

A 10-year-old patient with sickle cell disease comes to the physician for a routine visit. The patient has a history of multiple pain crises and pneumonias. He was started on hydroxyurea one year ago and has not had any further pain crises since then. He has had no recent illnesses or hospitalizations. His physical examination is unremarkable. The patient's laboratory results are shown below.

Complete blood count

Hemoglobin	9.0 g/L
Erythrocyte count	2.2 mln/mm <sup>3</sup>
MCHC	32%
MCV	105 fl
Reticulocytes	2.0%
Platelets	212,000/mm <sup>3</sup>
Leukocyte count	9500/mm <sup>3</sup>
Neutrophils	56%
Eosinophils	3%
Lymphocytes	36%
Monocytes	5%

Which of the following best describes the role of hydroxyurea in the treatment of patients with sickle cell disease?

- ☐ A. Removes sickled red blood cells from the circulation
- ☐ B. Lyses microthrombi in the circulation
- ☐ C. Protects against encapsulated bacterial infections
- ☐ D. Increases fetal hemoglobin
- ☐ E. Chelates iron to prevent iron toxicity

Submit



A 10-year-old patient with sickle cell disease comes to the physician for a routine visit. The patient has a history of multiple pain crises and pneumonias. He was started on hydroxyurea one year ago and has not had any further pain crises since then. He has had no recent illnesses or hospitalizations. His physical examination is unremarkable. The patient's laboratory results are shown below.

Complete blood count

Hemoglobin	9.0 g/L
Erythrocyte count	2.2 mln/mm <sup>3</sup>
MCHC	32%
MCV	105 fl
Reticulocytes	2.0%
Platelets	212,000/mm <sup>3</sup>
Leukocyte count	9500/mm <sup>3</sup>
Neutrophils	56%
Eosinophils	3%
Lymphocytes	36%
Monocytes	5%

Which of the following best describes the role of hydroxyurea in the treatment of patients with sickle cell disease?

- ☐ A. Removes sickled red blood cells from the circulation [3%]
- ☐ B. Lyses microthrombi in the circulation [2%]
- ☐ C. Protects against encapsulated bacterial infections [1%]
- ☒ D. Increases fetal hemoglobin [94%]
- ☐ E. Chelates iron to prevent iron toxicity [1%]

Proceed to Next Item



☐ C. Protects against encapsulated bacterial infections [1%]

☒ D. Increases fetal hemoglobin [94%]

☐ E. Chelates iron to prevent iron toxicity [1%]

Proceed to Next Item

### Explanation:

User Id: [REDACTED]

Hydroxyurea is a chemotherapy agent used in sickle cell disease to decrease vaso-occlusive pain crises as well as episodes of acute chest syndrome. Hydroxyurea works by increasing the amount of fetal hemoglobin in the circulation. Increasing the fetal hemoglobin dilutes the number of sickled cells in the circulation and reduces vaso-occlusive episodes. Hydroxyurea has been shown to decrease pain crises, the need for transfusions, and episodes of acute chest syndrome. Side effects occur because hydroxyurea suppresses the bone marrow. Leukopenia, anemia, and thrombocytopenia may occur. These effects are generally temporary and reversible but may predispose the patient to infection.

**(Choice A)** Hydroxyurea does not remove sickled cells from the circulation. The spleen is responsible for filtering out the sickled red blood cells.

**(Choice B)** Microthrombi caused by the sickled cells are the cause for vaso-occlusive pain crises. Hydroxyurea does not prevent or lyse these microthrombi, but decreases the proportion of sickled cells in the circulation.

**(Choice C)** Prophylactic penicillin is given to sickle cell patients to prevent infection with encapsulated organisms.

**(Choice E)** Patients with sickle cell disease often require transfusions. Frequent transfusions may lead to iron toxicity. Patients can be treated with iron chelators such as deferoxamine to help reduce the effects of the transfusion related hemosiderosis.

### Educational objective:

Hydroxyurea benefits patients with sickle cell disease by increasing fetal hemoglobin.

Time Spent: 2 seconds

Copyright © UWorld

Last updated: [10/17/2016]