Sickle cell disease (SCD) is a group of genetic disorders affecting 70,000-100,000 Americans, although exact numbers are unknown due to varied reporting and monitoring methods across states (Bahr & Song, 2015; Lentz & Kautz, 2017; Paulukonis et al., 2016). SCD manifests as chronic anemia, pain, stroke, vaso-occlusive crises (VOCs), organ failure, and premature death (Jenerette, Pierre-Louis, Matthie, & Girardeau, 2015; Lentz & Kautz, 2017; Shah et al., 2014).

SCD is most prevalent in people of African descent. Because of the demographics of affected persons, SCD also is associated strongly with racial health disparities (Adams-Graves & Bronte-Jordan, 2016). Approximately 1 in 400 African Americans and 1 in 16,000 Hispanic Americans have SCD (Centers for Disease Control and Prevention [CDC], 2017). An estimated 1 in 13 African-American babies is born with sickle cell trait (SCT); they carry one copy of the gene and, while not affected personally, can pass the trait on to offspring (American Society of Hematology [ASH], 2016; CDC, 2017). SCD also occurs in people of Mediterranean and Arabic, and South American and Central American backgrounds. Although the exact number of Americans or Americans by nationality affected with the disease is unknown, SCD is seen often in marginalized populations who experience significant health disparities complicated further by SCD (Adams-Graves & Bronte-Jordan, 2016; CDC, 2017).

Historically, individuals with SCD did not live into their 20s; in the 1970s, children with SCD were not expected to survive to age 15 (Paulukonis et al., 2016). However, over the next several decades, life expectancy of people with SCD increased to ages 40-60 (Adams-Graves & Bronte-Jordan, 2016). This increased life expectancy, coupled with a 3% decrease in pediatric mortality from 1979 to 2005, was a direct result of newborn screening for SCD, childhood penicillin prophylaxis, administration of blood transfusions, increased flu and pneumonia immunizations, and use of hydroxyurea (Porter, Wesley, Zhao, Rupff, & Hankins, 2017).

Although the decreased pediatric mortality rate and increased life expectancy for individuals with SCD may suggest a higher quality of care for all patients with SCD, this conclusion is inaccurate. While the pediatric mortality has decreased over the past 30 years, the adult mortality rate has increased slightly by 1% during this time (ASH, 2016). This slight increase in adult mortality is believed to be related to suboptimal care received by young adult patients with SCD rather than additional patients with SCD reaching young adulthood.
adulthood, or of disease progression and subsequent disease complications in adulthood (Adams-Graves & Bronte-Jordan, 2016; ASH, 2016). Although multiple interventions can be used to treat and even cure SCD, they are used infrequently. For example, as the only medication approved by the U.S. Food and Drug Administration to treat adults with SCD and known to reduce mortality in patients with SCD, hydroxyurea is underused. Although it has been shown to decrease pain, reduce the frequency and severity of VOCs and subsequently death, fewer than 25% of patients who meet the criteria for hydroxyurea receive it. The most likely reasons for failure to prescribe hydroxyurea are provider discomfort with use and prescription of the medication, and provider-perceived or actual patient nonadherence (ASH, 2016). This underuse contributes to poor quality of care and a subsequent increase in mortality in adults with SCD.

Other nonpharmacologic treatments for SCD are curative (e.g., bone marrow and stem cell transplantation), but these treatments are not used widely and are not considered the standard of care due to lack of donor availability, cost, and risk of patients’ nonadherence to post-transplantation medical regimens (ASH, 2016). However, failing to use curative treatments for SCD due to cost and fear of patient nonadherence highlights disparities in care experienced by some African Americans in the United States.

An additional problem is that young adults with SCD are aging out of pediatric settings (ASH, 2016; Porter et al., 2017). The adult healthcare system is not prepared to handle this influx (ASH, 2016). Thus, a shift in healthcare delivery at federal, state, and local levels is needed. Emphasis must be placed on health promotion in adolescents and young adults with SCD, and on education for nurses, physicians, and other providers, to improve outcomes of young adults who have aged out of pediatric health care (ASH, 2016; Porter et al., 2017).

To do this, many factors must be understood, including the overt and unconscious perceptions of healthcare providers regarding barriers to care for the young adult SCD population. The strengths and limitations of the current healthcare infrastructure also must be addressed, including issues of access to and affordability of care. Although research has found young adults with SCD have high hospital admission, morbidity, and mortality rates (Porter et al., 2017; Wilson & Nelson, 2015), little headway has been made to tackle these costly and detrimental concerns for young adults with SCD.

Barriers to care for young adults with SCD represent a long-term, disturbing issue in U.S. health care. Young adults participate in riskier behaviors and avoid preventive health care more often than their child, adolescent, and adult counterparts (Stroud, Walker, Davis, & Irwin, Jr., 2015). Risky behaviors coupled with chronic disease can lead to poor short- and long-term negative health and social outcomes as well as increased hospitalization and healthcare costs.

**Methods**

A literature search was conducted using PubMed Plus, CINAHL, and PsycINFO, with no date restriction. The oldest published paper related to policy affecting patients or young adults with SCD was from 1984 (Manly, 1984). Initial key terms used were sickle cell, sickle cell anemia, young adult, adolescent, provider, nurse, and transitions. Publications identified in the search were selected based on their relevance to the health care of young adults with SCD; further reviews of reference lists yielded additional publications. To obtain grey literature, Google and Google Scholar searches were used to obtain copies of *Healthy People 2020*, National Academies of Medicine reports, and reports from the American Society of Hematology. The search process was iterative and conducted over several years.

**Significance of Prioritizing SCD Care in the Adult Setting**

Patients with SCD begin to transition from pediatric to adult healthcare settings at ages 17-22, but this transition varies by patient and healthcare institution (Bermich-Stolz, Halanych, Howard, Hilliard, & Lebendesburger, 2015). Transitions are not considered successful when just young adults’ healthcare needs and medical records have moved to the adult setting; transition is successful only if the patient is able to function autonomously, establish trust with adult healthcare providers, and demonstrate self-efficacy (Andemarius et al., 2014; Sobota, Shah, & Mack, 2016).

Telfair, Alexander, Loooser, Alleman-Velz, and Simmons (2004) reported results of previous studies that identified barriers to successful transitioning to the adult setting for adolescents and young adults, including patient-provider attachment, and communication between the patient (or parents) and providers, specifically regarding expectations about the transition. Although over a decade has passed since this publication, barriers remain the same. Healthcare providers also may contribute to unsuccessful transitions and subsequently increased patient morbidity and mortality because of their discomfort caring for adult patients with SCD, unwillingness to provide care for a transitioning patient, and lack of knowledge about adult patients with a historically pediatric condition (ASH, 2016). Of note, the ASH report (2016) referred specifically to care provided by physicians. No literature was found that addressed the role of nurses in SCD care transitions.

**Emergency Department Utilization**

Receiving care in the appropriate setting is essential for young adults with SCD and healthcare systems to achieve continuity of care and lower healthcare costs (ASH, 2016). While many patients with
SCD are able to manage their pain primarily at home, some patients are consistently unable to do. These patients are hospitalized frequently and often receive the majority of their care in emergency departments (EDs) rather than from a consistent primary care or hematology provider (Brown, Weisberg, Balf-Soran, & Sledge, 2015). Brousseau, Owens, Mosso, Panepinto and Steiner (2010) provided the most up-to-date published statistics indicating approximately 20% of studied patients with SCD were in this high hospital-use subgroup and the majority of patients seen in EDs had Medicare or Medicaid as their insurance provider. Additional predictors of ED visits were severity of SCD symptoms, adolescence or young adulthood, and use of government-provided health insurance.

Frequent use of the ED by young adult patients with SCD is not an adequate replacement for care provided in primary or specialty care settings (Brousseau et al., 2010; Jenerette, Brewer, & Ataga, 2014). Lack of knowledge, fear of stigmatization, and lack of understanding of appropriate hospital use and symptom management by some patients with SCD contribute to increased ED use and perpetuate the cycle of hospitalizations and poor outcomes (Jenerette et al., 2014).

### Stakeholders
National and global stakeholders, including many specialists and professional organizations, seek to improve the overall health and wellness of Americans with SCD. Although their combined effort is important, understanding each organization’s role in the treatment of SCD and provision of patient services is also important. Table 1 provides a summary of national and global stakeholders as well as each group’s recommendations.

### Federal Legislation
Federal legislation plays a large role in quality care for persons with SCD. The Patient Protection and Affordable Care Act (ACA, 2010) was developed to expand Medicaid eligibility and allow affordable access to care; more than 30 million Americans became newly insured or remained insured (ACA, 2010; Blumenthal, Abrams, & Nuzum, 2015). Before passage of the ACA, more than half of all patients with SCD were covered by government health insurance (Brousseau et al., 2010). Enactment of the ACA likely provided more affected patients with healthcare coverage. Unfortunately, no literature could be found demonstrating this legislation’s direct effect on persons with SCD.

Before passage of the ACA, little national legislation was aimed at improving care of patients with SCD. A summary of the federal legislation and each statute’s respective influence on SCD is provided in Table 2. Of note, these legislative measures provided the impetus for creation of private organizations such as ASH with the goal of improving care of patients with SCD.

### Access to Care
Access to care involves ensuring patients with SCD receive the most up-to-date treatments, including hydroxyurea, vaccinations, and adequate screening for complications. Persons with SCD also should receive high-quality care, defined as non-stigmatized care from specially trained providers across the continuum of health care (ASH, 2016).
High-quality care for young adults with SCD begins with adequate newborn screening (NBS) for SCD in the context of reducing young adult morbidity and mortality. Early screening and monitoring are crucial for capturing an accurate prevalence of SCD and early identification of disease or trait, and for a national approach to improving care and outcomes for patients with SCD (Minkovitz, Grason, Ruderman, & Casella, 2016; Paulukonis et al., 2016).

States began screening all newborns for SCD in 1975; all states had NBS programs by 2006 (Minkovitz et al., 2016). However, no consistent structure or process exists for examining data related to NBS results; there is also no longitudinal follow up of patients and families with NBS results positive for SCD. This inconsistency in screening and reporting contributes to the inability to determine the number of Americans affected with SCD (Paulukonis et al., 2016). Because state requirements for follow up after NBS are variable, care of affected individuals also varies (Minkovitz et al., 2016). In an effort to correct this variation and subsequently improve reporting and care, Healthy People 2020 included a recommendation to improve the NBS process throughout the country (Minkovitz et al., 2016; Office of Disease Prevention and Health Promotion, 2019). Minkovitz and colleagues (2016) also reported variation in state lobbying and policy development, and a general lack of states’ involvement in policy decisions and advocacy related to SCD. From a national standpoint, variation in practice and policies among states makes it difficult to monitor outcomes across the patient lifespan and ultimately limits affected persons’ access to quality, evidence-based care.

### Disparities in Funding for SCD

If SCD is compared with other genetic diseases in which life expectancy has increased significantly over time (e.g., cystic fibrosis [CF]), the relatively little funding directed toward SCD is clear. The most up-to-date statistics regarding disparities in care are provided here. In 2003, private organizations raised $498,577 for SCD research and services combined; $152 million was raised for CF (Smith, Oyeku, Homer, & Zuckerman, 2006). In 2012, the National Institutes of Health spent $65 million on SCD research and $86 million on CF research (Bahr & Song, 2015). However, the number of patients with SCD in the United States is double or triple that of individuals with CF (Adams-Graves & Bronte-Jordan, 2016; Bahr & Song 2015). According to Smith and co-authors (2006), when private funding from independent donors and public funding are combined, the total support for a patient with SCD is $1,130 while the total support for a patient with CF is $9,340. Nearly nine times more funding is available to support research and provide treatments for patients with CF than for patients with SCD. Although other chronic pediatric diseases exist, SCD and CF are most comparable in terms of prognosis, life expectancy, genetics, and chronicity. One notable difference, however, is the demographic of affected persons. SCD primarily affects African Americans, while CF primarily affects Caucasians (Bahr & Song, 2015). The large discrepancy in both federal and private funding can be contributed directly to the

### Federal Legislation Related to SCD

<table>
<thead>
<tr>
<th>Federal Act</th>
<th>Influence on SCD Care</th>
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<tbody>
<tr>
<td>National Sickle Cell Disease Control Act (1972)</td>
<td>• Provided infrastructure for NIH to establish a national SCD program to improve outcomes (e.g., reduced morbidity/mortality, increased funding, improved provider education and overall patient care)</td>
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<tr>
<td>National Sickle Cell Anemia, Cooley Anemia, Tay Sachs, and Genetic Diseases Act (1976)</td>
<td>• Allowed federal funding to be used for genetic screening, counseling</td>
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<tr>
<td>Americans with Disabilities Act (1990)</td>
<td>• Prohibited employers from discriminating against employees with disabilities, including SCD</td>
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<tr>
<td>Sickle Cell Treatment Act (2003)</td>
<td>• Provided matching of federal dollars for SCD treatment, costs</td>
</tr>
<tr>
<td>Genetic Information Nondiscrimination Act (2008)</td>
<td>• Prohibited discrimination from insurance or employment based on genetics</td>
</tr>
<tr>
<td>Patient Protection and Affordable Care Act (2010)</td>
<td>• Provided state expansion of Medicaid and assess to EHB for insured persons • Allowed young adults to remain on parents’ insurance until age 26 • Ensured companies with over 100 employees provided them with insurance coverage</td>
</tr>
</tbody>
</table>

EHB = essential health benefits, NIH = National Institutes of Health, SCD = sickle cell disease

**Stigma**

Persons with SCD or SCT may be stigmatized by healthcare providers and communities. This can have a negative impact on their physical and psychological health (Adams-Graves & Bronte-Jordan, 2016; Porter et al., 2017; Wesley, Zhao, Carroll, & Porter, 2016). Stigmatization can prompt patients and families to distrust healthcare providers; it also can cause a failure to discuss these conditions within the community, and general lack of patient knowledge of SCD and SCT (Adams-Graves & Bronte-Jordan, 2016; Wesley et al., 2016). Discussion of these conditions within the African-American community is important to raise awareness and provide support to affected patients and families. Healthcare providers, particularly nurses, should examine their roles and responsibilities to affected patients.

**Nursing Implications**

The knowledge and skills of nurses have not been used fully to close the gap between recommended and actual care provided to young adults with SCD (Jenerette et al., 2015). Hospital-based nurses in all practice areas, including pediatric, medical-surgical, and critical care units, can play a crucial role in safeguarding patients with SCD from avoidable complications. They also ensure patients are prepared adequately for hospital discharge and remain free of hospital-acquired infections. Nurses manage patients’ pain and other symptoms throughout hospitalization. Hospital-based nurses should assess patients’ abilities to care for themselves and communicate with interprofessional team members. These responsibilities are magnified when nurses care for young adult patients with SCD who are vulnerable due to age, race, socioeconomic status, and disease symptoms.

High-quality care provided by hospital-based nurses not only promotes health and reduces complications in persons with SCD, but also reduces the risk of hospitals receiving federal penalties for readmissions and hospital-acquired conditions (Centers for Medicare & Medicaid Services [CMS], 2019). Nurses are most frequently the direct-care providers who manage pain, give education, and offer emotional support for patients with SCD (Jenerette et al., 2015). As persons with SCD live longer and more young adults are treated in adult settings, it will become increasingly important for nurses to be able to care appropriately for young adults with SCD.

A consistent model of care is needed across the United States to ensure young adults with SCD are prepared adequately for the transition from pediatric to adult health care, and from inpatient to outpatient settings. Nurses who focus on transitions in care have had a significant impact on preventing readmissions that occur with diseases primarily affecting older adults (Zhu, Liu, Hu, & Wang, 2015). The Transitional Care Model (TCM) is a nursing-focused framework used extensively to manage other adult chronic diseases across the continuum (University of Pennsylvania School of Nursing, n.d.). The TCM has not been used explicitly in adolescents with SCD, although transitions in care for these patients are a top priority for many national organizations (ASH, 2016). Medicare reimbursements for transitional care in older adults, but no Medicaid reimbursement option is available (CMS, 2016; University of Pennsylvania School of Nursing, n.d.). Medicaid reimbursements for transitional care would incentivize high-quality transitions to ensure improved health outcomes in young adults with SCD. Reasons for not employing a national transitions model such as the TCM for patients with SCD, as previously discussed, may be rooted in the lack of funding, disparities in care, societal constructs, and national lack of attention, knowledge, and understanding of the disease (Adams-Graves & Bronte-Jordan, 2016; ASH, 2016; Bahr & Song, 2015).

Literature suggests healthcare providers’ negative attitudes affect care and providers in general have negative attitudes toward patients with SCD (Jenerette et al., 2015). However, limited research has focused on nurses’ perceptions of barriers that may impede care of young adult patients with SCD (Jenerette et al., 2015; Jenerette et al., 2016). Focused studies of nurses who frequently care for young adult patients with SCD need to be conducted to identify nurses’ perceived barriers to high-quality care, transitions in care, prevention of hospital readmissions, as well as potential solutions for decreasing patient morbidity and mortality. Findings can inform national care models and provide insight into specific interventions that can be implemented on a national level to help nurses and other providers care for young adults with SCD.

Ensuring nurses and other healthcare providers are educated about the disease and treatments is paramount to treatment success (ASH, 2016). Many existing national recommendations, including those of ASH, focus on physicians and fail to recognize the potential impact of nurses. Failure to address nurses in these recommendations serves as a barrier to improved care of the young adult with SCD (Jenerette et al., 2015).

**Conclusion**

To ensure young adults with SCD live into older adulthood and receive the highest quality care possible, more attention to the disease and its optimal management are needed. Nurses in particular need to understand the history, socioeconomic and cultural aspects of SCD, federal policies that help and hurt young adults with the disease, and their own unique roles in caring for this population. Areas of opportunity for nursing education include lack of knowledge regarding the disease and plan of care, difficulty with interprofessional collaboration, difficulty caring for a marginalized
population, personal biases, and pain management for SCD (Jennerette et al., 2015). To ensure young adults with SCD receive the highest quality of care and improved outcomes, a complete body of research must be developed.

REFERENCES


National Sickle Cell Disease, 1972, 42 U.S.C. §§ 201-302


Patient Protection and Affordable Care Act of 2010, 42 U.S.C. §§ 1001-1011 (ACA)


