Abstracts
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IN VITRO SYNTHESIS OF HEMOGLOBIN IN BFU-e DERIVED COLONIES OF PERSONS WITH AN α OR A β-THALASSEMIA. A. E. Felice, C. A. Altay, A. L. Reese, B. Webber, and T.H.J. Huisman, Department of Cell and Molecular Biology, Medical College of Georgia, Augusta, GA 30912

The synthesis of α and non-α chains of human Hb was studied in 14-day peripheral blood BFU-e derived colonies harvested 24 hrs after the addition of ^{35}S-methionine (colonies) and in blood reticulocytes (retics) incubated for 2 hrs with ^{14}C leucine. The subjects were 7 β-Thal traits, 2 HPFH traits and 1 homozygote, 3 Hb G-Philadelphia traits of whom one was also homozygous for Hb S, 3 Hb G-Georgia traits, 1 Hb Leslie trait, and 2 Hb S traits. The proportions of the nascent α (Hb G) and β (Hb X) variants are markers for the presence of a mild or moderate α chain deficiency due to an α-Thal-2 heterozygosity or a homozygosity, respectively. The Σα/non-α ratios of β-Thal traits differed between retics (av. 1.89) and colonies (av. 0.88). Two HPFH traits gave values of 0.95 and 1.69 in retics and 0.87 and 0.95 in colonies. The α/γ ratios for the HPFH homozygote were 1.92 (retics) and 1.25 (colonies).

The Σα/non-α values in two Hb G-Georgia traits ($\alpha\alpha^G/\alpha\alpha$) were 0.90 and 1.01 (retics), and 0.75 and 0.86 (colonies) while the % Hb G was 39 (retics) and 13 (colonies). The father with Hb G-Georgia in association with an α-Thal-2 trait ($\alpha^0\alpha/\alpha\alpha^G$) had ratios of 0.62 (retics) and 0.88 (colonies), while the % Hb G was 62 (retics) and 13 (colonies). Another α-Thal-2 trait, who had an associated Hb G-Phil. trait ($\alpha^0\alpha^G/\alpha\alpha$) gave Σα/Σ non-α values of 0.72 (retics) and 0.93 (colonies) while the % Hb G was 35 (retics) and 36 (colonies). One α-Thal-2 homozygote with an associated Hb G Phil. trait ($\alpha^0\alpha^G/\alpha^0\alpha$; β^A/β^A) gave: Σα/non-α = 0.63 with 45% Hb G (retics) and Σα/non-α = 0.90 with 52% Hb G (colonies). Another α-Thal-2 homozygote who had sickle cell anemia ($\alpha^0\alpha^G/\alpha^0\alpha$; β^S/β^S) gave Σα/non-α = 1.04 with 56% Hb G (retics) and Σα/non-α = 1.14 with 55% Hb G (colonies). One Hb Leslie trait ($\alpha\alpha/\alpha\alpha$; β^A/β^L) had a Σα/non-α value of 1.69 and 71.5% Hb X (retics) and a Σα/non-α value of 0.76 with 33% Hb X (colonies). The two Hb S traits had an associated α-Thal-2 homozygosity ($\alpha^0\alpha/\alpha^0\alpha$; β^A/β^S). Although the percentages of Hb S were low, both in peripheral blood (30) and in colonies (31), the Σα/non-α values in colonies (0.89, 0.84) were higher than in retics (<0.74).

It seems that highly effective proteolytic mechanisms of immature erythrocyte precursors produced by cultured BFU-e cells could explain the balanced synthesis of α and β chains in β-Thal and in α-Thal colonies while the synthesis of γ chains may decline during the in vitro maturation of these cells.