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SHORT COMMUNICATION

Gastroenterology

Quality of life from childhood to adulthood: Perspectives from adult patients with pediatric-onset intestinal failure

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Abstract

Adult patients with pediatric onset short bowel syndrome (SBS) or intestinal failure (IF) have been described as a distinct population warranting further research. The aim of this exploratory study aimed was to offer initial insights into this population's navigation of childhood, adolescence, and transition into adulthood. Both quantitative and qualitative data were collected from a convenience sample of adults with pediatric-onset SBS/IF using a disease-specific pilot survey; 14 questionnaires were completed. Responses indicated childhood and adulthood were complex and marked by joys and trials, while adolescence was experienced by many as a particularly challenging time. As adults, numerous patients experienced barriers to accessing the medical care they desired and described difficulties finding experienced and knowledgeable providers who listened and offered individualized care. This study highlights the importance of further studying this unique patient population, suggesting it can offer critical insights to inform the development of interventions and transition programs.

KEYWORDS

care transition, parenteral nutrition, short bowel syndrome

1 | INTRODUCTION

Intestinal failure (IF) is a rare disease resulting from the inability of the gut to absorb the nutrients and fluids required to sustain health or support growth, most commonly due to short bowel syndrome (SBS), thus necessitating long-term supplementation with parenteral nutrition (PN) or intravenous fluids.[1] Due to substantial advances in SBS/IF treatment and management and the resulting reductions in mortality, interest has turned to long-term outcomes and the successful transition of pediatric patients into adulthood.[2] Research investigating the psychosocial implications of SBS/IF for pediatric and adult patients suggests the condition may impact all areas of life for affected patients[3–6] and their caregivers.[7–9] In a recent review comparing adult and pediatric IF, Belza and Wales suggest patients with pediatric-onset IF are a distinct population that must be better understood.[10] However, to our knowledge, no previous research has investigated the lived experiences of this patient population once they reach adulthood.

The purpose of this brief communication is to offer initial insights into the experiences and needs of adult patients with SBS/IF since childhood by highlighting results from an exploratory study. Using a mixed-methods, disease-specific pilot survey, we solicited patient perceptions of their quality of life (QoL) across multiple phases in the lifespan and their experiences receiving medical care.

2 | MATERIALS AND METHODS

2.1 | Study design and participants

The study design and recruitment processes were similar to those of a study on QoL for pediatric SBS

patients and their caregivers.[7,11] Briefly, the studies utilized a community-driven research design, with the core research team and a project stakeholder committee comprised of SBS/IF community members. A web-based cross-sectional questionnaire was shared with a convenience sample of stakeholder committee members who were at least 18 years old and had a pediatric diagnosis of SBS/IF. The survey was hosted on the institution's REDCap platform and could only be accessed via unique links sent to eligible committee members via email; data was collected from March to May 2021. The institutional review board at the University of Nebraska Medical Center approved the study, and respondents provided informed consent before beginning the survey. A final sample of 14 adult patients with SBS/IF since childhood completed all sections of the survey; data were stored in a secure database on a protected server at the institution.

2.2 | Measures

A mixed-methods disease-specific pilot survey was developed by an interdisciplinary study team to measure adult patient QoL (Supplement); this process paralleled the development of an SBS-specific QoL survey for caregivers of pediatric patients.[11] The pilot survey used adaptive questioning and included items about demographics and medical background. It also contained a matrix measuring the impact of disease-specific items on respondent well-being using a 5-point Likert scale. For items rated as having a strong negative impact, respondents were prompted to explain their answers in open-ended follow-up questions. Other open-ended questions asked about any additional items that negatively or positively impact well-being for respondents. Participants also answered questions about how their SBS/IF shaped their childhood and adolescence and their experiences receiving SBS/IF-related medical care.

2.3 | Data analyses

Quantitative analyses were conducted in Microsoft Excel and included examining survey responses for missing values and normality of distribution and basic descriptive analyses to assess survey and sample characteristics. To calculate the proportion of respondents who reported select disease-specific items as having a strong negative impact on their well-being, the number of respondents who rated an item a "4" or a "5" on the disease-specific item matrix was divided by the total number of respondents who endorsed the item as the denominator. Qualitative responses were coded thematically by a primary coder; a secondary coder then reviewed these codes. An interrater reliability of 0.73 was achieved between coders, and differences were resolved through discussion to reach consensus. Results were reported in accordance with the Checklist for Reporting Results of Internet E-Surveys.[12]

What is Known?

- Reductions in mortality for patients with short bowel syndrome (SBS) or intestinal failure (IF) have resulted in a growing interest in the condition's impact on patient quality of life (QoL).
- Research suggests children with SBS/IF experience poorer QoL than healthy peers.
- Programs designed to ensure the successful transition of pediatric SBS/IF patients into adulthood are urgently needed.

What is New?

- Adults with pediatric-onset SBS/IF participating in this study described their experiences both during childhood and in adulthood with nuance while adolescence was highlighted by many as particularly challenging.
- Respondents described barriers to accessing the knowledgeable and patient-centered medical care they desired in adulthood.

3 | RESULTS

3.1 | Respondent characteristics

Fourteen adult patients with IF since childhood completed all sections of the survey; a majority self-identified as female (79%) and non-Hispanic White (85%) and were between the ages of 19 and 39 (79%). Most participants (85%) had SBS specifically, most frequently due to volvulus (29%), necrotizing enterocolitis (29%), or gastroschisis (14%); diagnoses had been received in early childhood, before age 3, for 79% of participants. About a third of participants were currently receiving PN (29%) or had an ostomy (36%), while only 7% were currently receiving enteral nutrition (EN) via tube feeds; 85.7% of respondents reported at least one comorbidity (e.g., asthma, cerebral palsy, and anxiety disorder). Two respondents (14%) had previously received a transplant; both were receiving PN at the time of the study.

3.2 | QoL during childhood and early adolescence

In response to a set of open-ended questions about how IF shaped well-being during childhood,

participants offered nuanced perspectives (Figure 1). Some participants (36%) described positive aspects of their childhood including supportive relationships with their parents (21%) or perceiving their childhoods as "normal" (14%). Still, most (86%) felt their SBS/IF had affected their childhood negatively in some way. Just over a third (36%) described restrictions imposed by the condition or related therapies on their ability or willingness to participate in activities such as travel, sports, or sleepovers. Participants (43%) also recalled feelings of shame or "being different" from other children:

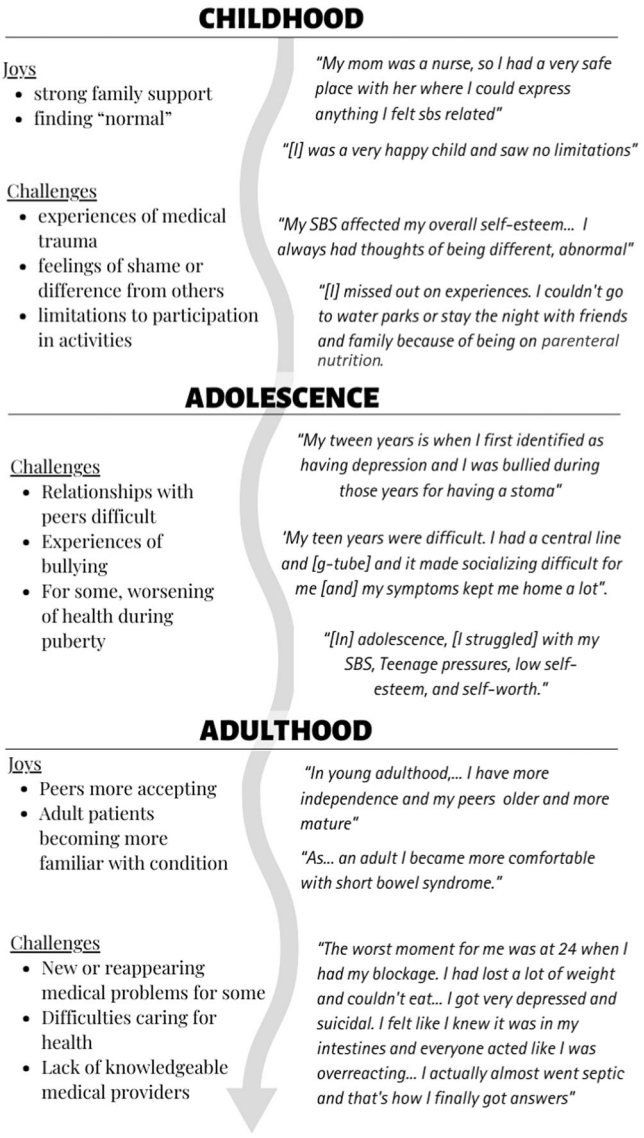

FIGURE 1 Patient quality of life across developmental phases based on qualitative responses to two open-ended survey questions: "Thinking about your childhood, in what ways did your SBS affect your overall well-being—that is, your sense of being comfortable, healthy, or happy?" and "Were there any phases in your childhood, adolescence, or young adulthood that your overall well-being was particularly high or low? If so, when were they?". PN, parenteral nutrition; SBS, short bowel syndrome.

> *I did not feel normal and was very embarrassed though my parents tried their best to make me feel normal*.

Respondents also described feeling unwell or experiencing physical symptoms including fatigue, stomach pain or discomfort (29%) as children; some noted that issues surrounding toileting (29%) were partly to blame for restrictions on social activities and feelings of shame:

> *I felt afraid to sleep over [at] people's homes and I [was] always running to the bathroom which made it difficult in school*.

Adolescence arose as a phase during which numerous respondents (43%) experienced significant challenges. These challenges could stem from worsening health during puberty, but most frequently came from difficulties navigating relationships with peers (21%). Some respondents explicitly described experiencing bullying because of their condition, which could have implications for mental health:

> *My tween years is when I first identified as having depression and I was bullied during those years for having a stoma*.

3.3 | QoL in adulthood

Survey responses offer evidence of the nuance and complexity that characterize participant QoL in adulthood. Responses to the matrix of disease-specific items measuring QoL over the previous year suggest respondents perceived many of the SBS/IF-specific items to negatively impact their overall well-being. Respondents utilizing medical technology or devices perceived their feeding tubes or ostomies (71%) or their central lines or PN (50%) to have a strong negative impact on their QoL. Additional items commonly reported by respondents to have a strong negative impact on their well-being were poor sleep (58%), fatigue (50%), pain or discomfort (46%), and issues related to toileting (42%). Notably, a substantial share of respondents indicated items within the mental and social well-being domain, including trauma associated with medical procedures (36%) and worries (29%), to have a strong negative impact on their well-being.

Qualitative follow-up responses provide additional evidence of the impact of these items on respondents' lives, including the limitations imposed by the condition, coupled with strategies to cope with these challenges:

> *The mid 30 s was a challenge as well. My treatments were failing and I was in constant pain. But I was able to visit with my doctor and develop a new treatment*

plan. Since then my quality of life has drastically improved.

I have stomach pain every day due to my condition… I use mental relaxation techniques and cannabis to cope.

I always feel fatigued… it has made me find other ways to cope for example online schooling versus public schools.

3.4 | Receiving medical care

Of the survey respondents, half described experiencing periods of not receiving optimal IF care, and only three (21%) were currently receiving multidisciplinary care via an intestinal rehabilitation program (IRP). One respondent was uninsured at the time of the survey and was receiving no medical care at all, while another was followed by a primary care physician. A third respondent, an adult in their late 20 s, still received IF-related care via a pediatric IRP. Over half of respondents (57%) had previously transferred their IF-related care from one provider or team to another, with the most cited reason for this transfer being the aging out of pediatric care (50% of all care transfers). These patients frequently experienced difficulties accessing care from providers who are knowledgeable and experienced with SBS/IF, as described by one respondent:

I was satisfied with my pediatric gastroenterologist. However, there are a lot of adult general practitioners and gastroenterologists who [have] never met an adult with SBS or [know] how to treat SBS.

Twelve participants provided open-ended responses to a question asking them to identify what medical professionals can do to support their overall well-being (Table 1). Over half described the importance of communication, most notably truly listening to patients. An additional common theme was the importance of an individualized approach to SBS/IF care. Two respondents specifically described desiring acknowledgment of their complex history, which could include previous traumatic experiences in the medical setting, on the part of clinicians. Other respondents described wanting providers to educate themselves about SBS/IF.

TABLE 1 IF patient desired medical professional supports for patient well-being ($N = 12$).

Theme 1: Thoughtful communication, including truly listening to patients ($n = 7$)
▪ *Listen!!! We know our bodies better than ANYONE else and we know when something is wrong!* ▪ *By not making assumptions and truly listening* ▪ *More communication on the reasoning behind the decisions made. If I understand why I'm taking this med, or we are doing treatments in this order, [then] I'm more likely to be compliant*
Theme 2: Individualized care ($n = 5$)
▪ *They need to understand that one size does not fit all* ▪ *It helps when they… take the time to understand your unique situation* ▪ *I think what's worked for me is that I talk to my doctor, nurses, and dieticians about the lifestyle that I live and they work around my nutrition so I can live that lifestyle better*
Theme 3: Understanding patient history ($n = 2$)
▪ *I have trauma medically from procedures in childhood that still haunt me* ▪ *Although I was very young when diagnosed and don't remember any surgeries, my childhood was different and greatly impacted how I am now*
Theme 4: Learn about IF ($n = 2$)
▪ *Try to learn more about short bowel and know that we are all different* ▪ *Educating themselves on the long term impact of SBS*

Abbreviations: IF, intestinal failure; SBS, short bowel syndrome.

4 | DISCUSSION

Findings from this exploratory pilot study highlight experiences of adult patients with SBS/IF since childhood and suggest the condition has implications across the lifespan. While some respondents perceived their childhoods as "normal," most described experiencing SBS/IF-related challenges beginning in childhood, largely due to limitations imposed by therapies and symptoms and difficulties related to peer relationships, particularly as they entered adolescence. Similar concerns have been previously highlighted by children with IF in other qualitative work.[13,14] As adults, the relationship of study respondents to their condition was multi-faceted; many had developed strategies for managing their condition that allowed them to participate in activities as desired (e.g., work and exercise) but also continued to experience challenges associated with therapies and medical devices, symptoms, and sleep disturbances.

One of the most common challenges in adulthood, as described by study participants, was accessing knowledgeable, specialized, whole-person medical care for their SBS/IF. Adolescents and young adults with SBS/IF must manage their complex, life-threatening condition while also navigating all of the standard challenges that come with entering adulthood. Without a gradual and intentional transition into the adult care setting, this population is at high risk for health complications and loss to follow-up.[2] This exploratory study provides

evidence of this phenomenon, with several respondents describing periods in adulthood during which they received no specialized care. For some, this gap in care had significant implications for their health and well-being. Even among those who had secured medical care for their SBS/IF as adults, many discussed feeling poorly understood and desired clinicians who were better educated and more experienced. Thus, these data suggest the integration of structured and multi-part healthcare transition programs into pediatric IF care as a critical need for this population, as has recently been recommended.[2]

Receiving SBS/IF care as adults, respondents described desiring clinician communication that was knowledgeable and patient-centered. Several study participants felt they had developed a level of expertize and experience from decades of living with the condition and wanted clinicians to see them as partners in care. These data align with the suggested utilization of a patient- and family-centered approach to care in the management of SBS or IF.[15]

5 | CONCLUSION

Due to the study's small sample size and utilization of an unvalidated data collection tool, we were unable to make generalizable inferences about QoL for adult patients with childhood-onset SBS or IF. Rather, our aim was to offer initial insights into the experiences of this previously unstudied patient population and suggest opportunities for future research. This pioneering population has accumulated decades of experience managing the physical, emotional, and psychosocial effects of SBS/IF from childhood through adulthood. Further engaging this patient cohort in research may offer valuable insights with the potential to inform care across the lifespan; it may also be informative for the development of programming aimed at supporting pediatric IF patients during adolescence and through their transition into adult care. Such programs are urgently needed in light of greatly improved clinical outcomes in pediatric SBS/IF, which is leading to a growing patient population that will soon transition into adolescence and adulthood.

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CONFLICTS OF INTEREST STATEMENT

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