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Young Adults Living with Sickle Cell Disease: Contextual Insights and Recommendations for Treatment

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Background and Objectives

The challenges related to transitioning from childhood to an adult for individuals living with a chronic disease has received increased attention in the literature.⁸ These challenges can be particularly pronounced among young adults living with Sickle Cell Disease (SCD) who not only must face the transition of health care (i.e. lack of adult-centric illness management resources, limited access and availability to medical providers with education to manage SCD in adults), but also face all the realities of transitioning into adulthood while living with a complex chronic illness.^{4,19,20,28} As a result of these challenges, many young adults living with SCD experience an increased mortality risk, barriers to attaining advanced education, difficulty in maintaining steady employment, decreased quality of life, problems with disease management and navigating increased autonomy, coupling and family planning dimensions of the family life cycle.^{1,16,28,29}

Currently, there is a paucity of literature that has explored how to assist young adults living with complex chronic illnesses navigate their transition into adulthood. Further, the literature specifically focusing on SCD populations is even more sparse. For BH providers working within these populations, insight and recommendations are needed to help guide their work when assisting these individuals and families. This presentation seeks to provide insights and recommendations from the extant literature to guide BH professionals working with young adults living with SCD. Recommendations for treatment were drawn from both literature surrounding treatment for patients with SCD as well as other literature looking at complex chronic illnesses.

At the conclusion of this presentation, participants will be able to...

- identify the challenges faced by young adults transitioning to adult care.
- identify the interplay between the biological, psychological, and social levels of individuals living with SCD.
- identify recommendations on how to effectively work with young adult patients living with sickle cell disease.

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Healthcare Considerations

Navigating the Transition from Pediatric to Adult Care:

As YAs transition from pediatric to adult care they may face challenges in navigating the healthcare system (e.g. limited knowledge of accessing resources within the healthcare system, provider attitudes and/or lack of knowledge of the disease). These challenges may result in poor disease management.^{4,19,20,28}

Family Planning:

YA must consider the implications for reproduction. When partnered with an individual who either has the genetic trait for a hemoglobin mutation or an expressed form of the mutated gene; resulting offspring face an increased likelihood of either being a trait carrier or having an expressed form of the mutation.^{15,16}

Psychosocial Considerations

Separation - Individuation:

The desire for autonomy and self governance is a natural part of the transition from childhood to adulthood. Health challenges, difficulties navigating/ accessing patient care, and environment factors (e.g. SES, geographic location) may make it difficult for individuals to maintain autonomy and engage in self-determination behaviors (e.g. Expanding relational system, launching).¹

Perceptions of Caregiver Burden:

Young adults may perceive that, they will be burdening the family/relational system when a pain crisis ensues. As a result they may have reluctance in utilizing relational support during periods of crisis and make attempts to manage pain independently often even without the assistance of healthcare resources.²⁰

Relationship Development:

The process of self-disclosure is a necessary and intimate part of the dating process. Despite the benefits of disclosure in establishing and deepening developing relationships; there are a number of factors that influence an individual's decision to disclose. Individuals with genetic disorders and complex chronic conditions are faced with determining when, how, and if to disclose their medical diagnosis during the development of intimate relationships. Additionally once in relationship individuals may continue to have hesitance in disclosing the experience of pain crisis or other symptomatology to intimate partners.^{12,33}

Employment and Education Attainment:

Young adults face challenges in their ability to reach employment and educational goals which may have implications for socioeconomic advancement. Maintaining employment or working to achieve certain educational goals may be complicated by the experience of pain crisis or other illness related symptomatology that may keep individuals out of work or school for significant amounts of time.^{18,24,22}

Transition Programs:

The utilization of Peer Mentors, Transitioning Programs/Care Teams and or Patient Navigators would assist YAs transitioning from pediatric to adult care. These programs and resources would help patients in preparing for the transfer to adult care, offer needed support in learning how to navigate the healthcare system, and provide a continuation of support in the months or years following their transition. BHP who may be working in these programs may be of use in addressing psychosocial and psychological challenges faced by patients.^{3,20}

Reproduction Education:

Reproduction Education Programs, with integrated BH providers could not only offer reproductive health knowledge to individuals with SCD and SCT, they would assist individuals in navigating the relational and mental health dimensions of such decisions, while also making an informed decision about reproduction.^{5,16}

Self-Management Programs and Self Efficacy Interventions:

Utilizing a Chronic Disease Self-Management program may have utility in assisting young adults in navigating this stage of development. These programs utilize a combination of strategies geared toward increasing self-efficacy and self-management behaviors for individuals with chronic illness. Additionally, health centers may be of particular use in offering interventions on health literacy and self-efficacy. BHP may be of particular utility in offering individual or family intervention for those who may be having trouble navigating this phase of life transition.^{2,27,30}

Caregiver Support:

While patients may fear that they are burdening members of their family or relational system, individuals such as caregivers or other systems of support may similarly feel burdened at times. BHP should assess for and offer interventions that focus on caregiver burnout as caregivers may experience feelings of anxiety, stress, fatigue, and depression.³⁰

Matining Relational Functioning:

Support from a family member or intimate partner is positively associated with coping and illness management. However, some individuals feel that choosing not to engage in the relationship formation process is perceived as a viable and rational choice when determining how to navigating their genetic condition in the context of relationship formation. BHP providers can play a vital role in assessing and assisting individuals in exploring and deconstructing factors (self concept, fears, and meaning associated with rejection) that ultimately hinder engagement in the couple process or shape relational functioning.³⁰

Employment and Education Adjustments:

Employers may consider offering flexible workplace accommodations (e.g. offering part time positions, time off for medical treatment) so that individuals can maintain employment. As it relates to education programs geared toward offering support and active coping strategies to assist individuals as they seek higher education may be of use. Given their knowledge of the psychosocial challenges faced by this population, BHPs may advocate at both the patient and policy level for workplace and/or educational accommodations.²⁴