

Feasibility, Reliability, and Validity of the Turkish Version of the Esophageal-Atresia-Quality-of-Life Questionnaires to Assess Condition-Specific Quality of Life in Children and Adolescents Born with Esophageal Atresia

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ABSTRACT

Background: This study reports the feasibility, validity, and reliability of the Turkish versions of the Esophageal-Atresia-Quality-of-Life (EA-QOL) questionnaires, which were originally developed in Sweden and Germany.

Methods: After translation from Swedish to Turkish and cognitive debriefings, 51 families of children aged 2-7 years (parent-reported, 17 items) and 54 families of children 8-17 years (child-reported and parent-reported, 24 items) responded to the EA-QOL questionnaires and a validated generic HRQOL instrument (PedsQL4.0). The medical records of the patients and the questionnaires were used to obtain clinical data. The Turkish version of the EA-QOL questionnaires were evaluated for feasibility (<5% missing item responses), reliability (internal consistency/retest reliability for 3 weeks), and validity (known groups/concurrent/convergent). The level of significance was $P < .05$.

Results: The feasibility of the Turkish version of the EA-QOL questionnaires was good. The internal consistency of all scales was satisfactory, as were the levels of agreement of EA-QOL scores between the field study and the retest study. Known-group validity and concurrent validity were achieved, since the EA-QOL questionnaires showed that esophageal symptoms and feeding difficulties were negatively associated with EA-QOL total scores, both in the age-specific versions (child-reported and parent-reported), and with respect to respiratory symptoms in the version for EA children 2-7 years (parent-reported). A higher number of respiratory symptoms decreased the EA-QOL total scores in both age groups (parent-reported, $P < .05$). Correlations between the EA-QOL total scores and PedsQL-4.0 total scores supported convergent validity.

Conclusion: The Turkish version of the EA-QOL questionnaires is feasible, valid, and reliable to assess condition-specific HRQOL in EA children.

Keywords: Esophageal atresia, health-related quality of life, dysphagia

INTRODUCTION

Esophageal atresia (EA) is the most common congenital anomaly of the esophagus, and has life-long consequences for feeding and respiratory functions. Dysphagia and gastroesophageal reflux disease (GERD) are the most frequent digestive symptoms, and may be associated with respiratory problems such as cough, wheezing,

and airway infections.^{1,2} As the survival rates of patients with EA exceed 90%, their long-term complications and health-related quality of life (HRQOL) have become the primary focal points of interest.¹

HRQOL refers to the patients' perception of the impact of disease and its treatment on physical, psychological,

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and social functioning and well-being. The HRQOL can be assessed using generic instruments, which enable application to broad populations and the comparison of results between patients and healthy references; and condition-specific instruments, which are more sensitive to clinical findings and issues relevant to the patients.³ According to the international standards, the input from pediatric patients and their parents for the development of a HRQOL questionnaire is paramount.⁴ Moreover, the accepted methods to assess the validity and reliability of such instruments are well defined.⁵

According to 2 systematic literature reviews covering articles from inception to December 28, 2019, HRQOL assessments of varying quality are used in pediatric patients with EA.^{6,7} Moreover, different generic assessments have revealed comparable overall HRQOL with healthy/general references,^{8,9} as well as worse overall HRQOL.^{10,11,12} The understanding of HRQOL in EA would benefit from a standardized tool which captures important content from these patients and enables international evaluations of treatment strategies and the long-term outcomes.^{6,7}

Recently, condition-specific HRQOL instruments for pediatric patients with EA were developed according to the current standards (the EA-QOL questionnaires), demonstrating good feasibility, reliability, and validity for pediatric patients with EA in Sweden and Germany.^{13,14} A child's perception of disease and treatment, as well as the understanding of its position in life, may be influenced by cultural norms. The extent to which the HRQOL concept is transferable from one cultural or country-specific context to another, is still unclear. Therefore, a condition-specific HRQOL-instrument needs evidence that the feasibility, validity, and reliability are adequately similar

between all language versions used in future studies.⁴ We aimed to report the feasibility, reliability, and validity of the Turkish version of the EA-QOL questionnaires in EA children and adolescents, and hypothesized that these measurement properties of the translated Turkish EA-QOL questionnaires would be adequate.

METHODS

The study procedure followed the standardized guidelines of translation, cognitive debriefing, and psychometric evaluation, complying with the international standards of patient-reported outcomes.^{5,14,15} The steps in translation and cognitive debriefing are overviewed in Figure 1.

Participants and Settings

The Ethical Committee (GO/2019/19-39) approved the study. From 2019 to 2020, families of children aged 2-17 years were recruited from 2 centers ($n = 105$). Informed consent was obtained from all patients and families prior to study inclusion. Three children were excluded due to cognitive impairments.

Patient and Clinical Data

The medical records of the patients were reviewed for birth features and surgical interventions. The sociodemographic data of the children and parents, as well as the children's feeding, digestive, and respiratory problems of the previous 4 weeks were collected from the parents' reports.

HRQOL Measures

Parents and children ≥ 8 years old responded to the EA-QOL questionnaires during hospital outpatient clinic visits, in the presence of doctors. To evaluate the test-retest reliability of EA-QOL questionnaires, the respondents replied to the questionnaires again under the same conditions, within 3 weeks after the first responses.

The EA-QOL questionnaires consist of a parent-reported 17-item questionnaire for children aged 2-7 years (scales: Eating, Physical health and treatment, Social isolation, and Stress) and a 24-item questionnaire for children aged 8-17 years, available as child-reported and parent-reported versions (scales: Eating, Social relationships, Body perception, and Health and well-being). The questions were answered based on a 4-week recall period and a 5-point Likert scale, with answers from never (1) to always (5).

The psychometric evaluation of the EA-QOL questionnaires was performed with a validated generic HRQOL

MAIN POINTS

- The Turkish versions of the Esophageal Atresia-Quality of Life (EA-QOL) questionnaires are feasible, valid, and reliable to assess condition-specific health-related quality of life in children with esophageal atresia (EA).
- The Turkish version of the EA-QOL questionnaire for children aged 8-17 years showed that children with a prior gastrostomy insertion had lower total scores.
- The Turkish version of the EA-QOL questionnaires for children 2-7 (parent-report) and 8-17 years old (child- and parent-report) reveal good discriminative ability with regard to digestive morbidity, reflecting that esophageal symptoms and feeding difficulties in EA children are associated with reduced EA-QOL total scores.

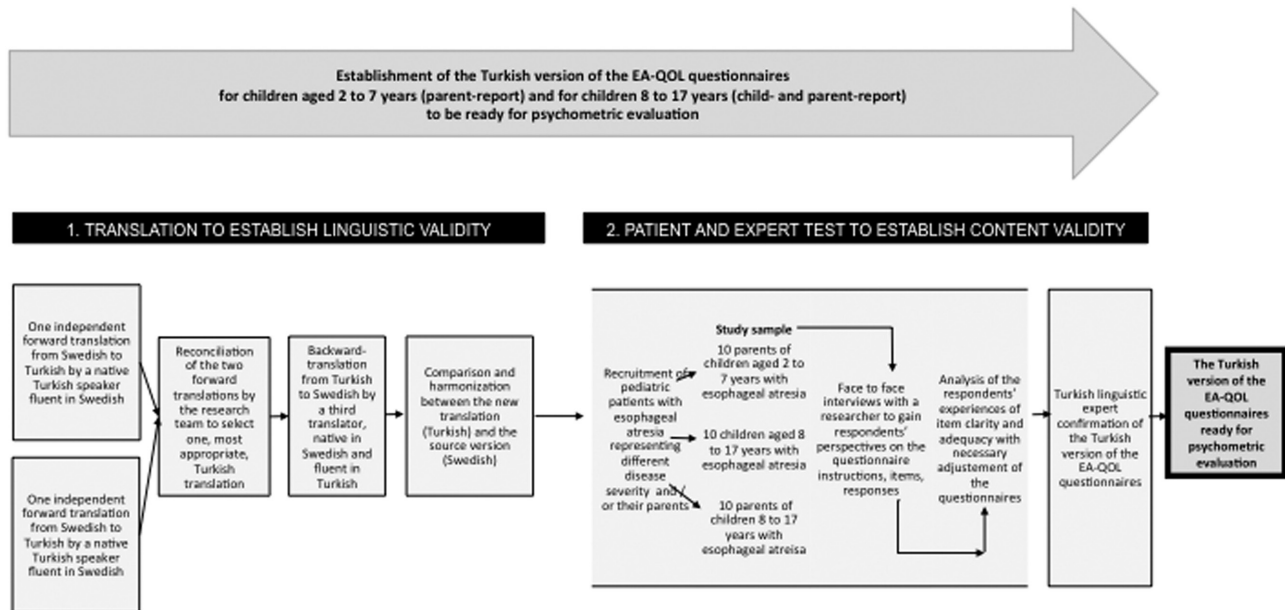


Figure 1. The steps in translation and cognitive debriefing to establish the Turkish version of the EA-QOL questionnaires, ready for psychometric evaluation.

questionnaire, and the Turkish version of the Pediatric Quality of Life Inventory 4.0 generic core scales (PedsQL 4.0) for children 2 to 18 years old.^{16,17}

Statistical Analysis

Data were analyzed using IBM SPSS Statistics for Windows (Version 23.0, Armonk, NY, USA: IBM Corp). The analysis of the Turkish version of the EA-QOL questionnaires complied with the Swedish-German evaluation.¹³ The item responses of the EA-QOL-questionnaires and PedsQL 4.0 were linearly transformed to a scale of 0-100, with higher scores reflecting better HRQOL. The level of significance in all tests was $P < .05$.

Descriptive Statistics: Median, min-max values, frequency, and percentages were given as descriptive statistics.

Feasibility: The feasibility of the EA-QOL questionnaires was determined from the low values of missing item responses (<5%), to indicate the respondent's ability to provide good-quality data regarding HRQOL.

Reliability: *Internal consistency* describes the extent to which multiple items within a scale are correlated, thus measuring the same concept. In this setting, it was evaluated using Cronbach's alpha coefficient within each of the EA-QOL questionnaire scales. An acceptable value of the Cronbach's alpha coefficient is 0.70 or above

(maximum 1.0), and this was used to determine the inclusion/exclusion from a scale. In other words, an item was deleted from the scale if we obtained an acceptable value for Cronbach's alpha by removing it.

Test-retest reliability refers to the stability of the instrument's scores over time. The stability of the EA-QOL questionnaires was evaluated as the level of agreement between the scale scores measured on 2 occasions with an interval of 3 weeks, using the intra-class correlation coefficient (ICC) and expecting moderate (0.5-0.75), good (0.76-0.9), and excellent (>0.90) levels of ICCs.

Group-comparison reliability was noted if the level of agreement between the child and the parent reports in the EA-QOL total scores were moderate to excellent, using ICCs as defined above.

Validity: Validity refers to the degree to which the instrument measures what it purports to measure.

Known-group validity evaluates the HRQOL scores for groups that would hypothetically differ. We analyzed the known-group validity by comparing EA-QOL total scores between EA children with and without esophageal, feeding, and respiratory symptoms respectively, using the Mann-Whitney *U*-test (in groups with at least 5 cases). We also compared the EA-QOL total scores between

surgical groups of EA children. The magnitude of the score differences between the groups was calculated using effect sizes (ESs) and Cohen's d classification (moderate (>0.5) and large (>0.8)), for a standardized interpretation of ESs.

Clinical concurrent validity refers to evidence that shows the extent to which the EA-QOL total scores are related to a substantial symptom burden in the patient. We expected that a higher number of different digestive symptoms, feeding difficulties, or respiratory symptoms in children with EA would decrease the EA-QOL total scores, using univariable linear regression analysis.

Convergent validity was confirmed if the EA-QOL total scores were related to the total scores of another questionnaire measuring a similar concept. It was examined using the Spearman's rho (r_s) correlation coefficient between the total scores of the EA-QOL questionnaire and of those of the PedsQL 4.0, expecting a moderate (0.40-0.59) to strong (0.60-0.79) correlation.

RESULTS

Overall, 105 families responded to the EA-QOL questionnaires; 51 parents of children aged 2-7 years, 54 parents of children aged to 8-17 years, and 47 children aged 8-17 years. The study sample is presented in Table 1.

Feasibility

The *Item feasibility* of the Turkish version of the EA-QOL questionnaires was good. All 105 parent-reported questions were answered, with no missing responses. Only 4 self-reports from children aged 8-17 years had missing items $>5\%$, out of all the HRQOL scales.

Descriptive Statistics and Reliability

The descriptive statistics, internal consistency, and test-retest reliability of the Turkish version of the EA-QOL questionnaires are shown in Table 2.

Internal Consistency

All scales in the parent-reported EA-QOL questionnaires showed acceptable levels of Cronbach's alpha (≥ 0.70 , Table 2). In the self-reported EA-QOL questionnaire for children aged 8-17 years, 1 item of the Eating scale (Can you eat at the same pace as other children your age?) and 2 of the Health and Well-being scale (Are you bothered by breathing difficulties if you exercise and play? Do you have trouble sleeping at night because of your

health condition, for example, due to acid reflux, heart-burn, or respiratory problems?) were omitted from their scales, because Cronbach's alpha was then improved and reached >0.7 .

Test-Retest Reliability

The test-retest reliabilities had mainly excellent results. Only the Health and Well-being scale (parent-reported) and the Eating scale (child-reported) showed moderate or good test-retest reliability respectively (Table 2).

Group-Comparison Reliability

In the EA-QOL questionnaire for children aged 8-17 years, the group-comparison reliability showed acceptable levels of child-parent agreements on the Eating (ICC, 0.86), Social relationships (ICC, 0.90), Body perception (ICC, 0.80), and Health and well-being (ICC, 0.64) scales, and the total scales (ICC, 0.93).

Validity

Known-Group Validity: The EA-QOL questionnaire for children 2-7 years old (parent-reported) and children 8-17 years old (child-reported and parent-reported) showed that the total scores were significantly lower in the hypothesized clinical groups of EA children, as detailed in Table 3.

In parents' reports of EA children aged 2-7 years, the child's problems with food impaction, dysphagia, painful swallowing, and vomiting were significantly associated with lower EA-QOL total scores. This was also the case for most of the investigated feeding difficulties and all the respiratory symptoms of coughing, wheezing, dyspnea, chest tightness, and respiratory infections ($P < .05$, moderate to large ESs).

In the EA-QOL questionnaire for children aged 8-17 years (child-reported), patients with prior gastrostomy insertion had significantly lower EA-QOL total scores. In both the child and parent reports, food impaction, dysphagia, the need to avoid certain foods, and eating small portions were significantly associated with lower EA-QOL total scores. In the parents' reports of children aged 8-17 years, the child's need to have >30 minutes to finish large meals; while in the children's reports, the child's need to increase fluid intake during meals, were both associated with lower EA-QOL total scores. Also, EA children aged 8-17 years with dyspnea or with chest tightness reported lower EA-QOL total scores, but only in the parents' reports ($P < .05$, moderate to large ESs).

Table 1. Data of the Families Which Responded to the Turkish Version of the EA-QOL Questionnaires

Demographics, n (%)	Children 2-7 Years Old (n = 51)	Children 8-17 Years Old (Child Report, n = 47)	Children 8-17 Years Old (Parent-Report, n = 54)
Child information			
Male	31 (60.8)	24 (51.1)	25 (50.0)
Gestational age (week, median: min/max)	37 (29-40)	37 (28-42)	37 (28-42)
Birth weight (g, median: min/max)	2400 (1260-3700)	2430, (1200-3900)	2700 (1200-3900)
Primary esophageal repair	38 (74.5)	43 (91.5)	46 (92.0)
Gross types,* n (%)			
A	19 (37.3)	8 (17.0)	9 (16.7)
B	2 (3.9)	1 (2.1)	1 (1.9)
C	30 (58.8)	37 (78.7)	42 (77.8)
D	0 (0)	1 (2.1)	2 (3.7)
Revisional surgery	5 (9.8)	1 (2.1)	2 (4.0)
Associated anomalies			
Cardiovascular	23 (45.1)	13 (27.7)	13 (26)
Anorectal	5 (9.8)	3 (6.4)	3 (6.0)
Uro-genital	8 (15.7)	4 (8.5)	4 (8.0)
Limb	7 (13.7)	2 (4.3)	2 (4.0)
Vertebral-skeletal	8 (15.7)	6 (12.8)	6 (12.0)
VACTER	13 (25.0)	5 (10.6)	5 (10.0)
Chormosomal anomaly	5 (9.8)	0 (0.0)	0 (0.0)
Postoperative complications			
Fistula recurrence	4 (7.8)	3 (6.4)	4 (8.0)
Anastomotic leak	6 (11.8)	3 (6.4)	4 (8.0)
Esophageal strictures**/dilatation	28 (54.9)	34 (72.3)	36 (72.0)
Parent information			
Mother	39 (76.5)	36 (66.7)	36 (66.7)
Parental age (years) (median: min/max)	33 (23-43)	39 (26-50)	39 (26-50)
Cohabitant partner	45 (88.2)	53 (98.1)	53 (98.1)
University education	9 (17.7)	21 (35.9)	21 (35.9)

*Gross types of esophageal atresia (EA): Type A, EA with no tracheoesophageal fistula (TEF); Type B, EA with proximal TEF; Type C, EA with distal TEF; Type D, EA with both proximal and distal TEF.

**The diagnosis of esophageal stricture was based on clinical findings, upper GI studies, and endoscopic interventions. Patients with esophageal strictures required at least 1 esophageal dilatation.

Clinical Concurrent Validity: As shown in Table 4, an increased number of digestive symptoms in EA children aged 8-17 years significantly decreased the EA-QOL total scores in both child and parent reports ($P < .001$), supporting clinical concurrent validity. Moreover, a higher number of different feeding difficulties in the child lowered the EA-QOL total scores in EA children aged 2-7 years (parent-reported) and 8-17 years (child-reported and

parent-reported) ($P \leq .001$). In parent reports, a higher number of different respiratory symptoms in the EA child significantly decreased the EA-QOL total scores of both the age-specific EA-QOL questionnaires ($P < .05$).

Convergent Validity: The EA-QOL total scores for children aged 2-7 years (parent-reported, $n = 51$, $r_s = 0.50$) and 8-17 years showed a moderate correlation with those

Table 2. Descriptive statistics, Internal Consistency, and Retest Reliability of the Turkish Version of the EA-QOL-Questionnaires

EA-QOL-Questionnaire Scores	Descriptive Statistics			Internal Consistency		Retest Reliability	
	Number of Items	Number of Respondents	Mean (SD)	Cronbach's Alpha	Number of Respondents	Level of Agreement, ICC	95% CI
Children 2-7 years old (parent-reported)							
Eating	7	51	65.7 (21.0)	0.72	51	0.88	0.80-0.93
Physical health and treatment	6	51	65.0 (21.8)	0.70	51	0.90	0.83-0.94
Social isolation and stress	4	51	64.8 (28.0)	0.75	51	0.90	0.84-0.94
Total scores	17	51	65.5 (18.0)	0.82	51	0.91	0.85-0.95
Children 8-17 years old (child-reported)							
Eating	7	47	83.0 (15.8)	0.78	47	0.98	0.96-0.99
Social relationships	7	47	88.8 (17.0)	0.82	47	0.99	0.99-1.00
Body perception	5	47	90.3 (15.7)	0.74	47	0.97	0.94-0.98
Health and well-being	2	47	88.8 (20.4)	0.74	47	0.96	0.923-0.98
Total scores	21	47	87.2 (13.3)	0.89	47	0.99	0.98-1.00
Children 8-17 years old (parent-reported)							
Eating	8	54	76.0 (17.5)	0.70	54	0.97	0.96-0.98
Social relationships	7	54	84.9 (20.0)	0.87	54	0.99	0.99-1.00
Body perception	5	54	88.3 (19.0)	0.85	54	0.99	0.98-0.99
Health and well-being	4	54	84.0 (18.4)	0.71	54	0.66	0.48-0.79
Total scores	24	54	82.5 (14.9)	0.88	54	0.97	0.95-0.98

Cronbach's alpha coefficient was acceptable if the value was 0.70 or above

ICC ≥ 0.9 was considered excellent reliability, $0.75 \leq \text{ICC} < 0.9$ good, and $0.5 \leq \text{ICC} < 0.75$ moderate.

EA, esophageal atresia; ICC, intra-class correlation coefficient; QOL, Quality of Life.

on the PedsQL 4.0 (child-reported, $n = 47$, $r_s = 0.42$; parent-reported, $n = 54$, $r_s = 0.42$), demonstrating convergent validity of the Turkish versions of the EA-QOL questionnaires.

DISCUSSION

The need for targeted HRQOL measures has been recognized in esophageal diseases^{18,19} and in pediatric conditions.³ The EA-QOL questionnaires for pediatric patients with EA were developed and tested in the 2 Northern European countries, Sweden and Germany.^{13,14} The rarity of EA, and the need to evaluate surgical treatments in complex cases and to understand long-term patient needs, emphasize the necessity of a standardized HRQOL measurement for patients with EA for cross-cultural use. Since HRQOL can be influenced by cultural contexts, it is necessary to evaluate the EA-QOL questionnaires further, according to the standards.⁴ This study showed that overall, the Turkish version of the EA-QOL questionnaires is feasible, reliable, and valid in children and adolescents with EA in Turkey.

The item feasibility of the Turkish version of the EA-QOL questionnaires showed good results, proposing that our Turkish sample of pediatric patients with EA and their parents were willing and able to provide good-quality data regarding the children's EA-QOL. As previously suggested, their young age may explain missing item responses in the child-reported version,¹³ which is similar to our findings.

The Turkish version of the EA-QOL questionnaires showed satisfactory internal consistency and retest reliability, similar to the Swedish-German results.^{13,14} However, 3 items were omitted from the child-reported EA-QOL questionnaire for 8- to 17-year-old patients, to increase the reliability. This was possibly needed because some children did not recognize the item content. However, these items also showed linguistic and content validity and can provide important narrative HRQOL information of the EA child. Statistics with more psychometric criteria, in a cross-cultural and larger study sample, are suggested to determine the final item exclusion.¹³ The retests were done 3 weeks after the first test, to consider whether the

Table 3. Comparison of Total scores on the EA-QOL Questionnaire for Children 2-7 and 8-17 Years Old Between Clinical Subgroups

	Children 2-7 Years Old (Parent-Reported)						Children 8-17 Years Old (Child-Reported)						Children 8-17 Years Old (Parent-Report)					
	Yes			No			Yes			No			Yes			No		
	N	Mean (SD)	P	Mean (SD)	N	ES [¶]	Mean (SD)	N	ES [¶]	Mean (SD)	N	ES [¶]	Mean (SD)	N	ES [¶]	Mean (SD)	N	ES [¶]
Surgical characteristics																		
Primary repair	38	65.4 (19.0)	13	66.1 (15.3)	.880	-0.039	43	87.1 (13.6)	4	88.1 (11.3)	NP	-0.072	49	82.6 (14.5)	5	81.0 (20.3)	NP	0.106
Prior Gastrostomy	25	64.7 (18.7)	26	66.3 (17.6)	.821	-0.090	15	78.5 (17.7)	32	91.3 (8.2)	.006	-1.068	18	76.0 (17.1)	36	85.7 (12.7)	.057	-0.682
Recurrent fistula	4	61.8 (23.8)	47	65.9 (17.7)	NP	-0.226	3	89.7 (5.9)	44	87.0 (13.7)	NP	0.197	4	84.1 (14.5)	50	82.4 (15.1)	NP	0.117
Anastomotic leaks	6	66.2 (19.3)	45	65.5 (18.0)	.943	0.040	3	80.6 (7.9)	44	87.7 (13.5)	NP	-0.533	4	71.9 (13.9)	50	83.3 (14.8)	NP	-0.778
Dilatation of esophagus	28	65.4 (20.1)	23	65.7 (15.4)	.712	-0.012	34	85.7 (14.1)	13	91.0 (10.9)	.146	-0.399	38	81.2 (15.6)	16	85.5 (13.2)	.343	-0.292
Digestive symptoms^a																		
Heart burn [*]	8	56.8 (22.6)	43	67.2 (16.8)	.153	-0.584	7	77.6 (23.2)	40	88.9 (10.3)	.178	-0.886	8	72.8 (19.3)	46	84.2 (13.6)	.096	-0.787
Vomiting ^{**}	21	58.3 (17.2)	30	70.6 (17.0)	.006	-0.717	4	66.4 (25.0)	43	89.1 (10.2)	NP	-1.933	5	54.6 (15.8)	49	85.3 (11.6)	NP	-2.564
Dysphagia ^{***}	22	56.6 (19.8)	29	72.4 (13.1)	.004	-0.969	13	74.3 (15.8)	34	92.2 (8.1)	<.001	-1.675	15	68.0 (14.3)	39	88.1 (10.9)	<.001	-1.683
Food impaction	27	60.1 (19.8)	24	71.7 (13.6)	.028	-0.676	13	74.5 (16.0)	34	92.1 (8.2)	<.001	-1.619	16	68.9 (14.6)	38	88.2 (10.9)	<.001	-1.604
Painful swallowing	8	47.4 (20.5)	43	68.9 (15.5)	.003	-1.316	4	62.5 (19.7)	43	89.5 (10.1)	NP	-2.448	5	57.3 (15.7)	49	85.1 (12.3)	NP	-2.202
Respiratory symptoms^a																		
Coughing	33	58.6 (18.1)	18	78.3 (8.0)	<.001	-1.286	20	87.6 (11.1)	27	86.9 (15.0)	.846	0.045	20	82.5 (12.5)	34	82.5 (16.3)	.615	0.002
Wheezing	31	58.9 (18.9)	20	75.9 (10.2)	.001	-1.059	16	85.6 (12.1)	31	88.1 (14.0)	.301	-0.186	16	79.1 (15.4)	38	83.9 (14.7)	.149	-0.323
Dyspnea	18	55.4 (20.9)	33	71.1 (13.6)	.007	-0.952	15	80.6 (18.8)	32	90.3 (8.5)	.097	-0.764	16	75.4 (18.5)	38	85.5 (12.2)	.038	-0.705
Chest tightness	7	45.2 (20.4)	44	68.8 (15.5)	.003	-1.461	12	78.3 (20.4)	35	90.3 (8.2)	.072	-0.972	12	74.0 (18.8)	42	84.9 (12.9)	.043	-0.758
Respiratory infections ^{****}	14	53.5 (18.1)	37	70.1 (15.9)	.003	-1.008	16	83.6 (17.6)	31	89.1 (10.3)	.431	-0.411	17	77.0 (17.6)	37	85.0 (13.0)	.058	-0.547

(Continued)

Table 3. Comparison of Total scores on the EA-QOL Questionnaire for Children 2-7 and 8-17 Years Old Between Clinical Subgroups (Continued)

	Children 2-7 Years Old (Parent-Reported)						Children 8-17 Years Old (Child-Reported)					
	Yes			No			Yes			No		
	N	Mean (SD)	P	Mean (SD)	N	P	ES [†]	N	Mean (SD)	P	ES [†]	N
Asthma ^{****}	16	59.7 (21.2)	.193	68.2 (15.9)	16	.048	-0.485	16	81.8 (17.6)	.137	-0.632	17
Feeding difficulties ^a												
Avoids certain foods	32	61.9 (18.6)	.048	71.7 (15.6)	13	.048	-0.558	13	75.9 (17.1)	.001	-1.369	16
Eats small portions	34	64.2 (16.8)	.238	68.2 (20.5)	15	.238	-0.218	15	78.1 (16.4)	.001	-1.130	18
Energy-enriched food	12	54.0 (19.1)	.017	69.1 (16.3)	2	.017	-0.886	2	54.8 (28.6)	NP	-2.948	2
Adjusted food consistency	13	50.3 (19.6)	.002	70.7 (14.3)	4	.002	-1.296	4	74.1 (27.5)	NP	-1.117	4
Needs more than 30 minutes to finish a large meal	24	59.7 (19.7)	.047	70.8 (14.8)	15	.047	-0.641	15	83.2 (16.7)	.177	-0.450	16
Increased fluid intake during meals	21	60.4 (18.5)	.083	69.1 (17.0)	18	.083	-0.492	18	81.5 (16.8)	.030	-0.734	18
Nutrition through gastrostomy	11	50.7 (19.0)	.002	69.6 (15.6)	5	.002	-1.161	5	87.6 (10.7)	NP	0.034	5
Nutrition through infusion pump	4	43.0 (12.2)	NP	67.5 (17.1)	2	NP	-1.448	2	86.9 (13.5)	NP	-0.024	2
Assistance by an adult during meals	15	57.6 (20.2)	.049	68.8 (16.2)	4	.049	-0.643	4	87.2 (10.0)	NP	-0.001	4

Values of $P < .05$ are significant. The number of patients included in the testing refers to patients with both available clinical data and EA-QOL-Total-scores.^aCohen's d used for a standardized interpretation; effect size >0.2 , small; >0.5 , moderate; and >0.8 , large.[†]Symptoms are reported to have occurred within the 4 last weeks.^{**}Patients were classified as suffering from heartburn if they were reported to experience this symptom more than once over the previous 4 weeks.^{***}Patients were classified as having vomiting problems if they were reported to experience vomiting associated with or after meals more than once times over the previous 4 weeks.^{****}Patients were classified as having dysphagia if they were reported to experience trouble swallowing, having food getting stuck in their throat, or choked on food more than once over the previous 4 weeks.^{*****}Patients were classified as having respiratory infection if they were reported to experience recurrent lower respiratory infection that required medical treatment.^{*****}Patients were classified as having asthma if they had hyper-reactive bronchial airway disease in the respiratory function test and had been diagnosed by a pediatric pulmonologist and/or an allergist.

EA, esophageal atresia; ES, effect size; NP, not performed; QOL, quality of life.

Table 4 . Clinical Concurrent Validity Using Linear Regression Analysis for Digestive Symptoms, Feeding Difficulties, and Respiratory Symptoms with Total EA-QOL Scores

The EA-QOL Questionnaires												
Symptoms	Children Aged 2-7 Years (Parent-Reported) n = 51				Children Aged 8-17 Years (Child-Reported) n = 47				Children Aged 8-17 Years (Parent-Reported) n = 54			
	β_0	β_1	R ²	P	β_0	β_1	R ²	P	β_0	β_1	R ²	P
Digestive symptoms*	NS	NS	NS	>.05	93.6	-7.4	0.53	<.001	89.6	-8.1	0.53	<.001
Feeding difficulties**	89.1	-4.4	0.24	<.001	92.3	-3.1	0.21	.001	88.5	-3.8	0.24	<.001
Respiratory symptoms***	84.6	-4.1	0.14	.008	NS	NS	NS	>.05	86.1	-2.0	0.08	.04
NS, not significant.												
*Dysphagia/heartburn/vomiting-problems.												
**Avoids certain food/eats small portions/energy-enriched food/adjusted food consistency/needs more than 30 minutes to finish a large meal/increased fluid intake during meals/nutrition through gastrostomy/nutrition through infusion pump/assistance by an adult during meals.												
***Coughing/wheezing/dyspnea/chest tightness/respiratory infections/asthma.												

NS, not significant.

*Dysphagia/heartburn/vomiting-problems.

**Avoids certain food/eats small portions/energy-enriched food/adjusted food consistency/needs more than 30 minutes to finish a large meal/increased fluid intake during meals/nutrition through gastrostomy/nutrition through infusion pump/assistance by an adult during meals.

***Coughing/wheezing/dyspnea/chest tightness/respiratory infections/asthma.

results may be contaminated by the respondents' memory. The excellent results could possibly be explained by the high response rate. Moreover, the child-parent agreements of the EA-QOL scores were good. In comparison, the parent-child agreement has been shown to be better regarding the EA children's condition-specific HRQOL, than generic HRQOL.²⁰

It is desirable for a condition-specific questionnaire to be able to define the role of clinical parameters on reduced HRQOL and discriminatively identify those in the population facing a larger burden and those facing a smaller burden.¹ Therefore, the EA-QOL total scores were, in terms of known-group and concurrent validity, investigated in relation to clinical variables indicating disease severity.

The Turkish version of the EA-QOL questionnaires for children 2-7 years old (parent-reported) and 8-17 years old (child-reported and parent-reported) showed good discriminative ability with regard to digestive morbidity, reflecting that esophageal symptoms and feeding difficulties in EA children are associated with reduced EA-QOL total scores. Therefore, the EA-QOL questionnaires can be understood as having acceptable known-group and clinical concurrent validity, similar to results in the Swedish-German field-test study.¹³ Esophageal morbidity is common in EA children¹ and may impair generic HRQOL, with risk factors like GERD⁸, hospitalization due to esophageal illness, gastrostomy insertion,⁹ and long-term follow-up for nutritional support.¹² A condition-specific HRQOL-instrument could further increase the knowledge of risk factors of HRQOL impairments in areas with proven importance to the EA patients, such as eating and aspects of social functioning. The implementation of such an instrument in a patient encounter during follow-up care could clinically add to the understanding of the patient's outcome, along with appropriate interventions.³

The Turkish version of the EA-QOL questionnaires for children 2-7 years discriminated well regarding the respiratory symptoms (known-group validity, concurrent validity). In the version for children aged 8-17 years, only the parent-reported version showed that an increased number of respiratory symptoms decreased the EA-QOL total scores. In the known-group analysis, dyspnea and chest tightness in EA children were associated with lower condition-specific HRQOL, similar to the Swedish-German evaluation¹³ and in line with studies demonstrating that severe respiratory disorders like pneumonia, dyspnea,¹⁰ and hospitalization due to respiratory illness may negatively influence generic HRQOL.

Respiratory problems may become less prevalent as the patients age,² and this has been described as an HRQOL concern primarily in young EA patients.²² In consistency, the EA-QOL questionnaire for children aged 2-7 years include more questions about respiratory problems than the version for older children.¹³ This can partly explain our findings.

The Turkish version of the EA-QOL questionnaire for children aged 8-17 years showed that children with a prior gastrostomy insertion had lower total scores, which was observed in EA children aged 2-7 years during the Swedish-German evaluation of the EA-QOL questionnaire.¹³ In our study, patients required gastrostomy insertion for reasons such as long-gap EA, revisional surgery, oropharyngeal dysphagia with severe aspiration, or associated anomalies, that is, with variable duration and causes. The results may therefore have to do with the underlying reason rather than the procedure. Notably, there were too few cases for a statistical analysis in several other surgical groups, limiting the statistical evaluation of the EA-QOL questionnaires.

Moreover, the convergent validity was good, showing that the measures of condition-specific and generic HRQOL were related in our investigation.

This study has limitations, which should be considered in relation to the findings. The clinical symptoms were parent-reported, and the standard definitions of dysphagia, heartburn, and feeding problems were lacking. The clinical heterogeneity of patients with EA may prevent an objective assessment and contribute to the study results. Although the data were obtained from 2 centers, the results reflected a Turkish cohort of patients with no standardized treatment and/or follow-up protocol. Moreover, future studies are warranted to decide on differences in EA-QOL scores between different countries.

Ethics Committee Approval: The Ethical Committee of Hacettepe University approved the study (GO/2019/19-36).

Informed Consent: Informed consent was obtained from all responders.

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