**SICKLE CELL DISEASE: Dispelling Myths, Making Clinical Progress**

**LEARNING OBJECTIVES**

After completing this continuing education activity, pharmacy technicians will be able to

1. Compare the cost savings of newer agents to the standard of care—hydroxyurea
2. Describe SCD’s different phenotypes and implications for therapy
3. Determine when to refer patients to the pharmacists for recommendations or referrals

**1. Which of the following is TRUE about sickle cell trait?**

A. People with sickle cell trait never have symptoms or health complications

B. People with one sickle cell gene and one normal gene have sickle cell trait

C. People with sickle cell trait have more symptoms than those with sickle cell disease

**2. Which of the following mutations can cause two forms of a disease?**

A. HbAS

B. HbSD

C. HbS beta-thalassemia

**3. Which of the following patients is MOST LIKELY to need treatment for SCD?**

A. A patient with one sickle cell gene

B. A patient with two sickle cell genes

C. A patient who is of Mediterranean heritage

**4. A physician calls and says that she is trying to find the prescribing information for a medication for sickle cell disease that is a P-selectin blocker. What would your next step be?**

A. Tell the physician she is referring to hydroxyurea and fax a package insert

B. Tell the physician you will transfer her to the pharmacist to discuss crizanlizumab

C. Tell the physician that the medication is rivipansel, and it’s investigational

**5. The guidelines from the National Heart, Lung, and Blood Institute (NHLBI) recommend initiating hydroxyurea in all children with SCD beginning at what age, regardless of clinical severity?**

A. 6 months old

B. 9 months old

C. 2 years old

**6. Which of the following lists medications from least expensive to most expensive?**

A. Hydroxyurea, L-glutamine, crizanlizumab, voxelotor

B. Crizanlizumab, hydroxyurea, L-glutamine, voxelotor

C. Voxelotor, L-glutamine, hydroxyurea, crizanlizumab

**7. A patient who has SCD has been taking hydroxyurea for several years, but she admits she is often nonadherent. She says, “Any excuse to skip a dose is OK with me.” When you question her, she has no explanation for her nonadherence at all. Which of the following explanations it the BEST ONE for you to provide to her to encourage adherence?**

A. “Your dose is too high. The pharmacist will contact your prescriber to reduce it by 5 mg/kg.”

B. “When taken consistently, hydroxyurea reduces your chances of being hospitalized.”

C. “If you don’t take your hydroxyurea, your prescriber will not be able to assess for toxicity.”

**8. A patient presents a prescription to you for an opioid analgesic. This patient also has prescriptions for hydroxyurea and a different opioid analgesic. The pharmacist you are working with grumbles about this patient’s frequent emergency room visits and multiple opioid prescriptions. He says, “These sickle cell patients all become addicts. We need to turn this away.” What barrier is the patient facing, and what should you do about it?**

A. Stigma; educate your colleague about SCD’s symptoms, the intense pain associated with vaso-occlusive crisis, and its tendency to last 6 or 7 days

B. Racial bias; educate your colleague that SCD is not just a problem for people of African American heritage, it affects people of all races

C. Negative provider attitudes; discussing these issues with people who have deep-seated beliefs is pointless, direct the patient to another pharmacy in town

**9. A patient is referred to a specialty pharmacy for crizanlizumab after not tolerating hydroxyurea, but she is uninsured. Which of the following options could a specialty pharmacy liaison exercise for the patient?**

A. Contact the prescriber to recommend they switch her back to hydroxyurea

B. Ask the pharmacist if she can have extra doses that other patients didn’t use

C. Research medication assistance programs through grant funding or 340B

**10. A patient who has been on hydroxyurea for many years asks if any new treatments will be approved in the next few years. What is the BEST response?**

A. “Four are in clinical trials, and they should be approved and reach the market soon.”

B. “Nothing is better than hydroxyurea, and you should expect to remain on it for life.”

C. “Several are in development, but some have had poor trial results or funding challenges.”