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Original abstract

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DENSE CELL FORMATION IN PEDIATRIC AND ADOLESCENT PATIENTS WITH SICKLE CELL DISEASE.

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Previous studies on the influence of α-thalassemia (α-thal) on dense cell formation were done mostly in adult SS patients without much concern to discriminate the independent effect of age, Hb F levels, and α-thal. In this report we studied 15 SS patients with α-thal-2 heterozygosity (α-/αα) and 21 non-thalassemic (αα/αα) SS patients of age 2-13. Red cell density distribution and the dense cells were determined by the phthalate ester method and in selected cases by the stractan method. When pooled data were used no significant difference was seen between the two groups. However, when the patients were divided into various groups according to age and Hb F levels, dense cells, D_{50} (median density), R60 (middle 60% density range) were significantly lower in the thalassemic group. The influence of α-thal on red cell density distribution was striking during 2 - 7 years of age and below Hb F levels of 10%. In both patient groups, Hb F levels showed an inverse relationship with D_{50} and % dense cells. Glycated Hb levels used as an index of red cell survival did not show any difference, but showed a direct relationship with the levels of Hb F and hemocrit. It is concluded that age, Hb F level, and α-thal influence dense cell formation in SS patients.