STUDY UPDATES #2

Forté S, Blais F, Castonguay M, et al. Screening for cognitive dysfunction using the Rowland Universal Dementia Assessment Scale in adults with sickle cell disease. *JAMA Netw Open.* 2021;4(5):e217039.

- Goal: To ascertain the prevalence of suspected dementia in multicultural adults with sickle cell disease (SCD)
- Adult outpatients (N=252) in 2 SCD comprehensive care centers in Canada were screened using the Rowland Universal Dementia Assessment Scale (RUDAS), which was specifically designed for cognitive screening in multicultural populations
- 29 adults (11.5%) had RUDAS scores suggestive of dementia
- Lower glomerular filtration rate and increasing age were associated with RUDAS scores suggestive of dementia; SCD genotype and disease severity were not
    - 8.7% of adults age 18 to 39 years
    - 14.5% of adult age 40 to 59 years
    - 36.4% of adults age ≥60 years

Friedman D, Dozor AJ, Milner J, et al. Stable to improved cardiac and pulmonary function in children with high-risk sickle cell disease following haploidentical stem cell transplantation. *Bone Marrow Transplant.* 2021;doi:10.1038/s41409.021-01298-7.

- Phase 2 trial of myeloimmunoablative conditioning followed by haploidentical stem cell transplantation in children (N=19; age 2 to 21 years) that were homozygous for hemoglobin S with ≥1 high-risk features
    - History of overt stroke, silent stroke, elevated transcranial Doppler velocity, multiple vaso-occlusive crises, and/or ≥2 acute chest syndromes
- At 2 years
    - Specific airway conductance (sGAW) was significantly improved (P<0.004)
        - Proportion with sGAW <80% predicted:
            - Baseline: 71.4%
            - 1 year: 50%
            - 2 years: 20%
    - No significant changes over time in the proportion of patients with forced expiratory volume over 1 second (FEV_1) <80% predicted, FEV_1/forced vital capacity <0.80, or diffusing capacity of lung for carbon monoxide <80% predicted
    - Left ventricular systolic function (fractional shortening) and tricuspid regurgitant jet velocity were stable

Nawaiseh M, Shaban A, Abualia M, et al. Seizures risk factors in sickle cell disease. The cooperative study of sickle cell disease. *Seizure.* 2021;89:107-113.

- Case-control study to compare clinical and laboratory parameters in pediatric and adult patients with SCD who experienced seizures with patients who did not
- 153/2804 (5.5%) pediatric patients experienced a seizure; mean age at first seizure 8.5 years
    - Mean follow-up: 8.5 years (seizures) vs 7.6 years (no seizure)
- 115/1281 (9.0%) adult patients experienced a seizure; mean age at first seizure 28.0 years
    - Mean follow-up: 8.6 years (seizure) vs 8.6 years (no seizure)

- Risk factors for seizures
  - Pediatrics: cerebrovascular accident (OR=5.7, 95% CI 2.9-11.0); meningitis (OR=3.6, 95% CI 1.8-7.2); eye disease (OR=3.4, 95% CI 1.5-8.0)
  - Adults: cerebrovascular accident (OR=7.5, 95% CI 3.5-16.0); meningitis (OR=5.6, 95% CI 1.5-20.0); nephrotic syndrome (OR=3.0, 95% CI 1.2-7.9); spleen sequestration (OR=2.7, 95% CI 1.1-6.3); pneumonia (OR=2.1, 95% CI 1.0-4.4)

Peterson RK, Williams S, Janzen L. Cognitive correlates of math performance in school-aged children with sickle cell disease and silent cerebral infarcts. *Arch Clin Neuropsychol.* 2021;36(4):465-474.

- To identify the cognitive underpinnings of math difficulties in children with SCD and silent cerebral infarcts (SCI)
- Youth (N=68) completed measures of attention, working memory, processing speed, math reasoning, and math fluency
- Overall intellectual functioning was in the low average range
- Math reasoning was significantly positively correlated with measures of working memory, processing speed, attention span, and executive functioning
  - Short-term attention—but not sustained attention—was associated with math performance
- Working memory deficits accounted for the greatest variance in untimed mathematic performance in youth with SCD, which is consistent with other populations with white matter dysfunction

OTHER PUBLICATIONS

Adeniyi AT, Okeniyi JAO, Adegoke SA, Oseni SBA, Smith OS, Abe-Dada AA. Clinical utilities of electrocardiography in the diagnosis of myocardial ischemia in children with sickle cell anemia: Correlation with serum cardiac troponin I. *J Pediatr Hematol Oncol.* 2021;doi:10.1097/MPH. 0000000000002230.

Chen-Goodspeed A, Idowu M. COVID-19 presentation in patients with sickle cell disease: A case series. *Am J Case Rep.* 2021;22:e931758.

De Araujo, JA, Rossi DAA, Valadao TFC, et al. Cardiovascular benefits of a home-based exercise program in patients with sickle cell disease. *PLoS One.* 2021;16(5):e0250128.

Hood AM, Nwankwo C, Walton A, et al. Mobile health use predicts self-efficacy and self-management in adolescents with sickle cell disease. *Transl Behav Med.* 2021;doi.10.1039/tbm/ibab041.

Moody KL. Paternal stress and child outcomes in youth with sickle cell disease. *J Pediatr Psychol.* 2021;doi:10.1093/jpepsy/jsab059.

Woodward KE, Johnson YL, Cohen LL, Dampier C, Sil S. Psychosocial risk and health care utilization in pediatric sickle cell disease. *Pediatr Blood Cancer.* 2021;e29139.