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Assessing Disease Knowledge and Self-Management in Youth with Sickle Cell Disease Prior to Transition

Barbara Speller-Brown, DNP, CPNP,
Children's National Health System, Washington, DC

The George Washington University, Washington, DC

Maureen Varty, BSN, RN,
MedStar Georgetown University Hospital, Washington, DC

Lisa Thaniel, PhD, LCSW,
Children's National Health System, Washington, DC

Marni B. Jacobs, PhD, MPH
Children's National Health System, Washington, DC

The George Washington University, Washington, DC

Abstract

Introduction: Transition of medical care from pediatrics to adult can be challenging and difficult. Until the 1970s, only half of patients diagnosed with SCD reached adulthood. As a result of patients living longer, there is a growing need to understand factors that influence readiness to transition. This descriptive study examined age-specific SCD knowledge, self-management skills of patients and education goals in a convenience sample of patients and their parents.

Methods: One hundred eighty-three transition surveys were distributed during scheduled hematology clinic visits. Surveys were analyzed with descriptive statistics to determine differences of knowledge between age groups, self-care skills, vocational and educational goals. There were 0–4 parent group (32), 5– 8 parent group (52), 9–11 parent group (12), 9–11 child group (24), 12–15 child group (31), and 16–21 adolescent and young adult group (32).

Results: Fifty percent of parents of the 0–4 age group and 33% of 5–8 age group parents knew their child's baseline hemoglobin. Only 38% of patients 16–21 knew their baseline hemoglobin. Seventy-nine percent of patients 9–11 years, 74% of 12–15 year olds and 78% of AYAs could name their hematology provider. Only 66% of patients ages 16–21 knew what symptoms required medical attention.

Discussion: Most patients and parents had adequate basic knowledge regarding sickle cell disease. AYAs lack the disease knowledge necessary to transition care away from parents to become more independent. An assessment for transition readiness should be ongoing to include disease specific knowledge and self-management skills.

Corresponding Author: Barbara Speller-Brown, DNP, CPNP, Children's National Health, System, 111 Michigan Avenue NW, Suite 4488, Washington, DC 20010, USA. bsbrown@childrensnational.org.

Keywords

Sickle cell disease; knowledge; transition; self-management

Introduction

As innovations have been made in healthcare for pediatric chronic diseases like sickle cell disease (SCD), the need has developed to care for those living after the age of 18 (American Academy of Pediatrics [AAP], 2002). Today, approximately 100,000 Americans live with SCD (Centers for Disease Control and Prevention, 2016; National Institutes of Health, 2016). Important advances including the Sickle Cell Treatment Act, universal newborn screening, penicillin prophylaxis, targeted vaccines, hydroxyurea therapy, and transcranial Doppler usage, have drastically improved life expectancy for these individuals (Gardner, Douiri, Drasar, Allman, Mwirigi, Awogbade, & Thein, 2016). In the 1970's, the expected lifespan for someone with SCD was 14 years, but as medicine has progressed, individuals with SCD are now living into their 40's and 50's (Hamideh & Alvarez, 2013). Over 93% of children with SCD are now surviving into adulthood (Quinn, Rogers, Mccavit, & Buchanan, 2010). With this increase in life expectancy, health care providers must develop transition strategies to support a life course approach to comprehensive sickle cell care.

Effective strategies for transition care have become increasingly important for this population, given that SCD patients experience increased hospitalization rates post-transition, increased complications related to SCD post-transition, and increased mortality shortly after transfer to adult health care (Blinder, Vekeman, Sasane, Trahey, Paley, & Duh, 2013; Hemker, Brousseau, Yan Hoffman & Panepinto, 2011; Brousseau, D. C., Owens, P.L., Mosso, A.L., Panepinto, J.A., & Steiner, 2010). These occurrences both increase healthcare costs and reduce individual quality of life, highlighting the need for improvements to the healthcare system to reduce risks to this population as they move through this challenging phase in their life.

Transition has been described as the "purposeful, planned movement of adolescents with chronic medical conditions from child-centered to adult-oriented health care" (Blum, 2002). This process is more complex than the adolescent and young adult (AYA) simply moving from seeing a pediatric health care provider to seeing an adult health care provider. Studies show that patients often feel anxious at the thought of transitioning to adult care and providing adequate preparation prior to transition is therefore critical (Fegran, Hall, Uhrenfeldt,Aagaard & Ludvigsen 2014; deMonatalembert & Guitton, 2014). As a process, there is a series of events that must occur for transition to be effective. This includes finding an adult health care team who can form a good relationship with the emerging adult. Transitioning also includes the development of skills and knowledge to not only navigate this change in health care providers, but also for individuals to learn how to self-manage their disease. The change requires the AYA to become more independent in health behaviors, as expected in the adult health care setting. Disease education should be a major component of the transition process, as adult primary care or emergency room providers may not be comfortable or knowledgeable about treating patients with SCD (Bemrich-Stolz, Halanych,

Howard, Hilliard & Ledensburg, 2015). Successful transition to adult care has been linked to improvements in self-care management, resulting in substantial reductions in morbidity and mortality (Lenoci, Telfair, Cecil & Edwards, 2002; Telfair, Alexander, Loosier, Alleman-Velez & Simmons, 2004)

Health care providers caring for the sickle cell population must assess various components of the transition process including transition planning, preparation, health care system design, and AYA and parental views on transition, as all of these facets play a role in producing a successful transition for an AYA with SCD (Bryant, Porter, & Sobota, 2015). Knowledge of SCD and its medical management are necessary to developing independence and self-management skills. Adult health care providers will expect that AYAs have some baseline knowledge and ability to perform health care behaviors. The primary goal of this descriptive study was to assess the knowledge of patients with SCD and their parents in order to develop an understanding of areas where disease knowledge and self-management were complete and where gaps existed.

Methods

All methods and procedures received approval from the Children's National Health System Institutional Review Board where the study was conducted. Parents and AYAs provided written informed consent and assent prior to participation. Surveys were developed using the National Hemophilia Foundation Transition guidelines (Mid III/ Mid-Atlantic Region Transition to Independence Survey). These were age-specific surveys with 4–6 ages grouped together. Each survey was tailored to fit the disease knowledge and self-management skills appropriate for patients with SCD.

Setting/Sample

The study was conducted in a large urban hospital, which provides comprehensive care to over 1700 SCD patients from birth to 21 years. One hundred eighty-three age-specific surveys were distributed during scheduled hematology clinic visits to a convenience sample of sickle cell patients and their parents. Surveys were then analyzed to determine knowledge and understanding of SCD disease management in different age groups and to identify knowledge gaps. We also evaluated vocation and educational goals. Of the 183 surveys completed there were: parents of 0–4 age group (n=32), parents of 5–8 age group (n=52), parents of 9–11 age group (n=12), patients 9–11 age group (n=24), patients 12–15 age group (n=31), and AYA 16–21 age group (n=32). A large age range was chosen so that changes in knowledge and self-management could be assessed across the life span.

Data Analysis

Descriptive statistics by patient age group were used to assess children, parents and AYA's knowledge of SCD and comfort level regarding self-care skills and disease management. For questions that had a correct response, answers were categorized as "correct" or "incorrect". If a participant left an answer question blank, then it was coded as "incorrect" during the

analysis. All questions were reported as frequency (n) and percent (%) among those who responded to the question. All summaries were completed in Excel.

Results

SCD Knowledge

Children, parents, and AYAs across age groups had varying levels of disease knowledge about SCD (Table 1). Thirty-two parents of children 0–4 years were surveyed. Ninety-four percent of parents knew their child's SCD phenotype, however only half knew their child's baseline hemoglobin. Ninety-seven percent knew that their child should avoid dehydration and cold weather, 91% knew how to palpate the spleen, 59% knew the importance of penicillin prophylaxis and 63% of parents knew at what temperature to bring their child to an ER for fever evaluation. The majority (66%) of parents also knew that the red blood cells are misshapen in SCD, 69% knew that every organ is affected, while 97% knew that SCD is inherited from both parents. Nine-one percent knew the risk factors associated with SCD. All parents felt comfortable recognizing signs and symptoms (infection, pain, and stroke) that required medical attention.

Fifty-two parents of children 5–8 years were surveyed. Thirty-three percent knew their child's baseline hemoglobin and 54% knew that SCD was inherited. Ninety percent of parents knew to get hematology clearance for their child before any surgical procedure.

Twenty-four children were surveyed in the 9–11 age group. Most children (88%) knew that SCD was a disorder of the blood, and 75% of children knew that SCD was a lifelong illness. When asked how they would treat pain, 96% knew to tell an adult right away, 92% responded rest, take pain medicine, drink more fluids, and 96 % would use heat and 71% would apply ice. In doing their part to stay healthy, 96% would drink plenty of fluids, 83% would treat pain early and go to the hospital with a fever and 92% would avoid the cold.

Twelve surveys were completed in the 9–11 parent group. Fifty-eight percent would have their child's eyes checked regularly for SCD complications and 92% thought it was important they receive an annual influenza vaccine. Seventy-five percent of parents knew about Hydroxyurea (a medication that helps to reduce pain crisis, acute chest syndrome and hospitalizations).

Thirty-one children and adolescents were surveyed in the 12–15 age groups. All knew SCD was a disorder of the blood. In response to pain interventions, none of them answered all of the questions correctly. Ninety percent would apply heat, however 68% would apply ice. In doing their part to stay healthy, 92% would drink plenty of fluids, 77% would treat pain early, 61% would avoid the cold and 65% would go to the hospital with a high fever.

Thirty-two AYAs were surveyed in the 16–21 age groups. Ninety-seven percent knew their SCD phenotype. Only, thirty-eight percent knew their baseline hemoglobin.

Disease Self-Management

Our sample reported a relative high degree of disease self-management skills (Table 2). Eighty-one percent of children and adolescents in the 12–15 age groups were familiar with sexual topics such as abstinence, safer sex practices and contraception/pregnancy. When running low on medications, all would let the parent now. Fifty-eight percent of those ages 12–15 responded they could always recognize the signs and symptoms of pain/poor response to treatment that required medical attention.

AYAs in the 16– 21 age group were asked about arrangements for travel. Eighty-eight percent would bring their medications, including pain meds, half would be able to name and know the location of the closest hospital and to be able to speak to their medical history, including surgeries and ICU hospitalizations, and arrange a way to contact the Children's hematology department if more information was needed. Sixty-six percent could always recognize the signs and symptoms of pain/poor response to treatment that required medical attention. When it was time to schedule their regular hematology clinic appointments, 34% would arrange the appointment for themselves, either while in the clinic or by calling, and 56% would have their parent make the appointment. Seventy-eight percent could name their hematology provider. Sixty-six percent knew how to make their own appointments with other doctors like pulmonology or ophthalmology. When running low on medications, 56% would call the hematology clinic to get a new prescription and 47% would let the parent now. Seventy-eight percent were unsure of how long their health insurance would remain in effect. Sixty-one percent of 12–15 year olds felt SCD would make a difference in their work choice. Eighty-eight percent of AYAs indicated their future educational plans included college. Thirty-eight percent knew about STRIVE, a psychoeducational support group for adolescents with SCD (Table 3).

Discussion

The transition from child-centered to adult-oriented health-care is often a challenging process for AYAs with SCD and their parents. Many AYAs lack important disease self-management skills to successfully transition to adult-oriented health-care (Jenson, Paul, LaCount, Peng, Spencer, et al. 2017; Wood, Sawicki, Miller et al., 2014). Transition programs that address medical, educational, and vocational needs of patients are essential to a seamless transition (Betz, Lobo, Nehring, & Bui, 2013; van Staa, van der, Stege, Jedeloo, Moll, & Hilberink, 2011). Understanding where there are knowledge deficits in these areas is critical to the development of transition programs. Providers have reported that they expect their patients to have disease knowledge after transition, making it important to understand their level of knowledge prior to transition (Telfair, Alexander, Loosier, Alleman-Velez & Simmons, 2004). In this study the majority of patients and parents across all age groups had a relatively high level of disease knowledge about SCD in most areas. However, growing older did not necessarily mean that the patient with SCD had adequate knowledge for disease management. Both AYAs and their parents had a high level of knowledge of how to assess for changes in their condition and what steps where needed to perform wellness behaviors to keep SCD under control. Understanding the etiology of SCD and how it can

impact the body is important knowledge to have when the AYA parent have conversations with their healthcare provider and when they are problem-solving during everyday life.

When reviewing the parent's answers in comparison to the AYA's responses to questions regarding SCD knowledge, the AYAs demonstrated that they had a higher than expected level of disease knowledge, which is a positive sign that as they age they will be able to assume management of their care. Even at a young age, AYAs understand that SCD is life long and that there are certain steps which must be taken to control their pain. However, there are some areas of concern as only about half of the AYAs were able to recognize signs and symptoms of pain/poor response to treatment that required medical attention. This finding is important to note as it demonstrates that healthcare providers should continue to assess knowledge levels overtime and should not make assumptions based upon age. After transition, these AYAs may have less parent involvement to aid them in decision-making, which means it is essential that they know when they must seek further medical attention. In addition, a small percentage of AYAs were not able to correctly choose which interventions would be best for their pain crisis, as they would apply ice (which can increase pain). As pain crises are the most common distinguishing issue they will face, it is critical that this population have proper knowledge of how to manage their pain, especially when they no longer have continual parental presence.

Our AYAs reported a relatively high degree of disease self-management skills. The majority of AYAs were familiar with sexual topics such as abstinence, safer sex practices and contraception/pregnancy and all knew what to do when running low on medications. While some studies report that AYAs and their parents lack important disease self-management skills (Sobota, Umeh, Mack, 2015), our findings did not completely support this. Our findings suggest in contrast to what has been reported previously, a majority of our AYAs have disease self-management skills and may be prepared for the transition to adult-oriented care, when appropriate. This is important as studies show that a higher level of disease self-management skills is associated with better readiness to transition to adult-oriented care (Treadwell, Johnson, Sisler, Bitsko, Gildengorin et al., 2015).

Vocation and educational goals are an important part of the transition process. Studies show that AYAs with SCD who are goal-oriented and motivated for the future often cope better with the transition to adult-oriented care (Smith, Lewis, Whitworth, Gold & Thornburg, 2011). Consistent with findings from the literature, the majority of our AYAs in the study showed a high motivation for the future despite their diagnosis of SCD. These results are similar to a study conducted by Sobota and colleagues (2015) where the majority of AYAs in the study reported that they had a plan for the future, and a majority said they knew the type of training/employment they needed for their career choice. Future research should be conducted to assess the health and social outcomes of having vocation and education support and motivation for the future in AYAs with SCD. Further research should also be conducted to understand what type of support may be beneficial for AYAs as they are developing independence in their disease self-management and preparing for transfer to care in the adult healthcare system.

Limitations

The generalizability of this study is limited by the small sample size in each group. There is the possibility of selection bias given that we used a convenience sample that may reflect those AYAs and/or parents who were most comfortable answering questions about their SCD. This was also conducted at a single site and may limit generalizability of the findings. Our findings were also limited by measurements that focused primarily on self-report, i.e. the ability to name their hematology provider and baseline hemoglobin level. These measures have also not been validated. Further research utilizing a larger sample and standardized measures is needed. Some participants did not respond to certain questions, and with the type of coding used to analyze the results, disease knowledge and disease self-management skills may have been underestimated. We recognize there maybe weaknesses in the tools used to survey participants as some responses that were yes/no or true/false lacked measures of validity and assessing a small area of basic SCD knowledge may not represent true objective measures of transition readiness.

Implications for practice

Sickle cell disease knowledge and self-care management skills have been linked to improved health outcomes, decrease emergency room utilization and hospital admissions. These results support the need to assess SCD knowledge and vocation and educational career goals. Healthcare providers should incorporate these essential components in a transition program designed to prepare AYAs for successful transition to adult-oriented care. Specific strategies for improvement in disease management and self-management include: starting transition readiness assessment early and incorporating specific SCD knowledge. Providing parent education and encouraging them to impart disease knowledge to their children should also begin early. We recommend measurable criteria, including those with test-retest reliability (which was not done in this study), with objective measures of transition readiness, that should be ongoing and utilized to assess youth and young adult's readiness to transition to adult care, with the goal of transitioning when developmentally ready and not based on chronological age.

Conclusion

The transition from pediatric to adult care can be a difficult and often challenging process. Successful transition depends on many factors; however, readiness, disease knowledge and disease self-management skills have been identified as key. Transitioning from pediatric to adult care is a process that occurs overtime. Disease education should be a major component of the transition process, as studies have shown that adult primary care or emergency room providers may not be comfortable or knowledgeable about treating patients with SCD. Education should begin early and continue until transition is complete. Readiness assessments should also be ongoing to include and foster disease specific knowledge. This population is at risk for decreased quality of life and life expectancy after transition to adult care. Results from this study suggested a SCD transition clinic that includes specific knowledge assessments and education as a major component of a systemic transition intervention.

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Table 1

Sickle Cell Disease Knowledge

Question	Group	N	Frequency/Correct Yes	Percentage Correct
My child has what type of SCD	0–4 Parent	32	30	94
I have what type of SCD	9–11 Child	24	22	92
	12–15 Child	31	31	100
	16–21 AYA	32	31	97
Avoiding cold/dehydration is important	0–4 Parent	32	31	97
Know baseline hemoglobin	0–4 Parent	32	16	50
	5–8 Parent	52	17	33
	16–21 AYA	32	12	38
Why it's important to take penicillin	0–4 Parent	32	19	59
How to palpate spleen	0–4 Parent	32	29	91
When to bring child to ER for fever	0–4 Parent	32	20	63
SCD causes red blood cells to be misshapen	0–4 Parent	32	21	66
Body parts that may be affected	0–4 Parent	32	22	69
SCD was passed down through	0–4 Parent	32	31	97
SCD can appear in family with no history of SCD	5–8 Parent	52	28	54
Risk factors associated with SCD	0–4 Parent	32	29	91
Recognize signs /symptoms of complications (Infection, pain, stroke)	0–4 Parent	32	32	100
SCD is a disorder of the blood	9–11 Child	24	21	88
	12–15 Child	31	31	100
I do my part to try and stay healthy by	9–11 Child	24	19	79
	12–15 Child	31	9	28
When I have pain I should:				
Apply ice	9–11 Child	24	7	29
	12–15 Child	31	21	68
Apply heat	9–11 Child	24	23	96
	12–15 Child	31	28	90
Drink more fluids	9–11 Child	24	22	92
	12–15 Child	31	27	87
Hydroxyurea helps reduce hospitalization, pain and acute chest in SCD	9–11 Parent	12	9	75

AYA= adolescent and young adult

Child=patient's response Parent=parent's response

Table 2

Disease Self-Management

Question	Group	N	Frequency/ Correct/Yes	Percent Correct
Familiar w/Sexual Topics	12–15 Child	31	25	81
Know Hematology Provider Name	9–11 Child	24	19	79
	12–15 Child	31	23	74
	16–21 AYA	32	25	78
Recognize s/s of pain /poor response requiring medical attention	12–15 Child	31	30	97
	16–21 AYA	32	30	94
When running low on medication I should:				
Let parent know	12–15 Child	31	31	100
	16–21 AYA	32	15	47
Call to get new prescription	16–21 AYA	32	18	56
When traveling I need to arrange:				
To bring medications	16–21 AYA	32	28	88
Know location/closest hospital	16–21 AYA	32	16	50
Speak to my medical history	16–21 AYA	32	16	50
Contact Hematology for more info	16–21 AYA	32	16	50
When it's time to schedule Hematology appointment:				
Parent/guardian schedules it	16–21 AYA	32	18	56
I arrange it	16–21 AYA	32	11	34
I know how to make own appointment with other doctors (i.e. Pulmonary/ Ophthalmology)	16–21 AYA	32	21	66

AYA= adolescent and young adult patients

Child=patient's own response, Parent=Parent's response

Table 3

Vocation and Educational Goals

Question	Group	N	Frequency Correct/Yes	Percent Correct
SCD will make a difference in my work choice	12–15 Child	31	12	39
Future education plans	16–21 AYA	32	28 College	88
Knowledge of after school program (Strive)	16–21 AYA	32	12	38

AYA=adolescent and young adult patients

Child=patient's response, Parent=Parent's response