

# CASE STUDY 5:

## School-age child

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### GENDER

F

### AGE

7

### SETTING

- Clinic

### ETHNICITY

- Mediterranean

### PREEXISTING CONDITIONS

- Beta-thalassemia

### COEXISTING CONDITIONS

- Hemosiderosis

### PHARMACOLOGIC

- Deferoxamine mesylate (Desferal)

### PSYCHOSOCIAL

- Effects of frequent and continued hospitalizations

### THE BLOOD

**Overview:** This case requires knowledge of beta-thalassemia, treatment and complications, growth and development, as well as an understanding of the client's background, personal situation (living with chronic condition), and parent-child relationship.

## Client Profile

**Adriana** is a 7-year-old who lives with her parents in a suburban community. Her parents brought Adriana to the United States from their homeland in Greece when she was 1 year old. At the age of 3, Adriana was in the 10th percentile for height and weight, pale, and her hemoglobin was 5.8 g/dL. Following further diagnostic studies, she was diagnosed with beta-thalassemia major. Over the course of the next 4 years, Adriana was hospitalized every 1–2 months so she could be transfused with packed red blood cells.

## Case Study

During a routine follow-up visit at the hematology clinic, Adriana's laboratory results were as follows:

Hemoglobin: 10 mg/dL

Total serum iron: 150  $\mu\text{g/L}$

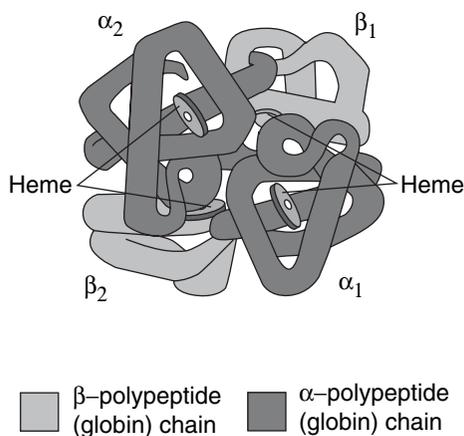
The hematologist discusses the planned treatment with Adriana and her parents.

## Questions

1. Discuss the significance of Adriana's family's geographical background to her diagnosis.
2. What is beta-thalassemia, its incidence, and etiology?
3. Discuss the clinical manifestations of beta-thalassemia.
4. Discuss the significance of Adriana's laboratory values.
5. Discuss the complications associated with the chronic blood transfusions associated with Adriana's condition.
6. Discuss the standards of care once Adriana reaches the parameters prescribed for annual packed red blood cell transfusions and how they affect the child's need for future blood transfusions.
7. What are the priorities of care for Adriana?
8. Adriana is diagnosed with hemosiderosis. What is hemosiderosis and how is it treated?
9. Adriana is prescribed an initial dose of deferoxamine mesylate 1 g IM and 400 mg SC each day for 5 days. Adriana weighs 15 kg (33 lb). Would the nurse question this prescription?
10. Discuss the options available for Adriana to prevent lifelong blood transfusions.

## Questions and Suggested Answers

1. **Discuss the significance of Adriana's family's geographical background to her diagnosis.** Beta-thalassemia is an autosomal recessive genetic disorder affecting hemoglobin chains and has its highest incidence in children



**Figure 5-1** *The structure of normal hemoglobin.*

of Mediterranean descent. The primary populations are Italian, Greek, and Syrian. With the increased influx of immigrants into the United States, the incidence of thalassemia is increasing.

**2. What is beta-thalassemia, its incidence, and etiology?** Normally the hemoglobin is made up of globin chains (Hb- $\alpha$  and Hb- $\beta$ ) (see Fig. 5-1). In beta-thalassemia the beta chains of the molecule of the hemoglobin are missing. This results in deficient hemoglobin and the development of fragile, microcytic, hypochromic erythrocytes. These erythrocytes are unable to carry adequate oxygen to the cells and tissue, leading to inadequate tissue perfusion and delayed growth and development. According to the Cooley's Anemia Foundation, there are three types of beta-thalassemia and their effects on the body range from mild to severe. Thalassemia minor, also referred to as thalassemia trait, lacks the beta protein but not sufficiently to interfere with the normal functioning of the body's hemoglobin. A person with thalassemia minor simply has the genetic trait. Thalassemia intermedia is a condition where the lack of beta protein in the hemoglobin is great enough to cause a moderately severe anemia and significant health problems, including bone deformities and enlargement of the spleen. The severity of this condition is measured by the number of blood transfusions the child requires. Generally speaking, clients with thalassemia intermedia need blood transfusions to improve their quality of life, but not in order to survive. Finally, beta-thalassemia major is the most severe form of this disease, in which the complete lack of beta protein in the hemoglobin causes a life-threatening anemia that requires regular blood transfusions and extensive ongoing medical care. Enlargement of both the spleen and liver, and jaundice resulting from

increased hemolysis of the abnormal red blood cells, is common. The stress placed on the bone marrow to increase production of erythrocytes for tissue perfusion causes hyperplasia of the bone marrow that leads to thickening of the bones throughout the body and delayed development.

Thalassemias are most dominant in certain Eastern European populations. Individuals of Mediterranean descent are more likely to have deficient beta-globulin thalassemia. This condition also is referred to as Cooley's anemia or Mediterranean anemia. Both Africans and African-Americans also have an increased incidence of thalassemia.

The incidence of thalassemia varies worldwide; however, more than 1,500 people in the United States are affected and 2 million carry the genetic traits for thalassemia. Thalassemia is an autosomal recessive genetic disorder, and those with the disease have a usual life span of 20–30 years.

**3. Discuss the clinical manifestations of beta-thalassemia.** The clinical manifestations are the direct result of the pathophysiologic process of thalassemia. Pallor is present because hemoglobin is a major factor in the normal skin color. Children with thalassemia are usually small for their age, reaching only the 5th percentile for growth. They experience recurrent severe anemia (hemoglobin < 6 g/dL) and hepatosplenomegaly. The stress on the bone marrow causes the bones to thicken and become less flexible (see Fig. 5-2). This leads to pathological fractures and pain.

**4. Discuss the significance of Adriana's laboratory values.** Adriana is experiencing anemia as indicated by her hemoglobin level of 10 g/dL; however, for children with beta-thalassemia, the goal of treatment is to maintain hemoglobin levels between 9 and 10 g/dL. The more disturbing value is her total serum iron of 150 µg/L. The normal level of serum iron for children is 50–120 µg/L. Adriana's value indicates iron toxicity.

**5. Discuss the complications associated with the chronic blood transfusions in Adriana's condition.** The primary complication of chronic blood transfusions is elevated iron levels that can lead to tissue and organ failure.

**6. Discuss the standards of care once Adriana reaches the parameters prescribed for annual packed red blood cell transfusions and how they affect the child's need for future blood transfusions.** The standard of care once Adriana reaches the parameters for annual red blood cell transfusions is the removal of the spleen. The spleen enlarges, resulting in increased hemolysis of red blood cells. This process increases the risk and progression of iron toxicity. A splenectomy results in a lifelong increased risk for infection, requiring the use of prophylactic antibiotics.

**7. What are the priorities of care for Adriana?**

- a. Ineffective tissue perfusion related to deficiency of hemoglobin secondary to disease process



**Figure 5-2** Face of a child with beta-thalassemia. Note the unusual features, including prominent and protruding forehead and flattened nose, caused by expansion of the facial bones to accommodate hyperplastic marrow.

- b. Risk for injury, tissue and organ damage related to elevated serum iron levels
- c. Deficient knowledge related to Adriana's condition, treatment, and home care

**8. Adriana is diagnosed with hemosiderosis. What is hemosiderosis and how is it treated?** Hemosiderosis is the term for elevated serum iron levels. It occurs as a result of chronic blood transfusions and is treated with the use of a chelating agent. Deferoxamine mesylate is the agent of choice to reduce serum iron levels.

**9. Adriana is prescribed an initial dose of deferoxamine mesylate 1 g IM and 400 milligrams SC each day for 5 days. Adriana weighs 33 lb. Would the nurse question this prescription?** As noted above, deferoxamine mesylate is the chelating agent of choice to treat iron toxicity caused by chronic blood transfusions. The normal loading dose of this agent is 1 g either intramuscularly or intravenously. It is then prescribed based on 20–40 mg/kg per day administered by subcutaneous mini-infusion pump over a period of 8–24 hours. The usual duration of treatment is 5–7 days. Adriana can receive

300–600 mg of this agent per day, so her dose is safe and although the nurse would evaluate the prescription based on the 7 rights of medication administration, she would not question this prescription.

**10. Discuss the options available for Adriana to prevent the need for life-long blood transfusions.** Either bone marrow transplantation (BMT) or cord blood transplantation is indicated to prevent the lifelong need for blood transfusions. BMTs are complicated by the need for the donor to be genetically matched to the recipient. Cord blood transplants are similar to BMTs except the stem cells are harvested from the placenta or umbilical cord of a suitable donor. The advantage of cord blood transplants is that the donor does not require as close a genetic match as for BMT and the probability of rejection is much lower.

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