Transfusion Management of Beta (β) Thalassemia
What to Transfuse: Specifications for Red Blood Cell Products

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 blood cell transfusions.

Healthcare providers can work with their blood bank partners to develop thalassemia-specific red blood cell transfusion protocols. In selecting red blood cell (RBC) products for patients receiving regular transfusions, it's important for the clinical team to carefully consider which products emphasize efficacy (hemoglobin response) while also minimizing risk of hemolytic and non-hemolytic transfusion complications.

What Type of RBC Product is Needed by People with β Thalassemia?

The optimization of RBC products is vital when transfusing people with β thalassemia. The Thalassemia Western Consortium recommends

- Leukoreduction of RBC units via filtration during the pre-storage period to prevent febrile non-hemolytic transfusion reactions
- Storage of RBC units in additive solutions (lower hematocrit and higher volume) or citrate phosphate dextrose-adenine (higher hematocrit and lower volume)
- Using non-apheresis-derived RBC units for transfusions in adults due to smaller hemoglobin increments with apheresis-derived units
- Washed RBC units for individuals who experience severe, recurrent allergic reactions
- RBC units that are less than 2 weeks old, when possible, to reduce the impact of the storage lesion on red cell survival
- Using non-irradiated RBC units, as routine use of irradiated RBC units may be detrimental

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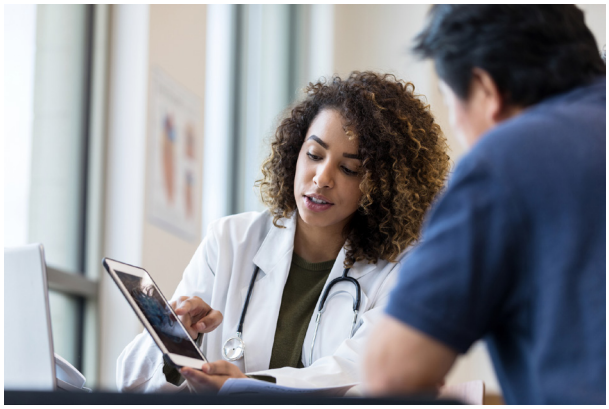
What Type of RBC Antigen Matching is Needed by People with β thalassemia?

Prevention of red cell alloimmunization is an important goal of transfusion management in thalassemia. Ideally, an individual's RBC genotyping is performed once the decision to initiate regular transfusions is made. The results of this typing determine their risk for developing alloantibodies and/or autoantibodies and determine the extent to which antigen-matched blood is needed. The Thalassemia Western Consortium recommends:

- Individuals without a history of alloimmunization are given RBC units matched to Cc, Ee, and K blood group antigens
- Individuals with one or more alloantibodies are given RBC units matched to Cc, Ee, K, Jka/b, Fya/b, and Ss blood group antigens



It is important for providers to maintain an accurate antibody history, as an antibody screen can become negative over time, and individuals with a history of developing antibodies need antigen-negative blood to prevent hemolytic transfusion reactions.

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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The original evidence-based report and recommendations were developed in 2021 by a multi-disciplinary committee, the Thalassemia Western Consortium, consisting of members from 10 hematology centers that was partially funded under a prior cooperative agreement CDC-RFA-DD14-1406. While the Consortium was supported with federal funding, their work on clinical recommendations for transfusion practices was independent of CDC, and the contents of the resulting published report are solely the responsibility of the authors and do not necessarily represent the official views of, nor an endorsement by, CDC or the Department of Health and Human Services.