The Daily Experiences of Adolescents in Lebanon With Sickle Cell Disease

Mia Atoui, MA, Lina Kurdahi Badr, PhD, RN, Tamara Drenttel Brand, MPH, Ruby Khoury, MD, Randa Shahine, RN, & Miguel Abboud, MD

ABSTRACT

Objectives: Despite the psychosocial and physical consequences associated with sickle cell disease (SCD), the daily lived experience of adolescents diagnosed with this disease is a phenomenon rarely described. The objective of this study was to explore the daily lived experience of adolescents with SCD living in Lebanon.

Method: Twelve adolescents with SCD between the ages of 12 and 17 years were interviewed with use of a semi-structured interview during a routine follow-up visit after they were assessed as being pain free. Interviews were transcribed verbatim, and thematic analysis was conducted.

Results: Adolescents with SCD experience a layered burden consisting of physical, emotional, and sympathetic pain that affects much of their daily personal and social lives. Nevertheless, they seem to claim normalcy and to downplay their pain and suffering in order to limit their caregivers' distress.

Conclusion: These findings can be used to assist health care providers in designing culturally sensitive interventions specifically designed for adolescents with SCD and their families to enable them to better cope with their illness. J Pediatr Health Care. (2015) *29*, 424-434.

KEY WORDS

Sickle cell disease, lived experiences, Lebanon

Sickle cell disease (SCD) is the most common single gene disorder worldwide, with different clinical manifestations in various ethnic groups and populations. The term refers to a collection of autosomal-recessive genetic disorders characterized by the hemoglobin S (HbS) variant of the β-globin gene that is resistant to malaria development and in which more than 50% the hemoglobin-β gene is replaced with HbS (El-Hazmi, Al-Hamzi, & Warsy, 2011; Saraf et al., 2014). SCD affects 1 in 500 African Americans and more than 30 million people worldwide, with predominance in populations of the sub-Sahara, India, and the Middle East (Inati et al., 2007; Khoriaty et al., 2014; McGann, 2014). There are different variants of the β-globin gene in SCD, including but not limited to sickle cell anemia, sickle-hemoglobin C disease, sickle beta-plus thalassemia, and sickle beta-zero thalassemia, with prevalence of certain variants types in particular ethnic groups (El-Hazmi et al., 2011; Saraf et al., 2014). In the Middle East population, different abnormal variants have been identified, with each country having a characteristic distribution and clinical presentation. The tradition of consanguineous marriage in Middle Eastern countries, which varies between 25% in Lebanon to 60% in Saudi Arabia and 90% in some Bedouin communities in Kuwait and Saudi Arabia, is a major contributor to the multiplication of SCD

Mia Atoui, Clinical Psychology Intern, Department of Psychiatry, American University of Beirut Medical Center, Beirut, Lebanon.

Lina Kurdahi Badr, Professor, Azusa Pacific University, Azusa, CA.

Tamara Drenttel Brand, Lactation Educator, American University of Beirut, Beirut, Lebanon.

Ruby Khoury, Physician, Rainbow Babies and Children's Hospital–Case Western Reserve University, Cleveland, OH.

Randa Shahine, Registered Nurse, Children's Cancer Center of Lebanon, American University of Beirut Medical Center, Beirut, Lebanon.

Miguel Abboud, Chair and Professor, Department of Pediatrics and Adolescent Medicine, American University of Beirut Medical Center, Beirut, Lebanon.

Conflicts of interest: None to report.

Correspondence: Lina Kurdahi Badr, PhD, RN, Azusa Pacific University, 701 E Foothill Ave, Azusa, CA 91702; e-mail: lbadr@apu.edu.

0891-5245/$36.00

Published online March 13, 2015.

http://dx.doi.org/10.1016/j.pedhc.2015.01.012

(El-Hazmi et al., 2011). Although it is inherited genetically, the symptoms and course of the disease (HbS) can be affected by environmental factors such as infections, nutrition, and socioeconomic status (Sanders, Labott, Molokie, Shelby, & Desimone, 2010). Among the hemoglobinopathies, SCD and β-thalassemia are the most common and have the highest impact on morbidity and mortality, with more than 330,000 infants diagnosed annually. The World Health Organization estimates that 21.7% of people in the Middle East region carry variants for either SCD or thalassemias (Modell & Darlison, 2008; World Health Organization, 2006). In Lebanon, a recent study found that among newborns, 2.1% were found to have an abnormal hemoglobin variant, with sickle hemoglobin being the most common variant (84%) and carrier rates being highest in Northern Lebanon (Khoriaty et al., 2014). These estimates may be lower than the actual incidence because of the early discharge of newborns and lack of newborn screening at many institutions. Compared with international data, the carrier rate of sickle cell and other hemoglobin variants in Lebanon is similar to Brazil (approximately 2%) but almost twice that of Italy (1.2%) and lower than Saudi Arabia (1.3% to 21.3%) and Bahrain (11.2% to 16.4%; Haber et al., 2010; Inati et al., 2007; Jastaniah, 2011; Makhoul et al., 2005).

SYMPTOMS OF SCD

The symptoms of SCD may occur during the first few months of age or appear later during childhood or adolescence; however, once they appear, the symptoms persist throughout life, and there is no cure. The clinical features of SCD are defined by chronic anemia, sepsis, hemolysis, and recurrent acute vaso-occlusive crises. The latter are characterized by pain and a systemic inflammatory response that may be severe, episodic, and unpredictable. Pain is the most common symptom in SCD that limits adolescents' social and personal initiatives and can affect their ability to perform on the same level as their healthy counterparts (Fosdal, 2015; Hildenbrand et al., 2014). The limitations placed on children with SCD as a result of their disease often hinders them from participating in sports and other school and recreational activities, compromising their ability to socialize and develop meaningful personal relationships. School absenteeism is another chief complaint in persons with SCD, with children missing an average of 12% of the academic year and more than one third of children missing at least 30 days. Furthermore, adolescents with SCD perform worse in school and on standardized tests compared with their peers and siblings (Schwartz, Radcliffe, & Barakat, 2009). Other manifestations include and are not limited to acute chest syndrome, bacterial sepsis, sequestration crisis, aplastic crises, ischemic stroke, fatigue, and exercise intolerability (Ameringer, Elswick, & Smith, 2014; Chakravorty & Williams, 2015; Masuda, Cohen, Wicksell, Kemani, & Johnson, 2011; Sanders et al., 2010). Some of the typical clinical manifestations of SCD such as jaundice, delayed puberty, or stunted growth distinguishes and separates these children from their peers and can contribute to low self-esteem, poor body image, and behavioral problems, especially in adolescence (Noll, Kiska, Reiter-Purtill, Gerhardt, & Vannatta, 2010; Wilson & Nelson, 2014).

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Although advances in medical technology have increased life expectancy for persons affected by SCD, the disease still greatly reduces life expectancy and results in long-term psychosocial problems, especially in adolescents. In the United States, mortality has decreased dramatically as a result of newborn screening and better comprehensive care. The median age of death in patients with SCD in the United States is 53 years for men and 58 years for women. A recent statement by the World Health Organization indicated that SCD contributes to the equivalent of 5% of deaths of children younger than 5 years in the African continent, more than 9% of such deaths in West Africa, and up to 16% of such deaths in individual West African countries. Mortality rates in Lebanon and the Middle East are not available; however, milder forms of the disease are present in the Arabian Peninsula, which results in lower mortality rates, and when patients receive comprehensive care as they do at the Children's Cancer Center in Lebanon (CCCL), the mortality rate is comparable with that of the United States.

QUALITY OF LIFE IN CHILDREN WITH SCD

A large body of literature has investigated the quality of life of this population (Barakat, Patterson, Daniel, & Dampier, 2008; Jackson, Lemanek, Clough-Paabo, & Rhodes, 2014; Menezes, Len, Hilário, Terreri &, Braga, 2013). An earlier study observed that children and adolescents with SCD spend most of their time enduring the chronic and acute pain events at home, tend to underreport their pain levels, and tend to dismiss the negative effects of SCD on their daily lives (Fuggle, Shand, Gill, & Davies et al., 1996). Family characteristics and dynamics can positively or negatively