

A 70-YEAR-OLD MAN WITH β -THALASSEMIA MAJOR

To the Editor:

We read with interest Dr. Thomas' case report, " β -Thalassemia Minor and Newly Diagnosed Polycythemia Rubra Vera in a 71-Year-Old Woman" in the April 2001 issue of *Hospital Physician*.¹ We were fortunate to care for one of the longest living patients with β -thalassemia major. This 70-year-old man was diagnosed with β -thalassemia major during infancy. He was of Italian descent, and his sister and brother had sickle-cell thalassemia and thalassemia trait, respectively. His surgical history included sternal surgery in 1936 (for marrow examination), splenectomy, bilateral cataract surgery, pleurodesis for repeated pleural effusions, and repeated procedures to correct bone fractures of the extremities. He was transfusion-dependent throughout his life and had bilateral macular degeneration, partial deafness, hypogonadism, dilated cardiomyopathy, and chronic renal failure. The patient had facies suggestive of Down syndrome, an enlarged heart with a grade 3/6 systolic murmur, and significant hepatomegaly. Electrocardiography during his last hospitalization showed atrial fibrillation and low-voltage recording, and radiography showed a mass in the posterior mediastinum, which was presumed to indicate extramedullary hematopoiesis. Laboratory evaluation at this time revealed the following levels: hemoglobin, 6.1 mg/dL;

serum iron, 78 μ g/dL; total iron binding capacity, 137 μ g/dL; and plasma ferritin, 3386 ng/mL. A peripheral blood smear showed nucleated erythrocytes, target cells, poikilocytosis, hypochromia, microcytes, acanthocytes, burr cells, and basophilic stippling.

The patient had multiple hospital admissions for exacerbated congestive heart failure and for blood transfusions. He had refused iron-chelating therapy for the past 15 years and had confined himself to his home to "avoid infections." Unfortunately, during his last hospitalization, he fell and sustained a femoral fracture. He soon developed a fever and sepsis and later developed multiorgan failure. He died at age 70 years.

It is interesting to note that although most patients with thalassemia major die in the second or third decade of life, our patient survived to 70 years without a bone marrow transplant.

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Reference

1. Thomas JP. β -Thalassemia minor and newly diagnosed polycythemia rubra vera in a 71-year-old woman. *Hosp Physician* 2001;37(4):78-83.

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