GAPS AND RECOMMENDATIONS

GAP 1: There is sometimes a lack of partnership between the disease specialist (i.e., the hematologist, oncologist, rheumatologist, or neurologist) and providers of comprehensive multidisciplinary pain programs.

- **RECOMMENDATION 1A:** Provide referrals to a comprehensive pain program early in the course of the chronic disease (e.g., MS, porphyria, systemic lupus erythematosus, migraine, Parkinson's disease, neuropathic pain syndromes) to determine the optimal approach to managing acute or chronic pain exacerbations, including potential non-opioid, alternative therapies and nonpharmacologic therapies. Establish a partnership between the disease specialist (e.g., the hematologist, oncologist, neurologist, or rheumatologist) and the pain team to optimize care.

ANNE'S STORY

PATIENT TESTIMONIAL

My name is Anne. I'm a 19-year-old girl and college sophomore. I have struggled with sickle-cell disease my whole life. My dad always told me that sickle cell does not have me — I have sickle cell. I have learned to persevere through the pain.

I found that my passions were the one thing that kept the pain away. Even though I would be in pain, I would still go out and dance. I would still sing. I was even a cheerleader at one point. I think that with sickle cell, it's also about what you can handle mentally.

I struggled with depression for a while and as recently as last February, I went through a period of depression. It was the hardest thing, but I kept telling myself, OK Anne, you're going to get better. Just keep pushing. Just keep pushing.

I have been through six surgeries in my 19 years. I have had brain surgery due to Chiari I malformation from sickle cell, which caused multiple migraines every day. I was shocked because I would still go to school. I would still get my homework done. I would still go out and have fun with my friends, even though I was still going through all this pain.

My brother told me that I am one of the strongest people he's ever met. And that was so touching because at that time I didn't believe I was a strong person. It was hard because my parents never dealt with a child with sickle cell. I'm their last child. And I've seen them cry. I've seen them persevere with me.

My family is one thing that keeps me going; I can't let anything stop me. I can't even let one little crisis stop me. I barely go to a hospital for my crisis now because I try to find ways at home to get rid of my pain.

2.7.7 Sickle Cell Disease

Sickle cell disease is a group of inherited disorders characterized by complex acute and chronic symptoms, including pain.[367] An estimated 90,000 people in the United States have SCD, which disproportionately affects minority populations, particularly African Americans.[368] Acute pain episodes, or "pain crises," associated with SCD are abrupt in onset and unpredictable, and they drive patients to seek care in the ED and inpatient unit, with estimated health care costs of almost $2 billion per year.[369,370] Chronic, severe, daily pain also occurs in approximately 30% to 40% of adolescents and adults with SCD, significantly impairing their functioning and increasing in incidence and severity with age.[371–373] Pain in SCD is unique in that it occurs throughout the patient's lifespan, from infancy to adulthood, and develops directly from the disease.[374] The biology of SCD pain is complex and varied; it likely arises from multiple mechanisms depending on whether an individual is suffering from acute or chronic pain.[375] Pulmonary,[376] orthopedic,[377] psychosocial,[378] and other comorbidities of SCD can also give rise to painful complications in adults and children.

GAPS AND RECOMMENDATIONS

GAP 1: There is a lack of evidence-based management guidelines for the treatment of acute and chronic pain in children and adults with SCD.

- **RECOMMENDATION 1A:** Develop comprehensive, evidence-based guidelines for the treatment of acute and chronic SCD pain in children and adults.
- **RECOMMENDATION 1B:** Conduct research to understand underlying mechanisms of acute and chronic pain, and develop mechanistic non-opioid pharmacologic therapies and nonpharmacologic approaches for SCD pain management.

GAP 2: Unpredictable, episodic exacerbations of acute pain pose a challenge for SCD pain management, and the majority of patients have failed non-opioid pain medications prior to presentation for acute care. Constraints on opioid treatment duration can make individualization of pain management difficult. Further, limited access to oral opioids at home for the treatment of unplanned acute pain can result in increased use of health care services that could have been avoided.

- **RECOMMENDATION 2A:** Protect access to the appropriate and safe use of opioids for patients with SCD, with consideration for exemption from prescribing guidelines and state prescribing laws that do not specifically address patients with SCD because of the complex nature and mechanism of acute and chronic sickle cell pain.
- **RECOMMENDATION 2B:** Consider the lowest effective dose of opioids to treat acute pain crises, and prescribe within the context of close follow-up and comprehensive outpatient pain care.
- **RECOMMENDATION 2C:** Develop an individualized approach to pain management that includes consideration of opioid and non-opioid therapies, such as behavioral health strategies and multimodal approaches.
- **RECOMMENDATION 2D:** Provide patient education on the risks and benefits of opioids.

GAP 3: The SCD patient population faces significant health care disparities that affect access to and delivery of comprehensive pain care and mental health services. Further, stigma, negative provider attitudes, and perceived racial bias are associated with SCD pain,[379,380] which may compromise care, thus leading to increased suffering from pain and pain care delivery.[381–385]

- **RECOMMENDATION 3A:** Develop comprehensive care delivery models for SCD pain management, including collaborative partnerships among pain medicine, SCD specialists and advocates, and multidisciplinary teams.
- **RECOMMENDATION 3B:** Develop outpatient infusion clinics/day hospitals for SCD pain management to decrease reliance on the ED for pain treatment.
- **RECOMMENDATION 3C:** Increase access to and reimbursement for mental health services for patients with SCD.
- **RECOMMENDATION 3D:** Provide education focused on stigma, negative provider attitudes, and perceived racial bias at all levels of health care to optimize delivery of pain treatment to patients with SCD.

2.7.8 Health Disparities in Racial and Ethnic Populations, Including African-Americans, Hispanics/Latinos, American Indians, and Alaska Natives

Considerable evidence exists documenting health disparities in racial and ethnic minority populations, particularly substantial disparities in the prevalence, treatment, progression, and outcomes of pain-related conditions.[386] These disparities in care are attributed to factors related to social disadvantage as well as factors within health systems.[387] Health disparities contributing to suboptimal pain management in these special populations may be related to such factors as barriers to accessing health care, lack of insurance, discrimination, lack of a PCP, lack of child care, a lower likelihood to be screened or receive pain treatment, and environmental barriers that impede effective self-management. Effective strategies and plans to address these issues specifically in these disparate communities are necessary to address these gaps to improve patient outcomes.