VDH VIRGINIA DEPARTMENT OF HEALTH

Division of Child and Family Health- Newborn Screening Follow-Up Program

Sickle Cell Disease (SCD)

Health Care Professional Fact Sheet

Sickle cell disease (SCD) is a group of inherited blood disorders characterized by the presence of two abnormal hemoglobin genes, with at least one being hemoglobin S. Specific mutations in the hemoglobin cause the red blood cells to become sickle shaped. As a result, red bloods cells are less functional, causing anemia and other vaso-occlusive complications. If SCD is left untreated, it can cause life-threatening infections, organ damage or even death. However, with early treatment, the child will most likely have a healthy life. Hemoglobin screening results are listed in order of the amount of hemoglobin present. Example: F>A>S (sickle cell trait) is not the same as F>S>A (sickle beta plus thalassemia disease)	
Incidence	Sickling syndromes affect: • Approximately 100,000 Americans • 1 in every 365 African American births • 1 in every 16,300 Hispanic-American births
Analyte Measured	Hemoglobin (HgB)
Normal Test Results	FA
Abnormal Test Results	FAS, AFS
Critical Test Results	FS, FSC, FSA, FSE, FSV
Signs and Symptoms *Please note: these findings may not be present in young infants or in milder forms of the disease*	SCD is a condition with multiple types; symptoms and complications are different for each type and person, and can range from mild to severe. Signs of the disease start to show usually around 5 to 6 months of age when the fetal hemoglobin levels decrease. **Early signs of SCD may include:** • Sleeping longer or more often • Tiredness • Difficulty breathing • Pain or swelling in the hands or feet (dactylitis) • Cold hands or feet • Pale skin
Next Steps	• **Contact family to notify them of the newborn screening result.** • **Arrange confirmatory testing of a second blood sample using a hemoglobin electrophoresis or repeat newborn screen (solubility tests are not recommended) preferably by 2 months of age, but no later than 6 months of age.** • **Refer family to regional pediatric hematologist.** • Provide parental education (see accompanying sheet). • Clinical assessment
Treatment (*if indicated*)	**Consult with regional pediatric hematologist.** Start penicillin by 2 months of age to prevent pneumococcal infections.
Additional Resources	• Baby's First Test *www.babysfirsttest.org* • Centers for Disease Control and Prevention https://www.cdc.gov/ncbddd/sicklecell/index.html • National Institutes of Health National Heart, Lung and Blood Institute https://www.nhlbi.nih.gov/files/docs/guidelines/sc_mngt.pdf • VDH Sickle Cell Program: http://www.vdh.virginia.gov/sickle-cell-programs/

Educational content adapted from www.babysfirsttest.org and https://www.cdc.gov/ncbddd/sicklecell/index.html

VIRGINIA Sickle Cell awareness PROGRAM

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Virginia Newborn Screening Services
Keeping Virginia's Babies Healthy – One Foot at a Time