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Health-related quality of life experiences among children and adolescents born with esophageal atresia: Development of a condition-specific questionnaire for pediatric patients

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ABSTRACT

Background/Purpose: The aims were to present the framework for the development a condition-specific health-related quality-of-life (HRQOL) questionnaire for children with esophageal atresia (EA) and to describe HRQOL experiences reported by children and by their parents.
Methods: Utilizing the well-established DISABKIDS methodology, standardized focus group discussions were held and transcribed. HRQOL experiences were identified, content analyzed and evaluated using descriptive statistics.
Results: 30 families (18 children 8–17 years, 32 parents of children 2–17 years) participated in ten focus group discussions. 1371 HRQOL experiences were identified referencing social, emotional and physical aspects of eating and drinking ($n = 368$), relationships with other people ($n = 283$), general life issues; physical activity, sleep and general health ($n = 202$), communicative/interactive processes of one's health condition ($n = 161$), body issues ($n = 109$), bothersome symptoms ($n = 81$), impact of health care use/medical treatment ($n = 78$), confidence in oneself and in the future ($n = 65$) and difficulties because of concomitant anomalies ($n = 24$). A basis of two age-related HRQOL questionnaires for children with EA (2–7 years, 8–17 years) was subsequently constructed.
Conclusions: EA interacts with various aspects of the child's life. In addition to HRQOL issues of eating and drinking, social dimensions like relationships and interactions with other people seem to be prominent condition-specific HRQOL parameters. The settings for the development of the first condition-specific HRQOL questionnaires for patients with EA are established.

Survival rates among children with esophageal atresia (EA) have improved to 95% [1]. However, chronic morbidity among children is common, ranging from dysphagia, gastroesophageal reflux and respiratory disorders [2,3] as well as growth retardation [4]. Children with EA may suffer from food impaction, oral aversion and vomiting [4] as well as from wheezing, dyspnea, barky or chronic cough and recurrent airway infections with subsequent need of medical care [3,5]. Moreover, additional morbidity may result from associated anomalies [6]. Although criteria to evaluate medical outcomes have become increasingly supplemented by patient-reported outcome standards [7], studies of health related quality of life (HRQOL) among children with EA are few ($n = 5$) [8–12]. Only two studies [8,9] have compared the overall HRQOL to healthy references, of which one demonstrated reduced overall HRQOL in pediatric patients with EA. Generic HRQOL measurements permit the advantage of comparison with healthy reference norms, condition-specific HRQOL questionnaires provide more sensitive information with regard to specific clinical characteristics [13]. Since no study using a condition-specific HRQOL questionnaire among patients with EA has been reported [14], knowledge of this field remains limited. The long-term aim is to advance knowledge of HRQOL among children with EA through the development of a condition-specific HRQOL questionnaire, in this study to describe the establishment of the conditions for such a questionnaire and the HRQOL experiences reported by children with EA and by their parents.

1. Materials and methods

Approval to conduct the study was obtained from the Regional Research Ethics Committee.

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1.1. Framework and assessment of condition-specific HRQOL experiences

This study was guided by the well-established methodology of the large European DISABKIDS project for children with chronic health conditions [15]. The methodology was preferred since the design comprises a distinction between generic, chronic-generic and condition-specific HRQOL measurements. Within the project, standardized pediatric condition-specific HRQOL measurements were developed. Moreover, a patient-derived nature of questionnaire construction is central [16] and in compliance with the Food and Drug Administration guidelines for construction of an HRQOL instrument [17]. The assessment of condition-specific HRQOL experiences is based on focus groups with children and their parents, composed according to the child's severity of disease, age and gender. The reported HRQOL experiences form the construction of questions for a preliminary questionnaire. This questionnaire undergoes a question reduction process and is evaluated through cognitive debriefing, a pilot test and ultimately a field test in a larger sample of the target population [15].

1.2. Focus group procedure

1.2.1. Selection of participants

Criteria were developed for two levels of severity of EA (mild to moderate and severe) including input from pediatric surgical expertise, QOL methodology expertise and from previous research of EA [5,12,18–22]. Medical records of 135 children born with EA in 1997–2013 were reviewed for clinical data. 73 children (54.0%) were categorized as cases of severe EA. Stratified for child gender, families of each five children with mild to moderate EA and five children with severe EA were selected in three age groups (0–7, 8–12, 13–17 years) and were invited to participate in the study. Children younger than eight years were represented by their parents (proxy-report). Patients were considered to have severe EA if one or more of four criteria were met (Table 1). Associated malformations were defined as severe if associated with patient disability [23].

1.2.2. Data collection

Informed consent to participate in the study was obtained. Separate focus group discussions with the children and their parents took place in 2014 and were digitally recorded. A questionnaire of child and family characteristics was completed. The DISABKIDS focus group manual, consisting of eight questions with a progressive focus on the main issues of HRQOL, formed a standard basis for discussions with both children and parents [15]. All participants were asked questions about the nature and extent to which the EA condition has continued to affect the child's daily life. Discussions were led and facilitated by a moderator who ensured that all participants had an opportunity to contribute. A research assistant was present during the child focus groups and was responsible for taking field notes of non-verbal communication and group interactions.

1.2.3. Data analysis

Focus group discussions were transcribed verbatim. HRQOL experiences were content analyzed as illustrated in Fig. 1. HRQOL experiences were extracted from the focus group text, merged with participant information into Excel 2010, formulated as a statement and card sorted into domains and overall HRQOL areas with a particular HRQOL content in common [15,16]. Descriptive statistical analysis of the clinical and sociodemographic data, the frequency and distribution of HRQOL statements according to domains, overall HRQOL areas, severity of EA, child gender, age group (0–7, 8–12, 13–17 years), child and proxy reports was performed using SPSS 22.0. The results were used to derive questions for the construction of a HRQOL questionnaire basis (adjusted for four weeks recall period and a five-point Likert scale from never to always). An unbiased categorization of HRQOL statements, as well as selection and reformulation into questions were ensured through consensus among several researchers.

Table 1
Inclusion criteria of patients in the severe esophageal group ($n = 15$).

	Number of patients (%)	Single inclusion criteria (%)
The primary anastomosis was delayed and/or EA replacement was accomplished	7 (47)[a]	3 (20)
Major surgical revision (open surgery) of the EA correction performed for causes as recurrent TEF or anastomotic leakage	5 (33)	1 (7)
Presence of a severe tracheomalacia or tracheobronchomalacia based on macroscopic estimation of an anteroposterior collapse documented as excessive, severe and/or of ≥75% without limitation of the child's age at the bronchoscopy [21][b]. If the child had been examined several times, the most recent bronchoscopy was considered the most valid for inclusion/exclusion	5 (33)	4 (27)
Presence of at least one other congenital health condition resulting in disability. The term disability was defined according to the ICF-CY. Disability is served as an umbrella term for impairments (problems in body function or structure as a significant deviation or loss), activity limitations or participation restrictions [23][b]	4 (27)[c]	3 (20)

EA, esophageal atresia; TEF, tracheoesophageal fistula; ICF-CY; International Classification of Functioning, Disability and Health-Child & Youth Version.
[a] Esophageal replacement was performed in three children.
[b] Reference number according to manuscript.
[c] The following associated malformations were considered as severe; anorectal malformation (two patients), severe urogenital malformations (one patient), central nervous system anomaly resulting in neurogenic bladder dysfunction (one patient), and congenital hypothyreos (one patient).

2. Results

2.1. Patient and parent characteristics

Ten focus group discussions were held (19.2 hours, mean 1.9 hours). All 30 families participated (100%) and were represented by 18 children (8–17 years) and 32 parents of children (2–17 years). As illustrated in Table 2, child gender and age were similar among patients with mild to moderate and severe EA. Morbidity and health care needs were present among children independent of severity of EA, but especially among patients with severe EA. In a majority of families, proxy-reports were provided by the maternal parent.

2.2. Condition-specific HRQOL experiences and the construction of a questionnaire basis for children with EA

1371 HRQOL statements/experiences were identified. A majority (66.9%) was generated from children with severe EA and their parents. Of the aggregate HRQOL experiences, 716 (52.2%) were child reports, and with respect to gender distribution, 730 (53.2%) and 641 (46.8%) were for males and females, respectively.

2.2.1. Characteristics and distribution of HRQOL domains

The HRQOL statements were categorized into 30 domains referencing emotional, social and physical perspectives of HRQOL experiences. Table 3 provides a description of each domain in descending order of statement frequency (116 to three). In Supplemental Material 1, each domain description is accompanied by a representative focus group quote as expressed by children and their parents.

2.2.2. Characteristics and distribution of overall HRQOL areas/domains

The allocation of 30 domains into nine overall HRQOL areas and their distribution according to severity of EA, child gender and child age