

A 16 year-old-girl with hereditary spherocytosis is scheduled for a splenectomy. She was previously managed with folate therapy and occasional blood transfusions, but her anemia became refractory to medical management alone. Before the operation, she is told that she will have an enhanced risk of developing pneumococcal sepsis. She then asks, "How long will this risk last?" What is the best response to her question?

- ☐ A. 2 weeks
- ☐ B. Up to 6 months
- ☐ C. Up to 2 years
- ☐ D. Up to 10 years
- ☐ E. More than 10 years

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- ☐ A. 2 weeks [5%]
- ☐ B. Up to 6 months [6%]
- ☐ C. Up to 2 years [3%]
- ☐ D. Up to 10 years [4%]
- ☒ E. More than 10 years [82%]

[Proceed to Next Item](#)

Explanation:

User Id: [REDACTED]

Hereditary spherocytosis is an autosomal dominant disorder. It is characterized by a lack of spectrin in the red cell membrane, which causes the cells to become spheres, instead of being normal, flexible and durable biconcave discs. The poorly flexible spherical cells are thus unable to pass through the small fenestrations in the splenic red pulp, and hemolysis takes place when the red cells are trapped within the spleen.

The treatment for most patients involves supportive care with oral folic acid and blood transfusions during periods of extreme anemia. Splenectomy is considered if patients have moderate to severe spherocytosis, or are refractory to medical management. The benefits of splenectomy must be balanced against the immediate and long-term risks of the procedure. Life-threatening anemia and the need for regular transfusions may be abolished by splenectomy, although a mild degree of anemia usually persists. Immediate risks (e.g., hemorrhage, postoperative infection, injury to nearby organs) are infrequent. The most feared long-term complication is overwhelming sepsis with encapsulated

✓ ☐ E. More than 10 years [82%]

Proceed to Next Item

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Studies have shown that the risk for pneumococcal sepsis is present up to 30 years and probably longer after splenectomy. To decrease this risk, current recommendations call for the administration of anti-pneumococcal, *Haemophilus*, and meningococcal vaccines several weeks before the operation, and daily oral penicillin prophylaxis for three to five years following splenectomy or until adulthood (for pediatric patients). In view of reported deaths from sepsis up to 30 years or more after splenectomy, a case can be made for lifetime penicillin prophylaxis. Alternatively, antibiotics can be made available at home for immediate treatment of any significant fever.

Educational Objective:

Studies have shown that the risk for sepsis is present up to 30 years and probably longer after splenectomy. Current recommendations state that patients should receive anti-pneumococcal, *Haemophilus*, and meningococcal vaccines several weeks before the operation, and daily oral penicillin prophylaxis for three to five years following splenectomy.

*Extremely important question for the USMLE step-2

Time Spent: 4 seconds

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