Transition Planning for Youth With Sickle Cell Disease: Embedding Neuropsychological Assessment Into Comprehensive Care

abstract

OBJECTIVE: Sickle cell disease (SCD) conveys a high risk of neuropsychological impairment due to chronic anemia, hypoxemia, cerebrovascular ischemia and stroke, in addition to causing pain and other biomedical complications. Few families accessed neuropsychological testing via the traditional referral system. In this paper, we describe a successful alternative strategy for addressing cognitive and behavioral needs of youth with SCD.

METHOD: The Sickle Cell Disease Program for Learning and Neuropsychological Evaluation (SCD-PLANE) was embedded in 2007 into the SCD Comprehensive Clinic of Children's Hospitals and Clinics of Minnesota. SCD-PLANE includes brief neuropsychological testing, individualized recommendations, patient education and advocacy, interdisciplinary collaboration, and follow-through to assure patients' access to recommended resources. The purpose of SCD-PLANE is to identify and recommend interventions for cognitive, academic, emotional, behavioral, or social problems affecting child development and transition to adulthood.

RESULTS: Access to neuropsychological testing improved dramatically and is well accepted by SCD patients and families. Test results reveal increased risk of problems with executive function, speed of processing, graphomotor function, academic achievement, and parent-reported anxious/depressed symptoms, despite normal intelligence and language abilities. Patients' needs had not been previously identified or served by schools or other community agencies.

CONCLUSIONS: The SCD-PLANE, providing brief neuropsychological testing embedded within routine annual visits to SCD Comprehensive Clinic, is an effective approach to detect unmet developmental, educational, and behavioral needs within an underserved high-risk population. As youth move towards adulthood, considering cognitive, academic, and mental health status facilitates successful health care transitions. Pediatrics 2010;126:S151–S159
Children and adolescents with sickle cell disease (SCD) are an underserved population on the basis of their high risk of neuropsychological problems (ie, cognitive, emotional, social, or behavioral problems related to developmental brain differences) in addition to biomedical complications, socioeconomic stressors, and demographically related health care disparities. In this article we describe our experience with the Sickle Cell Disease Program for Learning and Neuropsychological Evaluation (SCD-PLANE) at the Children’s Hospitals and Clinics of Minnesota, which makes neuropsychological services accessible and acceptable to families by embedding them within the Sickle Cell Disease Comprehensive Clinic (SCDCC). The purpose of the SCD-PLANE, which began in 2007, is to alleviate previously encountered obstacles to patient care; assess neuropsychological status efficiently and economically; identify any problems or special talents that may need to be addressed; recommend appropriate educational, psychological, and/or medical interventions; and help families locate and follow-through with treatment providers, as needed, by facilitating contacts with schools, hospital services (such as genetic counseling), or local community agencies (such as a psychological clinic or speech therapist). In contrast to a “traditional” model of care, the SCD-PLANE model has yielded a dramatic increase in patient access, participation, and follow-through by embedding neuropsychological services (introductions, testing, and “checkups”) into the annual SCDCC day. By addressing unidentified or unmet needs and “teaming” with parents to help them obtain services, the SCD-PLANE optimizes family-centered care for all children followed in the SCDCC and facilitates transition of older adolescents into appropriate adult health care, postsecondary education, and independent living.

SCD-RELATED MEDICAL AND NEUROPSYCHOLOGICAL COMPLICATIONS

The SCDCC hematology staff have always provided individualized health care–transition meetings for youth who are graduating to adult health systems. Physical complications associated with SCD (such as chronic anemia, pain, organ damage, and susceptibility to infectious illness) may present serious obstacles to young adults’ ability to work, pursue postsecondary education, and maintain adult relationships. Other SCD-related problems of particular concern during adolescence or young adulthood include persistent urinary incontinence, pulmonary hypertension, priapism, delayed puberty, small stature, bone necrosis, and retinal strokes. Health problems that are common for adolescents in the general population (such as inadequate nutrition, smoking, alcohol or drug use, and pregnancy) impose even greater medical risks for teenagers and young adults with SCD. Furthermore, maintaining adequate medical insurance coverage is a challenge when children outgrow their eligibility for coverage under their parents’ policies.

Neuropsychological functioning of individuals with SCD is affected by their history of chronic anemia, pain events and use of opioid pain medications in early childhood, susceptibility to infectious illnesses, airway disease, and cerebrovascular disorders.1–5 Of all children with hemoglobin SS SCD, 10% to 15% have overt strokes by the age of 15, and 20% to 25% have silent (subclinical) strokes.4,5 Neuropsychological testing is particularly sensitive for detecting silent strokes,4,6 which are associated with impaired executive function (concentration, working memory, planning and organization, and mental efficiency) and mildly diminished general intelligence. These symptoms are also common in children and adults with SCD who have normal neuroimaging test results.12 Common neuropsychological concerns among youth and young adults with SCD are impairment of executive function (ie, the skills required to initiate and sustain a systematic approach to accomplishing complex tasks) and slow processing speed (working more slowly than same-age peers). Youth with these difficulties may exhibit forgetfulness, distractibility, disorganization, inflexibility or overdependence on routine, slower task completion, and difficulty with multitasking or coping with time pressure.

Health care–transition planning for youth with SCD traditionally has not addressed individual variations in neuropsychological function, or familial/cultural factors, that affect health literacy, coping, resilience, and other cognitive-behavioral moderators of health status. Although studies of long-term outcomes for adolescents with SCD have been few, results of research on other populations with chronic medical conditions suggest that youth with neuropsychological impairment obtain poorer grades and academic achievement test scores than those with intact cognitive functioning and are more likely to be retained or drop out of school. Transitioning into adult life may be more problematic for these youth, perhaps because their problems with executive function compromise social problem-solving skills.27–9

OBSTACLES TO CARE UNDER THE TRADITIONAL MODEL

Before 2007, under the traditional model, our hematology staff typically recommended a neuropsychological evaluation if families complained of learning or behavior problems. Provid-
ERS faxed referrals to the neuropsychology service, where testing would be scheduled on some future date, in a different building, by a neuropsychologist whom most patients had never met and whose role and rationale they rarely understood. Most families of children with SCD did not reply to letters or calls from the neuropsychology service, never scheduled testing, or made an appointment but did not keep it. Those few who completed testing rarely kept follow-up appointments to discuss results and recommendations. Parents would be mailed a written report, but there was no reliable way to determine if they accepted the recommendations or understood how to implement them. Patient addresses and telephone numbers changed frequently, which made follow-up contact difficult.

Both psychology and hematologic providers felt dissatisfied with this model of care for children with such a high-risk condition. We perceived patient-provider relationship issues, as well as logistics, as obstacles to care. Almost all patients with SCD in our clinic, as in the United States, are people of color, and a majority of the health care providers are white. The families in the SCDC speak Spanish, Haitian Creole French, and a variety of African languages, as well as English. Racial, ethnic, or cultural differences in approaches to health care communication and self-advocacy can affect how patients access and use health care information. Social, cultural, and linguistic/communication problems and financial issues interfere with making and keeping appointments for traditionally scheduled neuropsychological testing for a child with SCD. These issues include mistrust of and unfamiliarity with neuropsychological tests and providers, unwillingness to take an extra day off school or work, transportation costs, difficulty negotiating with insurers, long waiting lists, complex forms to complete before testing (especially daunting for parents with limited education or English literacy), and concerns about stigmatization or racial bias in testing. Therefore, the importance of creating ongoing relationships of trust and respect was central to our thinking about the SCD-PLANE. As a clinic nurse put it, “Whatever else we do, we’ve got to have face time.”

EMPHASIS ON FAMILY-CENTERED CARE

Parents’ knowledge and perceptions of SCD affect their use of emergency and routine health care and serve as a model for self-care behaviors in youth and young adults. Therefore, family-focused programming throughout childhood is a key factor in successful youth transition. The program’s goal for teenagers is to promote a gradual transfer of responsibility and knowledge toward self-care and avoid the extremes of overdependence or alienation. As we and other investigators have reported, however, problems with executive function (“project management” of everyday life and school tasks) are more common among youth with SCD than in the general population. Youth who have significant difficulty with executive function seem particularly likely to need continued active parenting. Moreover, many families in our clinic, particularly those with strong African roots, identify with “collectivist” cultural norms, in which adolescents develop a secure, mature, individuated adult identity without ever radically separating themselves from their families of origin. Therefore, the SCD-PLANE strives to establish strong relationships with parents, educating and supporting them as advocates for their children, to foster development of strong relationships with adolescents, and to promote successful transitions into adulthood regardless of whether there is physical separation of the adolescent from his or her parents.

THE HOSPITAL-SCHOOL LIAISON ROLE

For children and youth with chronic medical conditions, hospital-based programs such as the SCD-PLANE provide a critical adjunct to school-based services. Special-education needs of youth with SCD often remain unidentified or underserved in schools. School personnel often are unaware that youth with SCD may qualify for accommodations and modifications of the standard curriculum, in either regular or special-education settings, under federal laws that pertain to persons with health-related disabilities. Furthermore, some school staff members are reluctant to identify a child as needing special-education services because of legitimate concerns about offending parents, being perceived as racially or culturally insensitive, contributing to the overrepresentation of children of color within special-education classes, or inaccurately labeling or stigmatizing the child. These issues are especially challenging in regard to families who are learning English as a second language. Finally, school psychologists may not be well informed about the educational implications of SCD-related medical complications or about strategies for supporting students with SCD. Therefore, the SCD-PLANE neuropsychologist functions as a hospital-school liaison. He or she advises schools about how the child’s medical condition affects learning and behavior, works with schools and families to develop appropriate educational and behavioral programming, and consults with medical providers as to how differences in learning and behavior may affect a child’s health status. Funding this part of the program is an ongoing concern. It is unfortunate that these hospital-school liaison services are not billable to medical
insurance or to school systems except in rare circumstances. So far, this aspect of the SCD-PLANE has been supported by hospital philanthropies.

METHODS

The SCDCC serves ~280 patients aged 0 to 21 years. Patients routinely receive an annual comprehensive checkup during the month of their birthday. Specifics vary with age and disease severity, but typically the patient’s day includes physical examination and consultation with a hematologist and pulmonologist; transcranial Doppler assessment of cerebral blood flow; an echocardiogram and pulmonary-function testing to assess pulmonary hypertension and asthma; and SCD-PLANE testing or follow-up visit (ie, 90-minute neuropsychological evaluation by SCD-PLANE staff, including a parent interview, age-appropriate individual testing, parent and teacher questionnaires, and review of relevant medical and school records). Lunch, parking, and/or transportation vouchers, funded by the hospital’s philanthropic foundations or by state social services, are available to those who request them.

For the SCD-PLANE, the goal is to evaluate every child of school or transition age (5–21 years) at least once; follow-up evaluations are performed annually or as needed to evaluate response to interventions, persisting needs, or (rarely) deteriorating function that may indicate intensified medical intervention (eg, a change from medication management to blood transfusion). Neuropsychological assessments are performed in conjunction with other annual laboratory tests, so parents and children need not miss an extra day of work or school and incur no additional transportation time or expense. Testing occurs in a trusted, familiar location. The neuropsychological test battery is briefer because it specifically targets areas of function most commonly affected by SCD, namely, executive functions such as attention, working memory, and processing speed. Additional measures sometimes are added to evaluate specific learning or emotional issues noted on previous testing, or concerns expressed by families.) With the assistance of a skilled psychometrist, the neuropsychologist is able to evaluate up to 4 patients on each of the 3 monthly comprehensive-clinic days. This increased volume enhances patient access to services and offsets costs of cancelled or failed appointments.

In addition to these logistic changes, the SCD-PLANE emphasizes personal introductions and face-to-face relationships to build rapport, respect, and trust. Hematology staff who have cared for children since birth introduce the neuropsychologist as a member of a multidisciplinary team, recommend testing, and help explain to parents how this information may benefit their child. The neuropsychologist currently begins testing when children are 5 years old, but we often meet the families of 3- to 4-year-olds to explain the SCD-PLANE. The neuropsychologist’s regular presence in the SCDCC facilitates introductions and follow-through, both of which build positive relationships that create a foundation for parents to accept, understand, and implement recommendations based on test findings. New patients who are not scheduled for testing are able to meet ad hoc with the neuropsychologist to follow-up on previous testing results or recommendations or to discuss future appointments. The neuropsychologist’s time for test interpretation, report-writing, and face-to-face treatment-planning with families is billable to medical insurance. The unbillable time for school visits and ad hoc meetings is supported by philanthropic and research grants.

After the testing, SCD-PLANE staff help families, school staff, therapists, or others who work with the youth, as needed, to design an individualized education program and nursing care plan and, if the child is in high school, a formal transition plan. Specific educational recommendations might include strategies for addressing warning signs of stroke and teaching strategies for problems with attention, organization, short-term working memory, or academic or processing-speed deficits. SCD-PLANE staff work with hematology providers to help schools and families design accommodations to address health-related concerns such as managing frequent unpredictable absences and infectious illnesses of children and youth with SCD; exposure to temperature extremes or dehydration, which can trigger acute pain episodes; decreased stamina and mobility, which may affect sports or physical education; and urinary frequency or enuresis. Youth who have emotional or behavioral problems are referred for mental health interventions that sometimes include medication. SCD-PLANE staff have attended court hearings as well as school meetings to provide information relevant to authorities’ decision-making and to assist parents to advocate effectively for their children. SCD-PLANE findings also help hematology providers identify needs for further neuroimaging, changes in medication, or consultation from other medical specialists (eg, developmental pediatrics, neurology, speech and occupational therapy, otolaryngology, genetic counseling, or sleep medicine).

When the time comes to transition from pediatric to adult health care (in our program, usually around the age of 21), young adults with SCD and their parents benefit from a series of closing interviews, during which they review pertinent health and developmen-
tal issues, and a personal introduction to the adult clinic by a health care worker. In addition to drawing on national transition-planning resources such as the Health Ready to Work National Center (www.hrtw.org) and the National Center for Secondary Education and Transition (ncset.org), the SCD-PLANE connects families with local transition-planning support agencies appropriate to the needs of the individual adolescent.

RESULTS

Access and Utilization of Neuropsychological Services

In 2004, 2005, and 2006, before the SCD-PLANE began, ~4 children per year completed neuropsychological evaluations related to SCD after being referred by hematology or pediatrics providers to the SCDCC. We do not have precise data on the total number of patients with SCD referred under the traditional system; however, existing records suggest that <20% were successfully tested. Several children had been referred repeatedly, but never successfully completed neuropsychological testing, before the SCD-PLANE began in 2007. Two of the children who had been tested were seen again for SCD-PLANE evaluations, and we found that their parents had not understood how to follow-through with obtaining recommended services through their school system despite having received a written report and in-person feedback meeting.

In contrast to the “nearly nil” accrual rate before 2007, the SCD-PLANE successfully tested 81 children during 2007–2008. The missed-appointment rate in that first 18 months was 21%, but a new scheduling and reminder system has decreased the missed-appointment rate to 10% during the first 6 months of 2009. Most patients (70%) are covered by Medicaid; the remainder of them have private insurance. SCD is equally frequent in both genders; in our relatively small sample, 57% of the children tested to date were male. To date, 37% of the children screened were aged 5 to 8 years, 35% were aged 9 to 12 years, and 26% were aged 13 to 18 years. Most children screened (69%) had hemoglobin SS SCD; 18% had hemoglobin SC, and 13% had hemoglobin Sβ⁰-thalassemia. All were black, and many were recent immigrants from African and Caribbean countries.

Obstacles to Access

Ongoing obstacles to completing neuropsychological evaluations include insurance denials, scheduling issues, and missed clinic appointments. Although 70% of our SCD population receives state medical assistance on the basis of low income, it requires frequent renewal. Children whose Medicaid eligibility has lapsed may not receive neuropsychological testing under current hospital policy. Some private insurance providers refuse to cover neuropsychological testing unless the child has evidence of neurologic deterioration, and others do not cover any psychological services at this hospital. A few insurers require detailed preauthorization forms several weeks in advance, but often the clinic scheduling staff is not able to reach the family to obtain needed details that far in advance of the appointment. On a few occasions, we had to cancel neuropsychological evaluations on the comprehensive-clinic date because the patient turned out not to have current insurance; that is a frustrating, time-wasting situation for everyone.

Of course, some patients fall ill on the appointment date, and some forget (despite an automated voicemail reminder system), decline, or fail to keep the appointment for other reasons. The rate of failed appointments for neuropsychological testing has declined dramatically with implementation of the SCD-PLANE. Under the traditional model, our no-show rate for patients with SCD was ~50% (in 2006, eg, only 4 of 9 scheduled patients kept their appointments). In contrast, the 19% rate of missed SCD-PLANE appointments (steady in 2008 and 2009) is at the median compared with reported “no-show” rates from a study of 135 family medicine clinics. When this program began, we anticipated that parents might decline routine neuropsychological testing, because it adds another 1 to 2 hours to their comprehensive-clinic day; however, few families have objected. Of the first 100 families offered the opportunity to schedule neuropsychological testing during their child’s clinic visit, only 3 declined. Those 3 families were concerned that their child might experience diminished self-esteem or that the child might be either stigmatized or “coddled” in school if testing revealed learning problems. Other families were reassured by knowing that results of the testing, and any recommended interventions, would not be shared with the school or other outside parties unless parents chose to use that information to seek educational support services for their child. Now that SCD-PLANE staff are a familiar presence in the clinic, it is common for patients and parents who are not scheduled for neuropsychological evaluation to greet us and inquire whether it is “their turn” for assessment.

Neuropsychological Test Profiles

Data for the first 65 youth tested by the SCD-PLANE in 2007–2009 show that most of their neuropsychological test scores are in the average range compared with published age-standardized test norms (the quantitative details of neuropsychological
tests are available from Dr Wills on request). As a group, these children with SCD had age-appropriate scores on tests of simple vigilance, verbal repetition, vocabulary and verbal fluency (naming items within a meaningful category), spatial perception and reasoning about visual analogies, and sight-word reading fluency. On the other hand, these children scored lower on most tests of executive function, including working memory (eg, repeating a number series in reversed order), response inhibition (voluntarily holding back from making a well-practiced response), and set-shifting (switching from 1 response or problem-solving approach to a different one, in reaction to changing cues). Consistent with individually administered clinical tests, there were also significant problems with parent-rated and teacher-rated questionnaires about executive function in home, school, and community life (ie, the “project management” skills involved in planning, organizing, and effectively carrying out self-directed, goal-oriented work).

Slow processing speed (a slowed-down tempo regardless of accuracy) was evident in hesitant visual-to-verbal association when naming an array of pictured objects or colors; slow silent-reading fluency for sentences and verbal fluency for unfamiliar words; slow speed and low accuracy of recalling basic math facts, and slow decision speed and writing, on the Wechsler coding tasks. Graphomotor skills (eye-hand coordination for drawing, copying, or handwriting) were scored below average even in an untimed format when copying geometric designs on the Visual-Motor Integration Test. Finally, parent-rated internalizing behaviors (ie, indicators of anxious or depressed mood) were significantly more common in this sample of children with SCD than in the general population of same-age, same-gender schoolchildren.

Identification and Referral Rates

Table 1 summarizes the rates of problem identification and referrals among 65 school-aged children seen in 2007–2009 who completed full neuropsychological testing including parent and teacher behavioral reports and completed follow-up visits to determine whether recommendations were accepted and implemented. Among these youth, 54% required psychosocial or educational interventions for attention, learning, executive function, or emotional-behavioral issues. The need for intervention was identified when a child scored ≥1.5 SDs below the published population norm on at least 2 measures of a particular area of functioning. Some children were referred for tutoring, speech therapy, or behavioral therapy, for specific problems. Some of them qualified for special-education programs in school. Special-education support services were recommended for those children who met Minnesota’s state criteria for educationally significant disabilities in learning, attention, or behavior. On the other hand, we identified 1 child as qualifying for an accelerated (“gifted”) program in school.

Of 35 children for whom we identified a need for educational support (ie, tutoring, Title I, accommodations in the regular classroom, or special-education programming), only 11 were already enrolled in appropriate school programming. For 17 children who were not receiving services, we met with parents and school personnel to develop plans to initiate or significantly extend special-education services. Parents of the remaining 7 children initially declined educational support services because they felt that their child either did not need such help or might be stigmatized by being identified as having special-education needs. Ultimately, 2 of these families pursued recommended educational support services after subsequent testing (at the following year’s comprehensive clinic) confirmed the child’s learning or behavioral problems. These measures show that a high number of patients with SCD (19 of 35) had been underscreened, unidentified, or underserved in the

<table>
<thead>
<tr>
<th>TABLE 1 Recommendations and Follow-through Based on the SCD-PLANE Neuropsychological Testing Results</th>
<th>n*</th>
<th>% of sample</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Educational referrals</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No special education recommended</td>
<td>30</td>
<td>46</td>
</tr>
<tr>
<td>Special education recommended</td>
<td>35</td>
<td>54</td>
</tr>
<tr>
<td>Actions taken at 3-mo follow-up:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Reviewed extant IEP or 504 plan, no change*</td>
<td>(11)</td>
<td>—</td>
</tr>
<tr>
<td>School conference to discuss IEP or 504 plan changes</td>
<td>(13)</td>
<td>—</td>
</tr>
<tr>
<td>Initiated new IEP or 504 plan</td>
<td>(4)</td>
<td>—</td>
</tr>
<tr>
<td>Parent declined recommended IEP or 504 plan</td>
<td>(7)</td>
<td>—</td>
</tr>
<tr>
<td><strong>Behavioral health referrals</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No behavioral referral recommended</td>
<td>39</td>
<td>60</td>
</tr>
<tr>
<td>Behavioral referral recommended</td>
<td>26</td>
<td>40</td>
</tr>
<tr>
<td>Actions taken at 3-mo follow-up:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Continue existing intervention</td>
<td>(8)</td>
<td>—</td>
</tr>
<tr>
<td>Initiated new intervention</td>
<td>(6)</td>
<td>—</td>
</tr>
<tr>
<td>Parent declined recommendation</td>
<td>(12)</td>
<td>—</td>
</tr>
</tbody>
</table>

IEP indicates individual education plan.

* Numbers in parentheses represent subtotals, broken down into specified categories, of the total number of cases referred for special education or total behavioral referrals.

* A 504 plan, required in Section 504 of the Rehabilitation Act of 1973, describes aids and services that meet the needs of a student with a disability as adequately as the needs of nondisabled students are met.
schools despite strong parental interest in seeking and receiving educational support services for an academically or behaviorally struggling child. Testing identified significant behavioral concerns (including oppositional-defiant, hyperactive-inattentive, anxious-depressed, or socially withdrawn behaviors) in 40% of tested children on the basis of parent or teacher behavioral rating questionnaires. Scores of >1.5 SDs from the population norms were considered to indicate behavioral concerns. Children’s problems with internalizing symptoms (unhappiness, loneliness, worrying) were consistent with reported findings of large multicenter studies. Children were referred for psychotherapy, behavior therapy, consideration of psychotropic medications, or some combination of these treatments. Although approximately half (54%) of the referred children followed through and received recommended psychological/behavioral interventions, the other half declined those services. Parents of 2 children expressed disagreement with and resentment about being referred, although 1 of those parents subsequently changed her mind and requested a therapist for her child. To date, only 1 parent has expressed sustained dissatisfaction (on the basis of a feeling that we had impugned her parenting skills by reporting that the child had “severe” behavior problems).

Recommended Interventions

Neuropsychological reports based on SCD-PLANE testing attempt to provide specific recommendations tailored to the needs of the individual child or adolescent; however, no research to date has addressed treatment of neuropsychological problems for people with SCD. We based our recommendations for the young people with these issues and their families on studies of other patient populations with similar neuropsychological difficulties, such as people with hydrocephalus, mild traumatic brain injuries, or attention-deficit/hyperactivity disorder. For example, suggestions in the Dawson and Guare book Smart but Scattered were relevant and helpful to many of our patients.

Recommendations vary, of course, depending on the nature and severity of cognitive, behavioral, or emotional needs. For example, youth with mild SCD-related neuropsychological disabilities may require academic accommodations in high school and college because of the increased demands for complex self-directed work, even if they had no apparent difficulties in elementary grades. We commonly recommend accommodations including extended time to complete tests; use of assistive technology such as voice recorders or computerized voice dictation, to reduce the stress of multitasking when taking notes and minimize the pressure of long written reports; and extra coaching to manage school projects.

Adolescents with moderate neurodevelopmental impairments may be challenged by learning to drive, managing finances, or multitasking at work. They often need a job coach, academic tutor, or extensive individualized accommodations at work or school. To develop adequate self-advocacy skills, they may require ongoing involvement from parents and other adults and adjustment of parental expectations for independent functioning. Understanding the young person’s neuropsychological profile, and knowing how to access relevant resources can assist many families in planning for adult health care needs including health literacy and self-advocacy, as well as postsecondary educational, vocational, leisure, legal, and financial matters. The SCD-PLANE encourages “person-centered planning” (see www.ncset.org for a description), which focuses on the young person’s interests and goals yet is also informed by identifying talents and deficiencies in learning or adaptive skills via interviews, emotional-behavioral ratings, and neuropsychological test scores.

Those with severe intellectual or learning disabilities may benefit from extended school programming (available through the age of 21 years), college programs with added student services, vocational rehabilitation programs, or some combination of them. The SCD-PLANE assists families with securing help with issues of postsecondary education, guardianship, finances, and housing through agencies such as the Client Assistance Project for legal rights of persons with chronic health conditions (for state listings, visit the National Disability Rights Network at www.napas.org).

LIMITATIONS

The major limitations or obstacles to success of the SCD-PLANE are the “3 F’s”: funding; follow-through; and fixing the problems. Funding is a problem because medical insurance does not fully cover the costs of this program. In particular, ongoing philanthropic funding is needed to support the hospital-school liaison aspect of the SCD-PLANE and the ad hoc (unscheduled) consultations by the neuropsychologist. Follow-through is much better in the SCD-PLANE than under the traditional model, because families stay more connected via multiple appointments with the SCDCD versus 1-time neuropsychology testing. Follow-through remains problematic, however, because so many families in our clinic face severe economic and social challenges (~70% receive state medical assistance). For example, women who single-parent a child with chronic illness while working or attending...
school can find it nearly impossible to take that child to weekly therapy appointments, and families with limited education or English proficiency may find it daunting to meet with school personnel to help develop an effective education plan. Addresses and telephone numbers change often, and so do children’s schools, therapists, and primary care providers. Programs established in 1 location may not transfer successfully to a new setting.

Furthermore, patient follow-through related to newly identified issues of behavioral or emotional health is markedly poorer than follow-through for newly identified educational issues. Parents typically have been eager to access school-based or agency-based educational services when SCD-PLANE testing identifies learning or attention difficulties. As noted earlier, many of these patients’ needs had not been identified or were underserved within their schools, consistent with results of research that showed that many students with learning disabilities related to SCD are underidentified.15 In our clinic, mental or behavioral health needs also are underidentified and underserved, but families are less likely to accept professional services. Indeed, only 9% of children with newly identified mental health needs accessed services such as behavioral therapy or psychiatric consultation. Several families, however, accepted referrals after problems were repeatedly identified over 1 or 2 subsequent annual SCD-PLANE visits. This experience illustrates that for mental or behavioral health issues in this population, 1-time screening may not be enough. Repeated annual testing builds trust, reveals problems that are persistent rather than transitory, and (sometimes) convinces parents of the need to intervene.

Finally, although we have good tools for identifying and describing neuropsychological problems, we lack satisfactory solutions to “fix the problems” that many patients present. Currently, the best that clinicians can do is refer children and youth for standard educational, vocational rehabilitation, or mental health services and confirm that they successfully access those services to ensure that their needs are addressed rather than neglected. We do not know whether the services currently available to these children are appropriate or sufficient to meet their needs, although we suspect this “status quo” is less than optimal. SCD-specific research is needed to provide evidence-based education, rehabilitation, and health care programming tailored to the needs of youth with SCD. Specifically, neuropsychologists need evidence-based information to prescribe strategies that will enhance happiness and self-reliance in adolescents with SCD as they transition to adulthood and will improve or compensate for problems with executive function and processing speed.

CONCLUSIONS

Children and youth with SCD face the double disadvantage of having a chronic illness and, in most cases, belonging to underserved racial and socioeconomic groups.20 The SCD-PLANE addresses the serious neuropsychological consequences of SCD and the sociocultural issues that impede care for these youth and helps prepare them for a smoother transition to adulthood. Compared with the traditional model of referring patients to a neuropsychology service, the SCD-PLANE has yielded many advantages for the hospital, as well as for families. It provides greater convenience, economy, and efficiency of access to neuropsychological services. It has been well accepted by patients’ families. We strive continually to create a model of care that is holistic, respectful, developmentally informed, relationship-based, family-centered, community-connected, and culturally competent.

We will monitor, as best we can in a clinical context, whether SCD-PLANE testing, identification, referral, and follow-through services effectively improve long-term physical, educational, or mental health outcomes for our patients.

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Transition Planning for Youth With Sickle Cell Disease: Embedding Neuropsychological Assessment Into Comprehensive Care
Karen E. Wills, Stephen C. Nelson, Jane Hennessy, M. Osita Nwaneri, Joyce Miskowiec, Elizabeth McDonough and Kristin Moquist

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