Transfusion Management of Beta (β) Thalassemia: Developing a Transfusion Plan

A Thalassemia Resource for Healthcare Providers

Key Facts About β Thalassemia

- β thalassemia is caused by β-globin gene variants that reduce the production of adult hemoglobin (HbA), which may cause anemia.
- Chronic anemia can have serious consequences for people with β thalassemia.
  - » Children: reduced activity, growth problems and delayed puberty, hepatosplenomegaly, osteopenia, cognitive impairment
  - » Adults: fatigue, decline in capacity to work or perform usual activities, cognitive impairment, osteopenia, fractures, hypersplenism, reduced quality of life

- The primary management of severe anemia in β-thalassemia is regular red cell transfusions.
- The Thalassemia Western Consortium recommends initiating regular transfusions if either of the following conditions are met:
  - » Hemoglobin <7g/dL on 2 occasions at least 2 weeks apart
    - β halassemia major: <7g/dL on 2 occasions, with or without severe symptoms
    - HbE β thalassemia: <7 g/dL on 2 occasions AND one or more severe symptoms
  - » Hemoglobin ≥7g/dL, with one or more severe symptoms

Once the decision to initiate regular transfusions is made, healthcare providers can develop a transfusion plan to determine the rate, frequency, and volume of transfusions that are needed to maintain a target pretransfusion hemoglobin level (the hemoglobin level at which the next transfusion should be administered). This transfusion plan may differ from person to person and may need to be adjusted over time.

What are the Components of a Transfusion Plan for People with β thalassemia?

Target Pretransfusion Hemoglobin Level

In general, the target pretransfusion hemoglobin level recommended by the Thalassemia Western Consortium for people with β thalassemia is

- 9.5–10.5 g/dL for people with β thalassemia major
- 9.0–10.0 g/dL for people with HbE β thalassemia

Maintaining these target pretransfusion hemoglobin levels has been previously determined to ensure normal growth, prevent fatigue, and suppress splenomegaly and marrow hyperactivity in most people with β thalassemia.

Rate

In general, blood is transfused at a rate of

- 5 ml/kg/hour for children
- 200–300 ml/hour, based on tolerance, for adults

The shortest time to complete a transfusion safely may differ between people.

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Frequency

In general, people with β thalassemia need transfusions

- Every 3 weeks for most older children and adults with β thalassemia major
- Every 4 weeks for younger children with β thalassemia major and most people with HbE β thalassemia

The frequency of transfusions may be changed to maintain the target pretransfusion hemoglobin level. A shorter interval between transfusions reduces hemoglobin level variability but requires more frequent visits to the infusion center. A person with β thalassemia who is on a 4-week transfusion schedule requires a larger volume of blood at each transfusion than a person who is on a 3-week transfusion schedule to attain the same target pretransfusion hemoglobin level.

Volume

In general, the volume of blood transfused for people with β thalassemia major is

- 4 ml/kg per gram increase in hemoglobin desired, up to 20 ml/kg at a single visit for children
- 2, 3, or 4 units per transfusion for adults. Common regimen: 3 units if the pretransfusion hemoglobin level is <10 g/dl; 2 units if the pretransfusion hemoglobin level is ≥10 g/dl

The volume of blood transfused during a visit may be changed to maintain the target pretransfusion hemoglobin level.

Are There Other Transfusion Considerations for People with β Thalassemia?

In general, it is important to consider the following when developing a transfusion plan for people with β thalassemia:

- People who have an intact spleen may need more blood during their transfusions than those who have had a splenectomy.
- Adults weighing more than 60 kg may need 4 units of blood at some visits.
- Higher target pretransfusion hemoglobin levels or more frequent transfusions are needed for people with
  - » Congestive heart failure
  - » Pulmonary hypertension
  - » Symptomatic extramedullary hematopoietic masses
  - » Occurrence of severe fatigue or bone pain in pretransfusion period
- People with congestive heart failure may also need a reduced transfusion volume and rate, depending on their cardiac function.

More information about target pre-transfusion hemoglobin levels can be found in the Thalassemia Management Checklist: Guidelines for Managing Transfusion Therapy for Thalassemia.

Reference: Lal A, Wong T, Keel S, Pagano M, Chung J, Kamdar A, et al. The transfusion management of beta thalassemia in the United States. Transfusion. 2021;1–13. https://onlinelibrary.wiley.com/doi/10.1111/trf.16640

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