

An 11-year-old boy with sickle cell disease is brought to the emergency department for worsening shortness of breath, weakness, and fatigue over the past 3 days. He has no fever, cough, or chest pain. His medical history includes many hospitalizations for treatment of acute pain and one hospitalization for acute chest syndrome. Physical examination shows a pale, tired-appearing boy with a III/VI systolic murmur heard throughout the precordium. Lungs are clear to auscultation. His abdomen is soft and nontender with no palpable liver or spleen. Laboratory results are as follows:

Hemoglobin	4.5 g/dL
Reticulocytes	0.1%
Platelets	200,000/mm ³
Leukocytes	10,000/mm ³

Which of the following is the most likely cause of this patient's symptoms?

- ☐ A. Acute chest syndrome
- ☐ B. Aplastic anemia
- ☐ C. Aplastic crisis
- ☐ D. Autoimmune hemolytic anemia
- ☐ E. Myocardial infarction
- ☐ F. Splenic sequestration crisis

Submit

An 11-year-old boy with sickle cell disease is brought to the emergency department for worsening shortness of breath, weakness, and fatigue over the past 3 days. He has no fever, cough, or chest pain. His medical history includes many hospitalizations for treatment of acute pain and one hospitalization for acute chest syndrome. Physical examination shows a pale, tired-appearing boy with a III/VI systolic murmur heard throughout the precordium. Lungs are clear to auscultation. His abdomen is soft and nontender with no palpable liver or spleen. Laboratory results are as follows:

Hemoglobin	4.5 g/dL
Reticulocytes	0.1%
Platelets	200,000/mm ³
Leukocytes	10,000/mm ³

Which of the following is the most likely cause of this patient's symptoms?

- ☐ A. Acute chest syndrome [11%]
- ☐ B. Aplastic anemia [15%]
- ☒ C. Aplastic crisis [56%]
- ☐ D. Autoimmune hemolytic anemia [3%]
- ☐ E. Myocardial infarction [2%]
- ☐ F. Splenic sequestration crisis [13%]

Proceed to Next Item

Explanation:

User Id:

Acute severe anemia in sickle cell disease		
Cause	Reticulocytes	Key features
Aplastic crisis	↓	<ul style="list-style-type: none">• Transient arrest of erythropoiesis• Secondary to infection (eg, parvovirus B19)

Explanation:

User Id: 

Acute severe anemia in sickle cell disease		
Cause	Reticulocytes	Key features
Aplastic crisis	↓	<ul style="list-style-type: none">• Transient arrest of erythropoiesis• Secondary to infection (eg, parvovirus B19)
Splenic sequestration crisis	↑	<ul style="list-style-type: none">• Splenic vasoocclusion → rapidly enlarging spleen• Occurs in children prior to autosplenectomy

©UWorld

Sickle cell disease (SCD) is characterized by chronic anemia with compensatory reticulocytosis. Patients with SCD are at risk for acutely worsening, **severe anemia**. An **acute** drop in hemoglobin accompanied by a **low reticulocyte** count ($<1\%$) **without splenomegaly** is most consistent with an **aplastic crisis**, which is characterized by a sudden halt in red blood cell production (erythropoiesis). The anemia is typically severe (<6 g/dL) and presents with pallor, weakness, and fatigue as well as a functional systolic murmur due to hyperdynamic blood flow. White blood cell and platelet counts are typically normal.

The most common cause of an aplastic crisis is **parvovirus B19**, which infects erythrocyte precursors. Aplastic crises generally present before age 15 as parvovirus outbreaks are most common in school-age children. Blood transfusions are the mainstay of treatment.

(Choice A) Acute chest syndrome is caused by pulmonary vasoocclusion or infection and is characterized by fever, chest pain, and a new infiltrate on chest radiograph. Acute chest syndrome is less likely in a patient without fever, cough, or chest pain and with clear lungs on auscultation.

(Choice B) Aplastic anemia, in contrast to aplastic crises, is characterized by pancytopenia (eg, thrombocytopenia, anemia, lymphopenia) due to bone marrow failure. Aplastic anemia can be congenital (eg, Fanconi anemia) or acquired (eg, drug-induced, autoimmune).

The most common cause of an aplastic crisis is **parvovirus B19**, which infects erythrocyte precursors. Aplastic crises generally present before age 15 as parvovirus outbreaks are most common in school-age children. Blood transfusions are the mainstay of treatment.

(Choice A) Acute chest syndrome is caused by pulmonary vasoocclusion or infection and is characterized by fever, chest pain, and a new infiltrate on chest radiograph. Acute chest syndrome is less likely in a patient without fever, cough, or chest pain and with clear lungs on auscultation.

(Choice B) Aplastic anemia, in contrast to aplastic crises, is characterized by pancytopenia (eg, thrombocytopenia, anemia, lymphopenia) due to bone marrow failure. Aplastic anemia can be congenital (eg, Fanconi anemia) or acquired (eg, drug-induced, autoimmune).

(Choice D) Both primary (eg, idiopathic) and secondary (eg, systemic lupus erythematosus) causes of autoimmune hemolytic anemia present with a dramatic decrease in hemoglobin. However, due to hemolysis, the reticulocyte count would be markedly elevated, and jaundice is typical due to hyperbilirubinemia from increased red blood cell turnover.

(Choice E) Patients with SCD are at risk for cardiac disease due to increased cardiac output secondary to chronic anemia and infarction of myocardial microvasculature. Infarction is more likely in adults and would also present with chest pain.

(Choice F) Splenic sequestration, a condition that occurs in patients with SCD whose spleens have not yet autoinfarcted, is caused by vasoocclusion and pooling of red blood cells in the spleen. Although splenic sequestration can also cause acute severe anemia, an elevated reticulocyte count and a rapidly enlarging spleen are also typical.

Educational objective:

Aplastic crises can cause acute severe anemia in patients with sickle cell disease and are characterized by a sudden drop in hemoglobin with a very low reticulocyte count (<1%) and lack of hepatosplenomegaly. Parvovirus B19 is the most common trigger.

References:

1. [Clinical presentations of parvovirus B19 infection.](#)
2. [Human parvovirus B19: general considerations and impact on patients with sickle-cell disease and thalassemia and on blood transfusions.](#)