



# Autologous blood transfusion and intraoperative cell salvage in a patient with homozygous sickle cell disease

JOHN S. FOX, BE, MD; L. AMARANATH, MD; GERALD A. HOELTGE, MD; JACK T. ANDRISH, MD

- **BACKGROUND** Autologous transfusion can eliminate the need for homologous transfusions. In addition, hypotensive anesthesia and devices that salvage red blood cells for return to the patient can reduce operative blood loss. However, blood from patients with sickle cell disease is difficult to store.
- **SUMMARY** A 16-year-old black girl with homozygous sickle cell disease needed surgery for progressive scoliosis. Her family's religious convictions precluded homologous transfusions. During surgery, 400 mL of autologous blood that had been successfully stored was transfused, as was 800 mL of blood salvaged using a cell-saving device, and 3800 mL of nonblood plasma expanders. Intravenous agents were used to maintain hypotension. However, following a rise in the patient's prothrombin and thromboplastin times, four units of homologous packed red cells were transfused with the permission of the patient's parents.
- **CONCLUSIONS** Patients with sickle cell disease can be given hypotensive anesthesia and autologous transfusions of blood donated before surgery and blood salvaged during surgery using a cell-saving device.

■ **INDEX TERMS:** BLOOD TRANSFUSION, AUTOLOGOUS; ANEMIA, SICKLE CELL; BLOOD PRESERVATION ■ *CLEVE CLIN J MED* 1994; 61:137-140

From the Departments of Orthopedics (J.S.F., J.T.A.), Anesthesiology (L.A.), and Blood Banking and Transfusion Medicine (G.A.H.), The Cleveland Clinic Foundation.

Address reprint requests to J.T.A., Department of Orthopedics, A51, The Cleveland Clinic Foundation, 9500 Euclid Avenue, Cleveland, OH 44195.

**P**ATIENTS WITH sickle cell disease who need elective surgery may undergo preoperative partial exchange transfusion, according to current recommendations. During surgery the anesthesiologist must take special care to maintain hydration, high oxygen tension, and normal body temperature.<sup>1-6</sup> To our knowledge, no one has reported using both a cell-saving device to return salvaged blood and hypotensive anesthesia during surgery in a patient with sickle cell disease.<sup>7</sup>

## CASE HISTORY

The patient was a 16-year-old black girl with progressive scoliosis and sickle cell disease. Her parents had deferred surgery because of religious concerns regarding blood transfusion. At presentation to this center, she weighed 37 kg. Her thoracic spine was curved 89° to the right and her lumbar spine was curved 100° to the left. Results of her pulmonary function studies were 50% of the predicted values. Her hemoglobin level was 9.6 g/dL, and her hematocrit measured

28.5%. Sickle cells were visible in the peripheral blood smear, and there was marked erythrocytic poikilocytosis. Results of hemoglobin erythrocytosis were consistent with homozygous hemoglobin S (hemoglobin S 95%, hemoglobin F 5%, hemoglobin A 0%). The serum bilirubin level was 3.6 mg/dL (normal range 0 to 1.4 mg/dL), and the serum lactate dehydrogenase (LDH) level was 765 IU/L (normal range 100 to 185 IU/L).

The family gave their consent for autologous transfusion and intraoperative return of salvaged blood. However, they requested that nonautologous blood be used only as a last resort. The patient donated 200 mL of blood 2 weeks before surgery and another 200 mL 1 week before surgery. The blood was collected in bags containing citrate-phosphate-dextrose-adenine (CPDA-1). Afterward, 100% oxygen, sterilized by filtration, was slowly bubbled through each of these whole-blood units for 30 minutes until the color of the unit approximated that of arterial blood. The first unit was reoxygenated daily for the first 3 days of storage; the second unit was oxygenated only once. Both units remained bright red during the storage interval. Two days before surgery, samples from the units were submitted for culture. Sterility and lack of visible hemolysis were assured before the units were released for transfusion. No sickled erythrocytes were detected microscopically in samples taken from each unit.

The patient underwent posterior spinal fusion with insertion of segmental instrumentation from T4 to L4. General anesthesia was administered, consisting of 70% nitrous oxide and 30% oxygen given via an endotracheal tube, and morphine, sufentanil, and d-tubocurarine given intravenously. Labetalol and nitroglycerine were given intravenously as well to maintain mean systolic arterial pressure between 60 and 70 mm Hg as measured by an indwelling cannula in the radial artery. The esophageal temperature was maintained at 37.2°C with warmed intravenous fluids and thermal blankets. Monitoring devices were used to continuously measure end-expired gas tensions, central venous pressure, oxygen saturation of the blood, and somatosensory evoked spinal cord potentials; arterial blood gas levels were measured intermittently. Arterial pH was maintained between 7.43 and 7.47 by deliberately inducing hyperventilation with carbon dioxide at 30 mm Hg and by intermittently giving sodium bicarbonate intravenously. As a prophylactic "antisludge" medication, 500 mL of low-molecular weight dextran was given.

The two autologous units were transfused at the beginning of the procedure. In accordance with the family's wishes, nonautologous transfusion was deferred until considered essential. Blood was salvaged using a cell-saving device (Cell Saver 4, Haemonetics Corp, Braintree, Mass), and when the hematocrit reached 10%, 800 mL of this blood was returned to the patient. The partial pressure of oxygen in this blood was 147 mm Hg. After the salvaged blood was returned, the prothrombin time was found to be 21.3 seconds (normal range 10 to 13 seconds) and the activated partial thromboplastin time was 53.4 seconds (normal range 21 to 31 seconds). At this point we obtained the consent of the patient's parents to give her a transfusion of homologous packed red cells, and we gave her four units to maintain the hematocrit at approximately 18%.

During the 4-hour surgical procedure, the patient lost an estimated 2500 mL of blood. Plasma-expanders that do not contain blood were given during surgery, including 1000 mL of hetastarch and 2300 mL of crystalloids. Intraoperative urine output was 500 mL. After the procedure, the patient was monitored overnight in the surgical intensive care unit. Morphine was given via epidural catheter for 2 days to manage pain satisfactorily. On the third postoperative day the serum LDH measured 350 IU/L and the total bilirubin level was 1.0 mg/dL. After an unremarkable postoperative course, the patient was discharged on the eighth postoperative day. Her hemoglobin level was 6.5 g/dL, and her hematocrit had increased to 20.4%.

---

#### DISCUSSION

---

The current recommendations for anesthesia under elective conditions for patients with sickle cell disease begin with optimizing the patient's medical status. This includes long-term administration of folic acid and prompt treatment of any infection. Some authors recommend partial exchange transfusions to increase the concentration of hemoglobin A to 40% and the hematocrit level to 35%.<sup>5,8</sup> To prevent hypoxia in the perioperative period, premedications that depress respiration are avoided.

Many authors believe that, provided the precipitating factors for sickling are avoided (ie, hypoxia, metabolic acidosis, stasis, hypothermia), any anesthetic technique is acceptable for patients with sickle cell disease.<sup>2,3,6</sup> To prevent sickling, one should induce controlled pulmonary hyperventila-

tion to maintain arterial oxygen tensions at normal to slightly higher-than-normal levels, maintain cardiac output to avoid inadequate regional ventilation-perfusion ratios, give alkaline buffers, position the patient carefully, and maintain the patient's temperature. Metabolic acidosis, stasis, and increased viscosity are compounded if the body temperature is allowed to fall. Meticulous attention should be continued in the postoperative period to avoid over-sedation and hypoventilation.

Although some authorities advocate erythropheresis<sup>9</sup> and partial exchange transfusions<sup>5,7</sup> to increase the minimum concentration of hemoglobin A to 40% and the hematocrit level to 35%, no controlled studies have established the smallest ratio of hemoglobin A to S (as measured by hemoglobin erythropheresis) required to maintain adequate blood flow. Furthermore, the possible decrease in perioperative morbidity after exchange transfusion has not been compared with the risks of exchange. Hence, exchange transfusion is usually reserved for crisis situations. Overly zealous transfusion can lead to an increase in the viscosity of the blood and can predispose to stasis.<sup>4</sup>

Storage of blood containing a high concentration of hemoglobin S is complicated by the risk of sickling and hemolysis *in vitro* under conditions of deoxygenation. It has been suggested that this risk is mitigated by the relatively young mean cell age of erythrocytes from patients with sickle cell disease.<sup>10</sup> Such cells maintain higher levels of 2,3 diphosphoglycerate than do cells containing hemoglobin A. Nevertheless, we thought it prudent to keep our patient's cells well oxygenated during liquid storage to minimize the risk.

Chaplin and colleagues<sup>11</sup> have described autologous transfusion in a patient with sickle cell disease and multiple alloantibodies. Eleven 600-mL donations at approximately 4-week intervals caused no ill effects and no apparent hematologic abnormalities. *In vitro* recovery of deglycerolized red blood cells ranged from 52% to 69% (average 66%) and was unrelated to the length of storage. This yield was 48% of that expected from an equivalent donation by a normal donor. Using chromium labeling, the authors demonstrated that there was no difference in survival between thawed deglycerolized frozen red cells and fresh red cells. Their patient showed no untoward symptoms, change in vital signs, or evidence of hemolysis after the transfusion of two

autologous units that had a recovered red-cell volume of 155 mL. One unit had been stored for 27 days, the other, 126 days. Another five autologous units were transfused when the patient was in a critical condition. These units had been stored an average of 30 days and had a total recovered red-cell volume of 418 mL. Again, there was no evidence of untoward signs or symptoms or of intravascular hemolysis.

Because our patient was scheduled for surgery in less than 5 weeks' time, and because the recovery of red cells following deglycerolization of frozen sickle cell disease units is poor, we did not store her blood as frozen units.

---

#### CONCLUSIONS

---

We tried to avoid or minimize the need for homologous transfusion by using hypotensive anesthesia and by giving autologous transfusions with blood donated before surgery or salvaged during surgery with a cell-saving device in a patient with sickle cell disease who underwent major spinal corrective surgery for scoliosis. There was no evidence of hemolysis or sickling in the autologous units donated before surgery, and there were no anesthetic or clinical complications relating to the return of this 400 mL of blood. An additional 800 mL of salvaged blood was returned to the patient from the cell-saving device.

During the course of the procedure, we observed a prolongation of the prothrombin time and of the partial thromboplastin clotting time. This was probably secondary to the early and intentional replacement of blood loss with crystalloids, the use of low-molecular-weight dextran, and the effect of heparin in the salvaged blood.

We had hoped that using hypotensive anesthesia would decrease the amount of blood lost. It didn't, but intraoperative use of nitroglycerin and labetalol as a part of the hypotensive anesthetic technique conceivably helped prevent sickling of red cells *in vivo* by improving microcirculatory flow.<sup>12</sup> There were no complications from the use of hypotensive general anesthesia.

In summary, a patient with sickle cell disease undergoing major elective surgery can successfully be given hypotensive anesthesia and autologous transfusions of blood donated before surgery and blood salvaged during surgery using a cell-saving device.

REFERENCES

1. **Aldrete JA, Guerra F.** The hematologic diseases. In: Katz J, Benumof J, Kadis LB, editors. *Anesthesia and uncommon diseases*. Philadelphia, Pa: W.B. Saunders, 1981:313–383.
2. **Griffin DR.** Sickle cell disease as it relates to anesthesia: Report of two cases. *Anesth Analg* 1966; **45**:826–828.
3. **Holzmann L, Finn H, Lichtman HC Harmel, MH.** Anesthesia in patients with sickle cell disease: a review of 112 patients. *Anesth Analg* 1969; **48**:566–572.
4. **McNiece WL.** Anemia. In: Stoelting RK, Dierdorf SF, editors. *Anesthesia and coexisting disease*. New York: Churchill Livingstone, 1983:523–529.
5. **Roizen MF.** Anesthetic implications of concurrent diseases. In: *Anesthesia*. 2nd ed. New York: Churchill Livingstone, 1986:255–357.
6. **Searle JF.** Anesthesia in sickle cell states. A review. *Anaesthesia* 1973; **28**:48–58.
7. **Cook A, Hanowell LH.** Intraoperative autotransfusion for a patient with homozygous sickle cell disease. *Anesthesiology* 1990; **73**:177–179.
8. **Lanzkowsky P, Shende A, Karayalcin G, et al.** Partial exchange transfusion in sickle cell anemia: use in children with serious complications. *Am J Dis Child* 1978; **132**:1206–1208.
9. **Wilhelm JL, Zakov ZN, Hoeltge GA.** Erythropheresis in treating retinal detachments secondary to sickle cell retinopathy. *Am J Ophthalmol* 1981; **92**(4):582–583.
10. **Castro O.** Viability and function of stored sickle erythrocytes. *Transfusion* 1980; **20**:695–703.
11. **Chaplin H, Mischeaux JR, Inkster MD, Sherman LA.** Frozen storage of 11 units of sickle red cells for autologous transfusion of a single patient. *Transfusion* 1986; **26**(4):341–345.
12. **Rodgers GP, Roy MS, Noguchi CT, Schechter AN.** Is there a role for selective vasodilation in the management of sickle cell disease? *Blood* 1988; **71**:597–602.

