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Development of a tool to describe overall health, social independence and activity limitation of adolescents and young adults with disability

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ABSTRACT

There is a need for research that focuses on the correlation between self-perceived quality of life (QoL) and the health outcomes of adolescents with disability transitioning to adulthood. To better understand the transition experience of adolescents and young adults with disability, we developed a questionnaire to assess the impact of disability on QoL. We recruited 174 participants who were 15–24 years old and diagnosed with Fragile X syndrome (FXS), spina bifida (SB) or muscular dystrophy (MD) and conducted an exploratory factor analysis to identify factors that characterize QoL. Five factors emerged: emotional health, physical health, independence, activity limitation, and community participation. To validate the tool, we linked medical claims and other administrative data records and examined the association of the factor scores with health care utilization and found the questionnaire can be utilized among diverse groups of young people with disability.

1. Introduction

There is a limited amount of research that focuses on the correlation between self-perceived quality of life (QoL), including emotional health, physical health, independence, activity limitation, and community participation, and the health outcomes of adolescents with disability transitioning to adulthood. The literature has summarized four tasks adolescents are

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expected to complete as they enter adulthood: establishing identity, forming relationships outside of the family unit, achieving independence from family, and finding a job (White, 1997). Adolescents and young adults may face challenges in completing these tasks depending on the nature and severity of their disability. While there is a growing body of literature surrounding the transition experience of people with disability, there is a need for an accessible and easily understood instrument that measures QoL of adolescents and young adults with disability.

In order to better understand the transition experience of adolescents and young adults with disability, we developed a questionnaire to assess the impact of disability on QoL. We administered the survey to people with three substantially different disabilities: fragile X syndrome (FXS), spina bifida (SB), or muscular dystrophy (MD). All three conditions are rare with prevalence below one per 10,000 people (Dicianno, Gaines, Collins, & Lee, 2009; Garber, Visootsak, & Warren, 2008; Hartley et al., 2011) The conditions cause some level of disability beginning in childhood, which can contribute to difficulties in the transition from adolescence to adulthood.

FXS is the leading cause of inherited intellectual disability (ID) and primarily impacts males. People with FXS do not generally have physical limitations that would require special accommodations needed by people with other types of ID (CDC, 2012a). SB is a congenital neural tube defect that frequently causes neurologic deficits below the level of the lesion, which may include paralysis. It is sometimes accompanied by hydrocephalus, which can result in neurodevelopmental complications (CDC, 2011). MD is a group of neuromuscular disabilities that include both childhood and adult onset. MD may involve progressive physical disability and declining mobility, cardiac and respiratory function (CDC, 2012b).

QoL questionnaires usually target either the general population or people with specific disorders. To be appropriate for large groups of people, generic QoL questionnaires do not include questions that would be of concern for persons with disability; this makes generic surveys inadequate for assessing the QoL of people with disability. In contrast, condition-specific surveys are sensitive to concerns of a particular population, but are difficult to use across populations (Dijkers, 1999; Guyatt et al., 1997; Liu et al., 2010; Rosenbaum & Saigal, 1996). The purpose of this study is to present the results of a validated QoL questionnaire that is general enough to be applied across all disability groups, but specific enough to address QoL concerns of individual disability groups.

2. Methods

The QoL survey for people with disability (specifically FXS MD, or SB) was developed in four phases: tool development, study recruitment, tool reduction, and tool validation.

2.1. Phase 1: tool development

The survey questions were selected from four validated and reliable surveys: the American Community Survey (ACS), the National Longitudinal Transition Study-2 (NLTS2), and the RAND-36 Measure of Health-Related Quality of Life (RAND-36).

The 14 demographic questions in our survey came from the ACS, which is part of the U.S. Census. Our survey also incorporated eleven questions from the RAND-36 designed to measure eight health concepts: physical functioning, role limitations caused by physical health problems, role limitations caused by emotional problems, social functioning, emotional well-being, energy/fatigue, pain, and general health problems. Nine questions that addressed mobility, medical care, condition type, use of help during the completion of the survey, and identification number to track responses were added by our research team. The remaining 120 questions included in our survey were taken from the NLTS2 in the sections identified as: social and leisure time activities, high school experiences, personal interests and activities, personal health, household, leaving high school, 2-year junior or community college, 4-year college or university, and jobs during last 2 years.

2.2. Phase 2: study recruitment

The same survey was administered to two different populations: US residents outside of SC and SC residents. To reach a national audience, we announced the survey through social media and a number of advocacy organizations. We enlisted a wide array of national organizations to post announcements in their newsletters and on their websites. The postings asked US residents who were 15–24 years of age, and diagnosed with SB, MD, or FXS to respond to an online questionnaire about their self-perceived health, social life, education, work experience and community participation. If needed, the participant was allowed to have help answering the questions. This was a convenience sample with no personal identifiers, but participants reported their age and state of residence. Only data collected on participants aged 15–24 was used in this analysis.

We conducted recruitment of South Carolina residents in a similar fashion, but also included medical providers in the recruitment process. We asked residents of South Carolina who had FXS, SB, or MD and were 15–24 years old to sign an informed consent form giving us permission to link their survey answers to data compiled as part of a larger study investigating the transition from adolescent to adult services for people with rare health conditions in South Carolina; details of which can be found in a methods paper (Royer et al., 2014). The data for this larger project including Medicaid and State Health Plan medical claims data is housed at the South Carolina Revenue and Fiscal Affairs Office, Health and Demographics (H&D). Data linkages and analyses were performed by H&D staff. South Carolina participants were compensated $50 for