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A Report from the GRNDaD Multi-site Registry for Sickle Cell Disease: Iron Overload is Under-recognized and Under-managed

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Introduction: GRNDaD is a prospective registry for people with SCD that opened to enrollment in 2016. Nine comprehensive SCD centers from across the United States are currently enrolling patients. The registry includes iron status and management data, important in SCD because chronic transfusion therapy is a mainstay of prophylactic management. Each unit of transfused blood introduces approximately 250 mg of iron into the blood, which can lead to systemic iron deposition, and untreated may lead to organ dysfunction or death.

Methods: GRNDaD currently contains prospective baseline and annual update information on approximately 1000 people with SCD. We analyzed ferritin levels relative to genotype, age, gender, treatment type, liver iron scan results, and chelation therapy history, using chi-squared and pearson statistics for discrete and continuous data, respectively.

Results: There were 783 adults in GRNDaD who had a non-crisis ferritin level from a routine follow-up visit. Nearly 1 in 3 of all participants (n=187, 31.4%) had a baseline ferritin ≥1500 mg/dL. More than a third of that group were not on chelation, and only a quarter had imaging

studies to assess iron accumulation. Ferritin levels were positively associated with liver enzymes, creatinine, and homozygous SCD phenotype.

Conclusion: A significant fraction of the adult SCD population in GRNDaD is living with iron overload, and management could use vast improvement. We speculate that undertreated iron overload is probably both widespread and under-recognized. We anticipate that GRNDaD may be a model for identifying and addressing deficiencies in current clinical practices for management of SCD.