Association of Hemoglobin H Disease With Hb J-Iran (β77 His → Asp): Impact on Subunit Assembly

By Samuel Rahbar and H. Franklin Bunn

A young Iranian female was found to be heterozygous for hemoglobin (Hb) J-Iran (β77 His → Asp) in combination with Hb H disease. The proportion of Hb J in the patient’s hemolysate was surprisingly high: 65% Hb J, 30% Hb A. Thus, the interaction of a negatively charged β subunit variant of Hb with α-thalassemia leads to a marked increase in the relative amount of the variant Hb within red cells. This observation provides further support for an electrostatic model of Hb subunit assembly.

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From the Department of Hematology and Bone Marrow Transplantation, City of Hope National Medical Center, Duarte, CA and Howard Hughes Medical Institute, Harvard Medical School, and the Hematology Division, Brigham and Women’s Hospital, Boston.

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Address reprint requests to Samuel Rahbar, MD, Department of Hematology and Bone Marrow Transplantation, City of Hope National Medical Center, 1500 E Duarte Rd, Duarte, CA 91010-0269.

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ASSOCIATION OF HbH DISEASE WITH Hb J-Iran

Fig 1. Effect of α-thalassemia on the proportion of negatively charged (O) and positively charged (Δ) β globin variants in heterozygotes. In all cases Hb A constitutes the great majority of the remaining Hb. In three-gene deletion α-thalassemia, a small amount of Hb H (\(\beta_4\)) is present. Modified with permission.1

In conclusion, the interaction of α-thalassemia with the negatively charged β globin variant Hb J-Iran provides informative and independent support for the importance of electrostatic attraction in the assembly of Hb subunits.1

REFERENCES

Association of hemoglobin H disease with Hb J-Iran (beta 77 His---- Asp): impact on subunit assembly

S Rahbar and HF Bunn