

Transfusion Dependent Thalassemia (TDT) Transfusion Therapy Guidelines

Goals of Regular Transfusion Therapy

1. Provide adequate hemoglobin for growth and development (children and adolescents), and allow normal activity without symptoms of easy fatigability and malaise in all patients.
2. Adequately suppress ineffective erythropoiesis, so as to ameliorate its consequences – medullary expansion and bone abnormalities, as well as extramedullary hematopoiesis (liver and spleen enlargement and nodules in the spine).

Transfusion Strategy

- When placing a patient on a regular transfusion regimen, a sample is sent to the blood bank for limited extended phenotyping. Antigens tested include C, D, E and Kell.
- The pre-transfusion hemoglobin target is ~10 g/dl.
- In infants who have just been diagnosed, transfuse 15 ml/kg packed red blood cells (PRBCs) at each visit. Initially transfusions may need to be performed at two – three weeks to reach a pre-transfusion hemoglobin of 10 g/dl. Once this has been achieved, transfusions may be spaced out to once per month (every four weeks).
- Toddlers and young children may continue monthly transfusions of 15 ml/kg at each visit until they are able to receive a full unit of PRBCs.
- If pre-transfusion hemoglobin is below the target of ~10 g/dl, transfusion may be increased to 17-18 ml/kg PRBCs, or the interval to the next transfusion may be reduced.
- When the volume to be transfused (15 ml/kg) exceeds one unit of PRBCs (typically, one unit has 250-300 mls), the interval between transfusions is reduced from four to three, and then to two, weeks. Split units are not recommended.
- When a patient reaches a weight of ~35 kg, he/she may receive two units at each transfusion visit, and the interval between transfusions may return to four weeks.
- As a patient's weight increases, the interval may need to be reduced again to three, and then two, weeks to maintain the target pre-transfusion hemoglobin level. Most adults receive two units every two – three weeks.
- At the New York Comprehensive Thalassemia Center, we do not generally transfuse more than two units at each visit.

Practical Considerations

- Transfusions are administered using an infusion pump. No saline is attached through a Y connection to keep the system closed and prevent contamination.
- The rate of administration is 3-5 mL/kg/hour in children. Each unit is usually administered over about 90 minutes in adults.
- A 22-gauge IV is preferred when the rate is to exceed 200 ml/hour, but a 24-gauge may be used in smaller children who have slower infusion rates.
- Premedication with acetaminophen (Tylenol®) and diphenhydramine (Benadryl®) is necessary if a patient has had allergic reactions in the past. If diphenhydramine is not tolerated, hydroxyzine (Vistaril®) may be used. If subjects have allergic reactions despite this, hydrocortisone may be added as a premedication.
- Washed PRBCs are only necessary for patients who have had multiple severe allergic reactions.
- For individuals who have been alloimmunized and have antibodies, the time required for all testing and cross-matching is longer, and patients may prefer to have their blood sample sent to the blood bank in advance of the day of transfusion.
- Young children with difficult venous access may require placement of a central line.