

Title: Beta-Thalassemia *GeneReview* – Hemoglobin Analysis

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Note: The following information is provided by the author and has not been reviewed by *GeneReviews* staff.

In vitro synthesis of radioactive labeled globin chains in affected individuals reveals the following

- β^0 -thalassemia: a complete absence of globin beta chains and a marked excess of globin alpha chains compared with globin gamma chains. The α/γ ratio is greater than 2.0.
- β^+ -thalassemia: a variable degree of reduction of globin beta chains resulting in severe (thalassemia major) to mild (thalassemia intermedia) clinical phenotypes. The imbalance of the α/β and γ ratio is similar to that in β^0 -thalassemia major.