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Transition from Pediatric to Adult Care by Young Adults with Chronic Granulomatous Disease: the Patient's Viewpoint

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Abstract

Purpose—Children with chronic illnesses are living longer, prompting health care provider attention to the transition from pediatric to adult care. Transition of care is successful when youth are independent in managing their health. The aims of this study were to identify the strengths and barriers to transition from pediatric to adult care and to determine strategies that could enhance the transition process.

Methods—A survey was administered via a structured interview to 33 young adult participants (19 to 27 years of age), living with Chronic Granulomatous Disease (CGD) all of whom transitioned from pediatric to adult care. The participants were predominately male (88%) and Caucasian (73%). Topics covered in the survey included understanding of disease and treatment, adherence, advance care planning, and barriers to transition. Data was analyzed using a conventional content analysis approach.

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Results—Seventy-six percent of the participants did not understand their disease process and only fifty percent understood their prophylactic medication regimen. Seventy-five percent of participants perceived their transition as uneventful. Ninety-four percent were independent in self-management skills such as making appointments and ninety percent in refilling prescriptions. More than half of the participants thought that the transition process needed improvement. Specific suggestions to create a practical approach to transition were offered.

Conclusions—Gaps in disease-related knowledge and transition planning were identified by adolescents and young adults living with CGD. The findings suggest the need for enhancing the transition process utilizing interdisciplinary collaboration to develop a transition policy and program.

Keywords

health care transition; health care self-management; pediatric onset chronic illness; adolescent and young adult; chronic granulomatous disease

Due to medical advances, children with chronic illnesses are surviving well into adulthood requiring eventual transfer from pediatric to adult health care.^{1–5} This change can be challenging for adolescents and young adults (AYA). Many chronically ill AYA face practical challenges, have remained more dependent on their parents than their peers, and lack independence in self-advocacy and self-care management.^{5–8} A poor transition can result in increased morbidity and mortality as well as poor social and educational outcomes.⁹ Many medical organizations, such as the American Medical Association and the American Academy of Pediatrics, recognize the significance of a successful transition to adult care. The transition literature addresses youth with HIV, cystic fibrosis, sickle cell disease and other chronic conditions, but an examination of the unique transition needs of youth living with Chronic Granulomatous Disease (CGD), a rare primarily X-linked genetic immunodeficiency affecting 1/200,000 to 1/250,000 live births, has not yet been published.^{5, 10–16}

There are over 2000 participants between the ages of 10 and 24 enrolled in active clinical trials in the National Institute of Allergy and Infectious Diseases (NIAID), National Institutes of Health (NIH). CGD is one of several immunodeficiencies being studied within the NIAID intramural program and 94 of these young clinical trial participants have CGD. This number represents approximately half of the total population of individuals with CGD followed at NIH at the time of the study. NIH follows the largest cohort of people with CGD in United States. CGD is a chronic disease characterized by frequent bacterial and fungal infections requiring close monitoring and prophylactic drugs. Treatment of infections often requires hospitalizations, IV medications, and sometimes surgery. Despite this large AYA population, there is no formal program to transition individuals with CGD or other immunodeficiencies from pediatric to adult care. Although most AYA have primary care physicians outside of the NIH Clinical Center (NIH-CC), many rely on the facility's providers for specialized care for their complex conditions, resulting in potentially unique transition needs for this population, not currently addressed in the literature. To better understand the transition experience of AYA with CGD, the authors designed an exploratory retrospective cross-sectional study to answer questions related to health literacy, self-

management, and the differences between pediatric and adult care. The study had two aims: (1) to identify and describe factors that AYA with CGD found to be strengths or barriers in the transition from pediatric to adult care in the Institute, and (2) to explore strategies that these AYA believe may enhance the process of transition from pediatric to adult care at the hospital center.

Methods

Participants

Ninety-four individuals enrolled in studies conducted within NIAID had a documented diagnosis of CGD and were between 18 and 24 years of age during the period of January 1, 2011 to February 28, 2014. All participants had to be currently enrolled in one of the Institute's three CGD protocols, have had a medical appointment at the facility prior to their 18th birthday, with at least one visit during the aforementioned time period, and be fluent in English. The study protocol was approved by the NIAID Institutional Review Board prior to participant recruitment. Thirty-nine AYA were eligible to participate and 33 enrolled in the study. Interviews were conducted between June 2014 and October 2015.

Measures

Since there was no validated measure available to assess the quality of a patient's transition from pediatric to adult care, the study team adapted a published transition readiness questionnaire used with HIV positive patients.¹² The questionnaire included open-ended and yes/no questions that assessed general disease knowledge, knowledge of medications, and self-management skills. Advance care planning was also explored in the questionnaire due to a lack of information in the existing AYA transition literature despite the fact that the Patient Self-Determination Act of 1990 requires that hospitals ask patients 18 years and older if they have an advance directive or would like information about one.¹⁷ Demographic information related to self-management skills were also obtained through the questionnaire.

Without using prompts, participants were asked to tell interviewers about their disease. Responses were scored according to a 5-point system developed in consultation with two expert physicians in the diagnosis and management of CGD. A participant received one point for verbalizing each of the following elements of CGD: 1) CGD is an immunodeficiency; 2) white blood cells do not function properly; 3) the disease leaves them susceptible to bacterial and fungal infections; 4) the condition is inherited; and 5) the disease predominately affects specific systems including the lungs, liver and the gastrointestinal system. Participants were designated as understanding their disease if they were able to identify three or more of these elements.

Medication understanding was measured by asking the AYA about their prophylactic CGD medication regimen. One point each was received for being able to identify the name and purpose of both their anti-fungal and anti-bacterial medications for a possible total of 4 points. All responses were weighted equally. A score of 4/4 was required for understanding medication prophylaxis. Lack of understanding and failure to follow the regimen may result in preventable infections.

Procedures

The investigators used a convenience sample to identify and recruit eligible subjects. The interview was conducted by phone or face-to-face at the time of initial contact after oral informed consent was obtained, if the participant was available. Otherwise, a future date and time to conduct the interview was established during the initial contact. The IRB waived the necessity of written informed consent, but oral consent was obtained at the beginning of each interview. Responses were recorded on paper by the interviewer. Answers to open-ended questions were read back to the participant to ensure accuracy. Interviews lasted between 26 and 73 minutes with a mean of 37 minutes.

Data analysis

Descriptive analyses of patient demographics and other categorical data were summarized by measures of central tendency. Two investigators (PD and RM) used a conventional content analysis approach to collect, organize, and analyze the qualitative data, since the aim of the study was to describe a phenomenon about which there was limited preexisting literature.¹⁸ The investigators used an open coding process to identify themes within the data and to establish a list of codes. The investigators then coded the data independently and met again to discuss the findings. All discrepancies were resolved through discussion and clarification.

Results

The average participant age at the time of the interview was 22.6 years (Range: 19–27). The sample was reflective of the larger CGD population followed at NIH in terms of gender distribution. The mean age for a CGD diagnosis was 3 years. Participants had been followed at the NIH-CC for a mean of 10 years (Range: 1–17 years). Seventy-six percent of the participants had some post-secondary education and 67% were working full-time or in school full-time at the time of the interview. (Table 1).

Three factors that fostered a successful transition were identified: (1) positive perceptions of transition, (2) self-management, (3) parental and medical team support

Perceptions of transition

Seventy-five percent (24/32) of participants perceived their overall transition from pediatric to adult care to be uneventful. Twenty participants had been inpatient on both the pediatric and adult units. Sixty percent (12/20) of these AYA described the transfer from the inpatient pediatric unit to adult units as uneventful. When addressing the change in inpatient units, one participant noted, “[It’s] not too much different. You have to go to a different area but everything else feels pretty much the same. I kind of expected to go to the adult unit at 18.” Some patients did have anticipatory anxiety prior to the transfer, “I was edgy about it because I knew all the nurses and the hallway and the games [on pediatrics]. I was nervous about having a roommate because they could be like 60 and that would be weird. I was nervous about getting a different level of attention.”

Self-management

All the participants had transferred to the adult care setting. Thirty-two of the 33 participants were independent in most self-management activities at the time of the study. The one participant who was not independent in self-management activities relied on his mother to manage all aspects of his care. Eighty-one percent (26/32) reported being independent in making air travel and lodging arrangements and assumed responsibility for these tasks on average at 18.5 years of age (range: 15.5 – 24 years). Three of the six participants who were not independent lived within driving distance of the hospital, so they did not develop these specific skills. For the remaining three, their parents continued to make arrangements. Ninety-four percent of participants (30/32) were making their own clinic appointments and 70% (21/30) began doing so before their 19th birthdays. Parents made appointments for the remaining two young adults. Seventy-two percent (23/32) traveled to the hospital and came to clinic visits independently with the majority beginning at age 18. Prior to their 18th birthdays 59% (19/32) had seen the healthcare provider alone. At the time of the study, 72% (23/32) were seeing their providers alone.

Two participants of the thirty-three were not taking any prescription medications at the time of the interview. Of those who were, 90% (28/31) reported refilling prescriptions on their own either by calling their nurse practitioner, a pharmacy or obtaining refills at a follow/up appointment. The average age for assuming this responsibility was 18.5 years (range: 10.5 – 25 years).

Eighty-two percent (27/33) reported having a home physician with ten participants still seeing a pediatrician. If they had a question or concern related to CGD, 61% (20/33) of the participants said they would call the medical team at NIH for assistance.

Parental and Medical Team Support

Participants learned to assume responsibility for medical self-management activities from parents, case managers, and physicians. (Table 2).

Seventy-four percent (23/31) of the participants reported that physicians educated them about their medications and 6.5% (2/31) were educated by their parents. Pharmacists and nurses were also mentioned as resources. Sixteen percent (5/31) of the participants reported teaching themselves by listening to discussions about other family members with CGD, researching on the Internet or simply absorbing information over time. One participant did not know how he learned about his medications.

Perceived barriers to transition to adult care included deficits in: (1) disease understanding, (2) medication regimen understanding, (3) knowledge about advance directives, and (4) preparation, planning and practice related to the transfer in care.

Knowledge Deficits

Participants' lack of knowledge about the disease was a major barrier in transition. Only 24% (8/33) of participants met the criteria for disease understanding. (Table 3).

Lack of medication regimen understanding was also identified by the study. Of the 33 participants, seven were no longer on a prophylactic drug regimen since they had undergone successful bone marrow transplantation. For the remaining 26 patients, fifty percent (13/26) understood their prophylactic drug regimen.

A knowledge deficit was identified in regard to advance care directives as well. Thirty-three percent (11/33) said that they knew what an advance directive was. Forty-two percent (14/33) of the participants reported that they had an advance directive. However, of these 14 only 57% (8/14) had an advance directive in their medical record. One participant who indicated he did not have one was found to have an advance directive in his record.

Lack of preparation, planning and practice

Nearly a quarter of participants (22.5%) reported that they did not feel included in planning the transfer of care or in the actual transition itself. One AYA said, *“There was no transition. It just kind of happened.”* At the same time, participants reported that they were informed that things would change once they turned 18. One participant noted that the medical team informed him that, *“Once I was 18...I would have to sign documents and that when traveling, I would have to come by myself.”*

The process was especially incomplete when it came to preparation and planning for an inpatient admission. Forty percent (8/20) of the participants who had been hospitalized on the pediatric unit were informed just the day before or the day of their admission of the admission to the adult unit. Only two participants were provided a tour of the adult unit before the admission. Forty-five percent (9/20) were made aware of the policy differences between the pediatric and adult units before they changed units.

Medication Compliance

Ninety-four percent (29/31) of participants reported that they had forgotten to take medications at some point, ranging from missing one dose every few months to forgetting all medications for a week at a time. Several participants articulated a difference between forgetting to take medication and making a conscious decision not to take it. One participant said, *“It’s less about forgetting and more about not feeling like it...the mental aspect of being on meds indefinitely. When you’re healthy, you just want to be healthy, but then you don’t take your meds and you get sick.”* Another said, *“I forget on purpose because I don’t want to take them. I’ve been taking medication so long that it’s irritating, annoying and I don’t want to take it. Everything should be fun at my age and I shouldn’t have to take 12 pills [a day].”*

Sixty-three percent (20/32) of the participants thought that the transition process needed improvement and provided insights on six themes: (1) age at which the process should begin; (2) the importance of peer support; (3) knowledge requirements; (4) methods familiarizing adolescents with the transition process; (5) the role of parents; and, (6) the role of the medical team. (Table 4).

Discussion

This study described the experience of AYA with CGD as they transitioned from pediatric to adult care and assumed greater responsibility for their own disease management. Participants perceived the transition from pediatric to adult care to be uneventful but that did not necessarily mean that they had a successful transition. Fair et al. described ten outcomes that are important for a successful transition, including understanding the characteristics of conditions and complications, knowing the names and purposes of medications, adherence to medications and/or other treatments and self-management of one's own condition¹⁹. While our results show that most of the participants were able to successfully navigate logistics independently, they had an underlying lack of knowledge of their disease, medications and advance directives. These findings highlight the discrepancy between the participants' perception of their transition and specific transition outcomes that suggest areas to improve transitions. Systemic barriers, including limited preparation and planning in the transition process by the medical team, were also found to have an impact on the AYA transition from pediatric to adult care. This deficit was especially prominent when it came to the transfer between the pediatric and adult inpatient care units and the failure to inform participants of policy differences between the two units. In the literature, other barriers related to transition program implementation include insurance and lack of provider training in childhood onset conditions, but neither of these issues were found to be barriers for the participants in our study.²⁰

Our findings reinforce the idea that family-centered care is a significant component of the transition process with the transition program involving AYA, their families and the entire medical team (physicians, nurses, social workers and pharmacists, etc.). It should build on the strengths that have been previously identified in this study including the participants' independence in making and attending appointments and refilling medications²¹. It is also crucial that the program assess and address the potential barriers that AYA encounter such as not having adequate knowledge about the disease process. As disease understanding is an important component of a successful transition, special attention must be paid to enhancing adolescent and young adult understanding of their condition.¹⁹

Medication understanding is another component that must be assessed in a successful transition program. For this study, we focused solely on the prophylactic antibiotic and antifungal drug regimen for CGD. Many patients were also on medications for CGD and other medical conditions. Given their limited understanding of the prophylactic regimen, they may not have adequate knowledge of their other medications. This may result in a lack of compliance and serious consequences including increased risk of morbidity and mortality.²²

While there is a body of literature on Advance Care Planning (ACP) with AYA, this topic is often overlooked in the transition literature.^{19,23,24} Though participants reported completing an advance directive, only slightly more than a half had advance care planning documented in their medical record. Moreover, the majority had limited knowledge of what an advance directive was and what that meant for their care. AYAs with chronic and acute illnesses have expressed wanting to share their values, beliefs and preference for treatment at end of

life.^{23,25,26} Therefore, it is important that AYA be able to make decisions about ACP, especially as they transition to adult care. As with any component of the transition process, readiness will need to be assessed on an individual basis, and support should be provided to families and providers to enhance their ability to assist in planning.

Finally, individuals with pediatric onset chronic illnesses fare worse than those without chronic illnesses in educational and vocational outcomes due to factors such as absenteeism from school or work.^{11,27} Twenty-one percent (7/33) of the participants in this study were not in school or working at all. For twenty-one percent (7/33), the highest educational level they had attained was a high school diploma or GED. These findings suggest that for AYA with CGD, a comprehensive transition process would benefit from collaboration among multiple systems—including educational and vocational systems—in addition to the medical and family systems. One way to begin to incorporate these other systems is for a transition program to include early assessment of vocational rehabilitation needs among participants.

Our findings suggest that within the hospital setting, successful transition from pediatric to adult care requires collaboration. The institution, the medical staff, the family and the AYA all have roles to play in the transition process.

Limitations

Several limitations must be noted. This study focused on one specific disease (CGD), had a small sample size and was conducted at a single institution; thus, the results may not be generalizable to all chronic conditions or settings. Furthermore, as the majority of our cohort was male, the results may not generalize to disease populations with a more equal gender distribution.¹⁵ Although the literature suggests that there may be gender-based differences in self-management among youth with chronic conditions, due to our small sample size, we were unable to compare the transition experience and outcomes based on gender in the present study²⁸. It is important to note that participants were asked about their disease through an open-ended question without prompts, which may have had an impact on the depth of the answers received. Perhaps if multiple-choice questions had been utilized, disease and medication understanding would have been assessed higher. Additionally, the interviews were not audiotaped. While the investigators felt confident that reading the answers to open-ended questions back to the participants allowed us to capture the participants' comments completely and accurately, additional information might have been captured if the interviews had been audiotaped. Furthermore, sexual and reproductive health is an important issue among young adults, this survey neither addressed the sexual and reproductive health consequences of CGD, nor did any participants raise the issue themselves. Ten participants were still cared for by pediatricians outside of NIH, which could have influenced the findings since they had not fully transitioned to adult care across all health care settings. Finally, along with the limitations associated with a retrospective cross-sectional design, on average, 5 years had elapsed since the transition, which may have biased participants' recall of their experience.

Conclusion

To the best of our knowledge, this is the first study that has explored transition from pediatric to adult care by AYA with CGD. Patient opinions suggest that transition planning was not difficult, but that a more structured transition program for adolescent patients with CGD would facilitate a smoother and more comprehensive process. Our data also suggest that patient education regarding their specific disease and medications needs to begin in early to mid-adolescence. Future directions include establishing a formal transition program for adolescents and young adults with CGD, followed by expanding the program to include adolescents with other chronic and rare conditions.

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Abbreviations

AYA	Adolescent and young adult
CGD	Chronic granulomatous disease
GED	General equivalency diploma
HIV	Human immunodeficiency virus
NIAID	National Institute of Allergy and Infectious Diseases
NIH	National Institutes of Health
NIH-CC	National Institutes of Health Clinical Center

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Implications and contributions

This study describes the strengths and barriers of transition to adult care as perceived by young adults living with CGD. The findings of this study will help inform the development of a transition policy and program that integrates young adults into adult health care.

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Table 1

Participant demographics

Demographic characteristic	N (%)
	Mean (Range)
Gender	
Male	29 (88)
Female	4 (12)
Race	
White	24 (73)
Black	6 (18)
Latino	3 (9)
Age at time of interview	Mean: 23 years (Range: 19–27)
Relationship status	
Single	22 (66)
Committed relationship	9 (27)
Married	2 (6)
Family History of CGD	
Yes	17 (52)
No	16 (48)
Had at least 1 inpatient admission on pediatrics	
Yes	25 (76)
No	8 (24)
Average number of pediatric admissions per year	Mean: 0.9 admissions (Range: 0–3.8)
Average number of pediatric inpatient days per year	Mean: 9 days (Range: 0–48)

Table 2

Educators of self-management skills

Activity	Educator			
	Parent n(%)	Medical team n(%)	Both n(%)	Self-taught n(%)
Travel & Lodging (N=25)	10 (40)	9 (36)	6 (24)	0(0)
Appointments (N=30)	4 (13)	17 (57)	7 (23)	2 (7)
Prescription refills* (N=28)	13 (46)	11 (39)	2 (7)	2 (7)

Table 3

Disease Understanding

Disease element & response type	N (%)	Example response
Immunodeficiency (N=33)		
Correct	17 (52)	<i>"I have an immune system but it's compromised and not working correctly."</i>
Incorrect *	16 (48)	<i>"Basically no immune system."</i>
Dysfunctional white blood cells (N=33)		
Correct	19 (58)	<i>"When the white blood cells don't produce hydrogen peroxide."</i>
Incorrect	14 (42)	<i>"Not enough white blood cells to fight infection."</i>
Susceptibility to bacterial and fungal infections (N=33)		
Correct	15 (45)	<i>"Can't fight certain infections—more prone to fungal and bacterial infections."</i>
Incorrect	18 (55)	<i>"You're vulnerable to airborne infections."</i>
Genetic (N=33)		
Correct	4 (12)	<i>"[It's] X-linked. My mother is a carrier."</i>
Incorrect	29 (88)	<i>"There's different types of CGD. I have the XY type."</i>
Primary systems affected (N=33)		
Correct	7(21)	<i>"Affects organs, lungs, kidneys, eyes, bones."</i>
Incorrect	26 (79)	Not addressed

* Includes responses in which element was not mentioned.

Table 4

Participant's strategies for improving transition

Thematic code	Representative quotes
Age <i>When should the transition process begin?</i>	<p>"Start young, like around 15 or 16, a few years before. Talk about their flights and appointments with their parents there but not doing it for them."</p> <p>"Have the doctor tell parents to talk to their kids at age 15 or 16."</p>
Peer support <i>Is peer support important?</i>	<p>"Have a support system with other patients. Talking to other patients about transition would help. I would be willing to talk to others."</p> <p>"I think it's best to have similar people your age ease you into the situation. Talking to someone personal would be really helpful. Someone to connect with on your level that knows what you're going through. I've found it useful to talk to other patients with CGD."</p>
Knowledge requirements <i>What do transitioning young adults need to know?</i>	<p>"Encourage patients to be an active participant. It's important to know what and why."</p> <p>"Make sure the patient is comfortable in the setting they are in. They need to 1) understand the importance of taking medicine, 2) if getting sick contact the doctor right away...4) Understanding the disease is highly important and 5) Know it is possible to live a full healthy lifestyle if staying on top of what they need to know."</p>
Methods of knowledge dispersal <i>What's the best way for transitioning young adults to learn what they need to know?</i>	<p>"Do more walk-throughs on the unit to get familiar with the unit and nurses."</p> <p>"A pamphlet outlining what goes into a visit starting with travel, reservations, clinic appointment."</p> <p>"Have a conference at NIH for adolescents with CGD."</p>
Parental role and involvement <i>How should parents be involved in the transition process?</i>	<p>"You gotta work with your parents to learn your disease and medications while you're growing up."</p> <p>"My parents always encouraged me to ask questions. The younger the better. My mom got scolded by the doctors for not taking a more active role but she said she wasn't always going to be around."</p>
Medical team role and involvement <i>How should the medical team be involved in the transition process?</i>	<p>"Sit down one on one with your doctor to talk about your meds and disease."</p> <p>"[The doctor] pesters me and she's persistent and I don't want to disappoint her. I finally got myself organized. The doctors motivated me in the past few years and I started taking care of things myself."</p>