusion

Your answer is correct.

**PREP Pearl(s)**

- Hyperleukocytosis, defined as a white blood cell count greater than 100,000/ $\mu\text{L}$  ( $100 \times 10^9/\text{L}$ ) is a medical emergency that needs prompt recognition and treatment.
- Individuals with hyperleukocytosis are at high risk for developing disseminated intravascular coagulation, tumor lysis syndrome, stroke, and leukostasis.
- Management of hyperleukocytosis includes cytoreductive therapy, hyperhydration, and control of uric acid production.
- Chemotherapy and leukapheresis are effective cytoreduction therapies.

**Critique**

Hyperleukocytosis is defined as a white blood cell count (WBC) greater than 100,000/ $\mu\text{L}$  ( $100 \times 10^9/\text{L}$ ). Hyperleukocytosis has been reported in 10% to 20% of children with acute myeloid leukemia (AML) and acute lymphoblastic leukemia (ALL), and in individuals with chronic leukemias and pertussis. Individuals with hyperleukocytosis have increased morbidity and mortality and are predisposed to developing leukostasis, tumor lysis syndrome, and disseminated intravascular coagulation (DIC).

Leukostasis occurs as a result of increased viscosity in the microcirculation caused by the increased number of larger and less deformable blast cells. Blast cells release proinflammatory cytokines and upregulate adhesion molecules, leading to increased migration of blast cells, transmigration into the tissues, and

endothelial and tissue damage. The elevation in WBC count does not correlate with symptoms. Symptoms are more frequently seen with AML than ALL. The lungs, brain, and kidneys are most frequently affected. Symptoms are nonspecific and include cough, dyspnea, hypoxemia, confusion, somnolence, dizziness, headache, delirium, focal neurological deficits, seizures, impaired vision, and tinnitus. Chest radiographs may show nonspecific patterns with bilateral diffuse interstitial or alveolar infiltrates. Brain imaging may show ischemic or hemorrhagic lesions that may be localized or diffuse.

Hyperleukocytosis is an emergency, and cytoreductive therapy should be initiated immediately. Leukapheresis (and exchange transfusion in younger children) has been found to be safe and effective in reducing the WBC count quickly. However, these therapies have failed to demonstrate improved survival rates or reduced adverse events as compared to early induction chemotherapy. Although no randomized controlled trials have been done comparing leukapheresis to chemotherapy, systematic reviews and meta-analyses suggest that routine use of leukapheresis for management of hyperleukocytosis is not beneficial. The American Society for Apheresis guidelines mention leukapheresis as a category III indication, either alone or in conjunction with other therapies, for symptomatic hyperleukocytosis or leukostasis (eg, coma, stroke). Supportive care includes hyperhydration with intravenous fluids for tumor lysis syndrome (with or without diuretics) and treatment with rasburicase or allopurinol for hyperuricemia. Transfusion of packed red blood cells should be avoided at this time as the resultant increase in hematocrit may worsen hyperviscosity. Platelet transfusion should not be required as a platelet count of  $58 \times 10^3/\mu\text{L}$  ( $58 \times 10^9/\text{L}$ ) should not be associated with significant risk of bleeding. Empiric antibiotic use is not indicated.

## Suggested Reading(s)

- Arad-Cohen N, Zeller B, Abrahamsson J, et al. Supportive care in pediatric acute myeloid leukemia: expert-based recommendations of the NOPHO-DB-SHIP consortium. *Expert Rev Anticancer Ther.* 2022;22(11):1183-1196. doi:[10.1080/14737140.2022.2131544](https://doi.org/10.1080/14737140.2022.2131544)
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- Shallis RM, Stahl M, Bewersdorf JP, Hendrickson JE, Zeidan AM. Leukocytapheresis for patients with acute myeloid leukemia presenting with hyperleukocytosis and leukostasis: a contemporary appraisal of outcomes and benefits. *Expert Rev Hematol.* 2020;13(5):489-499. doi:[10.1080/17474086.2020.1751609](https://doi.org/10.1080/17474086.2020.1751609)

## Content Domain

- Hematology, Leukocytes

## Learning Objectives

- Describe the complications and management of hyperleukocytosis

The correct answer is: hyperhydration with intravenous fluids

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**Question 6**

Correct

1.00 points out of 1.00

[Comment](#)

A previously healthy 6-month-old infant presents to the pediatric intensive care unit with severe respiratory distress and episodes of lethargy that culminate in brief periods of postprandial unresponsiveness. Over the past month, the infant exhibited diaphoresis and pronounced irritability with feeds, with concurrent poor weight gain. On examination, vital signs are heart rate of 180 beats/min, respiratory rate of 70 breaths/min, blood pressure of 70/45 mm Hg, and oxygen saturation of 92% in room air. The infant is pale with cool extremities and exhibits tachypnea and tachycardia. A faint pansystolic murmur is detectable at the left sternal border. Capillary refill time is delayed; there is no radiofemoral delay. Venous blood gas shows a pH of 7.28, PaCO<sub>2</sub> of 32 mm Hg, bicarbonate of 17 mmol/L, and lactate of 5 mmol/L. An electrocardiogram is performed (**Figure**).

**Figure.** Electrocardiogram of Patient in Vignette.



Courtesy of M. Rowin

Of the following, the MOST likely diagnosis is

- A. anomalous left coronary artery from the pulmonary artery ✓
- B. congenital dilated cardiomyopathy
- C. critical coarctation of the aorta
- D. septic shock

Your answer is correct.

**PREP Pearl(s)**

- Infants with an anomalous left coronary artery from the pulmonary artery (ALCAPA) may present after 2 to 3 months with symptoms such as feeding difficulty, diaphoresis with feeds, or tachypnea associated with activities that increase myocardial demand.

- Early diagnosis of an anomalous left coronary artery from the pulmonary artery (ALCAPA) is crucial since it can lead to myocardial ischemia and congestive heart failure.
- The hallmark of anomalous left coronary artery from the pulmonary artery (ALCAPA) is the coronary steal phenomenon, where blood preferentially flows into the pulmonary artery instead of the myocardium, leading to myocardial ischemia, most notably of the left ventricle.

## Critique

The infant in this vignette likely has an anomalous left coronary artery from the pulmonary artery (ALCAPA), a rare congenital cardiac anomaly which primarily manifests in infancy with clinical signs of myocardial ischemia and congestive heart failure. In this diagnosis, the left coronary artery anomalously emerges from the pulmonary artery, rather than its normal origin in the aorta. Embryologically, this defect is thought to arise from an aberrant division of the conotruncus into the aortic and pulmonary arteries, or due to the persistence of aortic buds, which ultimately become the coronary arteries. Although this anomaly predominantly presents as an isolated condition, it has been observed in conjunction with other congenital heart defects such as patent ductus arteriosus, ventricular septal defect, tetralogy of Fallot, or coarctation of the aorta. In fetal circulation, the pulmonary artery and systemic pressures are equal; this allows for sufficient global myocardial perfusion through the anomalous coronary artery. However, following birth, pulmonary vascular resistance falls rapidly, with concurrent significant increased metabolic demands of the left ventricle. During times of increased physiologic demands such as feeding or crying, the left ventricle is suboptimally supplied with deoxygenated blood from a low pressure system, thereby predisposed to myocardial ischemia.

Collateralization may develop between the right and left coronary arteries to provide perfusion to the myocardium of the left ventricle, but does not provide sufficient perfusion during times of increased physiologic demand. As pulmonary vascular resistance falls, a coronary steal phenomenon arises, with a diversion of blood away from the high-resistance towards the lower pressure pulmonary circulation; the coronary artery and the supplementary collateral circulation preferentially perfuse the pulmonary artery over the myocardium. This left-to-right shunting from the higher pressure left coronary arterial system to the lower pressure pulmonary arterial system leads to circulatory insufficiency, myocardial infarction, or life-threatening cardiac dysrhythmias. The sequelae of this maladaptive circulation leads to myocardial ischemia, specifically targeting the anterolateral wall of the left ventricle, predisposing it to infarction. The subsequent myocardial injury not only contributes to the enlargement of the heart, but also precipitates the onset of congestive heart failure, which is frequently exacerbated by mitral incompetence due to dilation of the mitral annulus or from necrosis of the papillary muscles from infarction. Notable electrocardiogram (ECG) findings in the **Figure** demonstrate Q waves in I and aVL, ST elevation in V3-V5, and inverted T waves in V6.

Neonates with ALCAPA are typically asymptomatic at birth, displaying a transient period of apparent well-being. Onset of symptoms generally occurs between 2 to 3 months of age, coinciding with the normalization of pulmonary arterial resistance. The clinical presentation may initially be characterized by acute episodes of distress during feeding, progressing to pronounced pallor, irritability, and diaphoresis with feeds or any situation that increases myocardial demand. The clinical spectrum of ALCAPA is diverse; a significant proportion of infants may instead present with hallmark features of congestive heart failure, including tachypnea, tachycardia, diaphoresis, compromised feeding efficiency, and inadequate weight progression as noted in the clinical presentation of this patient, making ALCAPA the most likely diagnosis.

An unrecognized dilated cardiomyopathy may present with a similar presentation of decompensated heart failure; however, the ECG findings are pathognomonic of myocardial ischemia associated with ALCAPA, and generally not seen in heart failure. Similarly, although patients may present in extremis with critical coarctation of the aorta, this is generally not associated with the aforementioned unique ECG changes. Moreover, there is no radiofemoral delay on physical examination in this vignette. Severe septic shock with subsequent myocardial dysfunction could theoretically lead to a similar presentation with metabolic acidosis,

tachycardia, and signs of poor perfusion. However, the specific ECG findings of myocardial ischemia and the lack of fever or other clinical signs of infection, make septic shock an unlikely primary diagnosis for this patient. In addition, sepsis-induced myocardial dysfunction would more likely cause diffuse ECG changes rather than the localized changes seen in this case.

## Suggested Reading(s)

- Bhushan R, Mallik M, Potey K, Grover V, Aiyer P, Jhahria NS. Anomalous origin of the left coronary artery from the pulmonary artery: a midterm experience of a rare entity at a tertiary care center. *J Cardiovasc Thorac Res.* 2023;15(3):181-185. doi:[10.34172/jcvtr.2023.31651](https://doi.org/10.34172/jcvtr.2023.31651)
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- Hoffman JIE. Electrocardiogram of anomalous left coronary artery from the pulmonary artery in infants. *Pediatr Cardiol.* 2013;34,489–491. doi:[10.1007/s00246-012-0599-7](https://doi.org/10.1007/s00246-012-0599-7)
- Peña E, Nguyen ET, Merchant N, Dennie C. ALCAPA syndrome: not just a pediatric disease. *Radiographics.* 2009;29(2):553-65. doi:[10.1148/rg.292085059](https://doi.org/10.1148/rg.292085059)

## Content Domain

- Physiology, Cardiovascular

## Learning Objectives

- Describe the anatomy and pathophysiology of ALCAPA
- Recognize the clinical presentation of ALCAPA
- Interpret diagnostic findings in ALCAPA

The correct answer is: anomalous left coronary artery from the pulmonary artery

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