

Transitioning to adult care among adolescents with sickle cell disease: A transitioning clinic based on patient and caregiver concerns and needs

Robert D Lutzman, PhD^{1,2}, Suvankar Majumdar, MD*³, Carolyn Bigelow, MD⁴, T David Elkin, PhD^{1,3}, Mary G Smith, MD³, Gail C Megason, MD³, Priya Srivastava, BA⁵ and Rathi Iyer, MD³

¹Department of Psychiatry and Human Behavior, University of Mississippi Medical Center, Jackson, Mississippi, ²Department of Psychology, Georgia State University, Atlanta, Georgia, ³Department of Pediatrics and ⁴Department of Medicine, University of Mississippi Medical Center, Jackson, Mississippi and ⁵University of Mississippi College of Medicine, Jackson, Mississippi, United States of America

Abstract

Transition from pediatric to adult health care has been identified as a very difficult time for the adolescent with SCD. While there is good evidence for the need of transitional programs, there is little data available on the outcome of such interventions. The current study assessed the general feelings and attitudes regarding transition of health care in patients with SCD, and the effectiveness of a transitional sickle cell clinic model with regard to psychological outcomes. Procedure: A questionnaire with overall measures of transitional-related concerns and needs and general feelings regarding transition from pediatric to adult care was administered to patients/caregivers attending a pediatric sickle cell clinic, a transitional sickle cell clinic and an adult sickle cell clinic. The transitional clinic is run in parallel with the adult clinic and staffed by a pediatric hematologist already known to the pediatric patients. Results: The number one area of concern for pre-transition patients was meeting new caregivers. Caregivers of pre-transition patients identified leaving behind the previous doctor as the number one area of concern. The most important concern for patients both in the transitioning and adult clinic was being seen in the adult emergency room. Compared to both pre-transitioning and adult clinic patients, transitional patients reported significantly lower levels of negative affect (fear and sadness; $p < 0.001$), and higher levels of positive affect for joviality ($p < 0.01$). Conclusion: The transitional sickle cell clinic model appears to be promising to help SCD patients cope with the transitioning process.

Keywords: Sickle cell disease, transition, adolescents.

Introduction

Sickle cell disease (SCD) is a chronic illness that affects about 80,000 individuals in the United States (1). It is an autosomal recessive genetic disorder resulting from a single base mutation in the beta-chain of the hemoglobin molecule that leads to sickling of

* **Correspondence:** Suvankar Majumdar, MD, Department of Pediatrics, Division of Pediatric Hematology/Oncology, University of Mississippi Medical Center, 2500 N State St., Jackson, MS 39216, United States. E-mail: smajumdar@ped.umsmed.edu

red blood cells. The classic clinical features are recurrent pain crisis from vaso-occlusion and chronic organ damage. Infants identified with SCD by the newborn screen are initially seen by the health care provider within the first few months of life, and patients who attend their scheduled appointments until adulthood will potentially be seen at least 40 times by the same group of providers, and even much more often if they have recurrent pain crisis (2). Consequently, over the years, patients as well as parents/families develop a sense of trust and understanding with their provider. However, there comes a time, usually not well defined and rather variable, when the patient is transferred to adult care.

As an increasing number of adolescents with SCD are surviving into adulthood and ready to transfer from pediatric to adult care, transition has become an important issue (3). Many adolescents and young adults with SCD living with chronic pain and anemia need additional supports to become independent in disease self-management/utilization of adult health services (3,4). As such, without adequate transitioning services, many SCD adolescents and families are left unprepared and potentially distressed, which may foster non-adherence with follow-up appointments with new adult health care providers (4). Moreover, adolescent SCD patients lost to follow-up during the transitioning process will likely receive inadequate medical care, eventually placing them at a greater risk of dying from the complications of chronic organ damage. Multiple studies have shown that individuals with SCD as well as other chronic illnesses not only have many fears and concerns related to transitioning to adult care, but more importantly have shown that there is a need for a formal transitional program in order to help individuals transition to adulthood (3,5-10).

In an attempt to begin to address the needs of SCD patients transitioning from pediatric to adult care, the University of Mississippi Medical Center has set up a transitional sickle cell clinic. Patients seen at the pediatric sickle cell clinic are transferred to the transitional sickle cell clinic at about the age of 16 years. The transitional sickle cell clinic is staffed by a pediatric hematologist with whom the patients/families are already familiar from the time they were seen at the pediatric sickle cell clinic. The current study represents a first step in evaluating this

clinic model by examining cross-sectional data comparing three groups of patients at various stages of transition. Specifically, the current study assessed the concerns and needs of patients regarding transition of health care from adolescence to adulthood in patients with SCD, and the general feelings regarding transition across three patient groups.

Methods

Participants included 71 African-American patients aged 14-to-26-years with SCD recruited over a 10 month period during their scheduled clinic visits. Participants were recruited from three clinics within a large university medical center in the Southern United States: The first group (n = 20; Mean age = 16.95 years; 40% female) were recruited from a pediatric sickle cell clinic (pre-transition); the second group (n = 27; Mean age = 18.21 years; 55.6% female) were recruited from the transitional (sickle cell) clinic; and the third group (n = 24; Mean age = 20.75 years; 41.7% female) were recruited from an adult sickle cell clinic consisting of patients who had recently transitioned to adult care but had not been seen at the transitional clinic. Data were also collected from 41 of the caregivers of the pediatric (n = 20) and transitioning (n = 21) patients. See table 1 for complete demographic information on the sample.

Although the pediatric clinic is held at a different location, the transitional and adult clinics are both run in parallel at the same clinic location and on the same day. The transitional clinic is staffed by the pediatric hematologist with one patient room next to the adult sickle cell clinic with six patient rooms. Transitioning patients are introduced to the adult hematologist at their scheduled transitional sickle cell clinic appointment. The same nurses and social workers manage both the transitional and adult sickle cell patients. In the pediatric clinic, the concept of transition to adult health care is introduced and discussed with the patient/caregiver by the nurse educator and pediatric hematologist at about 14 years of age. At clinic visits, patients/families are educated about the process of transition to adulthood and evaluated for transition readiness by one of the health care providers on an individual patient-to-patient basis that takes into consideration not only the age of

the patient but also their general knowledge about SCD, their health care practices and preparation for self-management of their health care, and if necessary, referred to other services such as the Children's Medical Program (CMP) and the Mississippi Sickle Cell Disease Foundation for other community resources to help with education and self-management of SCD.

Procedure

All study procedures were approved by the Institutional Review Board at the University of Mississippi Medical Center. Exclusion criteria included: history of stroke, use of psychoactive medication, inability to complete questionnaire, or any acute presentation requiring treatment (such as sickle cell pain crisis, fever), that were assessed via patient or caregiver report. Inclusion in the study was based on a known diagnosis of sickle cell disease, regardless of genotype and severity. Participants provided informed consent/assent before completing the questionnaire packet.

Measures

As part of the assessment battery, participant demographic information was obtained which included date of birth and gender. Participants and their caregivers were administered a SCD-appropriate modified version of the John Hopkins Adult Cystic Fibrosis Program Survey (11). The SCD Modified-JHACFPS consisted of 16-items across two categories: concerns related to transitioning and importance of various services within an adult sickle cell program. Fifteen of the 16 items were consistent with the original JHACFPS with only slight modifications (e.g., CF changed to SCD). Additionally, one additional item was written assessing concerns with regarding to being seen in the adult emergency room. Items regarding general views regarding transition as well as the opportunity to participate in the study were also administered but were not included in the current study. Participants responded on a 5-point Likert-type scale indicating their feelings with responses ranging from "very

slightly or not at all" to "extremely" for the concerns and importance items and from "very positive" to "very negative" for the general views on transitioning items. For a complete list of items please see figures 1 and 2.

Participants and their caregivers were administered the Positive and Negative Affect Schedule—Expanded Form (PANAS-X) to assess participants' general feelings regarding transitioning.(12) The PANAS-X is a 60-item list of adjectives that consists of scales measuring both broad domains, Negative Affect (NA) and Positive Affect (PA) as well as specific affects or emotions. NA reflects a situation-specific experience of negative emotion and PA reflects an individual's short-term, context-specific, experience of positive emotions. For the current study, we were most interested in these two broad dimensions and four of the specific affect scales most relevant to emotions related to transition: Fear (e.g., Afraid, Nervous), Sadness (e.g., Sad, Blue), Joviality (e.g., Joyful, Excited), and Serenity (e.g., Calm, At Ease). The other specific affect scales included on the PANAS-X were not as relevant (e.g., Attentiveness, Guilt). Participants responded on a 5-point Likert-type scale to indicate the extent to which they had felt a certain way regarding the "transition from pediatric to adult care." The PANAS-X scales are the most widely used measurement tool used to assess mood and affect, have been shown to be internally consistent and have demonstrated excellent discriminant and convergent validity (12). Internal consistency (Chronbach's alpha) in the current sample was good ranging from .90 to .93 for patient-and caregiver-reported broad dimensions and .72 to .95 for patient- and caregiver-reported specific affects.

Analyses

Analyses were conducted in two phases. The first phase of analyses consisted of examining patient and caregiver concerns and needs related to transitioning from pediatric to adult care. To examine the most important needs and concerns of patients and their caregivers, mean scores were calculated for each of the SCD Modified-JHACFPS items.

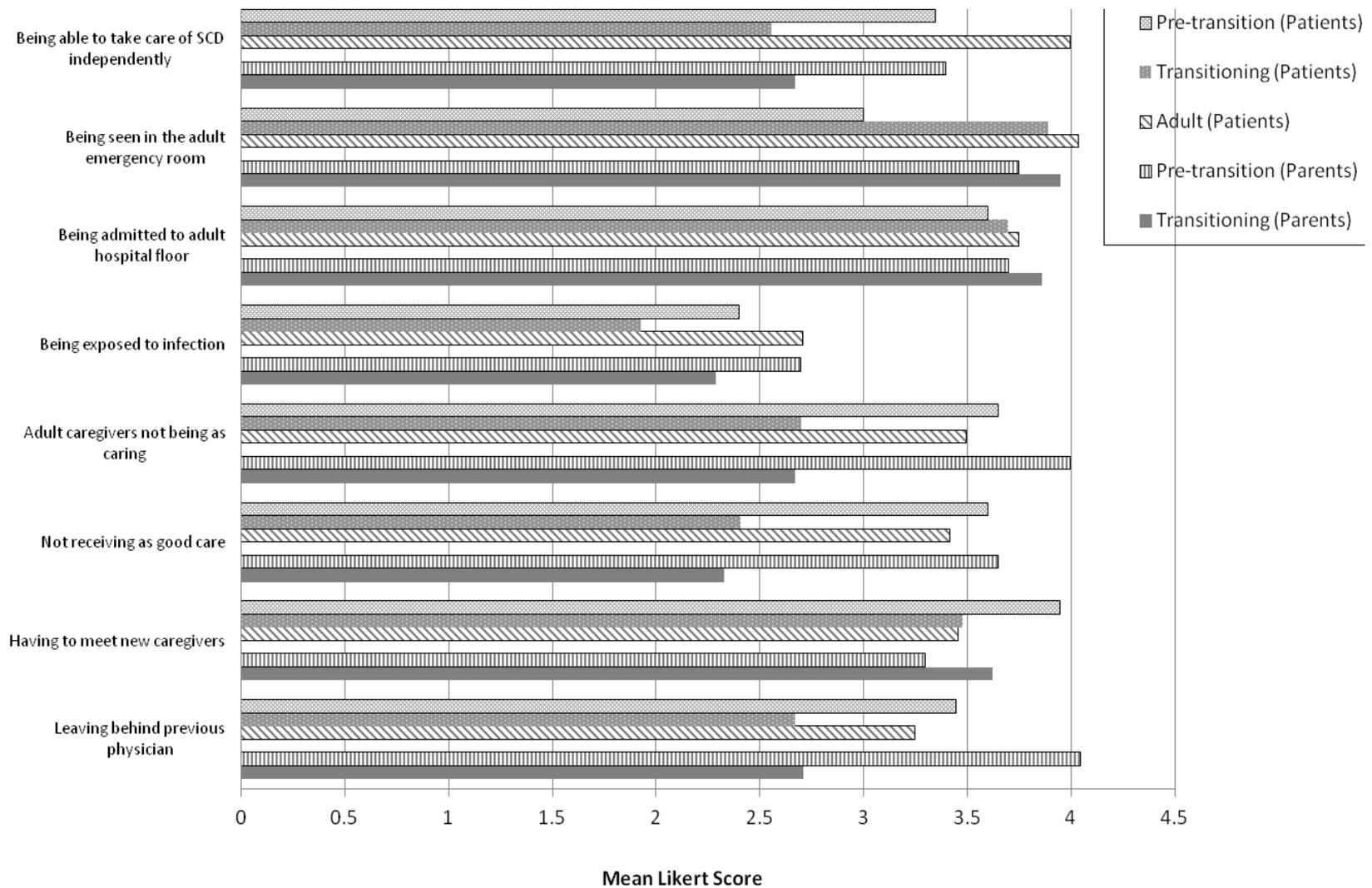


Figure 1. Participants response on a 5-point Likert-type scale indicating their feelings.

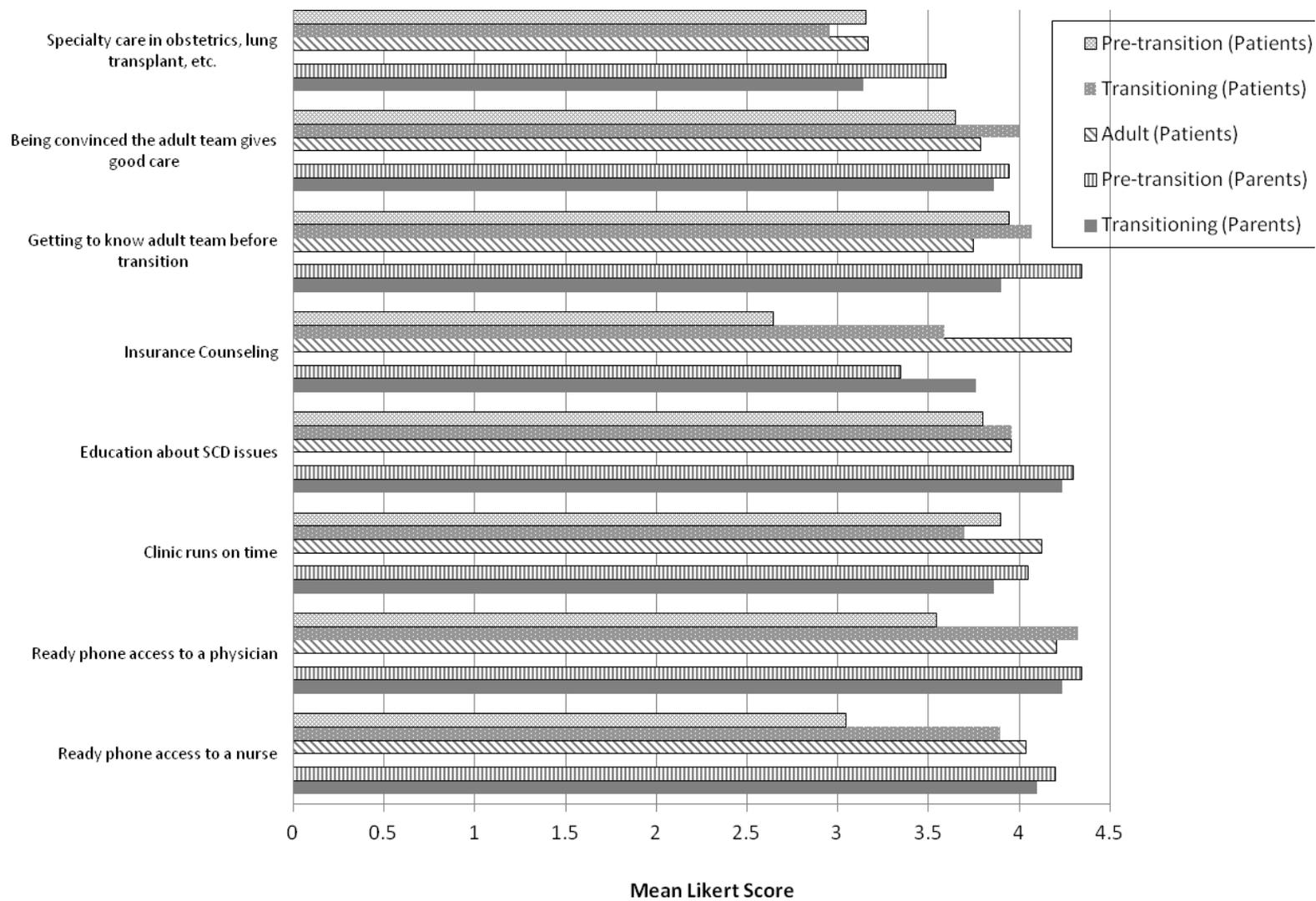


Figure 2. Participants response on a 5-point Likert-type scale indicating their feelings.

A one-way analysis of variance (ANOVA) was conducted to examine differences in total patient-reported concerns across the three patient groups and an independent samples t-test was conducted to examine total caregiver-reported concerns between the two groups for which caregiver-report was collected. The second phase of analyses consisted of examining the effects of the transitioning clinic on general feelings regarding transition, both positive and negative. To do so, patient reports of general feelings regarding transition across the three clinics were compared via a one-way analysis of variance (ANOVA). When a significant ANOVA was found, post-hoc pairwise comparisons utilizing a Tukey correction to account for Type I error were performed. For caregiver-reported general feelings regarding transition, independent samples t-tests were performed as only two groups had caregiver reports (the adult sample did not).

Results

Patient- and caregiver-reported areas of concern associated with transitioning to adult care for each group of patients are shown in figure 1 with patient-report first and caregiver-report second. When the various concerns were summed into a total score, groups did not significantly differ in terms of total concerns for either patient- ($F = 2.86, p > .05$) or caregiver- ($t = 1.80, p > .05$) report. The number one area of concern for pre-transition patients was meeting new caregivers ($M = 3.95 + 1.23$). Pre-transition patients also identified adult caregivers not being as caring ($M = 3.65 + 1.73$), not receiving good care ($M = 3.60 + 1.54$), and being admitted to the adult hospital floor ($M = 3.60 + 1.50$) as important concerns. Caregivers of pre-transition patients identified leaving behind the previous doctor ($M = 4.05 + 1.10$) as the number one area of concern. They also identified adult caregivers not being as caring ($M = 4.00 + 1.45$) and being seen in the adult emergency room ($M = 3.75 + 1.59$) as significant areas of concern.

The primary concern for patients in the transitional clinic was being seen in the adult emergency room ($M = 3.89 + 1.40$). Transitioning patients also identified being admitted to the adult

hospital floor ($M = 3.70 + 1.30$) and meeting new caregivers ($M = 3.48 + 1.01$) as significant areas of concern. Caregiver of transitioning patients identified being seen in the adult emergency room ($M = 3.95 + 1.36$) as the number one area of concern. Being admitted to the adult hospital floor ($M = 3.86 + 1.11$) and meeting new caregivers ($M = 3.62 + .97$) were also significant areas of concern.

For adult patients who did not go through the transitional clinic, the primary concern with transitioning was being seen in the adult emergency room ($M = 4.04 + 1.43$). These patients also identified being able to take care of SCD independently ($M = 4.00 + .93$) and being admitted to the adult hospital floor ($M = 3.75 + 1.36$) as significant areas of concern.

Relative importance of various services in an adult sickle cell clinic

The relative patient- and caregiver-reported importance of various services in an adult SCD clinic is shown in figure 2, again, with patient-report first and caregiver-report below for each item. Pre-transition patients identified getting to know the adult team before transition ($M = 3.95 + 1.10$) as the most important aspect of an adult clinic. The clinic running on time ($M = 3.90 + .97$) and education about SCD issues ($M = 3.80 + 1.20$) were also identified as important aspects of an adult clinic. Caregivers of pre-transitioning patients identified getting to know the adult team before transition ($M = 4.35 + .81$) and ready phone access to a physician ($M = 4.35 + 1.09$) as the most important aspects of an adult clinic.

The most important need in an adult SCD clinic identified by transitioning clinic patients was ready phone access to a physician ($M = 4.33 + .92$). Getting to know the adult team before transition ($M = 4.07 + 1.24$) and being convinced the adult team gives good care ($M = 4.00 + 1.14$) were also identified as important. Caregivers of patients in the transitioning clinic identified ready phone access to a physician ($M = 4.24 + 1.26$) and education about SCD issues ($M = 4.24 + 1.09$) as the most important aspects of an adult clinic.

For adult patients who did not go through the transitioning clinic, the most important aspect of an

adult sickle cell clinic identified was insurance counseling ($M = 4.29 + 1.23$). Ready phone access to a physician ($M = 4.21 + 1.14$) and a nurse ($M = 4.04 + 1.04$) were also identified as important.

Comparison of group difference in patient-reported feelings regarding transition

Mean levels of participants' general feelings regarding transition across the two broadband affect dimensions and the four specific affects are presented in table 2. Results of one-way ANOVA analyses showed a significant effect of group on all affect scales (all F values > 8.6 , all p values $< .001$). Post-

hoc comparisons utilizing a Tukey-correction for multiple comparisons indicated that transitional clinic patients, as compared to both pediatric and adult clinic patients, reported significantly lower levels of Negative Affect, the broadband dimension, and lower levels of the specific affect scales of Fear and Sadness. In addition to lower levels of Negative Affect, transitional clinic patients reported significantly higher levels across all positive affect scales. Specifically, transitioning patients reported higher levels of Positive Affect, the broadband dimension, and higher levels of Joviality and Serenity, the two specific affect scales (see table 2).

Table 1. Patient-report feelings regarding transition across groups

Pre-Transition	Transitioning	Adult	F	Post-Hoc Comparisons	
Negative Affect	2.42 (.99)	1.36 (.49)	2.29 (.86)	13.00	Peds > Trans < Adult
Fear	2.79 (1.34)	1.31 (.51)	2.48 (.97)	15.60	Peds > Trans < Adult
Sadness	2.12 (1.04)	1.30 (.52)	2.19 (.58)	9.68	Peds > Trans < Adult
Positive Affect	2.42 (.99)	3.33 (.96)	2.12 (.60)	13.61	Peds < Trans > Adult
Joviality	2.29 (1.15)	3.37 (1.16)	1.91 (.58)	14.73	Peds < Trans > Adult
Serenity	2.45 (1.18)	3.44 (1.06)	2.29 (.99)	8.63	Peds < Trans > Adult

Note. Values are mean (SD). All F 's significant at $p < .001$.

Table 2. Caregiver-reported feelings regarding transition

Psychological Outcome	Pre-transition	Transitioning	t
Negative Affect	2.29 (.91)	1.31 (.36)	4.55***
Fear	2.61 (1.18)	1.24 (.37)	5.06***
Sadness	1.95 (.93)	1.27 (.42)	3.09**
Positive Affect	2.52 (.82)	3.39 (.86)	-3.31**
Joviality	2.31 (1.11)	3.38 (1.04)	-3.18**
Serenity	2.59 (1.03)	3.24 (1.09)	-1.95

Note. Values are mean (SD). ** $p < .01$ *** $p < .001$.

Comparison of group difference in caregiver-reported feelings regarding transition

Mean levels of caregivers' general feelings regarding transition across the two broadband affect dimensions and the four specific affects are presented in table 2. Results of independent samples t -tests showed a significant effect of group (pediatric vs. transitional) on all affect scales (all t values > 13.11 , all p values $<$

.01) except for Serenity. Consistent with patient-report, caregivers of transitional clinic patients, as compared to caregivers of pre-transitioning patients, reported significantly lower levels of Negative Affect, the broadband dimension, and lower levels of the specific affect scales of Fear and Sadness. In addition to lower levels of Negative Affect, transitional clinic patients reported significantly higher levels of Positive Affect, the broadband dimension, and

joviality. No differences were found between the two groups for serenity (see table 2). Mean levels of caregivers' general feelings regarding transition across the six affect scales are depicted graphically in figure 2.

Discussion

Transition to adulthood from adolescence is considered to be an extremely difficult period as this is a time when the adolescent experiences much internal conflict about their role in society and the need for independence from childhood. Transition to adult health care becomes more difficult in individuals with chronic illnesses since they have to learn to assume primary responsibility for their condition, and make a change in providers. While several studies have identified the problems associated with transition in SCD and other chronic illnesses, to date there is little data available on the outcome of interventions such as a transitional clinic (13-18). The current study assessed patient and caregiver transitioning-related concerns/ needs, and a clinic model designed to address the difficulties related to transitioning from pediatric to adult care in SCD.

Our study provides insight into the specific concerns and needs of individuals and families of three transitioning patient populations: pediatric patients, transitional clinic patients, and adult SCD patients who had not been previously seen in the transitional clinic. Although the three groups differed in most important areas of concern and needs, a good deal of convergence emerged across groups and reporters. The most important concerns identified by pre-transition patients included: meeting new caregivers, adult caregivers perceived as not being as caring, and not receiving good care. Caregivers of pre-transition patients reported being most concerned about leaving behind the previous physician. Both pre-transition patients and caregivers identified getting to know the adult team before transition as the most important need. These findings are similar to the results of a national survey by Telfair et al (3), which showed that for 172 SCD respondents, the most common concerns were: lack of information about transition to adult care, fear of leaving the healthcare

provider with whom they were already familiar, and fear that new health care providers may not understand their needs.

The formulation of our transitional sickle cell clinic appears to address some of these concerns. While meeting new caregivers and leaving behind previous physicians was less of a concern reported by patients that attended the transitional clinic, the most important areas of concerns included: being seen in the adult emergency room, and being admitted to the adult hospital floor.

Although the transitional clinic model was not able to address all areas of concern (e.g., being seen in the adult emergency room), it does represent one of the first known attempts to begin to address the vital need for targeted approaches with SCD patients transitioning from pediatric to adult care. Our findings suggest that such an approach may lead to significantly improved feelings regarding this transition. Specifically, we found significantly lower levels of general Negative Affect, Fear, and Sadness and significantly higher levels of general Positive Affect, Joviality, and Serenity in terms of patient-reported feelings regarding transition for those patients in the transitional clinic as compared to patients in both the pre-transitional (pediatric) clinic and the adult clinic. These findings suggest that our clinic model may represent a promising approach to dealing with transition from pediatric to adult healthcare for patients with SCD. Additionally, this clinic model is likely more compatible with larger sickle cell centers which have a) both adult and pediatric hematologists and b) the relevant clinic space available to accommodate a pediatric hematologist running the transitional clinic in parallel with the adult clinics.

However, the current investigation is not without limitations. Due to the cross-sectional nature of the current study, causal conclusions regarding the transitioning clinic model are not possible. Rather, the current study provides a cross-sectional comparison of three groups of SCD patients at various stages of transition from pediatric to adult care. Additionally, there could be a disease-modifying effect that such a design is not able to detect. Specifically, adults with SCD may be sicker, and therefore experiencing higher levels of distress, though the lack of difference between the pre-transition and adult groups across all

PANAS-X scales makes this possibility less likely. Furthermore, while the current study provides essential data regarding the effect of a transitioning clinic on relevant psychological outcomes (i.e., feelings regarding transition), future research would benefit from the examination of other important outcomes such as number and severity of pain crises and visits to the emergency room. Lastly, SCD-related factors were not examined in the current study. As such factors are likely not independent of psychological outcomes (19), it will be important for future research to examine such disease-related factors and how they may interact with the transitioning process to result in more maladaptive versus adaptive outcomes.

In sum, the current investigation represents an important initial step in assessing the transitioning-related concerns and needs of patients with SCD and their families and evaluating a clinic model designed to address these concerns and needs. Results suggest that this clinic model that begins to address many of the concerns and needs of patients with SCD and their families, may result in lower levels of negative, and higher levels of positive feelings regarding transition from pediatric to adult care.

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