Procedural Panel has developed a new billing code (96127) for behavioral health screening that can be used to screen for cognitive impairment.[117] Physicians, nonpsychologist staff, and practitioners have also seen a dramatic increase in cognitive screenings conducted, indicating that surveillance and screening are possible in settings without ready access to specialists in psychological assessment.[118] Both the American Psychological Association and the National Academy of Neuropsychology have acknowledged that in many health care settings there is a need for medical doctors or nonpsychologist staff or practitioners to provide wider access to cognitive or behavioral health screenings, and these organizations have provided clarification on the distinction between screening and a more comprehensive psychological assessment requiring a specialist.[119,120]

The specific tests for cognitive screening in children and adults with SCD, and the optimal strategies to screen and implement rehabilitation for cognitive impairments, have not been established. Consequently, recommendations are extrapolated from non-SCD populations. Evidence-based practices for screening for cognitive impairments and rehabilitative interventions for cognitive function, specifically executive dysfunction, are well-established and endorsed practices in professional societies, such as the American Academy of Pediatrics, the American Academy of Neurology, and the American Congress of Rehabilitation Medicine. The guideline panel relied heavily on well-established evidence-based practices from these 3 prominent professional societies to identify strategies for screening and treatment of cognitive morbidities.[121-123]

Summary of the evidence. The systematic review did not identify any studies that compared screening to no screening. The review identified 48 studies that evaluated cognitive screening without a comparison group and reported outcomes of IQ score, the prevalence of cognitive impairment, developmental delay, or school performance. The evidence summary and EtD framework can be found online at: https://guidelines.gradepro.org/profile/72b18bf3a46115c0989f3068aa701629.

Rationale and expected benefits. The use of formal screening tools improves the ability to detect developmental delays or cognitive impairment compared with informal observational methods.[124,125] Furthermore, the combination of surveillance and screening appears to work better for identifying children in need of developmental services than either method in isolation.[126] Many screening tools can be used in primary care or community settings, require less administration time than full assessments, and can be administered by staff with less training, so that access is increased and burden to patients is decreased. See Tables 2 and 3 in supplemental File 5 for established initial screening tools and ongoing surveillance questions, respectively.

In the general population and in children and adults with SCD, identification of children and adults with developmental or cognitive delays increases likelihood of access to remediation services. In the United States and in most European countries, laws mandate interventions for children with significant developmental delay and conditions that impact learning in a public setting. If the child is <3 years of age, a significant developmental delay results in referral to early intervention therapy and the child and family undergo assessment for an individualized family service plan.[127] Services are typically provided in the home or "natural environment" and include occupational therapy, physical therapy, developmental therapy, and speech therapy. These services are provided to enhance the development of the child. If the child or young adults is between 3 and 21 years of age, and a significant impairment is identified, the local public school district is tasked with working with the family to develop an individualized education program. The program provides services in the public school educational setting including supplemental aides if indicated, learning accommodations, and modifications.[127] Both the individualized family service plan and individualized education plans include measurable goals, assessments, and meetings to review the children's progress. If a child is not making progress, the plans are typically modified to provide better support.

A recent systematic review of cognition across the lifespan of people with SCD confirmed that the magnitude of deficits increases as the injury to the brain increases. On average, performance declined from groups without infarct to those with silent cerebral infarct to those with overt stroke.[6] In addition, medium to large differences in cognition between children with SCD and their siblings or controls existed. Fewer adult studies were included, but small to large differences between the adults with SCD and controls were documented, with the greatest difference in processing speed.[6]

Vascular cognitive impairment is of particular concern in adults with SCD and requires ongoing efforts to identify cognitive symptoms and medical risk factors.[128] Detection of cognitive impairments is important for identifying treatable causes, helping patients and families to understand the cause of functional deficits, and discussing the prognosis to plan for future needs.[123] Outpatient services such as speech therapy, occupational therapy, or rehabilitation psychology may be indicated and available depending on the results of a full evaluation.[129]

In general, children and adults with SCD are a vulnerable population. The overwhelming majority of patients in the Unites States with this condition are African American[130] and living at or near poverty level[131-133]; associated social factors such as parental education attainment and measures of socioeconomic status impact cognitive performance.[134-137] In addition, in children with HbSS or HbSβ^0 thalassemia and disease-associated morbidity such as low oxygen hemoglobin saturation levels, silent cerebral infarcts and overt strokes are all associated with lower FSIQ scores.[134,138]

Summary of harms and burden. There are few downsides to surveillance and screening for cognitive impairment, developmental impairments, or both, as part of routine health care. Available cognitive-screening methods improve detection yet lack sufficient sensitivity to be used as a substitute for clinical judgment.[120] For cases in which the patient's history, risk factors, and functional complaints indicate a high risk for cognitive impairment, screening may be skipped in favor of directly referring for a comprehensive assessment. False-positive results from screening can also result in distress and inconvenience for patients and their families.[139,140] Such distress likely already exists for patients and families who already have concerns about developmental delays or cognitive impairment.

EtD criteria and implementation considerations. The quality of evidence demonstrating benefits of cognitive and developmental screening specifically in individuals with SCD is low. However, such screening is noninvasive and can lead to timely referral to address those with developmental delays and cognitive impairment. The harm of not addressing such deficits in children and adults is severe on a personal and societal level. Therefore, a low certainty in benefit but high certainty of harm (particularly from other populations) justified a strong recommendation.[141] When considering patient values and preferences, the panel members,