

## THE IMPACT OF PEDIATRIC GASTROINTESTINAL AND LIVER DISEASE ON PATIENTS' QUALITY OF LIFE: A SYSTEMATIC REVIEW

M. Hadi\*

*\*Faculty of Medicine, University of Malahayati, Indonesia*

**\*Corresponding Author:**

*mhadi.official2023@gmail.com*

---

### **Abstract**

**Aim:** *Identifies and evaluates all studies and instruments devised to measure quality of life (QOL) in children with gastrointestinal or hepatic diseases.*

**Methods:** *Using MEDLINE, EMBASE, CINAHL, and PsycINFO, pertinent articles published by the end of 2005 were identified through a search of the literature. These were examined by the two authors, and data were extracted using a standardized form. Articles were excluded if no attempt was made to measure QOL, if they did not pertain to minors 17 years of age, if they did not pertain to gastrointestinal or liver diseases, or if they were reviews. Identified quality of life instruments were evaluated based on proposed criteria.*

**Results:** *Following the first search, a total of 2379 publications were located; however, following the exclusion of 2309, only 70 research were considered for inclusion. These were classified as having one of the following conditions: inflammatory liver disease, numbering seven. These studies discuss the effects that bowel disease, with a sample size of  $n = 17$ , cystic fibrosis, with a sample size of  $n = 20$ , and surgery, with a sample size of  $n = 15$ , have on the quality of life of children who are afflicted with these conditions. Only five of these condition-specific quality of life instruments and one general instrument with a chronic disease module were found to meet the specified quality standards; hence, only five of these instruments may be recommended for use in the future.*

**Conclusion:** *Chronic gastrointestinal and liver illnesses can severely impact QOL for patients and their families. Several disease-specific paediatric QOL tools have been validated. Clinical practice and treatment evaluation should include quality of life. Research should focus on improving QOL in seriously handicapped youngsters.*

**Keywords:** *Pediatric quality of life, gastrointestinal disease, liver disease, quality of life's instrument, pediatric chronic disease*

**BACKGROUND**

According to the World Health Organization, health is "a state of complete physical, mental, and social well-being and not merely the absence of disease or infirmity".<sup>1</sup> Quality of life is a term used to describe a person's overall health. This encompasses all emotional, social, and physical aspects of a person's existence. When used in the context of medicine, the term is referred to as "Health-Related Quality of Life (HRQOL)".<sup>1</sup>

Approximately 12% of children and adolescents have abdominal pain predominant functional gastrointestinal diseases (AP-FGD).<sup>2</sup> Even though AP-FGD are not life-threatening, they are chronic and troublesome disorders that can have a significant impact on the lives of children who are affected. HRQoL becomes an essential objective measure of health status among children with AP-FGD in the absence of biological measures of disease activity. One study examined HRQOL and healthcare consultation among Sri Lankan 13–18-year-olds with abdominal pain-predominant functional gastrointestinal illnesses. AP-FGD children exhibit considerably worse HRQoL ratings for physical, emotional, social, and school functioning.<sup>3</sup>

Also, pediatric patients with liver disease and their parents had a poorer QoL than healthy children and parents. Cirrhosis lowers QoL in juvenile chronic liver disease patients.<sup>4</sup> 80 children with autoimmune hepatitis had worse physical, emotional, and school QoL than a healthy control group, according to Bozzini et al.<sup>5</sup> Tehranian et al. found worse health-related QoL in 55 chronic liver disease children than healthy controls.<sup>6</sup> Psychosocial health was most impaired in children chronic liver disease patients older than 10. Therefore, symptomatic alleviation, liver damage prevention, and psychological support in adolescents throughout long-term follow-up will improve patients' health-related QoL.<sup>4</sup>

Interviews or validated instruments, typically questionnaires, can be utilized to assess quality of life. Instruments for measuring quality of life can be disease-specific or generic. Generic instruments provide a comprehensive assessment of the child's QOL, while disease-specific instruments focus on issues related to a particular illness. Generic instruments can be used on healthy patients as well as those with any illness. Disease-specific instruments may be more sensitive to detect significant changes and include QOL components that are relevant to the patient population in question. Identifying problem-specific situations for individual children makes patient/treatment matching easier. DISABKIDS and KIDSCREEN are useful cross-cultural tools for measuring HRQoL in children and adolescents with chronic diseases. HRQOL instruments in public health surveys allow researchers to track population health over time, identify subgroups at risk of poor HRQOL, and evaluate public health interventions in a given population.<sup>7</sup>

**Methods**

We conducted a literature search using MEDLINE, EMBASE, CINAHL, and PsycINFO. The purpose of the search was to identify, up until the end of 2005, all publications addressing QOL in minors with gastrointestinal and hepatic diseases. We adapted Eiser and Morse's<sup>9</sup> search strategy, but instead of searching for chronic diseases, we looked for gastrointestinal and liver diseases. This strategy was used to search MEDLINE and was modified for the other databases.

1. "quality of life" OR "satisfaction" OR "happiness" OR "health status" OR "well-being"
2. Mesh/explode all subheadings with "quality of life"
3. #1OR#2
4. infant OR child, preschool OR child OR adolescents
5. "infant" mesh or exploded all of the subheadings
6. Make all subheadings mesh with or erupt under the headline "child, preschool"
7. Replace/explode all subheadings with "child"
8. "adolescent" should mesh/explode with all subheadings.
9. #4OR#5OR#6OR#7OR#8
10. gastrointestinal disease OR (gastr\* reflux) OR esophag OR oesophag\* OR gastr\* OR ulcer OR Hirschsprung OR intussusception OR (inflammatory bowel disease) OR crohn\* OR intolerance OR malabsorp\* OR celiac OR coeliac OR diarrh\* OR constipat\* OR (cystic fibrosis) OR pancrea\* OR liver OR hepat\*
11. "diseases of the digestive system" should mesh/explode all subheadings.
12. #10 OR #11
13. #3 AND #9 AND #12

We searched the reference lists of identified studies and the Patient-reported Outcome and Quality of Life Instruments Database<sup>10</sup> for additional studies and instruments, respectively. The abstracts of the identified publications were then independently reviewed by the authors. Publications were excluded if no attempt was made to measure QOL, if they did not pertain to minors under the age of 17, or if they did not pertain to gastrointestinal or hepatological disorders. Similarly, review articles were excluded. The remaining articles were obtained and evaluated using a modified data extraction form from Eiser and Morse.<sup>9</sup>

The Appendices at the conclusion of this article contain a listing of the excluded and included items. In Table 1 are enumerated all identified disease-specific instruments for paediatric gastrointestinal or liver disease. We used the following criteria, which are considered<sup>9</sup> to be to be essential aspects of pediatric QOL instruments: developed according to established procedures including child participation in item generation, validity and reliability tested in children with

the disease in question using established techniques, and able to be completed by child (or parent/caregiver when the child is unable to complete the instrument).

**Results**

The first search turned up a total of 2379 papers; however, of those, 2194 were disqualified from consideration for inclusion in the title due to the fact that they did not include pediatrics, gastrointestinal, or hepatic disorders. After looking at the abstracts of the 185 articles that were still in the running, an additional 73 were disqualified for the identical reasons. After reading the entire article, the current authors decided to eliminate 42 of the remaining 112 papers (see Appendix I), which resulted in just 70 of the research being considered for inclusion (see Appendix II). These were divided up into the following categories: inflammatory bowel disease (IBD), which accounted for 17, cystic fibrosis (CF), which accounted for 20, liver disease (which accounted for 11), surgery (which accounted for 15), and other, which accounted for 7. It was not feasible to do a meta-analysis of the results since the data were acquired in a variety of methods and with the assistance of a variety of tools.

**Table 1. Identified disease-specific instruments**

Instrument or Authors (ref)	Disease	Item generation from children	Validity reability and formally tested in children	Completed by children	Comments
Rabbett et al (14)	IBD	Partially	No	Yes	Superseded by IMPACT
IMPACT (16-18)	IBD	Yes	Yes	Yes	
Cystic Fibrosis Questionnaire (CFQ) (40-44)	CF	Yes	Yes	Yes	Child and adolescent/ adult versions
Cystic Fibrosis QOL Questionnaire (CFQOL) (45)	CF	Yes	Yes	Yes	Adolescents and adults only
DISABKIDS (46-47)	Generic and chronic illness module	Yes	Yes	Yes	Module for 7 chronic illnesses including CF
Zamberlan QOL Questionnaire for School Age Children after Liver Transplantation (ZQLQ) (57)	Liver transplantation	No	No	Yes	
Hanneman et al (67)	Hirschsprung disease and anorectal malformations	No	Yes, but problems	Yes	Better validity in adults than children
Ditesheim et al (65)	Imperforate anus	No	No	Parent	
Bai et al (68)	Anorectal malformation	No	No	Parent	
O'Neill et al (73)	Children with neurological impairments undergoing antireflux procedure	No	No	Parent	
Tawfik et al (75)	Children with severe disabilities with feeding problems	No	No	Parent	
Defecation Disorder List (79)	Constipation and functional nonretentive faecal soiling	Yes	Yes	Yes	

**Inflammatory Bowel Disease**

IBD was determined to have the greatest significant impact on a person's mental health in a meta-analysis of the effects of chronic illnesses.<sup>11</sup> In one research, 56% of children with inflammatory bowel disease (IBD) had a mental problem, compared to 18% of control patients who did not have IBD. These disorders were almost primarily emotional in nature.<sup>12</sup> A second study of children who suffered from chronic diseases found that 60% of children with inflammatory bowel disease (IBD), 30% of children with tension headaches, 20% of children with diabetes, and 15% of healthy controls suffered from psychological problems.<sup>13</sup> According to the findings of the first ever study to be published on quality of life in children with inflammatory bowel disease (IBD), conducted by Rabbett et al.<sup>14</sup>, a significant number of the children who were affected had difficulties attending school, participating in sports, going on vacations, and staying at the homes of friends. More depressed symptoms were seen in children who were undergoing steroid treatment. A recent study conducted in the Netherlands<sup>15</sup> found that younger children (ages 8 to 12 years) with inflammatory bowel disease had a quality of life (QOL) that was comparable to that of a reference population and better cognitive function; however, adolescents with inflammatory bowel disease had a QOL that was significantly impaired in four domains (body complaints, motor functioning, autonomy, and negative emotions).

The Quality of Life Assessment Tool for Children with Inflammatory Bowel Disease (IMPACT) was developed in Canada.<sup>16</sup> A list of the ways in which having IBD impacted the lives of these 82 children was developed over the course of conducting interviews with these children. These concerns were taken into consideration while developing an item reduction questionnaire, which was then given to 117 more children diagnosed with IBD. The patients rated how frequently each symptom disturbed them as well as how significant each item was to them. The items that scored the highest were utilized in the creation of the IMPACT questionnaire. IMPACT was shown to be a valid and reliable measure of quality of life in older children and adolescents with ulcerative colitis and Crohn disease, according to a second validation research that was conducted in Canada with 147 children ranging in age from 9.2 to 18.0 years.<sup>17</sup> Patients with IBD who did not have active illness had a considerably higher mean total IMPACT score than those who did have active disease. In addition to being translated into French, the initial measurement tool has also been reduced and verified in the Netherlands<sup>18</sup>, where it was developed. The worries of children diagnosed with IBD in the United Kingdom are found to be largely comparable to those of children diagnosed with IBD in Canada, according to a comparison that was carried out across cultural boundaries.<sup>19</sup> In addition, a computerized touch-screen version has been created<sup>20</sup>, and further validation is currently being conducted in the United Kingdom.

In a trial of children with active Crohn disease who were treated with exclusive enteral nutrition, 23 out of 26 children displayed clinical remission at 8 weeks, with improvements in QOL ratings evaluated using the IMPACT questionnaire.<sup>21</sup> The change in QOL score was predictive of achieving a clinical remission, but it did not indicate an improvement in histology status. A prospective study of children with inflammatory bowel disease (IBD) who attended a camp for one week showed a substantial increase in both the total IMPACT score as well as several of the specific domain scores.<sup>22</sup>

It has been established that adolescents who have inflammatory bowel disease who have closer social support networks have greater quality of life ratings. Those who had recently developed IBD were more likely to seek emotional support from their families than from their classmates, and they relied more on their parents' ability to cope than on their own. There was a strong correlation between adolescents' and their parents' ratings of stressful events<sup>23</sup>, and both groups' quality of life scores. It was discovered that parents are acceptable raters of the child's quality of life in terms of its objective components, but not in terms of its more subjective components.<sup>24</sup> It was discovered that pediatricians overestimate the severity of patients' physical symptoms.<sup>8</sup> When compared to their healthy peers, adolescents who had inflammatory bowel disease were more likely to utilize avoidant coping methods.<sup>25</sup> There was a correlation between the employment of a predictive coping style and a reduced usage of a depressed reaction pattern and improved quality of life in three out of the six areas.

A research that took a cross-sectional approach with Dutch youngsters found that adolescents with inflammatory bowel disease (particularly males) had a lower quality of life and exhibit more problem behavior that is internalizing compared with their healthy counterparts.<sup>26</sup> Self-esteem was a crucial factor in determining quality of life. In the United States, a cross-sectional research of long-term psychosocial outcomes in children with inflammatory bowel disease (IBD) found that 20% of the children with IBD had behavioral or emotional symptoms, which was comparable to the percentage of symptoms seen in healthy control participants.<sup>27</sup> An Italian research examining the long-term consequences of surgery on 21 children with ulcerative colitis found that 15 of the children had normal emotional status, and 14 of the children had normal social lives 21 years after the operation.<sup>28</sup>

In a study of the parents and siblings of children who had inflammatory bowel disease (IBD), the parents were concerned about the influence that IBD would have on their kid's future (for example, their child's chances of getting a job, getting married, and being independent) and were concerned about the difficulties that the ill child encountered at school.<sup>29</sup> Other concerns were the negative impact that the medicine was having on the patient and their feelings of guilt. Concern was expressed by siblings over the health of their brother or sister and the treatment that was being administered. They were also anxious about other children picking on them and the fact that their parents were keeping information about one of their siblings' illnesses from them. A study with a cross-sectional design was conducted in Australia with 46 parents of children diagnosed with IBD. The results revealed that the most prevalent worries were connected to the potential adverse effects of drugs and the child's future possibilities. Parents were pleased with several parts of the treatment provided at the IBD clinic; nevertheless, many of them expressed a need for additional employees, including counselors and educators, as well as continued education and simple access to information that is current.<sup>30</sup>

### **Cystic Fibrosis**

According to the findings of two research<sup>31,32</sup>, parents of children and adolescents diagnosed with CF evaluated their kid's quality of life worse than their child did themselves. The parents also stated that their child's quality of life was inferior when compared to that of children who were healthy or those who suffered from other chronic illnesses.<sup>31</sup> The quality of life of children who suffered from asthma, diabetes, and CF was lower than that of a sample of children from the community. During the course of the two-year trial, children with CF saw a deterioration in their physical condition; nevertheless, the ratings for physical and family activities were unchanged.<sup>33</sup> There was no correlation found between family evaluation of quality of life and teenage assessment of quality of life, according to a research.<sup>34</sup> An international study found that English adolescents with CF and healthy controls had a worse quality of life compared to German adolescents with the same condition in numerous different dimensions.<sup>35</sup> Compared to the norms for the population, a cross-sectional research conducted in Australia revealed reduced levels of psychopathology.<sup>36</sup>

In the early investigations of quality of life in children with cystic fibrosis, researchers utilized general tools such as the Quality of Well Being Questionnaire (QWB)<sup>37,38</sup> and the Child Health Questionnaire (CHQ).<sup>39</sup> According to a survey that was conducted in 2001, the vast majority (92%) of CF centers in the United States did not monitor patients' quality of life (QOL) for clinical or research purposes.<sup>40</sup> Since that time, a number of diagnostic tools that are unique to diseases have been created. France is responsible for developing and validating the CFQ, which is a disease-specific instrument. The CFQ 14 is intended for use with adults and older adolescents, whereas the CFQ Child P is intended for use with children between the ages of 8 and 13 years old.<sup>41</sup> They consist of three different modules that evaluate quality of life, symptoms, and one's assessment of their health. Physical functioning, energy/well-being, emotions, social restrictions, role, embarrassment, body image, food problems, and treatment load were the nine QOL aspects that were found. The items on the test were developed from a total of 33 interviews with patients, as well as interviews with the patients' parents. A survey that was conducted in a cross-sectional fashion with 393 patients and their parents served as the basis for item reduction as well as the assessment of internal consistency, convergent validity, and discriminant validity. The repeatability and responsiveness of the findings were validated in a second research that involved 124 patients and 85 parents. The

demonstration of every psychometric property was a resounding success. The following languages have had their versions of this instrument translated: American English<sup>42,43</sup>, German<sup>44</sup>, and Dutch<sup>45</sup>.

The CFQOL instrument is a disease-specific tool that was developed in the United Kingdom to assess QOL in adolescents and adults who have cystic fibrosis (CF).<sup>46</sup> Unstructured interviews, self-administered questionnaires, interaction with multidisciplinary professionals, a literature review, and an analysis of existing quality of life instruments were used to identify the areas of concern for adults and adolescents living with cystic fibrosis (CF). Validation, test-retest reliability, and sensitivity testing were subsequently carried out after the first testing was completed. The following nine dimensions were identified: chest symptoms, physical functioning, body image, interpersonal connections, career worries, relationships at work, and concerns for the future. Robustness was shown in both the discrimination of illness severity and the sensitivity to changes in health.

The Chronic Generic Module and Condition-Specific Modules for Children and Adolescents with Seven Different Chronic Illnesses, Including CF, Were Developed and Validated as Part of the European DISABKIDS Project.<sup>47,48</sup> Focus groups were used in the development of the condition-specific modules in order to design the pilot version, and factor analysis was used in order to discover domain structures and to reduce the number of items. After putting the final version through its paces in a pilot study with 360 participants, it was then put through its paces in a field test with 1152 participants across seven countries in Europe. Cronbach's alpha values that ranged from good to excellent provided confirmation of the study's internal consistency.

Several research have investigated whether or not there is an association between quality of life and respiratory function. Two different studies were conducted in the United States using generic measures to evaluate quality of life. According to the findings of the first research, quality of life (QOL) worsened in proportion to the severity of respiratory symptoms, the number of concurrent medical illnesses, and the total number of drugs used.<sup>40</sup> The validity of the QWB was demonstrated by a connection between the QWB score and respiratory function<sup>49</sup> in the second trial that was conducted in the United States. This study demonstrated that the QWB was sensitive to the treatment of an acute exacerbation. Both the CFQ and the CFQOL have demonstrated a link between the severity of the disease and quality of life in validation studies.<sup>42,46</sup>

The severity of the disease is not the only factor that affects quality of life; studies conducted on both children and adults found that individuals with poor coping abilities had a worse QOL, even when the Schwachman score (which measures the severity of CF) was good.<sup>50</sup> This study also shown that patients' subjective health judgments explained the differences in their quality of life measures. The most substantial correlations between quality of life and coping mechanisms were found.

The quality of life scores of those who were screened for cystic fibrosis and those who were not in a research conducted in the United States showed no significant difference between the two groups. The CHQ was completed by 36 patients aged 10 to 15.5 years old who were involved in either the screened or control groups of the Wisconsin CF newborn screening experiment.<sup>51</sup> This was a sub-section of a larger randomised prospective trial that looked at the advantages and disadvantages of using the CF neonatal screening test.

The Caregiver Quality of Life in Children with Cancer (CQOLCF) scale<sup>52</sup> was established in the United States of America, and it has been shown that the QOL of the caregiver declined as the severity of the child's condition grew.<sup>53</sup>

### **Liver Disease**

Quality of life (as judged by the Sickness Impact Profile) was good before treatment, worsened during treatment, and reverted to baseline after three months of treatment cessation in a research that looked at the effect of a-interferon therapy on children with chronic viral hepatitis.<sup>54</sup> These children were between the ages of three and fourteen years old. Using the Pediatric Quality of Life Inventory, a pilot trial of metformin was conducted in 10 children aged 10 to 17 years old who had biopsy-proven nonalcoholic steatohepatitis. The study found that the children's quality of life was deteriorated at baseline, but that it significantly improved after therapy.<sup>55</sup>

Using the Vineland Adaptive Behaviour Scales, a preliminary effort was made to investigate the quality of life of 25 Australian children aged 0.58 to 14.2 years who had undergone liver transplantation. The results revealed satisfactory mean composite and domain scores.<sup>56</sup> Zamberlan came up with an instrument to assess quality of life (QOL) in children who had received a liver transplant based on a study of the relevant research literature, clinical experience, and consultations with subject matter experts.<sup>57</sup> The instrument has not been verified in an official capacity. Zamberlan conducted interviews with a group of twenty children, ranging in age from five to twelve years old, three to six years after receiving a liver transplant. Delays in socializing, difficulty in forming peer connections, as well as feelings of loneliness and vulnerability, were experienced by those affected by this issue. The child's family provided the most significant amount of social support, and as a result, the family's dynamics restored to normal. As a side effect of their sickness and treatment, the children's looks caused them significant emotional suffering. Five of them were concerned that they might require another transplant. Children reported being content with their lives and optimistic about the future, but they also displayed increased anxiety and forgetfulness, performed less well in schoolwork, and struggled to form healthy

connections with their peers. Six of them had issues with their behavior when they were at home. Kita et al.<sup>58</sup> inquired about the quality of life of seventy-two Japanese youngsters after they had received a liver transplant. The majority of participants believed that their quality of life had significantly improved, and there was no discernible difference between the living-related and brain-death donor groups. According to the findings of a number of research<sup>59-62</sup>, the quality of life experienced by children following liver transplantation is lower than that of a reference population. Buchu-Vulas et al.<sup>63</sup> found that the patients' quality of life was comparable to that of youngsters suffering from other chronic illnesses. A research that was conducted by Cole and colleagues in 2004<sup>64</sup> found that one year following liver transplantation, there were substantial increases in all subscale scores, with the exception of global mental health. In the majority of the studies, the quality of life was evaluated by the parents. However, in the research conducted by Schultz et al.<sup>60</sup>, both the children and their parents were asked to complete the evaluation, and their evaluations of the child's QOL were quite different.

### **Surgery**

The first research to evaluate quality of life in children who had undergone gastrointestinal surgery made use of an unvalidated questionnaire in families of children who had undergone repair of high imperforate anus.<sup>65</sup> The majority of young children's quality of life was not determined by their ability to achieve faeces continence, while the quality of life of older children was determined by their ability to achieve faecal continence. In a subsequent research, it was shown to be possible and valid to administer a proxy version (completed by a parent) of the EuroQOL to children who were being treated for imperforate anus.<sup>66</sup>

A quality of life (QOL) scale that accounts for Hirschsprung disease and anorectal malformation has also been created.<sup>67</sup> There was good discriminant ability in adults, but not in children, and there were issues with the diet domains in children. The hypothesized domain structure was validated with strong internal reliability, there was good discriminant ability in adults, and there were difficulties with the diet domains in children. A research conducted in China on children who had surgery for anorectal malformation revealed behavioral issues, difficulty with peer interactions, limited diets, and absences from school.<sup>68</sup> The quality of life was poorer in patients as compared to the controls, and it was also lower in children who had difficulty maintaining faecal continence. A Chinese investigation examined quality of life changes following the Swenson treatment for Hirschsprung disease came to same conclusions.<sup>69</sup> According to the findings of a study conducted in the Netherlands, the quality of life of young children with anorectal malformation was negatively impacted significantly, whereas the quality of life of older children and adults was not significantly different from that of the general population.<sup>70</sup> Patients born with a congenital diaphragmatic hernia had results that were comparable to those described above.<sup>70</sup>

In patients with ulcerative colitis and familial adenomatous polyposis who had ileoanal pull-through surgery, there was no significant improvement in quality of life compared to population norms.<sup>71</sup> A research conducted in the United States on children undergoing ileal pouch/anal anastomosis found that the children's overall health scores were lower than the standards for the United States, but the children's scores on most psychosocial variables were normal.<sup>72</sup>

Following surgery for gastroesophageal reflux disease in neurologically impaired children, a retrospective assessment of caregiver satisfaction was conducted using an unvalidated questionnaire. The results of this survey revealed that both the child's and the caregiver's quality of life increased as a result of the procedure.<sup>73</sup> A following research that looked at the quality of life of carers of children with neurological impairments found that those who had oesophagogastric disassociation had a significantly better quality of life compared to those who had fundoplication.<sup>74</sup>

After the kid had a gastrostomy (with fundoplication if there was severe reflux), both the child's quality of life and the quality of life of the caregiver improved, according to the findings of another retrospective study that used an unvalidated questionnaire to collect data on children with severe impairments who had eating issues.<sup>75</sup> A recent research of maternal carers of children who were dependent on a gastrostomy found that these children required twice as much care time as caregivers of children who did not have a gastrostomy.<sup>76</sup> However, the caregivers of these children were not any more sad or less content with their life than caregivers of children who did not have a gastrostomy. The quality of life of caretakers of children diagnosed with cerebral palsy was evaluated by Sullivan et al.<sup>77</sup> before to the placement of gastrostomy tubes, as well as 6 and 12 months after the procedure. Caregivers reported a substantial decrease in the amount of time spent feeding their child, an improvement in the ease with which medications could be administered, and a reduction in their level of anxiety over their child's nutritional health. The mean scores on the Short Form-36 questionnaire improved in numerous categories after six months, and after twelve months, they were equivalent to normal reference standards. This was the case when the questionnaire was administered.

In an uncontrolled research of children with oesophageal atresia, parental reports indicated a superior quality of life in those who underwent gastrostomy, cervical oesophagostomy, and no effort at anastomosis compared with those who had previous attempts at reconstruction<sup>78</sup>; this was the case even though the study did not include any controls. In neither of the two groups did the children's evaluations of their quality of life differ from one another.

### **Miscellaneous**

Using established standards as a foundation, researchers in the Netherlands have devised a disease-specific instrument that may assess quality of life in children who suffer from functional nonretentive faecal soiling or constipation.<sup>79</sup> It is made up of 37 pieces that are divided into 4 categories. The validity was only modest when compared to the Dutch Tacqol

generic instrument (TNO-AZL), despite the fact that the reliability ranged from satisfactory to good. A research conducted in the United States found that children who suffered from chronic constipation had a poorer quality of life (QOL) than population norms and children who suffered from inflammatory bowel disease (IBD) and gastroesophageal reflux disease (GERD).<sup>80</sup> There was a correlation between the duration of symptoms and a lower quality of life. An improvement in quality of life was observed in a research conducted in Australia on children diagnosed with constipation and treated with an antegrade continence enema.<sup>81</sup> The quality of life of children with celiac disease was shown to be comparable to that of a reference population in a research conducted in the Netherlands.<sup>82</sup>

Two studies have been conducted on the quality of life of children who have GERD. According to the findings of Thomson et al.<sup>83</sup>, children who received an endoluminal gastroplication for severe GERD had increased quality of life scores (using an adult measure) 6 weeks after surgery, and these improvements were maintained for the next 12 months. Another trial that used a disease-specific pediatric asthma instrument found that acid suppression did not enhance quality of life in children who had both asthma and GERD at the same time.<sup>84</sup>

It has been demonstrated that children who suffer from chronic intestinal pseudoobstruction have a more difficult time attending school and taking part in social activities than children who are not affected by the condition.<sup>85</sup> They suffer from significantly higher levels of pain, sadness, and anxiety than children who are healthy or children who have juvenile rheumatoid arthritis. Additionally, their parents needed more time to care for their children and had a worse emotional state than average. Among a study of quality of life among parents of children receiving home parenteral feeding, there was evidence of mental illness, which was associated with worsening in social life, family life, sex life, and work.<sup>86</sup>

## Discussion

Children and their families who are impacted by chronic illnesses might experience significant reductions in their quality of life. Eiser and Morse, in their review of quality-of-life measures in chronic diseases of childhood, emphasized the need for the implementation of quality of life measures in pediatric research, the adoption of child-centered approaches to measurement, and the clarification of the relationship between child and proxy ratings.<sup>9</sup> In addition, Eiser and Morse highlighted the need for the adoption of child-centered approaches to measurement. In this comprehensive analysis, we made an effort to locate all disease-specific assessments (Table 1) as well as previously published research on quality of life in children who suffer from gastrointestinal and liver conditions.

The early attempts to define quality of life concentrated on functional issues.<sup>65</sup> Subsequent research employed unvalidated questionnaires or generic tools. Many of the studies that have been uncovered have relied on parents as proxy in order to evaluate their child's quality of life. There are inherent issues with the use of proxy evaluations, despite the fact that this may be the only feasible option to measure quality of life in young children or in people who have learning disabilities. This is due to the fact that proxy assessments do not always correspond strongly with patients' own judgments of their quality of life.<sup>9</sup> For Hirschsprung disease/anorectal malformation<sup>67</sup>, liver transplantation<sup>57</sup>, cystic fibrosis<sup>41-46</sup>, inflammatory bowel disease<sup>17</sup> constipation, and functional nonretentive fecal soiling<sup>79</sup>, disease-specific instruments have now been devised and may be seen in Table 1. Some of them were produced in the early days of pediatric quality of life research; nevertheless, they have not been developed from the perspective of the child, and they have not been properly verified. Only the IMPACT questionnaire for IBD, the CFQ, the CFQOL, the DISABKIDS, and the Dutch Defecation Disorder List fulfill all of these criteria and can be recommended for use in the relevant patient groups. Using the three criteria listed in the methods (item generation from children, validity and reliability formally tested in children, questionnaire completed by child), we found that only these five questionnaires were able to meet all of these requirements. It is essential to keep in mind that if these instruments are to be used in a country other than the one in which they were developed, not only do they need to be translated according to the globally accepted standards established by Guillemin et al<sup>87</sup>, but they also might call for further validation in the nation in which they are intended to be used.

There were several studies that were found to have been conducted that focused on the quality of life of the parents or carers of children who suffered from chronic diseases. The quality of life of caregivers of children who received home parenteral nutrition<sup>86</sup>, as well as the quality of life of seriously challenged children who struggled to eat<sup>75-77</sup>, was significantly diminished. Despite the fact that gastrostomy feeding may be associated with an enhanced quality of life for both the kid and the caregiver, it may be challenging for caregivers to decide whether or not to give their child a gastrostomy. Oral feeding may also be fun and is essential for maintaining communication and connection with the patient. There is still a lot of work to be done in order to assess quality of life in children who have chronic gastrointestinal and hepatological illnesses; nevertheless, there has been some progress achieved in this area. Several diagnostic tools that are tailored to a particular disease have been created or are now in the process of being developed. Many of the identified studies made use of generic questionnaires and/or proxy evaluations; however, in recent years, researchers have begun to place a greater focus on the perspectives of children and their families. Measuring a patient's quality of life may assist in raising the level of awareness among medical practitioners regarding problems that are vital to the patient, such as the emotional and psychological effects of a disease and the treatment for that sickness. Patients who have the impression that their input is being considered may be more likely to follow the recommendations made by medical experts.

It is essential to place a strong focus on the creation of rigorous disease-specific instruments to assess quality of life, but it is also essential to investigate methods in which quality of life may be improved. When evaluating potential novel

therapies, quality of life should be one of the major outcomes examined. Recent research has begun to investigate the connection that exists between coping methods and quality of life (QOL). If we educate youngsters more effective coping skills, perhaps this will contribute to an increase in their quality of life. In addition to this, we need to think of therapies that are particularly geared toward enhancing the quality of life of children whose QOL is substantially diminished. Counseling, psychological treatment, support groups, play therapy, education, or social events such as adventure activities and camps may all fall under this category.

## References

- [1]. World Health Organization: Handbook of basic documents. Geneva: World Health Organization. 1949.
- [2]. Devanarayana NM, Mettananda S, Liyanarachchi C, Nanayakkara N, Mendis N, Perera N, Rajindrajith S: Abdominal pain-predominant functional gastrointestinal diseases in children and adolescents: prevalence, symptomatology, and association with emotional stress. *J Pediatr Gastroenterol Nutr.* 2011;53:659-665.
- [3]. Devanarayana, N.M., Rajindrajith, S. & Benninga, M.A. Quality of life and health care consultation in 13 to 18 year olds with abdominal pain predominant functional gastrointestinal diseases. *BMC Gastroenterol.* 2014;14:150.
- [4]. Demiral M, Başarır Özkan T, Özgür T, Özkan B, Erdemir Eren G, Altay D. The Assessment of the Quality of Life in Children with Chronic Liver Disease. *Turk J Gastroenterol.* 2021;32(9):774-781.
- [5]. Bozzini AB, Neder L, Silva CA, Porta G. Decreased health-related quality of life in children and adolescents with autoimmune hepatitis. *J Pediatr (Rio J)* 2019;95(1):87-93.
- [6]. Tehranian S, Jafari S, Yousofi J et al. Health-related quality of life (HRQOL) in children with chronic liver disease in North East Iran using PedsQL™ 4.0. *Electron Phys.* 2015;7(4):1214-1219.
- [7]. Kourkoutas E, Georgiadi M, Plexousakis S. Quality of life of children with chronic illnesses: A Review of the Literature. *Procedia Social and Behavioral Sciences.* 2010;2:4763-4767 .
- [8]. Loonen HJ, Derkx BH, Griffiths AM. Pediatricians overestimate importance of physical symptoms upon children's health concerns. *Med Care* 2002;40:996 – 1001.
- [9]. Eiser C, Morse R. Quality-of-life measures in chronic diseases of childhood. *Health Technol Assess* 2001;5:1-157.
- [10]. Patient-Reported Outcome and Quality of Life Instruments Data- base. Lyon, France: MAPI Institute, 2005. Available at: [http:// www.proqolid.org](http://www.proqolid.org).
- [11]. Lavigne JV, Faier-Routman J. Psychological adjustment to pediatric physical disorders: a meta-analytic review. *J Pediatr Psychol* 1992;17:133 – 57.
- [12]. Steinhausen HC, Kies H. Comparative studies of ulcerative colitis and Crohn's disease in children and adolescents. *J Child Psychol Psychiatry* 1982;23:33 – 42.
- [13]. Engstrom I. Mental health and psychological functioning in children and adolescents with inflammatory bowel disease: a comparison with children having other chronic illnesses and with healthy children. *J Child Psychol Psychiatry* 1992;33:563-82.
- [14]. Rabbett H, Elbadri A, Thwaites R, et al. Quality of life in children with Crohn's disease. *J Pediatr Gastroenterol Nutr* 1996;23:528- 33.
- [15]. Loonen HJ, Grootenhuis MA, Last BF, et al. Quality of life in paediatric inflammatory bowel disease measured by a generic and a disease-specific questionnaire. *Acta Paediatr* 2002;91:348-54.
- [16]. Griffiths AM, Nicholas D, Smith C, et al. Development of a quality- of-life index for pediatric inflammatory bowel disease: dealing with differences related to age and IBD type. *J Pediatr Gastroenterol Nutr* 1999;28:S46-52.
- [17]. Otley A, Smith C, Nicholas D, et al. The IMPACT questionnaire: a valid measure of health-related quality of life in pediatric inflammatory bowel disease. *J Pediatr Gastroenterol Nutr* 2002;35:557- 63.
- [18]. Loonen HJ, Grootenhuis MA, Last BF, et al. Measuring quality of life in children with inflammatory bowel disease: the IMPACT-II (NL). *Qual Life Res* 2002;11:47-56.
- [19]. Richardson G, Griffiths AM, Miller V, et al. Quality of life in inflammatory bowel disease: a cross-cultural comparison of English and Canadian children. *J Pediatr Gastroenterol Nutr* 2001;32:573 – 8.
- [20]. Maity S, Thomas AG. Evaluation of a computerised touch-screen questionnaire to measure quality of life in children with inflammatory bowel disease. *Qual Life Res* 2001;9:272.
- [21]. Afzal NA, Van Der Zaag-Loonen HJ, Arnaud-Battandier F, et al. Improvement in quality of life of children with acute Crohn's disease does not parallel mucosal healing after treatment with exclusive enteral nutrition. *Aliment Pharmacol Ther* 2004;20: 167 – 72.
- [22]. Shepanski MA, Hurd LB, Culton K, et al. Health-related quality of life improves in children and adolescents with inflammatory bowel disease after attending a camp sponsored by the Crohn's and Colitis Foundation of America. *Inflamm Bowel Dis* 2005;11:164-70.
- [23]. MacPhee M, Hoffenberg EJ, Feranchak A. Quality-of-life factors in adolescent inflammatory bowel disease. *Inflamm Bowel Dis* 1998;4: 6-11.
- [24]. Loonen HJ, Derkx BH, Koopman HM, et al. Are parents able to rate the symptoms and quality of life of their offspring with IBD? *Inflamm Bowel Dis* 2002;8:270 – 6.
- [25]. Van der Zaag-Loonen HJ, Grootenhuis MA, Last BF, et al. Coping strategies and quality of life of adolescents with inflammatory bowel disease. *Qual Life Res* 2004;13:1011 – 9.
- [26]. De Boer M, Grootenhuis M, Derkx B, et al. Health-related quality of life and psychosocial functioning of adolescents with inflammatory bowel disease. *Inflamm Bowel Dis* 2005;11:400 – 6.
- [27]. Mackner LM, Crandall WV. Long-term psychosocial outcomes reported by children and adolescents with inflammatory bowel disease. *Am J Gastroenterol* 2005;100:386 – 92.



- [28]. Ceriati E, Deganello F, De Peppo F, et al. Surgery for ulcerative colitis in pediatric patients: functional results of 10-year follow-up with straight endorectal pull-through. *Pediatr Surg Int* 2004;20: 573–8.
- [29]. Akobeng AK, Miller V, Firth D, et al. Quality of life of parents and siblings of children with inflammatory bowel disease. *J Pediatr Gastroenterol Nutr* 1999;28:S40 – 2.
- [30]. Day AS, Whitten KE, Bohane TD. Childhood inflammatory bowel disease: parental concerns and expectations. *World J Gastroenterol* 2005;11:1028 – 31.
- [31]. Epker JV, Maddrey AM. Quality of life in pediatric patients with cystic fibrosis. *Int J Rehab Health* 1998;4:215 – 22.
- [32]. Britto MT, Kotagal UR, Chenier T, et al. Differences between adolescents' and parents' reports of health-related quality of life in cystic fibrosis. *Pediatr Pulmonol* 2004;37:165 – 71.
- [33]. Sawyer MG, Reynolds KE, Couper JJ, et al. A two-year prospective study of the health-related quality of life of children with chronic illness—the parents' perspective. *Qual Life Res* 2005;14: 395 – 405.
- [34]. Powers PM, Gerstle R, Lapey A. Adolescents with cystic fibrosis: family reports of adolescent health-related quality of life and forced expiratory volume in one second. *Pediatrics* 2001;107:E70.
- [35]. Abbott J, Baumann U, Conway S, et al. Cross cultural differences in health related quality of life in adolescents with cystic fibrosis. *Disabil Rehab* 2001;23:837 – 44.
- [36]. Szyndler JE, Towns SJ, van Asperen PP, et al. Psychological and family functioning and quality of life in adolescents with cystic fibrosis. *J Cyst Fibros* 2005;4:135 – 44.
- [37]. Kaplan RM, Anderson JP. A general health policy model: update and applications. *Health Service Res* 1988;23:203 – 35.
- [38]. Czyzewski DI, Mariotto MJ, Bartholomew LK, et al. Measurement of quality of well being in a child and adolescent cystic fibrosis population. *Med Care* 1994;32:965 – 72.
- [39]. Landgraf JL, Abetz L, Ware JE. *The CHQ User's Manual*. Boston: The Health Institute, New England Medical Center; 1996.
- [40]. Kotwicki RJ, Condra L, Vermeulen L, et al. Assessing the quality of life in children with cystic fibrosis. *WMJ* 2001;100:50.
- [41]. Henry B, Aussage P, Grosskopf C, et al. Development of the Cystic Fibrosis Questionnaire (CFQ) for assessing quality of life in pediatric and adult patients. *Qual Life Res* 2003;12:63 – 76.
- [42]. Quittner AL, Sweeny S, Watrous M, et al. Translation and linguistic validation of a disease-specific quality of life measure for cystic fibrosis. *J Pediatr Psychol* 2000;25:403 – 14.
- [43]. Modi AC, Quittner AL. Validation of a disease-specific measure of health-related quality of life for children with cystic fibrosis. *J Pediatr Psychol* 2003;28:535 – 45.
- [44]. Wenninger K, Aussage P, Wahn U, et al. German Cystic Fibrosis Questionnaire study group. The revised German Cystic Fibrosis Questionnaire: validation of a disease-specific health-related quality of life instrument. *Qual Life Res* 2003;12:77 – 85.
- [45]. Klijn PH, van Stel HF, Quittner AL, et al. Validation of the Dutch cystic fibrosis questionnaire (CFQ) in adolescents and adults. *J Cyst Fibros* 2004;3:29 – 36.
- [46]. Gee L, Abbott J, Conway SP, et al. Development of a disease specific health related quality of life measure for adults and adolescents with cystic fibrosis. *Thorax* 2000;55:946 – 54.
- [47]. Baars RM, Atherton CI, Koopman HM, et al., DISABKIDS Group The European DISABKIDS project: development of seven condition-specific modules to measure health related quality of life in children and adolescents. *Health Qual Life Outcomes* 2005; 3:70.
- [48]. Petersen C, Schmidt S, Power M, et al., The DISABKIDS Group- Development and pilot-testing of a health-related quality of life chronic generic module for children and adolescents with chronic health conditions: a European perspective. *Qual Life Res* 2005;14:1065 – 77.
- [49]. Munzenberger PJ, Van Wagnen CA, Abdulhamid I, et al. Quality of life as a treatment outcome in patients with cystic fibrosis. *Pharmacotherapy* 1999;19:393 – 8.
- [50]. Staab D, Wenninger K, Gebert N, et al. Quality of life in patients with cystic fibrosis and their parents: what is important besides disease severity? *Thorax* 1998;53:727 – 31.
- [51]. Kosciak RL, Douglas JA, Zaremba K, et al. Quality of life of children with cystic fibrosis. *J Pediatr* 2005;147:S64 – 8.
- [52]. Boling W, Macrina DM, Clancy JP. The Caregiver Quality of Life Cystic Fibrosis (CQOLCF) scale: modification and validation of an instrument to measure quality of life in cystic fibrosis family caregivers. *Qual Life Res* 2003;12:1119 – 26.
- [53]. Boling W. The health of chronically ill children: lessons learned from assessing family caregiver quality of life. *Fam Community Health* 2005;28:176 – 83.
- [54]. Iorio R, Pensati P, Botta S, et al. Side effects of alpha-interferon therapy and impact on health-related quality of life in children with chronic viral hepatitis. *Pediatr Infect Dis J* 1997;16:984 – 90.
- [55]. Schwimmer JB, Middleton MS, Deutsch R, et al. A phase 2 clinical trial of metformin as a treatment for non-diabetic paediatric non-alcoholic steatohepatitis. *Aliment Pharmacol Ther* 2005;21:871 – 9.
- [56]. Chin SE, Shepherd RW, Cleghorn GJ, et al. Survival, growth and quality of life in children after orthotopic liver transplantation: a 5 year experience. *J Paediatr Child Health* 1991;27:380 – 5.
- [57]. Zamberlan KEW. Quality of life in school-age children following liver transplantation. *Mat Child Nurs J* 1992;20:167 – 229.

- [58]. Kita Y, Ishimaru Y, Sugimoto N, et al. Quality of life in children undergoing liver transplants overseas or in Japan. *Transplant Proc* 1996;28:2406 – 8.
- [59]. Midgley DE, Bradlee TA, Donohoe C, et al. Health-related quality of life in long-term survivors of pediatric liver transplantation. *Liver Transpl* 2000;6:333 – 9.
- [60]. Schulz K, Hofmann C, Sander K, et al. Comparison of quality of life and family stress in families of children with living-related liver transplants versus families of children who received a cadaveric liver. *Transplant Proc* 2001;33:1496 – 7.
- [61]. Alonso EM, Neighbors K, Mattson C, et al. Functional outcomes of pediatric liver transplantation. *J Pediatr Gastroenterol Nutr* 2003;37:155 – 60.
- [62]. Manificat S, Dazord A, Cochat P, et al. Quality of life of children and adolescents after kidney or liver transplantation: child, parents and caregiver's point of view. *Pediatr Transplant* 2003;7: 228 – 35.
- [63]. Bucuvalas JC, Britto M, Krug S, et al. Health-related quality of life in pediatric liver transplant recipients: a single-center study. *Liver Transpl* 2003;9:62 – 71.
- [64]. Cole CR, Bucuvalas JC, Hornung RW, et al. Impact of liver transplantation on HRQOL in children less than 5 years old. *Pediatr Transplant* 2004;8:222 – 7.
- [65]. Ditesheim JA, Templeton JM Jr. Short-term v long-term quality of life in children following repair of high imperforate anus. *J Pediatr Surg* 1987;22:581 – 7.
- [66]. Stolk EA, Busschbach JJ, Vogels T. Performance of the EuroQOL in children with imperforate anus. *Qual Life Res* 2000;9:29 – 38.
- [67]. Hanneman MJ, Sprangers MA, De Mik EL, et al. Quality of life in patients with anorectal malformation or Hirschsprung's disease: development of a disease-specific questionnaire. *Dis Colon Rectum* 2001;44:1650 – 60.
- [68]. Bai Y, Yuan Z, Wang W, et al. Quality of life for children with fecal incontinence after surgically corrected anorectal malformation. *J Pediatr Surg* 2000;35:462 – 4.
- [69]. Bai Y, Chen H, Hao J, et al. Long-term outcome and quality of life after the Swenson procedure for Hirschsprung's disease. *J Pediatr Surg* 2002;37:639–42.
- [70]. PoleyMJ,StolkEA,TibboelD,etal.Shorttermandlongtermhealth related quality of life after congenital anorectal malformations and congenital diaphragmatic hernia. *Arch Dis Child* 2004;89:836–41.
- [71]. Shamberger RC, Masek BJ, Leichtner AM, et al. Quality-of-life assessment after ileoanal pull-through for ulcerative colitis and familial adenomatous polyposis. *J Pediatr Surg* 1999;34:163–6.
- [72]. Stavlo PL, Libsch KD, Rodeberg DA, et al. Pediatric ileal pouch- anal anastomosis: functional outcomes and quality of life. *J Pediatr Surg* 2003;38:935–9.
- [73]. O'Neill JK, O'Neill PJ, Goth-Owens T, et al. Care-giver evaluation of anti-gastroesophageal reflux procedures in neurologically im- paired children: what is the real-life outcome? *J Pediatr Surg* 1996;31:375 – 80.
- [74]. Gatti C, di Abriola GF, Villa M, et al. Esophagogastric dissociation versus fundoplication: which is best for severely neurologically impaired children? *J Pediatr Surg* 2001;36:677–80.
- [75]. Tawfik R, Dickson A, Clarke M, et al. Caregivers' perceptions following gastrostomy in severely disabled children with feeding problems. *Dev Med Child Neurol* 1997;39:746–51.
- [76]. HeymanMB,HarmatzP,AcreemM,etal.Economicandpsychologic costs for maternal caregivers of gastrostomy-dependent children. *J Pediatr* 2004;145:511 – 6.
- [77]. Sullivan PB, Juszczak E, Bachlet AM, et al. Impact of gastrostomy tube feeding on the quality of life of carers of children with cerebral palsy. *Dev Med Child Neurol* 2004;46:796 – 800.
- [78]. Ludman L, Spitz L. Quality of life after gastric transposition for oesophageal atresia. *J Pediatr Surg* 2003;38:53 – 7.
- [79]. Voskuijl WP, van der Zaag-Loonen HJ, Ketel IJ, et al. Health related quality of life in disorders of defecation: the Defecation Disorder List. *Arch Dis Child* 2004;89:1124 – 7.
- [80]. Youssef NN, Langseder AL, Verga BJ, et al. Chronic child- hood constipation is associated with impaired quality of life: a case-controlled study. *J Pediatr Gastroenterol Nutr* 2005;41: 56–60.
- [81]. King SK, Sutcliffe JR, Southwell BR, et al. The antegrade con- tinence enema successfully treats idiopathic slow-transit constipa- tion. *J Pediatr Surg* 2005;40:1935–40.
- [82]. Kolsteren MM, Koopman HM, Schalekamp G, et al. Health-related quality of life in children with celiac disease. *J Pediatr* 2001;138: 593.
- [83]. Thomson M, Fritscher-Ravens A, Hall S, et al. Endoluminal gastroplication in children with significant gastro-oesophageal reflux disease. *Gut* 2004;53:1745–50.
- [84]. Stordal K, Johannesdottir GB, Bentsen BS, et al. Acid suppression does not change respiratory symptoms in children with asthma and gastro-oesophageal reflux disease. *Arch Dis Child* 2005;90: 956 – 960.
- [85]. Schwankovsky L, Mousa H, Rowhani A, et al. Quality of life outcomes in congenital chronic intestinal pseudo-obstruction. *Dig Dis Sci* 2002;47:1965–8.
- [86]. Wong C, Akobeng AK, Miller V, et al. Quality of life of parents of children on home parenteral nutrition. *Gut* 2000;46:294 – 5.
- [87]. Guillemin F, Bombardier C, Beaton D. Cross-cultural adaptation of health-related quality of life measures: literature review and proposed guidelines. *J Clin Epidemiol* 1993;46:1417 – 32.