Sunday, December 8: Transforming Care for People with Sickle Cell Disease Across the Globe

See also ASH Press Release

Webcasts at the bottom

812 Chromatin Accessibility Mapping of Primary Erythroid Cell Populations Leads to Identification and Validation of Nuclear Factor I X (NFIX) As a Novel Fetal Hemoglobin (HbF) Repressor

Mudit Chaand, Chris Fiore, Brian T Johnston, Diane H Moon, et al.

CONCLUSION Collectively, these data have enabled identification and validation of NFIX as a novel repressor of HbF, a finding that enhances the understanding of beta-like globin gene regulation and has potential implications in the development of therapeutics for sickle cell disease.

613 Oral Arginine Therapy As a Novel Adjuvant in the Management of Acute Pain in Children with Sickle Cell Anemia in Nigeria: A Randomized Placebo-Controlled Trial

Richard Onalo, Peter Cooper, Antoinette Cilliers, Uche Nnebe-Agumadu, et al.

CONCLUSION Arginine deficiency plays a role in acute pain requiring hospitalization in Nigerian children with SCA, similar to what has been reported in the US. Plasma arginine levels significantly increased with arginine supplementation, and improved global arginine bioavailability was inversely associated with total analgesia and opioids used in VOE management. Total mean analgesia use and pain scores were lower, while time-to-crisis-resolution and LOS were shorter in children treated with arginine compared to placebo. No serious adverse events occurred in the arginine arm, while rates of adverse events were similar to placebo, providing further support for the safety of arginine therapy in children with SCA. Oral arginine is a promising adjuvant therapy for SCA-VOE management.

4667 Fragmentation of Care for Young Adults with Sickle Cell Disease in California

Ashley Shatola, Ann M Brunson, Theresa Keegan, Ted Wun, and Anjlee Mahajan

CONCLUSION Most young adult SCD patients (78%) received inpatient care at >1 facility. Of all age groups, children were most likely to be seen at only 1 facility, suggesting that fragmentation of care begins in early adulthood. Young adults without insurance, patients with more frequent admissions and those who did not always receive care at an SCD SC were at higher risk for fragmented care. Young adults with more frequent admissions were also at an increased risk of mortality. The effect of specialty centers and more consistent location of care on health-related outcomes for patients with SCD requires further study.