Published in final edited form as:

J Pediatr Hematol Oncol. 2014 July ; 36(5): 389–394. doi:10.1097/MPH.0000000000000110.

Self-Reported Transition Readiness among Young Adults with Sickle Cell Disease

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Abstract

Background—A growing body of literature addresses the need for transition programs for young adults with Sickle Cell Disease (SCD), however studies assessing transition readiness are limited and there are few validated instruments to use.

Objective—To conduct a pilot study to assess transition readiness of patients with SCD in our transition program and to evaluate a SCD specific assessment tool which measures five knowledge skill sets (medical, educational/vocational, health benefits, social and independent living), and three psychological assessment (feelings, stress, and self-efficacy).

Results—Of 47 SCD patients between the ages of 18–22 seen in our facility, 33 completed the assessment tool. The majority of patients reported good medical knowledge of SCD and said they were motivated to pursue a career despite the burden of living with the disease. We identified knowledge gaps in the area of independent living and health benefits skills sets. A majority of patients reported being worried that their SCD would prevent them from doing things in their life; however few respondents said they were worried or anxious about their transition to adult care.

Conclusion—Adolescents beginning a transition intervention program reported a high level of knowledge of their disease and demonstrated a positive attitude towards transition with good self-efficacy.

Keywords

Sickle Cell Disease; Transition

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Disclosures: Dr. Akinlonu's work on this project was supported by a grant from the Inavale Foundation. The other authors have no conflicts of interest or funding to disclose.

Introduction

Sickle cell disease (SCD) is a group of genetic blood disorders affecting close to 90,000 Americans which leads to significant health complications throughout life¹. Thanks to recent advances in medical treatment, almost all children with SCD are now surviving to adulthood² which brings with it a need to transition from pediatric- to adult-focused care. The goal of transition is to “maximize lifelong functioning and potential through the provision of high-quality, developmentally appropriate health care services that continue uninterrupted as the individual moves from adolescence to adulthood”³. This transition process typically culminates in a transfer of medical care. Many patients experience difficulty making this transition, with evidence showing an increased rate of emergency visits, re-hospitalizations, morbidity and early mortality for SCD patients as they leave pediatric care^{2,4,5}.

Transition has recently become a focus in SCD with a combination of medical transition as well as transition in work and school to improve the health outcomes of adolescents with SCD⁶. Process measures, such as having a transition plan, and counseling adolescents with SCD about transition were given the highest priority ranking in a recent study defining quality markers in SCD⁷. However despite the growing body of literature addressing the need for transition programs for children with SCD, there are limited studies that assess transition readiness among young adult patient. For example while a survey of sickle cell centers showed that most now have a formal transition program, only 70% reported they perform transition readiness assessment just before transfer⁸.

The current SCD transition program at Boston Medical Center consists of a monthly clinic held inside the adult hematology clinic and staffed by both adult and pediatric hematologists and social workers. Patients are seen in transition clinic for their routine SCD visits as well as transition focused education twice a year from age 18 until their transfer to adult care which typically happens at age 22. In order to focus our transition education on the needs of each patient we began using a disease specific transition assessment tool developed by Dr. Telfair and colleagues⁶ and used with his permission. We have been using the information from each patient on a case by case basis to provide targeted and developmentally appropriate interventions. In order to improve our practice and assess the efficacy of our transition program we sought to use this tool in a more formal manner by screening all patients in transition clinic. Our aims in doing so were to 1) determine the overall transition readiness of young adults as they entered our transition program, 2) pilot this sickle cell specific transition assessment tool, and 3) use the results to help focus the education provided in our transition clinic.

Materials and Methods

Over a period of 2 years (October 2010 – September 2012), we gave a self-administered Sickle Cell Transition Intervention Program Skills Checklists⁶ to a convenience sample of 33 patients seen in the SCD Transition Clinic at Boston Medical Center (transition clinic). The tool consists of five knowledge skills sets and three psychological checklists assessing

core areas show to be important in providing a successful transition program^{9,10}. This is, to our knowledge, the first use of the entire tool in a clinical setting. Patients were given this readiness assessment tool on one of their first visits to transition clinic. It was done by self-report in the presence of a social worker who was available to answer questions if needed.

Descriptive statistics were analyzed through Survey Monkey¹¹. As a measure of reliability Chronbach's alpha was calculated for each scale, with a value of 0.70 or higher indicating acceptable internal consistency¹². Because of the small sample size no subset analysis was conducted. Our study was determined to be exempt from further review by the BUMC Institutional Review Board.

Skill Sets

Medical—Eighteen yes or no questions assessing the respondent's knowledge of their disease (for example "I understand how I might pass sickle cell disease on to my child").

Educational/Vocational—Eleven yes or no questions assessing readiness for the future (for example "I plan to graduate from High School/or obtain a GED").

Health Benefits—Six yes or no questions measuring what individuals know about different types of health insurance (for example "I know how my age can affect my health benefits").

Social—Nine yes or no questions to evaluate if patients have the social supports they need (for example "I have friends that I can talk to about sickle cell disease").

Independent Living—Ten yes or no questions to assess if young adults are ready to live on their own (for example "I know how to fix my own meals").

In scoring each of the five knowledge skill sets a 'yes' answer was given 1 point and a 'no' or missing response was given zero.

Psychological checklists

Self-efficacy—Eight questions asking how sure patients are in dealing with their SCD on a day-today basis (eg "How sure are you that you can keep doing most of the things you do day-to-day?"). This was answered on a 5 point Likert scale (not at all sure, not sure, neither, sure, very sure) and scored from 1-5 with a higher score indicating a higher degree of self-efficacy. (note: the original self-efficacy checklist had 9 questions, however in our copy the first questions were inadvertently mis-printed and therefore not answered by any respondents).

Sickle Cell Stress—Eleven questions to determine how much stress individuals felt from having SCD (eg. "I worry about my illness getting in the way of school work/job"). This was answered on a 5-point Likert scale (strongly agree, somewhat agree, neither agree or disagree, somewhat disagree, strongly disagree) and scored from 1 to 5 with a higher score indicating a higher degree of stress.

Feelings about Transition—Twelve questions asking how patients think they will feel about leaving pediatrics for adult care. This was answered on a 5-point Likert scale (not at all, a little bit, moderately, quite a bit, extremely) and was scored from 1-5. Most of the questions were phrased negatively (for example “I will be anxious”), with a higher score indicating more negative feelings. Three of the questions were phrased positively (for example “I will be excited/happy”) so their scoring was reversed (scored from 5-1) with higher scores indicating less positive feelings.

Results

Over the 2-year period, 47 transition aged SCD patients (18–23 years) received care in our facility, 40 attended the transition clinic at least once, and 33 completed the assessment tool. This response rate represents 70% of transition age patients or 82% of patients seen in transition clinic.

Medical Knowledge

Almost all our respondents said they could explain what SCD was (97%) and knew how to take their medications without assistance (97%). The majority (97%) also said they understood how they had gotten SCD, and 94% reported knowledge of how it could be passed on to their children. Seventy-one % of the female respondents said they knew how SCD could affect their health during pregnancy. Only 30% of patients reported knowing what their baseline hemoglobin level is.

Educational/Vocational

The majority (91%) of patients indicated they had a plan for their future, and 94% said they knew the type of training/employment they need for their choice job. Fewer patients said they could identify the type of work that could cause problems related to SCD (76%) or knew where to find information about job training and opportunities (70%).

Health Benefits

A slight majority of patients (64%) said they understood the different types of health insurance available while 61% reported knowing how their age could affect their health benefits, and that they carry their own health insurance card.

Social

Many patients reported having good social support (97%) and hobbies/activities that they enjoy doing (94%). Somewhat fewer said they had friends they could talk to about SCD (70%) and only 48% knew about community-based sickle cell programs.

Independent Living

Almost all patients (91%) said they could get their prescription medications filled on their own, and 85% reported they knew how to make their own appointments. However, fewer reported they go to their doctor’s appointment alone even at the average age of 20 years (79%).

Self-efficacy, Sickle Cell Stress, and Feeling about Transition

At entry into our transition program, a minority of adolescents reported feeling “quite a bit” or “extremely” worried (25%) or anxious (9%) about transition and only 16% said they felt ‘not at all’ or ‘a little bit’ all right to move on to an adult health care setting. Self-reported self-efficacy was high, with 88% feeling ‘sure’ or ‘very sure’ that they can keep doing most of the things they do day-to-day, however about half (54%) of respondents were ‘sure’ or ‘very sure’ that they could manage their pain in ways other than by using medications. Patients did report high levels of sickle cell stress, with 76% saying they ‘strongly agree’ or ‘somewhat agree’ that they worry about their illness getting in the way of school or work and half (51%) of patients ‘strongly’ or ‘somewhat’ agree that SCD may keep them from doing things they enjoy. A very small fraction (9%) of patients however endorsed the worry that some doctors do not trust them with pain medication.

Chronbach’s alpha for each of the three psychological checklists was >0.8 , indicating good internal consistency. The mean score and Chronbach’s alpha for the self-efficacy scale (30.3, 0.83) were consistent with what has previously been reported¹³. Measures of internal validity for the knowledge skill sets were somewhat lower, ranging from 0.46 for social to 0.65 for independent living.

Discussion

To our knowledge this is the first published study of pre-transition readiness among SCD patients using a disease specific readiness assessment tool. The results highlight the gaps in knowledge that need to be addressed to help young adults make a successful transition to adult lives and an adult focused medical system¹⁴. Overall respondents demonstrated a good understanding of their disease and better preparation for transition, along with fewer negative feeling about transition than has been previously reported^{15,16}.

As youth transition into the adult health care system, they are faced with the enormous tasks of having a good knowledge of SCD, navigating the adult setting, and acquiring self-management and independent living skills. This change in health behavior takes time and ongoing preparation¹³. Most adult providers expect their patients to have an understanding of their disease during visits and communicate independently¹⁷. Our respondents showed promising competence in this area with almost all reporting a good knowledge of SCD.

As patients with SCD are living longer they have more opportunities for school and career. Our patients showed a high level of motivation for the future despite their diagnosis of SCD. We encourage patients to disclose their diagnosis at school and at work and to be aware of job accommodations for people living with SCD. Young adults should also be encouraged to make use of community resources on finding jobs and other social services that will foster a successful career.

The fear of being unable to pay for health care costs is known to be a barrier to a successful transition^{18,19}. The highest un-insured groups of people are young adults between 18–24 years²⁰, and the type of insurance a patient has strongly affects his/her accessibility to specialty care in the adult setting¹⁷. Consistent with findings from the literature, our

respondents reported low levels of understanding about health insurance²¹. Since gaps in coverage can lead to a loss of medical care, it is important to provide anticipatory guidance about health insurance before adolescents become adults.

Several studies have highlighted the importance of patients being able to manage their own medication prior to transfer to adult care²². It is encouraging therefore that our patients reported being able to independently manage their own appointments and medications. We can encourage parents to allow their children to display this independence by going to medical appointments alone starting a younger age.

Overall, a larger fraction of our respondents claimed they had control over their SCD at this point in the transition process. They said they could control their pain and manage their day-to-day lives while dealing with the frustrations of having the disease. In addition, almost all of our respondents reported they had a good social support system which is a good predictor to adaptation to chronic illnesses²³. However a good number of patients still say they do not have friends they can talk to about their SCD, which highlights the social isolation patients can face dealing with a disease that is still very much stigmatized.

Limitation

Our study has a few limitations. Firstly, we used a convenience sample of SCD patients which could pose a threat to validity because of the selection bias. However, respondents reflected the majority of transition-aged SCD adolescents in our practice so is likely to be representative of our patients. Since our sample size was small, further studies using a larger number of patients are needed to validate the assessment tool we used. We did not ask patients to demonstrate any skills, simply to report them, which may have led to over-reporting of positive results.

Despite these limitations, we have been able to show that our young adult patients beginning a transition program claim satisfactory knowledge and self-management skills as they begin to prepare for a transfer to adult health care. These results predict promising outcomes at the end of a transition intervention program.

Conclusion

Our transition program already has many promising features in place. For example every patient meets the adult hematologist prior to being transferred to their care, which is not common among transition programs, but has been associated with improved post-transition access to care^{24,25}. Our transition readiness assessment identified gaps in knowledge that we have taken steps to fill through the creation of a transition specific guidebook that is given to each patient at their first clinic visit²⁶. Next steps in research include examining associations between the sickle cell specific assessment tool used in this study and patient outcomes, and applying it longitudinally to measure knowledge improvement over time.

Acknowledgments

Dr. Akinlonu's work on this project was supported by a grant from the Inavale Foundation.

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Figure 1. Patients self-reported feeling of a) feelings about transition, b) sickle cell stress, and c) self-efficacy
 *scales reversed for questions regarding positive feelings rather than negative/neutral feelings

Table 1

Demographic characteristics of respondents.

Variables	N (%)
Gender	
Male	18 (55)
Female	15 (45)
*Race	
African American	23 (82)
Hispanic	5 (18)
*Sickle cell genotype	
SS	20 (74)
SC	6 (22)
Sβ ⁺ Thalassemia	1 (4)
*Education	
Completed High School	8 (44)
College/Technical	10 (56)
Age	
18–20	22 (67)
>20	11 (33)
Average age (years)	20

* some respondents did not answer all questions

Table 2

Summary of responses to the 5 knowledge skill sets

Variable	N (%)	N (%)
Medical	Yes	No
I can tell someone what SCD is	32 (97)	1 (3)
I know what my hemoglobin level is	10 (30)	23 (70)
I know how to take my medications without assistance	32 (97)	1 (3)
I understand how I got SCD	32 (97)	1 (3)
I understand how I might pass SCD on to my child	31 (94)	2 (6)
I understand how SCD could affect my health in pregnancy	10 (71)	4 (29)
Educational/Vocational		
I have a vision for my future	30 (91)	3 (9)
I plan to attend college or obtain post high school training	33 (100)	0 (0)
I know what type of training and/or employment I might need for my choice job	30 (94)	2 (6)
I know the type of work situations that could cause problems related to SCD	25 (76)	8 (24)
I know where to find information about job training and opportunities	23 (70)	10 (30)
Health Benefits		
I understand the different types of health insurance available	21 (64)	12 (36)
I know how my age can affect my health benefits	20 (61)	13 (39)
I carry my own copy of health insurance card	20 (61)	13 (39)
I have, or am working on a portable medical history form	4 (13)	27 (87)
Social		
I have hobbies or activities that I enjoy	31 (94)	2 (6)
I have friends that I can talk to about SCD	23 (70)	10 (30)
I have good social support system	32 (97)	1 (3)
I know about community based SCD programs	16 (48)	17 (52)
Independent Living		
I have held a full or part-time job	24 (73)	9 (27)
I know how to get my prescription medications filled	30 (91)	3 (9)
I know how to make my own doctor's appointment	28 (85)	5 (15)
I go to my doctor's appointments on my own	26 (79)	7 (21)

Table 3

Descriptive Statistics and Internal Consistency Rating for each Scale

Scale	Mean (SD)	Range	Chronbach's alpha
Sickle cell stress	39.6 (9.6)	21–55	0.86
Feelings about transition	26.0 (9.5)	8–46	0.86
Self-efficacy	30.3 (6.2)	16–40	0.83
Independent living skill set	8.58 (1.7)	4–10	0.65
Medical skill set	14.1 (2.2)	9–17	0.60
Health benefits skill set	3.45 (1.5)	0–6	0.56
Educational/vocational skill set	7.52 (1.8)	2–10	0.55
Social skill set	7.45 (1.4)	4–9	0.46