

Sickle Cell Disease

Condition Description

Sickle cell disease is a genetic abnormality found in ~ 1/400 individuals of African descent. It is a life-long condition whose course is quite variable. The oxygen-carrying red blood cells can become sickle shaped (like a crescent moon) and block blood vessels causing pain and organ damage. Childhood complications can include pain crises (often requiring hospitalization), stroke, silent strokes, growth failure, and even death.

Treatment

Sickle cell disease is one of the hemoglobinopathies detected by newborn screening. Early detection of the disorder allows for treatment with antibiotics to prevent specific infections. Specialists monitor for complications including severe anemia and strokes and educate families about strategies to prevent pain crises.

Range of Outcomes

As Schatz summarized in 2006 ~ 1/3-1/4 affected children have specific cognitive deficits and academic difficulties. Risks for stroke increase after the first year of life ultimately affecting 12% of the children (Day). In addition to physical sequelae of strokes Wang showed that average IQ falls with a WISC score of 76.9. Silent infarcts (MRI changes but no physical signs) found in 20% of kids resulted in WISC scores of 77.2 in Wang's series of 373 patients. Schatz and others also have demonstrated neurocognitive changes in children without MRI changes especially in executive function, auditory discrimination, and attention span. These changes may explain why 2 studies (Steen & Chua-Lim) showed children with sickle disease scored worse on kindergarten readiness testing than matched controls. Cognitive deficits together with pain crises, increased absenteeism, and socioeconomic factors explain reports that over half of the children with sickle cell disease are either held back in school or placed in special education (Schatz, Day). Multiple hospitalizations and pain crises can become stressors for the child's social-emotional functioning and the parents' coping skills.

For More Information

This fact sheet was developed for use in determining eligibility for early intervention services only. For more complete information, the following resources might be useful:

MedlinePlus

MedlinePlus brings together authoritative information from NLM, the National Institutes of Health (NIH), and other government agencies and health-related organizations. Preformulated MEDLINE searches also give access to medical journals.

<http://medlineplus.gov/>

GeneReviews

Information on genetic testing and genetic disorders is available in the GeneReview section of the GeneTest website. www.genetests.org

References

Chua-Lim, C et al, Deficiencies in School Readiness skills of Children with Sickle Cell Anemia, Southern Medical Journal 1993, 86(4); 397-402.

Day, S and Chismark, E, The Cognitive and Academic Impact of Sickle Cell Disease, The Journal of School Nursing 2006, 22(6); 330- 335.

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Steen, RG et al, Cognitive Deficits in Children with Sickle Cell Disease, Journal of Child Neurology 2005, 20(2); 102-107.

Wang, W, et al, Neuropsychologic performance in school-aged children with sickle cell disease: A report from the Cooperative Study of Sickle Cell Disease, The Journal of Pediatrics 2001, 139 (3); 391-397



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