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Transition From Pediatric to Adult Care in Sickle Cell Disease: Perspectives on the Family Role

Jerlym S. Porter PhD, MPH [a,*], J. Carolyn Graff PhD, RN [b], Alana D. Lopez PhD [a], Jane S. Hankins MD, MS [a]

[a] *St. Jude Children's Research Hospital, Memphis, TN*
[b] *University of Tennessee Health Science Center, Memphis, TN*

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Transition from pediatric to adult care poses challenges for adolescents with sickle cell disease (SCD). This study explored the transition perspectives of adolescents with SCD, their siblings, and caregivers. Focus groups were conducted with 12 African American families. Adolescents, siblings, and caregivers demonstrated awareness of transition and need for disease management responsibility. Siblings' and caregivers' concerns included adolescent medication adherence. Family concerns included leaving the pediatric environment and adult providers' lack of knowledge. Families recommended more transition preparation opportunities. Family members' perspectives are valuable in informing transition planning. Family-focused interventions designed to prepare and support families during transition are necessary.

THE MORTALITY RATE among children with sickle cell disease (SCD) has substantially decreased in the past three decades. Because of medical advances, such as routine administration of antibiotic prophylaxis, immunizations, and disease-modifying therapies, such as chronic transfusion and hydroxyurea, greater than 90% of children with SCD reach the age of 21 years, and the majority are expected to live beyond their 40s (Hassell, 2010; Quinn, Rogers, McCavit, & Buchanan, 2010). However, a longitudinal cohort study of 940 children with SCD found an increase in mortality after 18 years of age and soon after the transfer to adult care with a mean time of 1.8 years following transfer to an adult provider (Quinn et al., 2010). Further, health care utilization is highest around the time of transition among young adults with SCD ages 18–30 (Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010; Hemker, Brousseau, Yan, Hoffmann, & Panepinto, 2011). As a result, transitioning youth receive less treatment-related services (transfusions, hydroxyurea, chelation therapy) but have higher healthcare costs and more SCD complications compared to pediatric patients (Blinder et al., 2013). For this reason, securing a successful transition from pediatric to adult care becomes essential to maintain continuity of services and appropriate disease management for those living with SCD (Treadwell, Telfair, Gibson, Johnson, & Osunkwo, 2011; Wang, Kavanagh, Little, Holliman, & Sprinz, 2011). The transition process is particularly important for ensuring continued adherence to treatment to avoid higher morbidity and mortality associated with SCD (Ballas & Dampier, 2004; Quinn et al., 2010).

Although a rationale for transition care has been established, obstacles to effective implementation remain. The nature of transition itself is a challenge as it should ideally be a fluid, ongoing process that requires active participation from patients, family members, and health care providers (Reiss, Gibson, & Walker, 2005; Telfair, Alexander, Loosier, Alleman-Velez, & Simmons, 2004). The transition to adult care has been studied among pediatric and adult chronic illness populations, demonstrating similarities in transition experiences and perceptions irrespective of disease. A study involving semi-structured interviews with 24 young adults aged 15–22 years with different chronic illnesses (hemophilia, diabetes, spina bifida,

* Corresponding author: Jerlym S. Porter, PhD, MPH.
E-mail address: jerlym.porter@stjude.org.

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congenital heart disorders, cystic fibrosis, juvenile rheumatoid arthritis, and sickle cell disease), 24 parents, and 17 providers found the following themes: (1) patients, parents, and providers realized that the transition to adult care was necessary; (2) systemic differences between pediatric and adult care impacted the transition process; (3) preparation for the systemic differences and changes in roles and responsibilities for disease self-management was needed for both patient and parent; and (4) more collaboration among pediatric and adult providers was essential (van Staa, Jedeloo, van Meeteren, & Latour, 2011). Tuchman, Slap, & Britto (2008) conducted semi-structured interviews about transition experiences and expectations with 22 adolescents and young adults aged 15–21 years with chronic illness (cystic fibrosis, sickle cell disease, juvenile rheumatoid arthritis, and inflammatory bowel disease). Adolescents and young adults expressed concerns related to transition such as timing of the transfer of care, ability of adult provider to care for complex medical and psychosocial needs, and their ability to advocate for themselves in the medical setting (Tuchman et al., 2008).

A comprehensive systematic review of the literature examining the perspectives of adolescents and young adults with special health care needs on health care transition yielded the following themes: (1) reflections on the transition experience, (2) recommendations for improving transition services, (3) obstacles experienced during the transition process, (4) expectations of adult care, and (5) attitudes toward self-management skills (Betz, Lobo, Nehring, & Bui, 2013). These studies taken together highlight the complexities and challenges experienced during the transition process.

Studies also have examined patient and caregiver perspectives of the transition from pediatric to adult SCD care. Specific pediatric and young adult concerns identified in the SCD literature include leaving a familiar provider and environment, being seen by a provider who may not have knowledge of SCD, establishing independence from caregivers, and having adequate health insurance (Hauser & Dorn, 1999; McPherson, Thaniel, & Minniti, 2009; Telfair, Ehiri, Loosier, & Baskin, 2004; Telfair, Myers, & Drezner, 1994). Caregivers have reported similar concerns, such as fear of their child being treated in an unfamiliar environment, changes in their role in their child's health care, and their child's ability to advocate and manage his or her disease (Hauser & Dorn, 1999; Telfair et al., 1994).

Although the extant literature includes studies examining patient and caregiver perspectives about transition, no studies in SCD to date were found which included the perspectives of other family members, such as siblings. Most research involving healthy siblings of patients with SCD has examined their general psychosocial adjustment, adjustment in the context of their brother's or sister's SCD, communication about SCD within the family, and relationship with their brother or sister with SCD (Gold, Treadwell, Weissman, & Vichinsky, 2011; Graff et al., 2010; Noll et al., 1995). Within the context of transition, siblings' unique viewpoints may offer additional insight into the transition to adult SCD care, particularly as they can provide valuable emotional support to the transitioning patient. Additionally, patient, caregiver, and sibling perspectives may provide information about each individual's expectations regarding his or her role in the transition process. A better understanding of siblings' transition perspectives as well as each family member's role expectations can yield important information to inform current transition programming and optimize family involvement and adjustment to transition. Thus, the purpose of this qualitative study was to describe the perspectives of adolescents with SCD, siblings closest in age to the adolescent, and their caregivers regarding the transition from pediatric to adult SCD care and to identify recommendations for improving the transition process.

Theoretical Framework

Bronfenbrenner's bio-ecological theory is based on his original theory of human development, which posited that a person's development occurred within the context of interactions of systems (Bronfenbrenner, 1979). The theory was altered and revised into the current bio-ecological theory with the process–person–context–time (PPCT) model (Bronfenbrenner, 2005). According to the PPCT model, the primary mechanism of development involves proximal processes in which individuals interact with others, objects, and symbols in their immediate environment. As individuals develop, these processes become more complex and are deemed more effective for intellectual, emotional, social, and moral development when they occur regularly over a period of time (Bronfenbrenner, 2005). The impact of the proximal processes on development varies as a function of the other three elements of the PPCT model: person characteristics, context, and time. Bronfenbrenner identified three person characteristics: demand characteristics (age, gender, physical appearance), resource characteristics (past experiences, skills, social and material resources), and force characteristics (temperament, motivation, persistence). Context, based on Bronfenbrenner's original theory, involves the interaction of four systems: microsystem (immediate environment such as family), mesosystem (interaction between systems), exosystem (environments with indirect influence such as the parent's workplace), and macrosystem (culture and societal norms) (Bronfenbrenner, 1979). Time, the final element of the PPCT model, includes historical events and changes that occur within the larger context of society and community over time which also can influence human development.

The Bronfenbrenner PPCT model provides a useful framework to examine transition to adult care among the sickle cell population. Transition and preparation for transition represents a dynamic process involving interaction