

Effective Iron Chelation Practice for Patients With β -Thalassemia Major

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Chronic blood transfusion is the only treatment for severe anemia in patients with β -thalassemia major. However, red blood cell transfusions lead to iron overload and subsequent organ damage because of the toxic effects of iron. The heart is particularly vulnerable to iron toxicity, and heart failure is the leading cause of death among these patients. Iron chelation therapy prevents or reverses iron loading, thereby reducing the risk of complications from excess iron. Serum ferritin and liver iron concentration often are used to gauge the risk of organ iron overload, but these measurements may not correlate well with cardiac iron load. Magnetic resonance imaging (MRI) is a noninvasive diagnostic tool that can provide a more direct measure of iron concentration in both the heart and liver. Cardiac iron determined by MRI is expressed as a function of $T2^*$, in which higher values represent lower concentrations. Changes in $T2^*$ are used to assess the effectiveness of iron chelation and to adjust therapy. Early treatment and compliance are keys to successful therapy. Nursing strategies to optimize chelation therapy include identifying patients who are at risk for developing organ damage, developing chelation plans, promoting compliance, and educating patients. The efficacy and safety of iron chelators, as well as nursing best practices, are reviewed.

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Thalassemia is an inherited disorder characterized by the inability or reduced ability to produce adult hemoglobin (Hb). The various thalassemias are named according to the subunit of the Hb molecule, or globin chain, affected by a genetic defect. Thus, α -thalassemia is named for altered α -globin production and β -thalassemia for altered β -globin production (Cunningham, 2008).

β -thalassemia major is caused by the inheritance of two β -thalassemia alleles, resulting in reduced synthesis of Hb (Hoffman et al., 2008). The disorder has a prevalence of about 1,000 cases in the United States (Centers for Disease Control and Prevention, n.d.). The symptoms of β -thalassemia major appear at age 4–6 months of life, coinciding with the replacement of fetal Hb by adult Hb, which requires β -globin chains (Hoffman et al., 2008). The disease causes clinically severe anemia, as opposed to β -thalassemia intermedia or thalassemia minor, which are less severe diseases.

β -thalassemia major requires lifelong blood transfusions and likely is fatal in early childhood if untreated; however, treated patients who also remain compliant with chelation therapy can approach normal life expectancy (Cunningham, 2008). Thalassemia is detected in newborn screening, and the thalassemia clinical team is able to determine exactly when to begin chronic red blood cell transfusions. The requirement for chronic transfusion places patients at risk for organ damage from iron overload, a predictable and preventable side effect of transfusion. Each unit of blood contains 200–250 mg of iron, yet the human body has no mechanism for excreting excess iron (Andrews, 1999). Therefore, the heavy iron influx from transfusion leads to rapid iron loading of vital tissues. The liver is one of the major storage sites for excess iron (Kohgo, Ikuta, Ohtake, Torimoto, & Kato, 2008). However, in the presence of a large excess iron pool, the pituitary gland, pancreas, and heart also store iron (Kohgo et al., 2008). With iron accumulation, the