SCD-Related Cerebrovascular Disease

What it covers

- Screening and treatment guidelines for SCD complications affecting the brain, including risk of strokes and other neurological complications for both children and adults.

Why it matters

- Neurologic complications such as stroke and silent stroke are a major cause of disability and death for children and adults living with SCD.
- Effective screening for stroke risk in SCD may help prevent or reduce the risks associated with these types of complications.
- "Silent strokes" – those without obvious neurological symptoms – affect 1 in 3 children and at least 1 in 2 adults with SCD, and often go undetected. They are linked to cognitive impairment potentially impacting school performance, and are risk factors for future strokes, which can be life-threatening. Silent strokes occur in a region of the brain that does not affect motor skills, which is why they are often not detected without a brain scan.
- Clear, updated guidelines can help emergency room physicians quickly manage and treat children who may have had a stroke. In these settings, the faster a child or adult with SCD is treated, the better the outcome.
- Most children born with SCD live in low-to-middle income countries, which have the largest global population of SCD, but are frequently without access to stroke-prevention practices.

Who it affects

- **Hematologists:** Hematologists are looked to as experts on SCD and therefore have a role to play in ensuring neurologists and other specialists are implementing the most current clinical practice guidelines.
- **Primary care providers:** Many primary care providers have limited experience managing SCD, but are often caring for people living with SCD. The guidelines will serve as an easy, accessible reference to support them in preventing and treating neurological complications of the disease.
- **Neurologists:** Neurologists are the experts in acute and long-term management of strokes; however, they have not been traditionally part of the care team of children and adults who have had a stroke. The guidelines will integrate the known management of strokes in the general population with the management of strokes in those with SCD.
- **Emergency room physicians:** In the emergency room setting, caregivers need to act quickly and navigate complex issues to prevent further brain damage for a patient experiencing stroke. This includes knowing when to call a hematologist and neurologist, order an image of the brain, and give a patient a transfusion.
- **Parents:** Parents can seek professional help in assessing whether their child has had or is at risk for strokes or silent strokes. Knowing this information will help their child get needed support.
- **Educators, employers, and family members:** Educators, employers, and family members should understand that children and adults with SCD may have experienced silent strokes and not know it. They may need or require additional educational, employment, or family support to maximize their potential.
- **Policymakers:** Policymakers will be informed of the knowledge gaps that exist in SCD evidence so they can direct research funding to fill in the gaps. In this regard, these guidelines may help shape policy.

SCD-Related Cerebrovascular Disease

What are the highlights

- Adults and children should be screened via brain scans to assess their risk for a silent stroke. Based on the screening:

  > Children can receive regular blood transfusions to reduce the risk of a new stroke, another silent stroke, or both. In addition, screening can identify children who have already had a silent stroke and are therefore eligible for school-based resources and other types of educational support.

- *In children age 2-16 years with the most common type of SCD who have abnormal brain scan measurements:* receiving monthly blood transfusion therapy - over no transfusion - is recommended in order to decrease the 1 in 10 risk of having a stroke in the next 12 months.
- *In children age 2-16 years with SCD, living in low-resource settings with abnormal ultrasound brain scan measurements, and unable to receive regular blood transfusion therapy:* hydroxyurea therapy - over no therapy - is recommended to decrease the 1 in 10 risk of having a stroke in the next 12 months.
- *In children and adults presenting with acute strokes:* physicians should consider performing a simple red blood cell transfusion without delaying for sub-specialty consults or a definitive diagnosis of stroke via brain scan.

Total number of panel recommendations: 18

REFERENCE

DeBaun MR, Jordan LC, King AA, et al. American Society of Hematology 2020 guidelines for sickle cell disease: prevention, diagnosis, and treatment of cerebrovascular disease in children and adults. *Blood Adv.* 2020;4(8):1554-1588.