Transient Disappearance of Sickle Hemoglobin After Transfusion

To the Editor:

An 18-year-old woman with sickle cell anemia was first observed by us on November 16, 1993. She had a stroke 15 years ago and was regularly transfused. Her last transfusion was received on October 26, 1993, 3 weeks before the first blood sample in our clinic was obtained. In the 10-month interval before her transfer, she received transfusions monthly to maintain her sickle hemoglobin (Hb S) level less than 30%. Measurements of the percent Hb S, determined at various times in relation to blood transfusion, showed levels between 4.5% and 35%.

Upon examination, she felt well and had only mild left-sided weakness. Hb S levels, reticulocyte counts, and hemoglobin determinations during a 1-year period, commencing with her first visit to our clinic, are shown in Fig. 1. Our first two blood samples, taken 1 week apart and examined by acid and alkaline electrophoresis and isoelectric focusing, showed only normal hemoglobins. Hb S was undetectable 4 weeks after her last transfusion. By 5 weeks, about

Fig 1. Variation of the (♦) percent Hb S, (■) hemoglobin concentration, and (□) reticulocyte count during 1 year of observation and seven transfusions. Arrows indicate the time of blood transfusions.
2.5% Hb S was present. The Hb S level then increased rapidly, reaching 50% within 5.5 weeks. Leukocyte and platelet counts increased from 8.3 and 284 × 10⁹/L when Hb S was absent and reticulocytes were less than 1% to 21.5 and 553 × 10⁹/L just before resumption of transfusions, respectively. B19 parvovirus IgG titer was greater than 10 RIA units in November 1993, January 1994, and March 1994, but IgM anti-B19 antibody was not detected, suggesting that acute parvovirus infection was not the cause of bone marrow suppression.

Because of regular transfusions, our patient’s blood contained a preponderance of normal erythrocytes when her marrow was suppressed. Their normal survival accounts for the slow decrease in hemoglobin concentration and the absence of severe anemia in the presence of erythroid aplasia. In untransfused sickle cell anemia, blood counts decrease when erythropoiesis fails. Very low reticulocyte counts during her first clinic visits suggested that no new sickle cells were accumulating. Rapid destruction of Hb S-containing cells remaining in the circulation accounted for the “normal” hemoglobin electrophoresis during her first visits. During the first year of observation in our clinic she received seven transfusions totaling 14 U of packed red blood cells. Once the Hb S concentration decreased to 22% immediately after transfusion and then decreased further, reaching a nadir of 11% in 10 days.

Some chronically transfused patients with sickle cell anemia have greater repression of endogenous erythropoiesis than others. Of 11 chronically transfused patients with sickle cell anemia receiving similar amounts of packed red blood cells, the Hb S level 3 to 4 weeks posttransfusion in 3 was 6.1% ± 0.6%, compared with 23.0% ± 2.1% in the remaining 8. The reasons for this variation were unclear. Whether Hb S ever totally disappeared from the blood was not reported. Our patient is among those individuals who appear to have more intense marrow suppression when chronically transfused. Yet, the Hb S levels after transfusion were usually predictable. This finding suggests that the disappearance of Hb S when she was first seen and occasional decreases of Hb S to lower than predicted levels were the result of an unknown exogenous event. Most likely this event was related to blood transfusion.

Martin H. Steinberg
VA Medical Center
University of Mississippi
Jackson, MS

REFERENCES

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MH Steinberg