Health Related Quality of Life in Children with Intestinal Failure

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Abstract

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Introduction: The life expectancy of infants with intestinal failure (IF) has increased in the past decade. Now children with IF survive well into their childhood years with a strong potential for long-term survival. However, data on their health-related quality of life (HRQOL) are lacking. We hypothesized that HRQOL would be lower among children with IF and their families compared to that of healthy children.

Methods: We performed a cross-sectional study of HRQOL of children and their parents enrolled in the outpatient intestinal rehabilitation program at Seattle Children's Hospital using the PedsQL^{TM} 4.0 Generic Core Scales parent proxy-report and the Family Impact Module questionnaires. We compared PedsQL^{TM} scores with previously published normative data and examined associations between PedsQL^{TM} scores and patient demographic and clinical characteristics. Parents were asked two open-ended questions pertaining to the suitability and completeness of the PedsQL^{TM} to assess their and their child's HRQOL.

Results: Parents of 23 children with IF completed the questionnaires. Compared to norms for healthy children, parents reported significantly lower total PedsQL$^{\text{TM}}$ scores for children ages 1-2 years (mean difference= 13.16, p=0.003) and 3-4 years (mean difference= -17.63, p=0.001). Scores were also lower for children younger than 1 year (mean difference= -6.43) and children aged 2-3 years (mean difference= -11.96), though neither test reached statistical significance. The Family Impact Module scores of parents were similar to previous scores of parents caring for children with chronic illnesses (mean difference= -1.19, p=0.852). No measured demographic or clinical characteristics were associated with HRQOL in this sample. The majority of parents (65%) said the PedsQL$^{\text{TM}}$ failed to address important impacts of IF on children and their families. In particular, the PedsQL$^{\text{TM}}$ did not address challenges parents face nurturing their child or supporting their child's attainment of developmental milestones.

Discussion: Children with IF and their parents have a decreased HRQOL compared to healthy children as measured by the PedsQL$^{\text{TM}}$. The majority of parents identified additional challenges associated with IF that affect their child's and family's HRQOL. A disease-specific module or separate HRQOL questionnaire is needed for a more comprehensive assessment of patient and family HRQOL of children with IF.

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INTRODUCTION

Health-related quality of life (HRQOL) is a multidimensional concept that consists of physical, psychological and social health dimensions of an individual's well-being (1). Preferred measurements of HRQOL reflect patients' self-assessment of the effects of their chronic disease and treatment on their functioning and well-being (1). HRQOL is considered a helpful adjunct outcome measure of the health of the pediatric population and their families, especially for children with chronic illnesses (2-3).

Over the past decade the surgical and medical management of pediatric intestinal failure (IF) has evolved such that long-term survival is common, with patients living well into their childhood years even though they may remain dependent on parenteral nutrition (PN) (4-5). Recently reported long-term survival rates from tertiary care pediatric centers range from 73% to 90%, and there are contemporary data that suggest a survival benefit for children treated in dedicated, multidisciplinary intestinal rehabilitation programs (6-9). These programs are based on Wagner's chronic care model in which the community, the health care system, and the provider organization all collaborate to optimize the care of patients with chronic disease (10-11). Thus, IF now fits the model of a chronic disease with potential long-term effects on child growth and development.

Few studies to date have explored the HRQOL of children with IF (12-14). Two studies of children receiving intravenous nutrition secondary to primary intestinal problems reported no negative impact on HRQOL relative to healthy children (12-13). In contrast, a third study demonstrated a significant degree of distress in children receiving PN at home (14). None of these studies were conducted in the U.S. and the measure of HRQOL differed in each. One study used interviews with open-ended questions (12), and another used different questionnaires that, although validated, were not consistent throughout age groups (13). The third study used measures of psychological distress and social adaptation, but not specifically quality of life (14).

The PedsQL^{TM} questionnaires are a reliable and valid method to asses HRQOL in children and infants (3, 15-21). PedsQL^{TM} scores accurately distinguish between healthy, acutely ill and chronically ill children, with lower PedsQL^{TM} scores representing lower quality of life (19-20). PedsQL^{TM} versions exist for children ages 1 month to 18 years. The PedsQL^{TM} Infant Scales are used for children 1 month to 24 months and the PedsQL^{TM} 4.0 Generic Core Scales are used for children 2 to 18 years. Parent-report versions are available for all ages. PedsQL^{TM} items are grouped into several domains that reflect different aspects of a child's life. Each domain includes 3 to 12 questions answered on a Likert scale. Three domains: Physical Functioning, Emotional Functioning and Social Functioning, are common to all the PedsQL^{TM} Generic Core Scales questionnaires. The questionnaires for children 2 years and older include a School Functioning domain. The questionnaires for children from 1 to 24 months include a Physical Symptoms domain and a Cognitive Functioning domain. Internal consistency and reliability for the PedsQL^{TM} Infant Scales and all the age-group versions of the PedsQL^{TM} 4.0 Generic Core Scales has been demonstrated in prior studies and is high (19-21).

In addition to the age-specific generic PedsQL^{TM} questionnaires, there are a number of PedsQL^{TM} disease-specific modules. Disease-specific modules address symptoms, treatments, and complications unique to particular disease states that may be overlooked by generic questionnaires. Many of these disease-specific modules, including the asthma module, diabetes module, and arthritis module, have been shown to be effective and accurate in evaluating the HRQOL of children with these chronic conditions (22-24). To date, there is no disease-specific questionnaire available for individuals with pediatric IF.

The primary aim of this study was to explore the HRQOL of children with IF and their parents as reflected by the PedsQL^{TM} Generic Core Scales or Infant Scales and Family Impact Module questionnaires and to compare these scores with normative data for healthy children. A secondary aim was to learn if there are HRQOL concerns that affect children with IF or their families that are not captured by the PedsQL^{TM}. We hypothesized that children with IF and their families would have a lower HRQOL compared to healthy children and their families.

METHODS

Participants

We performed a cross-sectional survey of children 6 months to 6 years of age and their parents attending the outpatient Intestinal Failure Program at Seattle Children's Hospital (SCH-IFP) from August, 2011 to March, 2012. Inclusion criteria consisted solely of active outpatient enrollment of the child in the SCH-IFP. Exclusion criteria included inpatient status of the child, age less than 6 months, surgery other than central venous line placement or removal within one month of the study, and discharge from the hospital for the patient's first admission within three months of the study. The purpose of these exclusions was to assure parents had adequate, recent experience at home with their children at a stable standpoint in their disease process from which to respond to the survey questions.

Measures

The quality of life study described in this paper was carried out using the PedsQL^{TM} developed by Dr. James W. Varni. Parent participants were asked to complete the PedsQL^{TM} 4.0 Generic Core Scales questionnaire for parent proxies or the PedsQL^{TM} Infant Scales, and the PedsQL^{TM} Family Impact Module (19-20, 25). We chose to use the parent proxy-report because there are no child self-report versions for children younger than age 5 and few of the patients eligible for this study were older than 5 years of age.

The PedsQL^{TM} questionnaires for children from 1 to 24 months are divided into 5 domains: Physical Functioning (6-9 items), Physical Symptoms (10 items), Emotional Functioning (12 items), Social Functioning (4-5 items), and Cognitive Functioning (4-9 items). Taken together, the Emotional Functioning, Social Functioning and Cognitive Functioning domains yield the Psychosocial Health Summary. The Physical Functioning and Physical Symptoms domains can be combined to yield the Physical Health Summary (20).

The parent proxy PedsQL^{TM} questionnaires for children older than 24 months are comprised of the following 4 domains: Physical Functioning (8 items), Emotional Functioning (5 items), Social Functioning (5 items), and School Functioning (3-5 items). Combined, the Emotional Functioning, Social Functioning and School Functioning domains yield the Psychosocial Health

Summary, and the Physical Functioning domain is equivalent to the Physical Health Summary (19).

The Family Impact Module, used with all ages and completed by parents, consists of 8 domains: Physical Functioning (6 items), Emotional Functioning (5 items), Social Functioning (4 items), Cognitive Functioning (5 items), Communication (3 items), Worry (5 items), Daily Activities (3 items), and Family Relationships (5 items). Combined, the Physical Functioning, Emotional Functioning, Social Functioning, and Cognitive Functioning domains constitute the Parent Functioning Summary. The Daily Activities and Family Relationships domains yield the Family Functioning Summary (25).

All of these questionnaires are answered based on the frequency of which a specific event has occurred in the last month in the child's or the parent's life. The response options are: 1) never, 2) almost never, 3) sometimes, 4) often, and 5) almost always.

This study included two open-ended questions asked by the study investigator after parents completed the generic and Family Impact Module PedsQL^{TM} questionnaires. These questions were: "What other things affect your child's and/or your family's quality of life not asked in this questionnaire?" and "How do these other things affect your child's and/or your family's quality of life?"

Demographic and clinical characteristics describing the children participants in this study were obtained by chart review. These included age, gender, diagnosis leading to IF, length of time since event leading to diagnosis of IF, PN dependence, ability to be feed enterally by mouth or through a gastric tube, and total inpatient length of stay at SCH since IF diagnosis.

Procedures

Written informed consent to participate in the study and consent for investigators' access to the child's medical file was obtained from parents in writing. Following this, the questionnaire data were obtained. Parents had several options to complete the PedsQL^{TM} questionnaires. They could complete the forms independently, at home or in clinic. If they chose to do so in clinic, they were allowed time after they provided consent to fill out the forms. If they chose to do it at home they were provided with an addressed and stamped envelope so they could mail the questionnaires back. They also had the option to have assistance to fill out the questionnaires, with the investigator reading the questions aloud and marking the parent's answers on their behalf. When the questionnaires were filled out in this manner, it was always done in person. There is no evidence to suggest that responses obtained by these two modes differ (19).

The open-ended questions were asked after the parents completed the PedsQL^{TM} questionnaires. Depending on the parents' choice for answering the questionnaires, the open-ended questions were asked in person or over the phone. A study investigator knowledgeable of the care of patients with IF and with excellent typing skills recorded all the parents' answers verbatim and then read them back to the parents to assure accuracy.

All aspects of this study were approved by the Seattle Children's Hospital Institutional Review Board (IRB application # 13660).

Data Analysis

The PedsQL^{TM} questionnaires were scored according to published instructions (19-20, 25). Items were reversed scored and linearly transformed to a 0-100 scale, so that higher PedsQL^{TM} scores indicated better HRQOL. To reverse score, we transformed the 0-4 scale items (0 = never a problem, 1 = almost never a problem, 2 = sometimes a problem, 3 = often a problem, 4 = almost always a problem) as follows: 0=100, 1=75, 2=50, 3=25, 4=0. To create domain scores, the mean was computed as the sum of the items divided by the number of items answered. If more than 50 percent of the items in the scale were missing, the domain score was not computed. In cases where 50 percent or more of the items were completed, we used the individual's mean score for the scale (based on non-missing items) to fill the missing values.

We calculated the Physical Health Summary and Psychosocial Health Summary according to published scoring guidelines. Scores were computed separately for children aged 1-12 months, 13-24 months, 2-3 years, and 3-4 years. We also created Total Scale scores for the different age groups. From the Family Impact Module, we calculated the Parent Functioning Summary and the Family Functioning Summary, in addition to the Total Scale score. We computed one-sample t-tests to compare the scores of our cohort to previously published scores obtained from healthy children (20-21). We also used student t-tests to compare the scores obtained by parents in the Family Impact Module to previously published scores obtained from parents taking care of chronically ill children at home (25). We used simple linear regression to investigate potential associations between PedsQL^{TM} total scores, Physical Health Summary scores, and Psychosocial Health Summary scores and patient's demographic and clinical characteristics.

Content analysis was used to identify themes in parents' responses to the two open-ended questions (26). The conceptual framework of the PedsQL^{TM} was used as a start-list of domains; responses that did not fit within the PedsQL^{TM} framework were grouped thematically to form new domains. A third investigator independently coded parent's answers using the final coding scheme that consisted of both the PedsQL^{TM} and new domains.

RESULTS

Parents of 23 children with IF completed the PedsQL^{TM} questionnaires and answered the two open-ended questions. There were no missing answers in the questionnaires. The mean age of the study children was 28.54 months (SD= 16.87); 69.6% were girls. The distribution of race/ethnicity was as follows: 52.2% White non-Hispanic ethnicity, 21.8% Hispanic, 13% Asian or Pacific Islander ethnicity, and 3% mixed ethnicity. The diagnoses leading to IF varied and included: necrotizing enterocolitis (43.6%), gastroschisis (34.9%), midgut volvulus (4.3%), intestinal atresia (4.3%), Hirschprung's disease (4.3%), meconium peritonitis (4.3%), and congenital short bowel syndrome (4.3%). The mean length of time with IF was 28.32 months (SD= 13.96). Sixteen of the 23 children (69.6%) were PN dependent at the time of the study; 20 of them (86.96%) were able to feed enterally, either by mouth, through a gastric tube, or both. The mean length of total inpatient hospital stay was 141.12 days (SD= 101.63) (Table 1).

Results of the linear regression analyses showed there were no statistically significant associations between any of the clinical or demographic characteristics tested and the total PedsQL^{TM} HRQOL scores (Table 2). Likewise, there were no statistically significant associations between any of the patient characteristics tested and the Physical Health Summary (Table 3). Mixed race/ethnicity was the only characteristic associated with a decreased Psychosocial Health Summary score.

PedsQL^{TM} 4.0 Generic Core Scales and Infant Scales Results Compared to Existing Norms

Compared to existing norms for healthy children, subjects aged 1-12 months (n= 7) scored lower on all domains of the PedsQL^{TM} (Table 4) (20). The mean differences ranged from 2.12 points lower on the Cognitive Functioning domain to 12.54 points lower on the Physical Functioning domain. The differences that reached statistical significance were all related to physical health: Physical Functioning (p= 0.004), Physical Symptoms (p= 0.006), and Physical Health Summary scores (p= 0.002).

Children aged 13-24 months (n= 4) also scored significantly lower than existing norms for healthy children in all aspects of physical health (20). The differences ranged from 11.29 to 18.79 points lower in the Physical Functioning (p <0.001) and Physical Symptoms (p= 0.02) domains (Table 4). In addition, children in this age range scored significantly lower in the Cognitive Functioning domain, resulting in a significantly lower overall Psychosocial Health Summary score (p= 0.012).

Children aged 2-3 years (n= 3) also scored lower in all domains of the PedsQL^{TM} (21). The mean difference ranged from 1.88 points lower in the Social Functioning domain to 26.50 points lower in the Physical Functioning domain compared to healthy normative values (Table 4). Because none of these children were enrolled in school, their parents did not complete items in the School Functioning domain. Similar to children aged 1-12 months, only the Physical Functioning domain score was significantly different from that of healthy children (p= 0.003).

Children aged 3-4 years (n= 7) had scores 7.77 to 27 points lower than scores published for healthy children of the same age (21). All differences in domain scores and summary scores reached statistical significance except for the Emotional Functioning domain (Table 4).

In this sample, there was only one child between 4 and 5 years of age and only one child between 5 and 6 years of age. Given confidentiality concerns, we choose to not report individual test scores for these two study subjects.

Comparison of Family Impact Module Results with Scores Reported by Other Parents of Children with Chronic Illness

Parents' responses on the Family Impact Module were compared with scores obtained in a study of 23 parents of children with chronic illness living either at home or at a long-term care facility since there are no previously published Family Impact Module scores for parents of healthy children (25). Differences ranged from 14.60 points lower in the Family Relationships domain to 12.34 points higher in the Communication domain. There were no statistically significant differences between our sample and the prior published scores (Table 5).

Parents' Opinions of the Adequacy of the PedsQL^{TM} to Describe Their and their Child's HRQOL

Eight of 23 parents (35%) reported that the items contained within the PedsQL^{TM} questionnaires provided an adequate description of the HRQOL of their child and family. The majority (65%)

said the PedsQL™ failed to address important impacts of IF on their children and families and offered additional information. Most of the information elaborated concepts contained within the PedsQL™ but with examples unique to IF (Tables 5 and 6). For instance, several parents mentioned decreased mobility or decreased independence secondary to their child's tunneled central line or gastrostomy tube. Some examples included were: "She was not able to crawl until they got rid of her g-tube" and "He can't carry all his TPN bags and lines, so that limits his independence". Six parents (40%) described a decreased or delayed ability of their child to achieve normal developmental milestones, specifically in potty training, through statements such as: "Potty training is very hard, he doesn't poop a few times a day like normal kids, it can be up to 10 times a day sometimes". Both of these themes pertain to the PedsQL™ domain of Physical Functioning.

Several parents also expressed concern regarding their child's inability to do things that other children of the same age could do. Statements associated with this theme, which is reflected in the PedsQL™ Social Functioning domain, included: "She can't eat the same things that every other kid around her can" and "Other kids are munching on stuff at this age and she can't". Seven parents (47%) reported a significant impact on HRQOL associated with diarrhea in statements such as "Sometimes he just explodes, the diaper doesn't hold it all in" and "Every time, the minute he would eat, he would poop". This type of concern is present in the PedsQL™ Physical Symptoms domain for children younger than 2 years, but not present in PedsQL™ questionnaires for older children.

Other comments, offered by four parents (27%), are consistent with the PedsQL™ Family Impact Module domain of Worry. Examples often included worry about their child injuring themselves with their central line or feeding tubing. One said it this way: "We worry he will strangle himself with all the cords that are always around him". Two parents (7%) shared concerns about the impact of their child's condition on the family budget, noting the daily expenses of specialized diapers and the expense of numerous loads of laundry or special meal planning. Seven parents (47%) provided additional information about impacts on their own Daily Activities and Social Functioning related to IF causing their children to stool and vomit often. Specific examples affecting their daily activities included the time required for frequent, large loads of laundry, bathing and changing their children's clothes several times a day, and numerous diaper changes. Examples affecting their social functioning mainly related to the social embarrassment associated with their child's unpredictable stooling and vomiting.

Our qualitative coding of parents' discussion of the impacts of IF on quality of life identified one new domain not reflected in the PedsQL^{TM} questionnaires (Table 6). The themes that typify this domain reflect how the complex symptoms of IF interfere with parents' ability to nurture and bond with their child. Five parents (22%) discussed this challenge with examples such as: "She loves baths and she gets very excited about them, and I do too, but we can't do them because of the risk of infection"; "Instead of taking her to the playground, what I end up doing is taking her to the hospital instead"; and "We want to feed him what he wants when he wants, but we don't want him stooling out".

DISCUSSION

Pediatric IF has evolved from a terminal illness to a chronic disease over the last decade. Thus, HRQOL is now an aspect of pediatric IF health outcomes that can guide medical providers and caregivers in patient management decisions. With this in mind, the purpose of this study was to evaluate the HRQOL of children with IF and their families through well-established measuring tools and investigate whether these tools were adequate to perform this function.

The results of this study showed that children with IF have a lower HRQOL than healthy controls as measured by the PedsQL^{TM} Infant Scales and 4.0 Generic Core Scales. This is consistent with our hypothesis that IF is a chronic health condition that affects patients' HRQOL similar to other chronic health conditions (27-30). This is also in line with a previously published study that found a significant level of distress in children receiving home PN (14). Conversely, two other prior studies of the HRQOL of PN-dependent children failed to show a difference between their HRQOL and that of healthy children (12-13). Our data is more consistent with previous data concerning HRQOL in adult patients with IF, for which multiple studies have reported a significant impairment in HRQOL (31-34). Additionally, our study found that parents of children with IF reported a lower HRQOL, with scores comparable to that of other parents caring for children with chronic health conditions (25).

As expected, the greatest discrepancy in HRQOL in children with IF compared to healthy children relates to physical functioning. This was true for all age groups, as indicated by lower scores on the Physical Health Summary (Table 4). Many patients with IF have central venous access lines and gastrostomy tubes, along with the tubing necessary to connect these devices to bags containing PN or formula. It is not surprising that this equipment interferes with the child's ability to perform normal physical functioning tasks, such as crawling, walking, bathing or playing. The reported inability to physically perform at the same level as their healthy peers appeared greater for older children with IF than it was for younger patients, although this trend did not reach statistical significance (Table 3). This is consistent with the increase in physical activity in children seen as they move from infancy into the toddler stage. Thus, a larger sample size might be able to uncover an association between age and the Physical Health Summary scores of children with IF.

Our findings in the psychosocial health realm were less predictable. There was a wide range of variability and no specific trends in the average scores obtained for each age strata. Nonetheless, children with IF invariably scored lower in the Psychosocial Health Summary compared to normative values for healthy children (Table 4). This is in line with historical data relating chronic illness with decreased psychosocial functioning and with a prior study reporting a large degree of psychological distress in PN dependent children (14, 35-36). Thus, it is likely that, with a larger sample, this difference would have reached statistical significance. Mixed race/ethnicity was associated with lower Psychosocial Health Summary scores, although the basis for this association is unclear. Previous studies have reported a decreased HRQOL in children of ethnic minorities with chronic illnesses, specifically in the psychosocial realm (37-38). Mixed race could be considered a minority and our findings may be explained by the complex relationships between minority race and socioeconomic status, differences in disease incidence, and differences in cultural views. However, we did not find an association between Hispanic ethnicity, also a minority, and HRQOL in this population. Thus, further research is needed to elucidate the sources of decreased HRQOL in mixed race children with IF.

None of the patient or disease variables we examined were associated with child or parent HRQOL total scores or child Physical Health Summary (Tables 2 and 3). Thus, we did not have any concrete characteristics to be included in a multivariable regression model. However, a number of variables examined can presumptively be associated with HRQOL in children with IF. Specifically, a longer length of time with IF, regardless of the way a child is nutritionally supported, could be associated with higher HRQOL if, with time, patients and parents learn how to cope with the condition and children advance in their intestinal rehabilitation. Similarly, PN dependence in itself could be related to a lower HRQOL. Multiple studies in adult patients have shown that PN dependent patients with IF have a lower HRQOL than enterally fed patients with IF (34, 39-41). On the other hand, being able to receive feeds by either mouth or gastrostomy tube could be associated with an improved HRQOL, even if a patient remains dependent on PN while taking enteral feeds. There is no previous data in children investigating these potential associations and it is likely that our study lacks the necessary power to detect them. However, these potential associations unique to IF are plausible and warrant further investigation.

While our study was able to show a low HRQOL in children with IF and their parents, we found that there are additional challenges for these families that are not adequately reflected in the generic PedsQL^{TM} questionnaires. For example, the PedsQL^{TM} Infant Scales has a specific domain regarding physical symptomatology affecting HRQOL that is not present in the questionnaires for children aged 2 years and older. Children with IF have physical symptoms that affect their HRQOL and persist for the duration of their disease, such as diarrhea, vomiting, and gastric tube leakage (Table 6). Fortunately, there is a PedsQL^{TM} Gastrointestinal Symptom Module that addresses some of these concerns currently undergoing field testing (42). Likewise, parents reported significant concerns regarding potty training (Table 6). This is not directly addressed in the PedsQL^{TM} questionnaires and deserves additional consideration in this population. The inability of children with IF to eat as healthy children do, a central facet of pediatric IF, is also something that should be considered in a HRQOL questionnaire designed for this population. While the PedsQL^{TM} has a Social Functioning domain in which it addresses a child's inability to do the same things other kids their age can do, eating is a basic human function that appears to be of important significance in children with IF.

Many parental concerns with regards to their own HRQOL are present in the PedsQL^{TM} Family Impact Module including feelings of frustration, worry about treatment side effects and lack of time for social activities. However, we identified a number of issues these parents experience that could be highlighted in an IF-specific questionnaire (Table 7). For example, social embarrassment related to increased stooling, vomiting or gastrostomy tube problems could be added to the Social Functioning domain. Also, concerns about the increased financial cost of managing their child's illness could be highlighted within the Worry domain of the Family Impact Module. This module specifically addresses the impact of caring for a chronically ill child on every day activities such as completing chores and preparing for family activities. However, parents of children with IF stressed the increased time and effort required specifically to take care of their child. This function is missing from the Family Impact Module and should be reflected prominently in a future questionnaire directed towards families dealing with IF.

Parents' responses to the open-ended questions revealed an additional challenge that is not contained within any of the existing PedsQL^{TM} domains. Parents reported significant distress associated with constraints on their ability to nurture their child (Table 7). This was demonstrated in statements expressing how parents felt unable to do simple, common things they would usually do with their children if they did not have IF, such as bathing them, taking them to the park, or indulging their food cravings. The barriers were directly related to the child's IF, either due to treatment requirements of their condition (i.e.: central lines preventing baths) or symptomatology (i.e.: hesitancy to take the child outside the home due to frequent diarrhea). This would be a key domain to include in a future HRQOL questionnaire for children with IF and their parents.

The greatest limitation to this study is its small sample size. The SCH-IFP is the only intestinal rehabilitation program in a large catchment area, receiving referrals from 5 different states for the management of patients with IF. However, IF remains a rare condition and the number of patients being actively cared for at SCH with IF is approximately 20-30 at any given time. Fortunately, this was a low risk study and the majority of families being cared for at the SCH-IFP during the study period enrolled in our study. However, this still left us with relatively low numbers of patients in total (n=23), and specifically in the 13-24 month (n=4) and 2-3 year (n=3) age range. A multi-institutional study with a larger sample of patients would increase the power to detect any associations between HRQOL and patient characteristics thought to affect HRQOL scores. Furthermore, this is a cross sectional study and as such, these findings ought to be substantiated through a longitudinal study. Given the improved long-term survival in pediatric IF, it would be interesting to evaluate whether HRQOL measurements change over time as these children grow and develop.

Another limitation to our study relates to referral practices to intestinal rehabilitation programs. While data demonstrate that IF is best managed at multidisciplinary programs such as the one at SCH, these programs are relatively new and providers in remote areas might be unaware of the benefits of referring their patients with IF early in the disease course. Thus, it is possible that the SCH-IFP cares primarily for more complicated patients with long-standing IF refractory to feeding advancement. This could bias our results towards more complex patients who may experience a poorer HRQOL than would be true for the whole IF population and limit the generizability of our results.

Finally, we were unable to audio-record the parent answers to our open-ended questions. Collecting this information by typing parents' responses as they speak could create bias if the study investigator selects to write down some comments and not others. We minimized the risk of this bias by having a study investigator with excellent typing skills knowledgeable of the language associated with the care of patients with IF typing parents' responses and reading them back to the parents to assure their accuracy.

In conclusion, children with IF, similar to other children with chronic health conditions, appear to have a lower HRQOL than healthy children. Their parents also withstand a significant impact from their child's condition that adversely influences their own quality of life. These children and parents have concerns expressly related to IF that are not effectively addressed in the PedsQL^{TM} generic questionnaires. This population would benefit from a disease-specific HRQOL questionnaire to more thoroughly investigate the extent to which this disease affects their and their families' lives.

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Table 1: Descriptive Statistics for Demographic and Clinical Characteristics of 23 Study Participants

Characteristic	Mean ± SD or N (%)
Age (months)	28.54 ± 16.87
Gender	
Girls	7 (30.4%)
Boys	16 (69.6%)
Race/Ethnicity	
White Non-Hispanic	12 (52.2%)
Black Non-Hispanic	0
Hispanic	5 (21.8%)
Asian/Pacific Islander	3 (13.0%)
Other	3 (13.0%)
Diagnosis	
Gastroschisis	8 (34.9%)
Necrotizing Enterocolitis	10 (43.6%)
Midgut Volvulus	1 (4.3%)
Intestinal Atresia	1 (4.3%)
Hirschprung's Disease	1 (4.3%)
Meconium Peritonitis	1 (4.3%)
Congenital Short Bowel	1 (4.3%)
Length of Time with IF (months)	28.32 ± 13.96
PN Dependent	16 (69.6%)
Enterally Fed	20 (87%)
Feeding by Mouth	16 (70%)
Feeding by Gastrostomy Tube	13 (56.5%)
Length of Inpatient Stay (days)	141.12 ± 101.63

Table 2: Linear Regression Analyses Testing Associations between Patient Characteristics and PedsQL^{TM} Generic Scales or Infant Scales and Family Impact Module Total Scores

Characteristic	PedsQL^{TM} Generic Core Scales or Infant Scales			PedsQL^{TM} Family Impact Module		
	Coef	95% CI	p-value	Coef	95% CI	p-value
Age (months)	-1.6	-6.1, 2.9	0.473	-0.7	-6.7, 5.3	0.819
Gender						
Girls	(reference)					
Boys	-4.9	-17.3, 7.6	0.427	-4.3	-20.9, 12.3	0.594
Race/Ethnicity						
White Non-Hispanic	(reference)					
Hispanic	-1.0	-17.3, 15.3	0.904	2.8	-24.0, 70.2	0.840
Asian/Pacific Islander	-11.0	-29.1, 7.1	0.232	-12.8	-36.6,11.1	0.295
Mixed Race/Ethnicity	-8.3	-19.9, 3.3	0.163	7.1	-13.1,27.3	0.493
Diagnosis						
Necrotizing Enterocolitis	(reference)					
Gastroschisis	2.3	-11.3, 16.0	0.736	-9.8	-25.5, 5.9	0.220
Other*	6.7	-7.4, 20.7	0.353	-5.3	-31.4, 20.7	0.689
Length of Time with IF (months)	-0.1	-0.5, 0.4	0.775	0.1	-0.4, 0.7	0.715
PN Dependent	2.4	-10.2,15.0	0.696	-7.6	-23.9,8.8	0.348
Feeding by Mouth	6.8	-6.5, 20.2	0.313	2.3	-16.2, 20.7	0.810
Feeding by Gastrostomy Tube	-3.3	-15.4, 8.7	0.587	3.5	-11.5, 18.5	0.649
Length of Inpatient Stay (days)	-0.0	-0.1,0.0	0.404	0.0	-0.1,0.1	0.986

*Other: Congenital short bowel, Hirschprung's disease, intestinal atresia, meconium peritonitis and midgut volvulus.

Table 3: Linear Regression Analyses Testing Associations between Patient Characteristics and PedsQL^{TM} Physical Health Summary Scores and Psychosocial Health Summary Scores

Characteristic	PedsQL^{TM} Physical Health Summary			PedsQL^{TM} Psychosocial Health Summary		
	Coef	95% CI	p-value	Coef	95% CI	p-value
Age (months)	-0.6	-5.7, 4.6	0.829	-1.3	-7.1, 4.6	0.672
Gender						
Girls	(reference)					
Boys	-1.2	-18.2, 15.9	0.893	-5.8	-18.0, 6.4	0.353
Race/Ethnicity						
White Non-Hispanic	(reference)					
Hispanic	15.5	-3.1, 34.0	0.103	-8.0	-25.9, 9.9	0.380
Asian/Pacific Islander	-10.3	-32.7, 12.0	0.364	-12.8	-29.2, 3.6	0.127
Mixed Race/Ethnicity	3.4	-15.4, 22.3	0.720	-14.2	-23.4, -4.9	0.003
Diagnosis						
Necrotizing Enterocolitis	(reference)					
Gastroschisis	-0.9	-22.5, 20.8	0.938	5.2	-6.2, 16.6	0.369
Other*	6.7	-14.0, 27.5	0.525	6.1	-10.0, 22.3	0.458
Length of Time with IF (months)	-0.2	-0.8, 0.5	0.628	0.1	-0.3, 0.5	0.746
PN Dependent	3.0	-18.1, 24.0	0.782	2.2	-8.0, 12.4	0.669
Feeding by Mouth	8.7	-11.4, 28.7	0.397	5.6	-8.7, 19.9	0.442
Feeding by Gastrostomy Tube	-6.4	-22.5, 9.7	0.438	-3.3	-15.9, 9.3	0.609
Length of Inpatient Stay (days)	-0.1	-0.2, 0.1	0.256	0.0	-0.1, 0.0	0.759

*Other: Congenital short bowel, Hirschprung’s disease, intestinal atresia, meconium peritonitis and midgut volvulus.

Table 4: PedsQL^{TM} Generic Scales or Infant Scales Mean Results for Children 1-12 Months, 13-24 Months, 2-3 Years, and 3-4 Years Old Compared to Existing Norms

Parameter	Mean (SD)	Mean (SD)	Mean Difference	p-value	95%CI
	IF Infants 1-12 Months (n=7)	Varni et al Data (n=246) (20)			
Total	76.04 (9.54)	82.47 (9.95)	-6.43	0.093	-13.93, 1.07
Physical Health Summary	73.21 (14.89)	84.98 (9.45)	-11.77	0.002	-19.03, -4.51
Physical Functioning	75.00 (17.18)	87.54 (11.16)	-12.54	0.004	-21.10, -3.98
Physical Symptoms	72.14 (15.51)	83.45 (10.39)	-11.31	0.006	-19.27, -3.35
Psychosocial Health Summary	75.54 (8.06)	80.47 (12.64)	-4.93	0.306	-14.40, 4.54
Emotional Functioning	72.32 (10.33)	76.59 (13.71)	-4.27	0.415	-14.57, 6.03
Social Functioning	87.50 (20.09)	89.62 (14.87)	-2.12	0.713	-13.46, 9.22
Cognitive Functioning	73.21 (12.87)	83.11 (20.65)	-9.90	0.209	-25.37, 5.57
	IF Infants 13-24 Months (n= 4)	Varni et al Data (n=141) (20)			
Total	72.39 (4.85)	85.55 (8.74)	-13.16	0.003	-21.86, -4.46
Physical Health Summary	74.01 (8.62)	88.84 (7.68)	-14.83	<0.001	-22.55, -7.11
Physical Functioning	71.53 (12.70)	90.32 (8.96)	-18.79	<0.001	-27.86, -9.72
Physical Symptoms	76.25 (15.34)	87.54 (9.29)	-11.29	0.020	-20.77, -1.81
Psychosocial Health Summary	68.99 (3.88)	83.12 (11.02)	-14.13	0.012	-25.07, -3.19
Emotional Functioning	77.08 (13.50)	78.60 (12.80)	-1.52	0.815	-14.36, 11.32
Social Functioning	91.25 (11.81)	91.14 (10.77)	0.11	0.984	-10.71, 10.93
Cognitive Functioning	45.83 (19.31)	84.65 (15.76)	-38.82	<0.001	-54.70, -22.94
	Toddlers 2-3 Years (n= 3)	Varni et al Data (n=946) (21)			
Total	76.18 (20.28)	88.14 (12.11)	-11.96	0.089	-25.73, 1.81
Physical Health Summary	63.54 (30.83)	90.04 (15.07)	-26.50	0.003	-43.66, -9.34
Psychosocial Health Summary	82.50 (15.67)	86.83 (12.41)	-4.33	0.547	-18.4, 9.76
Emotional Functioning	78.33 (20.21)	84.55 (14.00)	-6.22	0.443	-22.13, 9.69
Social Functioning	86.67 (12.28)	88.55 (16.08)	-1.88	0.840	-20.13, 16.37
	Toddlers 3-4 Years (n=7)	Varni et al Data (n=907) (21)			
Total	70.33 (15.09)	87.96 (11.63)	-17.63	<0.001	-26.31, -8.95
Physical Health Summary	66.07 (26.91)	90.07 (14.24)	-24.00	<0.001	-34.69, -13.31
Psychosocial Health Summary	72.28 (11.46)	86.41 (12.16)	-14.13	0.002	-23.18, -5.08
Emotional Functioning	75.71 (12.39)	83.48 (14.52)	-7.77	0.158	-18.57, 3.03
Social Functioning	70.00 (25.33)	88.89 (14.13)	-18.89	0.001	-29.49, -8.29
School Functioning	68.75 (10.49)	88.31 (16.05)	-19.56	0.015	-35.33, -3.79

Table 5: PedsQL^{TM} Family Impact Module Mean Results Compared to Scores Reported by Other Parents of Children with Chronic Illnesses

Parameter	Parents of Children with IF (n=23)	Varni et al Data (n=11) (25)			
	Mean (SD)	Mean (SD)	Mean Difference	p-value	95%CI
Total	61.30 (17.32)	62.49 (17.26)	-1.19	0.852	-14.11, 11.73
Parent Functioning Summary	63.48 (15.64)	62.94 (19.83)	0.54	0.932	-12.23, 13.31
Physical Functioning	62.34 (17.83)	53.03 (22.83)	9.31	0.203	-5.27, 23.89
Emotional Functioning	64.13 (19.23)	64.48 (26.59)	-0.35	0.965	-16.63, 15.93
Social Functioning	60.05 (26.96)	61.93 (25.99)	-1.88	0.849	-21.79, 18.03
Cognitive Functioning	66.96 (21.78)	74.09 (18.95)	-7.03	0.360	-22.76, 8.50
Communication	64.49 (26.97)	52.15 (24.67)	12.34	0.209	-7.28, 31.96
Worry	61.74 (21.25)	56.82 (25.52)	4.92	0.558	-12.01, 21.85
Family Functioning Summary	57.61 (20.09)	68.81 (24.11)	-11.20	0.163	-27.20, 4.80
Daily Activities	46.38 (29.92)	51.89 (31.48)	-5.51	0.625	-28.22, 17.20
Family Relationships	64.35 (20.69)	78.95 (27.62)	-14.60	0.094	-31.83, 2.63

Table 6: Parents Answers to Open Ended Questions Affecting their Child’s HRQOL

PedsQL™ Domain	Theme	Example
Physical Functioning	Problems crawling/walking	“She was not able to crawl until we got rid of the g-tube” “He can’t carry all his TPN bags and lines so that limits his independence”
	Problems toilet training	“Potty training is very hard, he doesn’t poop a few times a day like normal kids” “Right now we are trying to potty train her and it is so hard…”
Physical Symptoms	Diarrhea	“Every time, the minute he would eat, he would poop” “Sometimes he just explodes, the diaper doesn’t hold it all in” “Her skin is macerated from how much she stools” “He has a large number of stools a day”
	Gastric tube leakage	“There is always broken skin around the g-tube and the area is always wet and irritated” “The g-tube leaks all the time”
Emotional Functioning	Feeling angry/frustrated	“It is frustrating for him because he can’t do what other kids can do, and he realizes that” “He doesn’t talk in the hospital, he gets grumpy, it is very hard for him” “Having to make adjustments for diarrhea or vomiting in the middle of the day is something that does bother him when he is at school”
	Requiring increased attention	“He is very needy” “He has gotten used to us being with him all the time and when we are not he gets really upset”
Social Functioning	Not able to do things other kids his/her age can do	“She can’t eat the same things that every other kid around her can” “Other kids are munching on stuff at this age and she can’t” “He can’t do everything that my other son can do” “There are limitations on what he can eat and cannot eat, like he can’t have sugar, or a bottle normally”

Table 7: Parents Answers to Open Ended Questions Affecting their Own HRQOL

PedsQL^{TM} Domain	Theme	Example
Physical Functioning	Feeling tired	“I get no sleep” “One or the other parent is always tired”
Emotional Functioning	Feeling frustrated/ anxious	“His feeding goal keeps going up and it’s frustrating, it feels like he is never going to get rid of his TPN” “It causes a lot of anxiety not being able to get her to eat different foods outside of what she eats” “I have issues not knowing what to feed her, it is not something you can find in a textbook” “Approaching each developmental step, such as potty training, is stressful because we always question how and when to do it in this situation”
Social Functioning	No time for social activities	“It is hard to live a life outside of this”
	Social embarrassment	“You leave poop everywhere, all over the floor, yourself, the restaurant…” “There is gastric contents all over the place” “The Mic-key is very obvious, it sticks out a lot” “He smells like formula all the time”
	Inability to do social activities	“It is an issue to be gone for long periods of time because his feeding pump needs to be on at specific times” “It is not easy to find a baby sitter”
Communication	Others don’t understand	“Mom and Dad don’t really understand what it is like, nobody can relate to it” “There is no way to put in words how difficult this is on a family” “Health care workers have no idea what it is like to be covered in poop and vomit all the time”
Worry	Worry about side effects	“We worry he will strangle himself with all the cords that are always around him” “We worry about his lines getting infected or breaking” “We are always worried that he is going to pull his g-tube or central line out” “We are concerned about feeding, do we feed him? do we not feed him?”
	Worry about letting child go outside	“We want him to go to school but there are a lot of germs in a school and we worry about it” “We couldn’t let him go outside, we weren’t about to let him crawl on the grass”
	Financial worries	“The diapers I use are $13 a piece, that is expensive” “We make huge investments in meal planning and then there is more food that gets wasted than eaten”

Table 7 continued

Daily Activities	Chores/caring for the child taking a lot of time/effort	“It takes about 30 minutes each time to change a poopy diaper” “We change the bed at night about 3-4 times, each night, the complete bed, and her clothes too” “It takes a long time to plan to go anywhere because of his central line and TPN dependence” “We change his clothes 2-4 times a day because he is always soiling them with how much he poops” “In the morning she HAS to get a bath and be scrubbed down because of all the stool she puts out and how often she vomits, and her bed has to be scrubbed down every day as well”
Family Relationships	Conflicts/stress tension between family members	“This can definitely destroy a marriage”
New Domain	Theme	Example
Caregiving	Challenges to nurturing or bonding with the child	“She loves baths and she gets very excited about them, and I do too, but we can’t do them because of the risk of infection” “Instead of taking her to the playground what I end up doing is taking her to the hospital instead” “We want to feed him what he wants, but we don’t want him stooling out” “There are things that are very hard to do, but we try anyway, like going camping” “We don’t let him do all the same things [as other kids]”