Differential Response to Hydroxyurea and Incidence of Stroke in Sickle Cell Disease

Study's intent, design and background

The purpose of the study is to identify genetic variants associated with the degree of HbF induced by hydroxyurea. Response to hydroxyurea is variable, and the amount of HbF induced is the best predictor of clinical response on the drug. Sibling and twin studies suggest that HbF response to hydroxyurea is heritable.

Represent/describe the source of the data (e.g., questionnaires, interview forms, protocols)

The data was collected as part of two NIH supported clinical trials, HUSTLE, in which patients with SCD were treated prospectively with hydroxyurea, and SWiTCH, in which patients in the experimental arm were transitioned to hydroxyurea from chronic transfusion therapy for secondary stroke prevention. HUSTLE is Hydroxyurea Study of Long-Term Effects (NCT00305175); SWiTCH is Stroke with Transfusions Changing to Hydroxyurea (NCT00122980).

Which question(s)/section(s) of the documents are associated with each variable.
Start age is age they initiated the drug, hydroxyurea, in years.
HU mg/kg is the maximum tolerated daily dose of hydroxyurea in miligrams per kilogram
BL F/F+S is the baseline fetal hemoglobin (HbF) before starting hydroxyurea. It's (%HbF/%HbF+ %HbS)*100, a corrected value in case they had been transfused with normal blood at that time point.
MTD F/F+S is the HbF at the maximum hydroxyurea dose, adjusted as above.
Delta HbF is the MTD F/F+S-BL F/F+S.

Methods (e.g. derivations/algorithms) that were used to obtain the final data values especially if data values of one variable were derived from more than one source/question/ measurement or variable.

The data was collected from patient's clinical charts. Fetal hemoglobin levels were adjusted for the possible presence of transfused blood with the equation {%HbF/(%HbF +%HbS)} X100.