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Original article

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 (19–27 years of age), living with chronic granulomatous disease 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 were analyzed using a conventional content analysis approach.
Results: Seventy-six percent of the participants did not understand their disease process and only 50% 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 90% 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 chronic granulomatous disease. The findings suggest the need for enhancing the transition process utilizing interdisciplinary collaboration to develop a transition policy and program.

IMPLICATIONS AND CONTRIBUTION

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

Owing 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 (AYAs). Many

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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 2,000 participants between the ages of 10 and 24 years 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 the United States. CGD is a chronic disease characterized by frequent bacterial and fungal infections requiring close monitoring and prophylactic drugs. The treatment of infections often requires hospitalizations, intravenous 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, 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 AYAs 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 before 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 before participant recruitment. Thirty-nine AYAs 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 five-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 antifungal and antibacterial medications for a possible total of four points. All responses were weighted equally. A score of 4/4 was required for understanding medication prophylaxis. The 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 institutional review board 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 (P.D. and R.M.) 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 pre-existing literature [18]. The investigators used an open coding process to identify themes