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Comprehensive Infant Clinic for Sickle Cell Disease: Outcomes and Parental Perspective

Brenda M. Martin, MSN, CPNP, Lisa N. Thaniel, DSW, LICSW, Barbara J. Speller-Brown, DNP, CPNP, & Deepika S. Darbari, MD

ABSTRACT

Introduction: Comprehensive care for children with sickle cell disease (SCD) includes penicillin prophylaxis, pneumococcal immunization, hydroxyurea therapy, and transcranial Doppler screening for stroke prevention. Along with caregiver education, these strategies have been shown to be effective in reducing early morbidity and mortality in this population. The subspecialty Infant Sickle Cell Clinic was initiated to improve access, education, patient outcomes, and family satisfaction.

Method: Telephone surveys were conducted with parents to assess satisfaction with the Infant Sickle Cell Clinic, compliance with guidelines, and comfort level with managing their child's SCD.

Results: This quality improvement project reported high levels of parent satisfaction and improved outcomes with the proposed approach but also presents areas for improvement.

Discussion: Our report presents a unique model of providing care to families with infants newly diagnosed with SCD. The group format serves as a useful model to allow families an interactive educational session with guest speakers. J Pediatr Health Care. (2018) ■■, ■■-■■.

KEY WORDS

Education, infant, sickle cell, parent

Brenda M. Martin, Nurse Practitioner, Division of Hematology, Children's National Health System, Washington, DC.

Lisa N. Thaniel, Social Worker, Division of Hematology, Children's National Health System, Washington, DC.

Barbara J. Speller-Brown, Nurse Practitioner, Division of Hematology, Children's National Health System, Washington, DC.

Deepika S. Darbari, Attending Physician, Division of Hematology, Children's National Health System, Washington, DC.

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Correspondence: Brenda M. Martin, MSN, CPNP, Children's National Medical Center, Center for Cancer and Blood Disorders, 111 Michigan Ave., NW, Washington, DC 20010; e-mail: BMmartin@childrensnational.org

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INTRODUCTION

Sickle cell disease (SCD) is the most common genetic disorder affecting 1 in 365 African American births and 1 in 16,300 Hispanic births (Centers for Disease Control and Prevention, 2016). In the United States, about 2,000 children are born with SCD each year (National Institutes of Health et al, 2002), and approximately 100,000 individuals with SCD are living in the United States (Hassell, 2010). In the 1970s, the average age of life expectancy among individuals with SCD was less 20 years (Lanzron, Carroll, & Haywood, 2013), with a median age at death of approximately 14 years, primarily caused by infections (Hamideh & Alvarez, 2013; Paulukonis et al., 2016). SCD-related deaths among children younger than 4 years has decreased by 42% from 1999 through 2002 (Quinn, Rogers, McCavit, & Buchanan, 2010; Yanni, Grosse, Yang, & Olney, 2009). In the United States, 95% of children born today are expected to survive into adulthood (Chaturvedi & DeBaun, 2016).

In 1975, when the first statewide newborn screening program for SCD was initiated, widespread acceptance and implementation of newborn hemoglobinopathy screening was difficult to implement because of concerns about potential effectiveness of early diagnosis and treatment (Vinchinsky, Hurst, Earles, Kleman, & Lubin, 1988). The Prophylactic Penicillin Study (PROPS) showed that initiation of twice daily

oral penicillin prophylaxis reduced the incidence of serious infection in young children with SCD by 84% compared with placebo (Gatson et al., 1986). These findings led to widespread adoption of newborn screening programs for SCD to identify these infants early for treatment to prevent mortality from overwhelming sepsis (Falletta et al., 1995). In addition to newborn screening and starting penicillin prophylaxis, other advances in medical treatment and interventions such as vaccination against pneumococcus, screening for stroke risk by transcranial Doppler (TCD), and hydroxyurea therapy, in conjunction with parental education, have led to improved outcomes for children with SCD (Quinn et al., 2010; Sergeant, 1995; Yawn et al., 2014). Although the infant mortality rate for young children with SCD has improved and comprehensive care has been proven to be the link, a significant proportion of children may not receive all the age-appropriate screenings and immunizations. Although annual TCDs are recommended to assess children at risk for stroke (starting at 2 years of age), many children across the United States do not receive this screening (Neunert, Gibson, Lane, & Synder, 2016; Reeves, Madden, Freed, & Dombkowski, 2016), and young children with SCD remain at risk for morbidity and mortality (Booth, Inusa, & Obaro, 2010).

The current SCD management guidelines suggest that newborns with SCD begin penicillin prophylaxis and attend their initial hematology appointment by 3 months of age (Kavanagh, Wang, Therrell, Sprinz, & Bauchner, 2008). Children's National Health System provides care to children affected by SCD in the Washington, DC, metropolitan area. This is one of the largest programs in the United States, where approximately 80 new infants with SCD are seen annually. In 2007, it was recognized that families of infants newly diagnosed with SCD had long wait times for the next available appointment and that the SCD education given was not standardized. To overcome this problem, in 2008, the subspecialty Infant Sickle Cell Clinic (ISCC) within the hematology division was established with the quality improvement goals of seeing all newborns with SCD within 8 weeks of diagnosis based on the newborn screening. The ISCC provides services to families of infants with SCD from birth to 2 years of age. Families are educated about SCD through a standardized education program. The ISCC team includes one hematologist, two pediatric nurse practitioners, one social worker, a genetic counselor, and a clinical care coordinator. The clinic is held once every 2 weeks. The ISCC staff provides medical care, SCD-related education, genetic counseling, social work services with psychosocial evaluations, and developmental screenings. Since the initiation of the ISCC clinic, administrative staff has been instructed to schedule all infants with the new diagnosis of SCD to be seen in the ISCC within 8 weeks of the diagnosis. Standardized education, which includes discussion of SCD basics; pathophysiology, fever, and pain management; spleen palpation; infection risk and immunizations; appropriate screenings; and anticipatory guidance is provided one to one to the families. In addition, family group education sessions are held with invited guest speakers who have expertise in a wide variety of areas, such as genetics, neuropsychology, pain management, bone marrow transplantation, nutrition, transfusion, and emergency medicine. Treatment options such as hydroxyurea and bone marrow transplantation for SCD are also discussed. The visits allow families of patients with SCD an opportunity to interact and learn from one another and increase provider efficiency and productivity (Jaber, Braksmajer, & Trilling, 2006). The primary purpose of this quality improvement project was to assess parents' knowledge of SCD and their level of satisfaction with the education in the ISCC. The second aim was to evaluate the efficacy of the ISCC.

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METHODS

The clinic team developed a telephone survey of 10 questions to evaluate the impact of establishing the ISCC and assess the experience of families with SCD infants. Telephone interviews were conducted on a convenience sample of 100 parents of infants with SCD who had been seen at least once in the ISCC between years 2012 and 2014. Participants were randomly selected, and parents of children between 2 months and 5 years old were called by the clinic staff (patients attended the ISCC only from birth to 2 years). A telephone survey, which included 10 questions, was conducted with the parents or legal guardians (see Box 1).The clinic staff was instructed to ask only the questions in the survey and not to interpret the question for the parent. Questions were designed to capture knowledge and understanding as it relates to SCD and overall satisfaction with the ISCC. The families were informed about the quality improvement nature of the project and that responding to questions was voluntary. They were also informed that the information would be kept confidential and deidentified. Parents were asked to answer the questions on a 5-point Likert scale, with 1 being