STUDY UPDATES #3

Connolly ME, et al. Cognitive functioning and educational support plans in youth with sickle cell disease. *J Pediatr Hematol Oncol.* 2021;43(5):e666-e676.

- Goal: To determine if cognitive functioning in youth with sickle cell disease is associated with educational support
- Youth (age 7 to 16 years) (N=91) with sickle cell disease completed the Wechsler Intelligence Scale for Children
- Caregivers reported educational support (504 Plan/Individualized Education Program) and completed the Behavior Rating Inventory of Executive Function
- Results
  - 58% received educational support
  - Youth with a full-scale intelligence quotient (FSIQ) <90 were significantly more likely to have educational support than youth with FSIQ ≥90
  - Youth with FSIQ ≥90 and FSIQ = 80 to 89 were significantly less likely to have educational support than youth with FSIQ ≤79
  - Relative weaknesses in processing speed and behavioral aspect of executive functioning were associated with educational support
- While youth with significant deficits in intellectual functioning, processing speed, and executive functioning were more likely to receive educational support, youth with more subtle deficits were less likely to receive educational support

Uter S, et al. Measures to reduce red cell use in patients with sickle cell disease requiring red cell exchange during a blood shortage. *Blood Adv.* 2021;5(12):2586-2592.

- Goal: Assess whether implementation of 2 measures could reduce blood use in patients with sickle cell disease requiring chronic red cell exchange
- Measure 1: obtain pretransfusion hemoglobin S results by transfusion start time, thereby allowing calculation of exact red cell volume needed to achieve the desired post red cell exchange hemoglobin S (HbS) level
- Measure 2: identify patients for whom increasing the targeted end procedure hematocrit up to 5% higher than the pretransfusion level (≤36%) was not medically contraindicated, with the goal to enhance suppression of endogenous erythropoiesis, thereby reducing the number of red cell units needed to maintain the same target HbS%
- Implementation of both measures 1 and 2 reduced by 18% (351 units) the number of red cell units transfused to 50 patients undergoing chronic red cell exchange over 6 months
- Pretransfusion HbS% target goals were maintained without a significant change in the transfusion interval; net iron accumulation was low

OTHER PUBLICATIONS

DeMartino P, et al. A budget impact analysis of gene therapy for sickle cell disease. The Medicaid perspective. *JAMA Pediatr.* 2021;175(6):617-623.

Guenther CS, et al. SNAP: Supportive noninvasive ventilation for acute chest syndrome prevention in children with sickle cell disease. *Pediatr Blood Cancer.* 2021;68(8):e29136.

Rashkin SR, et al. Generalization of a genetic risk score for time to first albuminuria in children with sickle cell anaemia: SCCRIP cohort study results. *Br J Haematol.* 2021;doi:10.1111/bjh.17647.

Wright LA, et al. Pain and QOL in pediatric sickle cell disease: Buffering by resilience processes. *J Pediatr Psychol.* 2021;doi:10.1093/jpepst/jsab034.