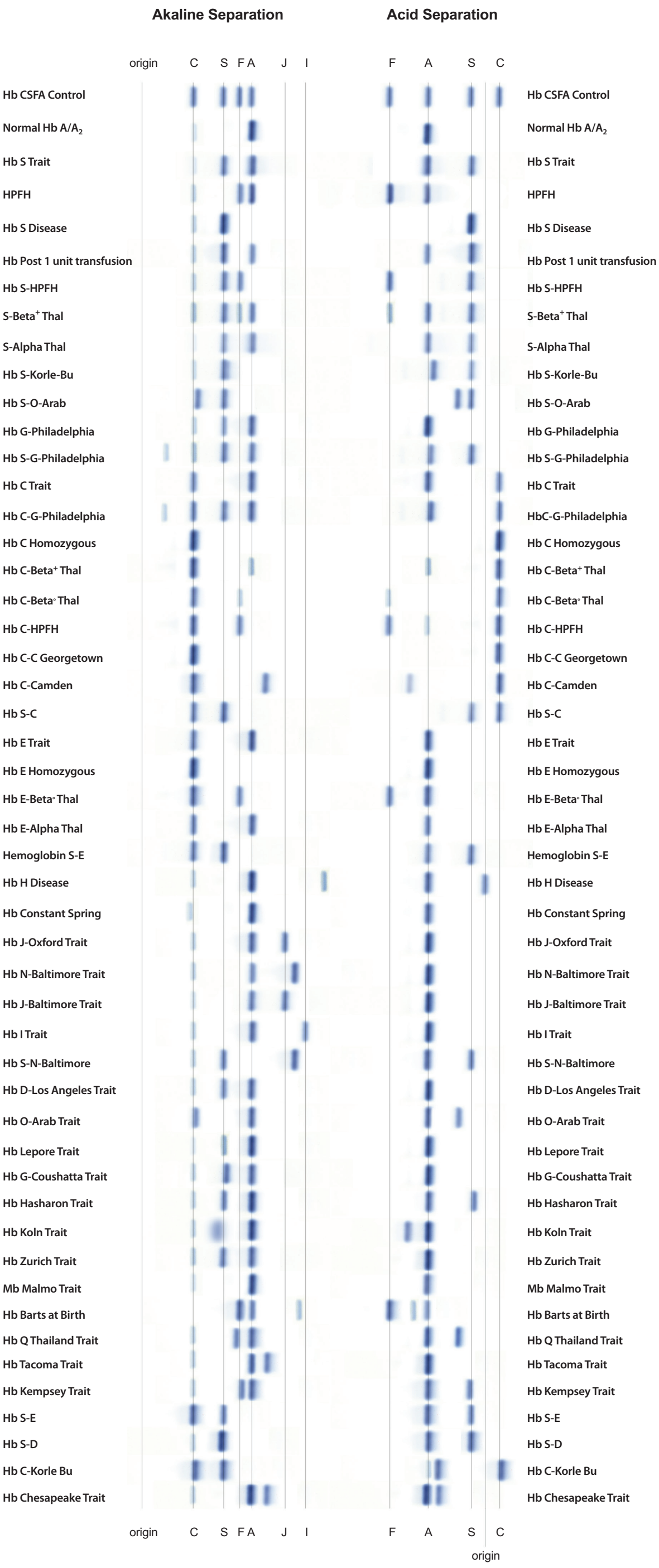
Relative Hemoglobin Mobilities by Gel Electrophoresis[1,3,4]



COMMON HEMOGLOBIN ABNORMALITIES:

Sickle Trait: In alkaline buffer, HbA ~60%, HbS ~35% and HbA_2 <4%.

Sickle Cell Disease: In alkaline buffer, HbA 0%, HbS ~96%, HbA_2 <4% and maybe a small amount of HbF.

Homozygous Beta0 Thalassemia (Beta Thalassemia Major): Beta chain production is completely stopped; HbF 100%.

Beta+ Thalassemia Trait: Beta chain manufacture is reduced but not eliminated. In alkaline and acidic buffer, no abnormal bands are seen. HbA_2 may be elevated 4-8%. Peripheral blood smear shows microcytosis, target cells, hypochromia and basophilic stippling. The degree of symptoms is inversely proportional to the amount of beta globin chain present.

HbS/Beta+ Thalassemia: In alkaline buffer HbS will be greater than 35% depending on how much of HbA is being made by the thalassemic gene.

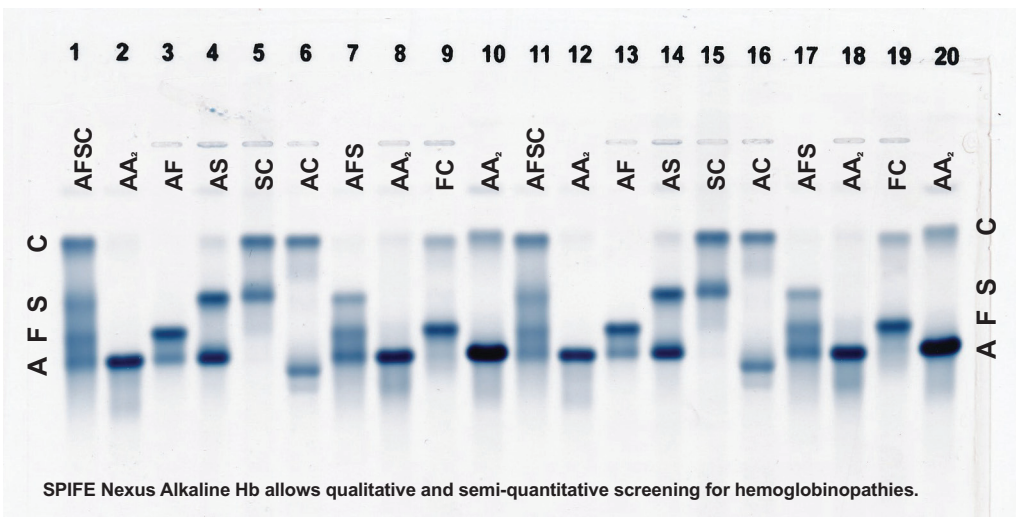
Gel electrophoresis and peripheral blood smears are routinely used to evaluate hemoglobin disorders. Initial electrophoretic separation using an alkaline buffer allows the differentiation of HbA, F, S and C and creates a framework to compare other hemoglobin variants. Further testing using an acidic buffer assists in differentiating variants with similar alkaline electrophoretic mobilities. The SPIFE Touch and SPIFE Nexus gel systems provide clear separations with greater automation than traditional methods. Fifty alkaline and acid electrophoretic patterns are presented here.[1]

Several hundred hemoglobinopathies with one or more amino acid substitutions in one of the four types of globin chains have been identified. These mutations can result in formation of a variant hemoglobin in lieu of the normal hemoglobin A or A_2, or it may cause thalassemia by inhibiting the manufacture of normal hemoglobin.

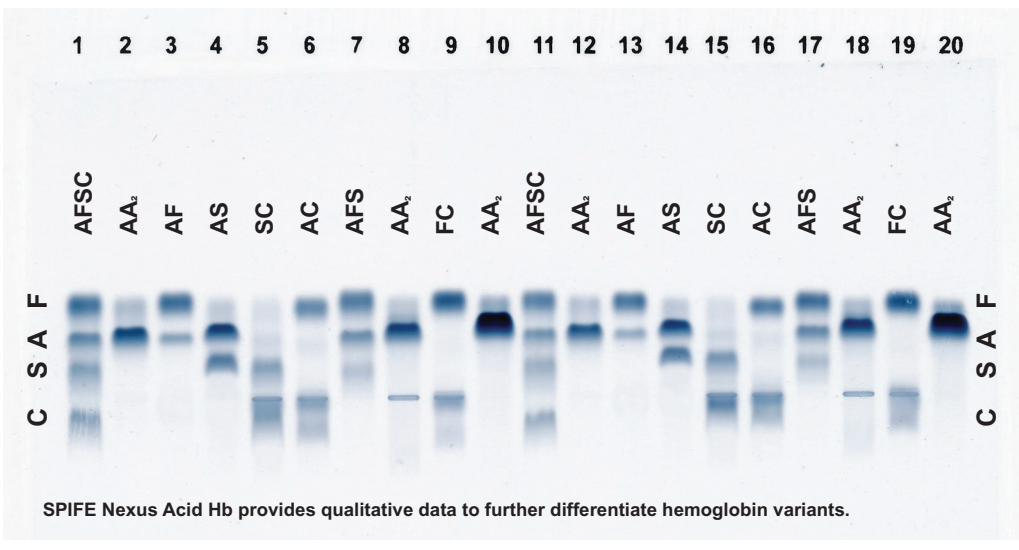
Almost a third of African Americans have some type of deletional α-thalassemia[2] and double mutations are quite possible. Decrease in the production of gamma or delta chains is clinically silent since neither HbA_2 nor HbF are produced in large amounts in an adult. The severity of α or β thalassemia is a function of the number of gene loci deleted and how much functional hemoglobin remains. Deletion of one α loci decreases output of αδ, αβ and αγ hemoglobin bands. At less than one year of age, HbF is still present and the gamma bands without an alpha pair may form up to 2% Barts (γ4). Two α loci deleted may increase Barts to 5% and three α loci deleted results in HbH of 10-15% and the presence of up to 10% HbF. Because α chains are represented in both HbA_2 and any beta chain variant, the percentage of the beta variant fraction is not helpful in identifying the presence of α thalassemias.[3]

Beta+ thalassemia only decreases the percentage of the beta bands present. This means since there are no beta bands in HbA_2, the percentage of this band may be increased up to 10% compared to other hemoglobins present but HbA_2 may be normal in the presence of low iron levels. Low levels of small bands such as HbA_2 and HbF should be quantitated by a more precise method if the quantity is important for interpretation.

For a thorough discussion of thalassemias see Color Atlas of Hemoglobin Disorders by Hoyer, JD and Kroft, SH and published by CAP.[3,4]



SPIFE Nexus Alkaline Hb allows qualitative and semi-quantitative screening for hemoglobinopathies.



SPIFE Nexus Acid Hb provides qualitative data to further differentiate hemoglobin variants.

Bibliography:

1. Schmidt, R. M., Rose G Schneider, and Ronald Barwick. "Table 1: Relative Electrophoretic Mobility." In *Handbook Series in Clinical Laboratory Science* Vol 1, Hematology., 90–91. Boca Raton, FL: CRC Press, 1979.
2. Fairbanks, Virgil F. In *Hemoglobinopathies and Thalassemias*, 20–20. New York, NY: Brian C Decker, 1980.
3. Hoyer, James D, and Steven H Kroft, eds. "Cases 2-3." In *Color Atlas of Hemoglobin Disorders*, 15–28. Northfield, IL: College of American Pathologists, 2003.
4. Bain, Barbara J. *Haemoglobinopathy Diagnosis*, 3rd ed., 91–95. Hoboken, New Jersey: John Wiley & Sons, Inc. 2020.

For more information on the detection of hemoglobinopathies, call Helena Laboratories at 800-231-5663.

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