Transition and Sickle Cell Disease

This is the 10th article in our series, “Transitions to Adult Care.”

abstract

Sickle cell disease (SCD), the most common genetic disease screened for in the newborn period, occurs in ~1 in 2400 newborns in the general population and 1 in 400 individuals of African descent in the United States. Despite the relative high prevalence and low pediatric mortality rate of SCD when compared with other genetic diseases or chronic diseases in pediatrics, few evidence-based guidelines have been developed to facilitate the transition from pediatrics to an internal medicine or family practice environment. As with any pediatric transition program, common educational, social, and health systems themes exist to prepare for the next phase of health care; however, unique features characterizing the experience of adolescents with SCD must also be addressed. These challenges include, but are not limited to, a higher proportion of SCD adolescents receiving public health insurance when compared with any other pediatric genetic or chronic diseases; the high proportion of overt strokes or silent cerebral infarcts (~30%) affecting cognition; risk of low high school graduation; and a high rate of comorbid disease, including asthma. Young adults with SCD are living longer; consequently, the importance of transitioning from a pediatric primary care provider to adult primary care physician has become a critical step in the health care management plan. We identify how the primary care physicians in tandem with the pediatric specialist can enhance transition interventions for children and adolescents with SCD. Pediatrics 2012;130:926–935

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KEY WORDS
transition, adolescent, young adult sickle cell

ABBREVIATIONS
AAP—American Academy of Pediatrics
CCM—chronic condition management
SCA—sickle cell anemia
SCD—sickle cell disease

www.pediatrics.org/cgi/doi/10.1542/peds.2011-3049
doi:10.1542/peds.2011-3049

Accepted for publication Jul 3, 2012

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PEDIATRICS (ISSN Numbers: Print, 0031-4005; Online, 1098-4275).
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FINANCIAL DISCLOSURE: The authors have indicated they have no financial relationships relevant to this article to disclose.

FUNDING: Funded by the Burroughs Wellcome Foundation and the HRSA SU38MC22220-02-00.
In the United States, the prevalence of sickle cell disease (SCD) is estimated to be ∼70,000 to 100,000.1 The life span for children with SCD has increased dramatically in the past 50 years.2 No single scientific advancement has contributed to the increase in survival; however, several noticeable improvements greatly increased the survival rate of infants and children with SCD. Among the key advancements, daily penicillin prophylaxis started shortly after birth3 and continued up to at least 5 years of age, combined with the use of conjugated and unconjugated pneumococcal4–6 and Haemophilus influenzae vaccination,7 have greatly decreased the rate of infection related mortality and morbidity in children with SCD. Consequently, children with SCD are expected to survive to adulthood with eventual referral to adult primary care providers. This new emerging young adult population will require the standard medical care offered by the adult hematologist or specialist in SCD, coupled with routine and preventive medical measures provided by primary health care teams.

The reluctance of pediatric providers and adolescents with SCD to transfer to adult primary care and specialty care (eg, hematology) providers has been based on several factors, including the perception, with some factual basis, that the transfer may result in an increase in morbidity and mortality. Brousseau et al provided an indirect measure of increased morbidity in the age group most affected by transfer of care, 18- to 30-year-olds.8 Using a large retrospective study design from the 2005 and 2006 Healthcare Cost and Utilization Project State Inpatient Databases and State Emergency Department Databases, the investigators showed that the highest rate of acute care encounters and rehospitalizations was in the 18- to 30-year-olds,8 and not the older group as would be expected because of increasing disease morbidity with age.9 Additionally, in a single-center longitudinal cohort of children with SCD with 940 participants followed for 8857 patient-years, Quinn et al demonstrated the majority of deaths occurred after 18 years of age and after transfer to an adult provider (85%, 6 of 7).2 The mean time to death after transfer to adult provider was 1.8 years (median, 1.2 years; range, 0.2–5.3 years).2 Taken together, these data support the reluctance of pediatric providers, adolescents, and their parents to transferring young adults to adult providers. Stronger data are required to determine whether recent transfer of a young adult from a pediatric provider to an adult provider is associated with an increased SCD related morbidity and mortality. This review includes the best available evidence, coupled with expert opinion about key elements of a transition program for adolescents with SCD addressing their underlying disease process and their emotional and socio-cultural development.

**UNIQUE FEATURES OF YOUNG ADULTS WITH SCD**

Historically, areas of the world with a high prevalence of SCD include countries that border the Mediterranean Sea, sub-Saharan Africa, the Middle East, and select portions of India. Secondary to migration (both forced and unforced), SCD is also prominent in North America, South America, and the Caribbean. In the United States, the unique social and demographic profiles of individuals with SCD are unlike any individuals with other chronic diseases; most individuals with SCD are of African descent. In addition, on average, 60% of the pediatric SCD population has public health insurance (Medicaid).8 The combination of being both black and poor for the majority of children with SCD in the United States presents a distinct set of challenges when the goal is to transition the young adult from a pediatric to an adult health care provider. No specific data exist regarding the proportion of uninsured young adults with SCD. However, in 2008, the National Health Interview Survey demonstrated that the highest uninsured group when compared with any other age group were young adults aged 18 to 24 years at 29%.10 On the basis of the high rate of children with SCD receiving Medicaid, we can only postulate that the uninsured rate among young adults with SCD that require transitioning to an adult health care provider is at least 29%. The absence of health insurance is a major concern for adolescents with SCD. Telfair et al note that young adults with SCD, when preparing for transition, are concerned about health insurance status before or after transfer.11 Given the importance of insurance status to health care access, developing a plan for anticipated health insurance when adolescents with SCD are no longer eligible due to age is a critical part of the transition process.

Access to specialty care for children with SCD has not been systematically evaluated. For the general pediatric population, evidence strongly suggests that the type of health insurance, public versus private, greatly influences access to specialty care. Specifically, in a prospective study, 273 pediatric specialty clinics were called twice for an appointment with a pediatric specialist.12 The only difference between the 2 calls was insurance status, public (Medicaid) versus private (Blue Cross Blue Shield). A significant difference in access to pediatric subspecialty care existed based on the insurance status. When Medicaid was mentioned as the insurer, 66% were denied an appointment, compared with 11% of privately insured callers. In addition, the wait time for an actual appointment was 22 days longer when Medicaid was stated.
Unfortunately, no corresponding data exist for young adults with SCD. However, given the high proportion of children with SCD on Medicaid, we postulate that access to specialty care for young adults will be equally challenging. In summary, before transferring adolescents with SCD to an adult provider, identifying a network of adult primary and specialty care providers that will offer health services for adolescents with their current or anticipated health insurance is a fundamental requirement in any SCD transition program.

Another distinctive attribute specific to SCD, is the high prevalence of cognitive deficits that will have an impact on educational attainment and employment. Overt strokes still occur in children with SCD and are a major cause of cognitive impairment and ongoing cerebral injury even when receiving regular blood transfusion therapy.13 Upon transfer to adult providers, individuals with strokes should continue to receive blood transfusion therapy. Despite not completely preventing progression of cerebral infarcts, cessation of the transfusion is associated with increased rate of overt strokes.14 and most likely is associated with ongoing silent cerebral infarcts. Although no randomized clinical trial has proven the added benefit of continuing blood transfusion therapy for adults that had strokes in childhood, all evidence suggest that once an ischemic stroke has occurred, ongoing cerebral injury may be progressive,15 particularly in individuals with moyamoya, a severe form of cerebral vasculopathy.15 If transfusions are to be stopped after a stroke, a clear benefit-to-risk ratio must be discussed with the young adult, including an increased risk of a new overt or silent stroke. In some cases, young adults with strokes may elect to stop the transfusion therapy and pursue hydroxyurea as an alternative for secondary stroke prevention. Results from a phase III clinical trial indicated that regular blood transfusion therapy was superior to hydroxyurea for secondary prevention of strokes.16 Specifically, in the trial none (0 of 68) of the participants who were randomly allocated to blood had a second stroke, when compared with the hydroxyurea group in which 10% (7 of 67) had second strokes.16 Thus, if the young adult elects to change from regular blood transfusion therapy to hydroxyurea therapy for secondary stroke prevention, the increased absolute and relative risk of this change in therapy must be explicitly discussed and acknowledged. If the young adult elects to change from regular blood transfusion therapy to hydroxyurea therapy for secondary stroke prevention, the increased absolute and relative risk must be explicitly discussed and acknowledged, along with the fact that the use of hydroxyurea would be considered departure from standard care.

The rate of overt strokes in children with sickle cell anemia (SCA) has dropped ~10-fold with the routine use of transcranial Doppler assessment to identify children with SCD with elevated measurements, coupled with regular blood transfusion17; silent cerebral infarcts have become the most common cause of neurologic injury. Unfortunately, children with silent cerebral infarcts will not have any findings on physical examination18 and can only be detected with a magnetic resonance image of the brain.18 In a single-center study, Bernaudin et al demonstrated that at least 35% of the patients with hemoglobin SS will have a silent cerebral infarct by their 14th birthday.18 These data are consistent with a multicenter trial in which the prevalence of silent cerebral infarct was 31% with an average age of enrollment of ~9 years of age.20 The high prevalence and associated cognitive morbidity associated with silent cerebral infarcts highlights the importance of knowing the infarct status in young adults before transition from pediatric to adult health care providers.21 Regardless of the presence of cerebral infarcts, all students and young adults with SCD are at considerable risk for decreased cognitive functioning when compared with ethnic-matched control subjects. Vichinsky et al evaluated adults with SCA who did not have any evidence of overt strokes and demonstrated that this group had lower global IQ scores when compared with age-, ethnic-, and gender-matched controls.22 Summarizing the rigorous cognitive studies in children with SCA, Hogan et al demonstrated a consistent gradient in global IQ of students with SCD compared with age- and ethnic-matched controls.23 Siblings without SCD controls have a higher global IQ compared with siblings with SCD but without silent cerebral infarcts. Individuals with silent cerebral infarcts had a higher IQ than individuals with overt strokes. Furthermore, Schatz et al demonstrated that students with SCD and with silent cerebral infarcts had a much higher rate of grade failure or requirement for special classroom support compared with sibling controls (30% vs 6%).24 Factors that may contribute to lower cognitive performance in students with SCD include, but are not limited to, the higher prevalence of depression compared with controls,25 a lower baseline hemoglobin level26 compared with individuals with SCD, and medical complications such as vaso-occlusive crises that may contribute to an increased rate of school absenteeism.27 In one small single-institution study, students with SCD who had at least 3 hospitalizations for vaso-occlusive pain episodes missed an average of 38.4 school days in the academic year.27 Regardless of the etiology, an inventory of the educational attainment of adolescents with SCD, at the least, should be completed before
transition. For patients with significant school problems, overt strokes, or silent cerebral infarcts, a cognitive battery may be indicated before transfer to the adult team. At least 1 SCD center has successfully integrated cognitive assessment as part of its transition program. Without such an educational attainment assessment, the young adult could be placed in an unfortunate position in which chronological age may not match cognitive abilities, resulting in failure to comply with expected self-management strategies.

**TRANSITION TO ADULT CARE**

A critical, but often missing component of current research done on transition in SCD is the perspective of the primary health care provider team. Although not commonly recognized, primary care teams, not hematologists, manage the bulk of the children with SCD. Recent recommendations for SCD transition programs are based on expert opinion or single-center nonrandomized studies and not the perspective of the primary health care team. Before more definitive clinical or comparative effectiveness trials are implemented for transition in SCD, better relationships must be forged between the primary care physicians in pediatrics and internal medicine or family practice with practical strategies for primary care providers. For the foreseeable future, an ongoing effort will be required to educate a new cadre of adult primary health care providers, including nurse practitioners and physician assistants about the medical care of young adults with SCD, as well as other chronic diseases such as cystic fibrosis and congenital heart disease, that were initially believed to be the exclusive domain of pediatrics.

For most individuals with SCD, having the condition is a part of their life, despite common perception of SCD being a chronic, debilitating disease. In developing the appropriate context for a transitional care program for adolescents and young adults with SCD, there exists a limited but growing body of literature from the policy and research on youth with special health care needs, reports from work supported by federal funders such as the US Department of Health and Human Services Health Resources and Services Administration have been recently made available. Key findings from this literature make clear that consideration must be given to (1) how adolescents and their family view the transfer of the care, (2) how physicians and other health care providers view the transfer of care, (3) social and cultural context of transition care, and (4) strategies needed to improve the system for care (quality). Evidence from empirical studies is critically important in developing a comprehensive program of transition.

By definition, transition is a multifaceted, vibrant process initiated with a child- and parent-focused orientation and ending with an adult-focused health orientation. Health transition coincides and is expected to facilitate transition in other areas as well (eg, work, community, the school). Transition proceeds at different rates for different individuals and families (and programs). A transition program for adolescents should include not only physical transfer of medical care from one facility and provider to another, but also address the needs of the developing adolescent, such as completion of high school or vocational education and strides toward becoming fiscally independent.

Despite recognizing the importance of transition as a continuum of health care for adolescents with SCD, a paucity of established effective strategies exists. In a recent study, McPherson et al found that adolescents reported an inadequate level of preparation for transition to adult care; the readiness increases with age but was still considered less than optimal. In another study, Telfair et al found that most providers agree a transition program is necessary; however, only a few providers did anything to demonstrate their involvement in the transition process. When asked, adolescents and adults indicate a range of issues and concerns about transitioning to adult care and settings. The most common concerns were (1) fear of leaving the health care provider with whom they were already familiar; (2) fear that adult care providers might not understand their needs; (3) fear of being treated as adults, and (4) concerns about payment for the cost of care. Ultimately, a successful SCD adolescent transition program will require an open discussion about the anxiety and concerns that the adolescent may have about transferring their care. These discussions should begin at 12 years of age and continue annually with clear benchmarks describing the annual incremental progress toward transition well before the anticipated transfer date.

Exceptional opportunities exist to develop evidence-based transition strategies in SCD. Given the unique population demographics of SCD, coupled with the high prevalence of comorbidities, core principles of a successful transition program in SCD will likely be generalizable to other transition programs for young adults with chronic diseases. Understanding the perspective of the young adult, and not the parent as a spokesperson for the adolescent, will be a necessary next iteration of the SCD transition research. Equally important is understanding the perspective of the parents and the pediatric provider so that both real and false perceptions may be addressed before, during, and after the transition process.

Transition to adult care for adolescents and young adults with SCD must be considered within the larger (holistic) context of the life of the youth (Fig 1).
should be coordinated on macro- and meso-levels, with the youth and adolescents remaining the core focus of the effort. The transition process should include an open invitation to address patients', caregivers', and providers' concerns or questions regarding the transition process. Reiss and Gibson proposed 5 recommendations to facilitate transition: (1) have an orientation that is future focused and flexible, (2) anticipate change and develop a flexible plan for the future, (3) foster personal and medical independence and creative problem solving, (4) anticipate future needs by developing personal life maps, and (5) celebrate transitions as they occur with graduation ceremonies, certificates of completion, and other rites of passage.

MEDICAL GUIDELINES FOR TRANSFER OF CARE

Evidence-based guidelines for the appropriate completed diagnostic studies before transferring the medical care to an adult health care provider have not been established. As noted earlier, young adults with SCD have a high prevalence of other comorbidities, such as asthma, avascular necrosis of the long bones, restrictive lung disease, retinopathy, cardiac dysfunction, and renal dysfunction. Based primarily on the high prevalence of comorbid conditions in adults with SCD, we believe a series of medical evaluations should be performed before the transition. A complete evaluation will allow both the young adult with SCD and the new physician to start with full knowledge of past, current, and potential future health problems. Coordination of these medical evaluations before transfer of the patient is strongly encouraged so that replication of diagnostic studies will not be done shortly after the referral. If possible such medical evaluations would preferably be completed at the medical facility where ongoing adult care will be delivered to establish an appropriate baseline for longitudinal evaluations. A secondary benefit is to familiarize young adults with the new medical system. We recommend that a comanagement strategy occur between the referring pediatric hematologist, primary care provider, and adult primary care physician at least 1 year before the referral; therefore, the diagnostic studies logistics will be scheduled. If an adult hematologist is available, we also recommend that care is also coordinated with both the adult primary care provider and adult hematologist. Unfortunately, the vast majority of locations in the country do not have adult hematologists willing to provide medical care for adults with sickle cell disease. Thus, transition planning for young adults, at minimum, must include a primary care team. The laboratory evaluation that could be considered when transferring the medical care from the pediatric facility to the adult facility is based on evidence of ongoing end-organ injury with age. Specifically, we recommend the evaluations shown in Table 1 occur before transfer because evidence of end-organ disease may change the management of the patient upon transfer.

NAVIGATING THE HEALTH CARE SYSTEM FOR TRANSITION CARE: A NEW ROADMAP

In the absence of proven and well-established benchmarks for transition programs focused on SCD, practice variation is wide, and consensus and expert opinion is the most reasonable starting point. In 2011, the American Academy of Pediatrics (AAP), the American Academy of Family Physicians, and the American College of Physicians–American Society of Internal Medicine, released a clinical report, Supporting the Health Care Transition From Adolescence to Adulthood in the Medical Home.
The concept of the medical home, which delivers family-centered care throughout the life span, has become a cornerstone of primary care delivery in pediatrics. The medical home includes preventive care, acute illness management, and chronic condition management (CCM). The consensus group developed an algorithm for transition planning to start at 12 years of age and continue up to the point of transition, 18 to 21 years (Fig 2). Most important, for children with special health care needs that require CCM, a distinct branch was created in the algorithm.

On the basis of the Consensus Statement for children that require CCM, we have created a modified version for children with SCD. Given the high prevalence of overt and silent cerebral infarcts in SCD, an active and early decision must be made between the parent and the providers about the ability of the youth to eventually become independent. If the patient is deemed not likely to become independent, the appropriate arrangements must be undertaken for a Power of Attorney to be secured before his or her 18th birthday. Another recommendation, consistent with the CCM, is for a written care plan for management of pain for the patient, family, and the provider. For adolescents with SCD, clear and written expectations regarding the management of pain should be considered as standard care and part of the transfer packet to the adult primary care physicians. Although not proven, these strategies should dramatically decrease the perceived adolescent bias concerning issues of pain management and differences between pediatric and adult providers, along with the different paradigm of care for family members.

Given the complexity of the medical care for individuals with SCD, with a heavy emphasis on age-specific primary and secondary preventive strategies, coupled with the consensus recommendation for transitional activities, a registry or database for ongoing assessment of the milestones should be strongly considered. Similar strategies have already been introduced in the complex management of children with cancer in which a therapeutic roadmap is standard care. The local guidelines for transition must take into consideration the hospital policy for management of adults >21 years of age, teenage mothers, adolescents who have been incarcerated, and adults with significant cognitive delay. Unfortunately, no consensus metrics have been developed and validated for the transition of SCD care from a pediatric to an adult health care team. On the basis of our standard care, we have elected to start transition care at 12 years of age with the medical, social, and educational roadmap shown in Table 2.
Medical care plans with coordinated care have become another standard in the medical home. As is the case with asthma action plans, SCD care plans, in part, should be directed toward active management for the patient and family. Although efficacy has not been demonstrated for SCD, we recommend a written care plan based on the World Health Organization stepwise approach for increasing intensity of pain corresponding with increasing strength of pain medication. For individuals with SCD, a pain action plan uses clear written instructions on previously agreed-on management (both medicinal and nonmedicinal based) for increasing intensities of vaso-occlusive pain. The pain action plan is recorded in the electronic medical record and available to the emergency department staff when reviewed.

A novel concept endorsed by the AAP Consensus Statement was that CCM visits should be focused on the transition

TABLE 2 Medical, Social, and Education Milestones

Medical milestones: should be assessed annually and should be done before transfer to an adult health care provider; requires some coordination with specialty and primary health care teams.

1. Ophthalmology evaluation to assess retinopathy and other ophthalmic complications
2. Dental evaluation to evaluate dental hygiene
3. Pulmonary function tests to include spirometry and lung volumes
4. Cardiac evaluation for assessment of elevated tricuspid regurgitant velocity and measurement of brain natriuretic protein to assess pulmonary hypertension
5. Urine albumin/creatinine ratio to assess microalbuminuria and sickle cell disease nephropathy
6. Magnetic resonance of the head to assess for silent cerebral infarct and, if indicated, cognitive testing
7. Level of education achieved (GED, high school, college, etc)
8. Immunization status
9. Assessment of partner’s trait status (if applicable)
10. Assessment of knowledge on safe-sex practices, STDs, and pregnancy
11. Assessment of ability to make future independent decisions as a young adult; if deemed not likely, then conversations should begin regarding conservatorship or power of attorney by 18 y of age

Social milestones: should be addressed sequentially and require action on the part of the adolescent

1. Ability to conduct a health care visit with the guardian in the room but with the majority of the interview, assessment, and plan directed toward the patient
2. Documentation that instructions for medical care, such as taking medication or making a medical appointment, are accomplished initially with parental support and subsequently without parent support
3. Ability to conduct part or the entire health care visit without the guardian in the room, with assessment and plan directed toward the patient and subsequently validation of the assessment and plan with the guardian who accompanied the minor to the visit
4. Ability to conduct a health care visit without the guardian at the health care facility, but adolescent must be of legal age
5. Ability to make appointments and follow through on self-management medical care plan (appointments, prescription refills, etc) without dependence on guardian
6. Visit to adult health care team office before transition
7. Ability to seek out and obtain acute medical care independently or with minimum supervision by guardian
8. Ability to determine health insurance status, before transition, with knowledge about benefits and copays (may require visit to financial administrator)

Education milestones

1. Document knowledge about the following:
   - SCD phenotype
   - SCD pain plan including how to take long- and short-acting opioids
   - Ability to manage pain according to plan
   - Preventive measures for SCD complications
     - Pain (extreme climate conditions, environmental tobacco smoke, other self-activators)
     - Fever; importance of seeking out prompt medical attention
   - Symptoms of stroke
2. Knowledge of current primary care health team
3. Documentation that instructions for medical care, such as taking medication or making a medical appointment, are accomplished initially with parental support and subsequently without parent support
4. Ability to describe current medications, allergies to medications, or medications that have not proven effective for patient’s intended consequence
5. Ability to articulate most important components of medical history including, but not limited to, surgeries, life-threatening medical complications, and knowledge about challenges with blood transfusion therapy
6. Ability to seek out medical care with minimal parental supervision
7. Knowledge of future adult primary health care team
8. If considering offspring, awareness of partner’s hemoglobinopathy trait status
9. If partner has been tested for hemoglobinopathy trait status
10. If partner has been tested for hemoglobinopathy trait status
11. If partner has been tested for hemoglobinopathy trait status

The milestones in this table should be considered as part of the transition planning that starts at age 12 years for all children with SCD and continued annually until completion of transition to an adult health care provider. The progress with each milestone is independent of age and primarily dependent on ability of the adolescent. Assessments of the milestones are best handled in annual transition visits. GED, general educational development test; STD, sexually transmitted disease.

a Clinical utility of tricuspid regurgitant velocity has not been established in pediatrics and if performed should be done in conjunction with adult hematologists or adult primary care provider with a clear plan as to how the results may alter therapy.

b Recommendation is specifically for individuals with hemoglobin sickle cell anemia (most severe). Less evidence supports MRI of the head for young adults with hemoglobin sickle cell disease (less severe) or hemoglobin sickle cell beta thalassemia, and a case-by-case decision should be made as to whether a MRI of the head should be obtained before transfer to the adult provider.
and the anticipated steps that must be undertaken by both the patient and the health care team. Among the steps for health care providers is an explicit approach for comanagement between the current pediatric primary health care team, current pediatric specialist care, and the corresponding adult providers. Establishing these comanagement teams to facilitate transition is not customary in the management of SCD, but is clearly endorsed by the Consensus Statement and will require considerable effort and perhaps event trial and error before the process becomes smooth. Until more data are available to facilitate transition, the Consensus Statement provides a reasonable starting place to guide the primary care physician and specialist on how to establish or enhance their transition program for SCD.

One of the biggest challenges in coordinating care is the reimbursement for this time-intensive process. As per the AAP Consensus Statement, payment of services related to chronic care management is feasible. For transition planning providers can schedule specific visits and bill the visit with the Current Procedural Terminology codes of 99214 or 99215 if >50% of the visit is counseling. On the basis of these AAP guidelines, along with the milestones mentioned in Table 2, our multidisciplinary team has elected to schedule annual visits focused solely on addressing the transition milestones. Also, per the AAP consensus guidelines, coordination of the transition care, referred to as care plan oversight, can be billed. More specifically, this effort may include calls to adult primary care providers and even conversations with the adolescent and family to coordinate before, during, and after the transition. These activities can be billed using Current Procedural Terminology codes 99374 (15–29 minutes) or 99375 for monthly cumulative time spent on care plan oversight. The ability to improve transition of care with scheduled coordinated care visits must be coupled with effective reimbursement strategies.

**SUMMARY**

Recent advances in the medical management of SCD have resulted in a new and growing population of young adults. Despite the increasing population, few, if any, evidence-based strategies exist for transitioning an adolescent with SCD from a pediatric provider to an adult provider. In the United States, most children and young adults with SCD will receive their medical care from primary care providers including physician assistants and nurse practitioners, not hematologists. A focus on the medical home model with a primary care team when transitioning the young adult patient with SCD is not only practical but, in many cases, the only option in the region. Based primarily on best practices, transition for young adults with SCD should be a gradual process with a flexible timeline of goals in partnership with the patient, family, and future primary care provider. The process requires a starting point with the ultimate goal of completing the transfer of care. Benchmarks for each stage should be clearly identified so that they can be tracked as part of the medical record. In the absence of a validated and generalizable tool specific for SCD, some assessment of a baseline and ongoing individual assessment of the adolescent, caregiver’s, and provider’s needs and level of readiness to transition must be a core component of the transition program. Annual discussions of transition should be starting by 12 years of age with ongoing patient, family, and caregiver education about the disease and navigating the medical system. Given the disproportionate number of individuals with SCD receiving public insurance and requiring subspecialty care, identification of not only the adult primary care provider but adult specialists who are willing to provide services, regardless of the type of insurance, is a necessary and critical step in transition. Federal support for clinical trials and comparative effectiveness research is needed if we are to deliver the best and most efficient transition care to this vulnerable population.

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MODELS FOR THE REAL WORLD: The other night, I was in my hotel room flipping through one of the free magazines, and noticed they seemed to consist mainly of advertisements for local and national clothing designers. The models were all quite young and wore expensive clothes. However, rather than thinking about the clothes themselves, I could not escape the fact that none of the women looked anything like most of the women I know. After all, how many 18-year-old, 5 feet 9 inch tall, 115-pound women are hauling kids to school, working at the hospital, watching children play soccer, or in hotels attending business meetings? While some modeling agencies have adopted age and weight guidelines for their models, most models are quite young and thin. As reported in CNN (Living: July 28, 2012), however, the tide may finally be changing—but not because of rules. A few designers and fashion labels have opted to feature women who better represent the population. One label recently selected 11 individuals of various ages—not associated with the fashion or modeling world—to represent their line. While sometimes seen in local newspapers, the use of ordinary (and older) people to showcase clothes has been rare in the fashion world. Some model selections by designer labels have been quite dramatic. The designers say they want people who are interesting beyond their clothes. For example, one designer chose a 60-year-old woman to be the face of a new campaign, while another chose an infant with Down syndrome to represent a new kids line. Furthermore, some designers have realized that clothes have to look good on someone other than an archetypal model. A message that beauty can be associated with a few wrinkles resonates with readers. As for me, the use of people of all sizes, colors, and ages to model clothes is a welcome change. Maybe now, I won’t be thinking how silly the clothes or person looks, but how interesting both are—and may be more inclined to purchase something.
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*Pediatrics* 2012;130;926; originally published online October 1, 2012;
DOI: 10.1542/peds.2011-3049

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/content/130/5/926.full.html