Table 4: Evidence Supporting Anticipatory Guidance Regarding School Attendance/Performance in Children with Sickle Cell Disease

TYPE OF EVIDENCE	KEY FINDINGS	LEVEL OF EVIDENCE (USPSTF RANKING*)	CITATIONS
Clinical guidelines (from agencies or groups)	NHLBI guidelines suggest that: Unless children have impairments related to stroke, they have normal intelligence and should be encouraged to reach their full potential. Teacher and other school officials who interact with children with SCD should be provided with educational materials, and school staff should meet with parents to set realistic educational goals. Illness often interrupts school and extracurricular activities, so tutoring and other assistance may be needed. School-age children should participate in PE but be allowed to rest if they become tired, and they should be encouraged to drink fluids after exercise. While addressing special needs and routine childhood health care, allowances must be made for regular school attendance (by flexible scheduling of appointments), for activity (by offering alternatives to inappropriate sports), and for learning (by providing support and education to school staff).	III	National Heart Lung and Blood Institute. The Management of Sickle Cell Disease. National Institutes of Health. Bethesda, MD, 2002.
Clinical guidelines (from agencies or groups)	The AAP sections on Hematology/Oncology and the Committee on Genetics suggest that: School performance should be monitored for evidence of neurodevelopmental problems. Comprehensive evaluations also provide an ideal setting providing age-appropriate family and patient education and for evaluating and addressing psychosocial issues. Psychosocial care for school-age children should include reviews of school attendance and performance and discussions about whether to consider formal neurocognitive testing; and offers to help educate school personnel about SCD.	III	American Academy of Pediatrics Section on Hematology/Oncology and Committee on Genetics. Health supervision for children with sickle cell disease. *Pediatrics* 2002; 109(3):526-535.
Clinical overview	In young children, poor educational performance may indicate early ischemia of the central nervous system; neurocognitive testing may be appropriate. For adolescents, ongoing counseling is required for issues around school performance.	III	Claster S, Vichinsky EP. Managing sickle cell disease. *BMJ* 2003; 327:1151-1155.

TYPE OF EVIDENCE	KEY FINDINGS	LEVEL OF EVIDENCE (USPSTF RANKING*)	CITATIONS
Clinical overview and guidelines	School personnel should be educated about SCD and encouraged to accommodate frequent and unpredictable absences. Poor school performance may indicate the need for neurocognitive testing. Important stresses that can affect a family's ability to cope with SCD include the economic and educational consequences of time lost from work and school.	III	Lane PA, Buchanan GR, Hutter JJ, et al. Sickle cell disease in children and adolescents: diagnosis, guidelines for comprehensive care, and care paths and protocols for management of acute and chronic complications. 2001; Annual Meeting of the Sickle Cell Disease Care Consortium, Sedona, AZ. Nov. 10-12, 2001. http://txch.org/wp-content/uploads/sickle-cell-disease-guidelines-complications.pdf; accessed March 20, 2014.
Clinical overview	The compromised intellectual performance of school-age children with SCD demonstrates the need for effective intervention to prevent this subtle but profound complication of SCD. Patients with SCD may benefit from special education programs that provide assistance in targeted areas of learning disability and from tutorial programs for those who fall behind in school. More essential, however, are interventions that address the underlying pathophysiologic mechanisms that result in silent infarcts and deterioration in neuro-psychometric performance. Such interventions, which might include hydroxyurea (a promoter of Hb F), stem cell transplantation, or chronic transfusion therapy, are needed before the early years of primary school.	II	Wang W, Enos L, Gallagher D, et al. Neuropsychologic performance in school-aged children with sickle cell disease: A report from the Cooperative Study of Sickle Cell Disease. *J Pediatr* 2001; 139(3):391-397.

Note: USPSTF criteria for assessing evidence at the individual study level are as follows: I) Properly powered and conducted randomized controlled trial (RCT); well-conducted systematic review or meta-analysis of homogeneous RCTs. II) Well-designed cohort or case-control analytic study. III) Opinions of respected authorities, based on clinical experience; descriptive studies or case reports; reports of expert committees.