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ABSTRACT

Purpose: Assessing quality of life (QoL) after esophageal replacement (ER) for long gap esophageal atresia (LGEA).
Methods: All patients after ER for LGEA with gastric pull-up (GPU n = 9) or jejunum interposition (JI n = 14) at the University Medical Center Groningen and Utrecht (1985–2007) were included. QoL was assessed with 1) gastrointestinal-related QoL using the Gastrointestinal Quality of Life Index (GIQLI)), 2) general QoL (Child Health questionnaire CHF87-BREF (children)/World Health Organization questionnaire WHOQOL-BREF (adults)), and 3) health-related QoL (HRQoL) (TNO AZL TACQoL/TAAQoL). Association of morbidity (heartburn, dysphagia, dyspnea on exertion, recurrent cough) and (HR)QoL was evaluated.
Results: Six patients after GPU (75%) and eight patients after JI (57%) responded to the questionnaires (mean age 15.7, SD 5.9, 12 male, two female). Mean gastrointestinal, general and health-related QoL total scores of the patients were comparable to healthy controls. However, young adults reported a worse physical functioning ($p = 0.02$) but better social functioning compared to peers ($p = 0.01$). Morbidity was not associated with significant differences in (HR)QoL.
Conclusions: With the current validated QoL most patients after ER with GPU and JI for LGEA have normal generic and disease specific QoL scores. Postoperative morbidity does not seem to influence (HR)QoL.
Type of Study: Prognosis Study.
Level of evidence: III.

Esophageal atresia (EA) is a rare congenital disorder characterized by absence of esophageal continuity. In most patients, a primary anastomosis can be performed. However, if the distance between the two esophageal remnants is too wide for primary repair, esophageal replacement (ER) strategies may have to be deployed. Replacement with jejunum [1–3], colon [4], or stomach [5] have all been advocated.

Gastrointestinal and respiratory morbidity have been investigated after primary anastomosis for EA [6–11]. Long term morbidity after primary EA repair has been considered to be moderate and QoL in adult patients has been demonstrated to be excellent [12–13]. However, long term morbidity for long gap esophageal atresia (LGEA) appears to be significant. Only a few studies have investigated QoL after ER and mostly without using validated tools. QoL after jejunum interposition has never been analyzed before. We hypothesized that the long term QoL will be diminished in patients who underwent ER in comparison to healthy controls. For optimal care of children after ER and their transition from pediatric to adult healthcare, we should have knowledge of their medical, as well as psycho-social status. Therefore, this study aims to investigate QoL after ER for LGEA in children and young adults and analyze whether morbidity might influence patients' well-being.

1. Patients and methods

A cross-sectional cohort study was performed. All patients that had undergone a gastric pull-up (GPU) at the University Medical Center Groningen (UMCG) between 1985 and 2006 and jejunal interposition (JI) at the University Medical Center Utrecht (UMCU) between 1988 and 2007 for LGEA were included. At the time of the study, GPU was the preferred method at the UMCG and a JI was the preferred method at the UMCU. The participating centers did not perform colon interposition, which is a procedure scarcely encouraged in Europe since it is reserved as last option for esophageal replacement [14,15]. In this cohort, patients were diagnosed with LGEA if a primary end-to-end

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anastomosis was not feasible due to the distance between the proximal and distal esophagus measured under fluoroscopy.

Primary endpoint of the present study was the assessment of HRQoL and QoL outcome in LGEA patients after JI or GPU.

Secondary endpoint was the evaluation of morbidity parameters associated with (HR)QoL.

1.1. Ethical approval

This assessment was conducted in accordance with the local medical ethics review boards of the University Medical Center Groningen (UMCG, Ref. M14.159735) and University Medical Center Utrecht (UMCU, Ref. WAG/om/15/001186).

2. Measurements

Patient characteristics were collected from the medical records. Sociodemographic aspects were assessed using structured questions on marital status; education and occupation.

2.1. Quality of Life measurements

QoL was assessed using validated questionnaires. The QoL measures were self-report measurements. Three areas were investigated: Disease-Specific QoL using the Gastrointestinal Quality of Life Index (GIQLI), general QoL using the CHF87-BREF (children) and WHOQOL-BREF questionnaire (adults), and health-related QoL using the TACQoL (children 6–15 years old) and TAAQoL (patients aged 16 years and older).

2.2. Disease-specific QoL

The GIQLI, introduced by Eypasch et al. [16], is a validated tool to assess HRQoL in patients with gastrointestinal (GI) disease and especially in those who underwent surgery. The questionnaire contains 36 items, each with five response categories concerning gastrointestinal disease-related symptoms, physical status, emotions and psychosocial functions. The questionnaire is developed with 5-point Likert scale, ranging from 0 to 4, with 4 implying the least complaints (a higher score represents a better QoL). The theoretical maximum score is 144 points. A GIQLI score less than 105 indicates that the responder experiences persistent GI symptoms. Patients with a total score of less than 105 were therefore considered as symptomatic.

2.3. General QoL

The Child Health Questionnaire Child Form (CHQ-CF87) [17] measures psychosocial and physical well-being in patients of 5 to 18 years of age. It provides a qualitative assessment of overall health status across multiple domains. It consists of 87 items divided into 10 multi-item scales, per scale items are summed up and transformed into a 0 (worst possible score) to 100 (best possible score) scale. Reference data were obtained from 444 subjects, mean age 12.8 (9–17), SD 1.7.

The WHOQOL-BREF [18] is a QoL assessment developed by the WHOQOL group for adults. It consists of 26 items in four different domains and a general QoL facet. The domains are physical health, psychological health, social relationships, and family/social environment. The response scales are 5-point Likert scales. A higher score represents a better QoL. Reference data were obtained from 11.830 subjects, mean age 45 (12–97), SD 16.

2.4. Health-related QoL

HRQoL is a combination of health problems and emotional responses towards these health problems. It reflects the subjective perception of health and is increasingly recognized as a relevant 'patient-reported outcome' since it measures the emotional impact of self-reported functional problems [19–20].

HRQoL was assessed using TACQoL/TAAQoL [21–24] questionnaires developed by The Netherlands Organization (TNO) for Applied Scientific Research and the Academic Hospital in Leiden (LUMC), which explicitly offers respondents the possibility to differentiate between their functioning and the way they feel about it.

The TACQoL (for children 6–15 years old) contains 7 domains: social functioning, autonomous functioning, physical complaints, motoric functioning, cognitive functioning, positive emotions and negative emotions. Reference data were obtained from 1253 subjects, mean age 13.4 (12–15), SD 1.0.

The TAAQoL (for patients aged 16 years and older) consists of 12 domains: gross motor functioning, fine motor functioning, cognition, sleep, pain, social contacts, daily activities, sex, vitality, happiness, depressive mood and anger. Items are scored on a 0–4 point Likert scale. Scales are transformed to a 0–100 scale, with higher scores representing a better HRQoL. Reference data were obtained from 4410 subjects, mean age 47.5 (16–97), SD 16.9.

2.5. Parameters of morbidity and QoL

Relation between (HR)QoL measurements and post-operative symptoms such as heartburn, dysphagia, dyspnea on exertion, recurrent pneumonia and cough and post-operative surgical re-intervention (anastomotic revision and esophageal dilatations) were investigated.

3. Statistical analysis

Data were entered into a SPSS database and statistical analysis was performed using SPSS (SPSS version 23 9SPSS Inc., Chicago, IL). Data were expressed as mean ± SD for continuous variables, group differences were analyzed using one sample t-test, and two sample t-test for the CHQ-CF87. To examine differences in (HR)QoL between GPU and JI, the means of the two groups were compared using two sample t-tests. Because children completed either the TACQoL or the TAAQoL, depending on age, age-appropriate z-scores of the two were compared. (HR)QoL measurements of patients reporting a specific complain at last follow-up (e.g. heartburn) were compared with those of patients not presenting that symptom using Mann–Whitney U-test. Statistical differences were considered as significant for p-value <0.05.

4. Results

In total nine GPU and 15 JI patients had undergone ER for LGEA at the UMCG and UMCU respectively. One JI patient with trisomy 21 died at the age of 10 years most likely as a result of massive aspiration. Six GPU and eight JI patients had responded to the questionnaires and could be evaluated for this study. Mean age of the 14 responders was 15.7 +/−5.9 SD (12 male, two female).

No differences were found in patient characteristics between responders and non-responders (Table 1a). Characteristics of patients joining the study are shown in Table 1b. Sociodemographic factors did not differ within the two groups (see Table 2), almost 50% of the patients ever flunked a year at school. The median follow-up duration after surgery was 12 years (4–24): 12 years (4–17) after GPU and 14 years (7–24) after JI (Tables 8a and 8b), all but one patient (GPU) were on full oral diet and did not require nutritional supplements. No differences were found in morbidity between the patients who participated in the study and the patients who did not.

4.1. Gastrointestinal QoL (GIQLI)

There was no significant differences between the total mean score of both patients groups ($n = 14$) and helathy controls (124.2, SD 11.0 vs 125.8, SD 13.0, $p = 0.6$). One JI patient reported a total score of less

Table 1a
Responders vs non-responders patient characteristics. GPU (gastric pull-up), JI (jejunum interposition).

	Responders *(n = 14)*	Non-responders *(n = 9)*	p Value
Gestational age *(weeks)*	35.2 *(+/−2.9)*	34.4 *(+/−3.2)*	0.5
Weight at birth *(gr)*	2150 *(+/−755)*	2154 (+/−740)	0.8
Type atresia A	5	1	0.3
Type atresia B	8	7	0.4
Type atresia C	1	1	1
Age at surgery *(days)*	124 (+/−104)	100 (+/−89)	0.4
Any VACTERL anomalies	8 *(57%)*	5*(55%)*	1
Cardiac	4	2	1
Renal	2	3	0.3
Anorectal	2	1	1
Vertebral	3	3	1
GPU	6 *(66%)*	3 *(33%)*	1
JI	8 *(57%)*	6 *(43%)*	1

Table 1b
Patient characteristics.

	Total *(n = 14)*	GPU *(n = 6)*	JI *(n = 8)*	p Value
Gestational age *(weeks)*	35.2 *(+/−2.9)*	34.6 (+/−3.6)	35.6 (+/−2.5)	0.6
Weight at birth *(g)*	2150 *(+/−755)*	2054 (+/−685)	2221 (+/−842)	0.8
Type atresia A	5	4	1	0.09
Type atresia B	8	1	7	0.02
Type atresia C	1	1	0	0.4
Gastrostomy	14	6 (100%)	8 (100%)	1
Age at surgery *(days)*	124 (+/−104)	140.5 (+/−90)	111.8 (+/−118)	0.3
Any VACTERL anomalies	8 (57%)	5(83%)	3(37%)	0.1
Cardiac	4	2	2	1
Renal	2	2	0	0.1
Anorectal	2	1	1	1
Vertebral	3	3	0	0.05
Anastomotic leak requiring re-intervention	3 (21%)	0	3 (37.5%)	0.2

Table 2
Sociodemographic factors.

	Total *(n = 14)*	GPU *(n = 6)*	JI *(n = 8)*	p Value
Mean age	15.7 +/−5.9 (6–28)	17.7 +/− 5.5 (8–28)	14.3 +/− 6.2 (6–25)	0.4
Still student	43% (6)	33% (2)	50% (4)	0.6
Ever flunked	50% (7)	66.7% (4)	37.5% (3)	0.5
Additional job	21.4% (3)	33% (2)	12.5% (1)	0.5
Finished with studies and unemployed	- (0)	- (0)	- (0)	-
Currently full time job	14.3% (2)	16.7% (1)	12.5% (1)	1
Partner	7% (1)	- (0)	12.5% (1)	1
Living alone	28.6% (4)	16.7% (1)	37.5% (3)	0.5
Living with partner	- (0)	- (0)	- (0)	-
Living with parents	71.4% (10)	83.3% (5)	62.5% (5)	0.5
Having children	- (0)	- (0)	- (0)	-

than 105 and was considered symptomatic (Table 3). No significant differences were found between the different domains of the GIQLI.

4.2. Generic QoL

There was no significant differences between the total mean score of the children after ER and healthy controls (Table 4). Three children after ER (21%), had a very low mean score (<-2SD) in the domains pain, general behavior and emotional functioning.

There was no significant difference between the total mean score of the young adults after ER and healthy controls. In the domain physical functioning, young adults scored significantly lower compared to healthy controls (16.9 (SD 1.5) vs 18.3 (SD 3), p = 0.02). In the domain environment, mean scores were higher compared to healthy controls (17.2 (SD 1.7) vs 15.9 (SD 2.8), p = 0.05). None of the young adults scored below -2SD (Table 5). No statistically significant differences were found between GPU and JI in QoL measurements, the mean z-score of QoL after GPU was 0.0015 (SD 0.9) and after JI was 0.09 (SD 0.7), p = 0.6.

Table 3
Disease specific QoL evaluated using GIQLI.

	GPU *(n = 6)*		JI *(n = 8)*		
	Mean	SD	Mean	SD	p Value
Physical well being	23	5.1	23.5	3.5	*0.9*
Gastrointestinal symptoms	65.8	4	63.1	8.7	*0.8*
Social well being	19.3	1	18.7	17.3	*0.3*
Emotional well being	18	1.6	17.4	1.9	*0.4*
Total	126.1	10.9	122.7	13.1	*0.6*

4.3. HRQoL

Children after ER scored significantly higher than healthy controls in both the positive (15.6 (SD 0.5) vs 13.0 (SD 2.8), p = 0.00) and negative (13.6 (SD 1.6) vs controls 11.6 (SD 2.5), p = 0.01) emotion domains.

Table 4
QoL evaluated using CHQ.

	Patients (n = 7)		Controls		
	Mean	SD	Mean	SD	p Value
Physical functioning	97.3	3.5	96.8	5.4	0.7
Role functioning-emotional	90.4	20.7	92.3	16.8	0.8
Pain	75.7	26.9	78.2	19.5	0.8
General behavior	82.1	16.5	83.6	10.2	0.8
Self esteem	76.7	5.2	75.4	12.5	0.5
General health	65.2	11.7	74.6	15.9	0.07
Mental health	84.3	8.6	78.2	13	0.1
Family cohesion	86.4	18	75.7	23.1	0.1

Table 5
QoL evaluated using WHOQoL.

	Patients (n = 9)		Controls		
	Mean	SD	Mean	SD	p Value
Physical functioning	16.9	1.5	18.3	3	*0.02*
Psychological functioning	16.3	1.6	16.1	2.8	*0.6*
Social Relationship	16.5	2.2	15.8	3.3	*0.3*
Environment	17.2	1.7	15.9	2.8	*0.05*

Table 6
HRQoL evaluated using TACQoL.

	Patients (n = 9)		Controls		
	Mean	SD	Mean	SD	p Value
Physical functioning	26.0	3.2	23.6	5.3	*0.07*
Motor functioning	29.6	2.6	29.7	3.2	*0.9*
Cognitive functioning	27.2	3.5	27.5	4.1	*0.8*
Autonomy	30.7	3.5	31.0	2.9	*0.8*
Positive moods	15.6	0.5	13.0	2.8	*0.00*
Negative moods	13.6	1.6	11.6	2.5	*0.01*

One child after JI scored <-2SD in the domain autonomy. In the other domains no differences were found (Table 6).

In the domain social functioning, young adults scored significantly better than controls (95.8 (SD 7.5) vs 83.7 (19.2 SD) p = 0.01). More aggressive emotions (98.1, SD 4.5) were reported by young adults compared with healthy controls (87.6, SD 16.8, p = 0.002). In the other domains, no differences were found. One young adult after JI scored <-2SD in the domain sleep (Table 7). No statistically significant differences were found between GPU and JI in HRQoL measurements, the mean z-score of HRQoL after GPU was 0.409 (SD 0.62) and after JI was 0.171 (SD 0.82), p = 0.077.

Table 7
HRQoL evaluated using TAAQoL.

	Patients (n = 7)		Controls		
	Mean	SD	Mean	SD	p Value
Cognitive functioning	89.5	10.2	82.7	22.8	*0.1*
Sleep	67.7	21.8	73.8	26.1	*0.5*
Pain	82.2	18.7	73.2	24.2	*0.2*
Social functioning	95.8	7.5	83.7	19.2	*0.01*
Daily activities	86.4	20.3	83.4	24.8	*0.7*
Sexuality	87.5	13.6	84.4	25.7	*0.6*
Vitality	54.1	18	63.8	23.9	*0.2*
Positive emotions	76.3	14.3	64.5	21.8	*0.8*
Depressive emotions	81.9	13.3	77.9	20.6	*0.4*
Aggressive emotions	98.1	4.5	87.6	16.8	*0.002*

Table 8a
Postoperative morbidity.

	GPU (n = 6)	JI (n = 8)	TOTAL (n = 14)
Heartburn	1 *(16%)*	1 *(12%)*	2 *(14%)*
Esophageal dilatation	3 *(50%)*	1 *(12%)*	4 *(28%)*
Episodic dysphagia	3 *(50%)*	4 *(50%)*	7 *(50%)*
Asthma-like symptoms	2 *(33%)*	0 *(−)*	2 *(14%)*
Recurrent pneumonia	1 *(16%)*	2 *(25%)*	3 *(21%)*
Dyspnea on exertion	3 *(50%)*	2 *(25%)*	5 *(35%)*
Recurrent cough	2 *(33%)*	3 *(37%)*	5 *(35%)*
Re-operation	0 *(−)*	3 *(37%)*	3 *(21%)*

Table 8b
Postoperative morbidity responders vs non-responders.

	Responders (n = 14)	Non-responders (n = 9)	p Value
Heartburn	2 *(14%)*	1 *(11%)*	1
Episodic dysphagia	7 *(50%)*	4 *(44%)*	1
Dilatations	4 *(28%)*	6 *(66%)*	0.1
Asthma like symptoms	2 *(14%)*	2 *(22%)*	1
Recurrent pneumonia	3 *(21%)*	3 *(33%)*	0.6
Dyspnea on exertion	5 *(35%)*	4 *(44%)*	1
Recurrent cough	5 *(35%)*	4 *(44%)*	1
Reoperation	3 *(21%)*	3 *(33%)*	0.6
Full oral diet	13 *(93%)*	7 *(77%)*	0.5

Table 9a
Relation between morbidity and HRQoL measurements in patients up to 15 years old (TACQoL). Data are reported as p value. A p value <0.05 indicates a symptom associated with significant lower HRQoL measurement.

	Physical function	Motor function	Cognitive function	Autonomy	Positive moods	Negative moods
Heartburn	1	0.5	0.8	0.5	0.8	0.3
Esophageal dilatation	1	0.6	0.4	0.2	0.6	0.7
Dysphagia	0.5	0.1	0.7	0.3	0.6	0.1
Asthma-like symptoms	0.4	0.5	0.7	0.6	0.3	0.1
Recurrent						

pneumonia10.50.80.50.80.3**Dyspnea on exertion**0.40.20.70.60.30.4**Recurrent cough**10.40.10.20.60.2**Re-operation**0.10.090.40.60.20.7

4.4. Parameters associated with QoL

Re-intervention due to anastomotic leakage and esophageal dilatations were not associated in a change in (HR)QoL. Post-operative symptoms were not associated with significant differences in (HR)QoL measurements (Tables 9a, 9b, 10).

5. Discussion

This study investigated (HR)QoL in children and young adults after ER for LGEA. It is the first study on (HR)QoL after JI in children and young adults. We found that generic and disease specific QoL in the majority of patients after ER is comparable to normal QoL scores as measured in healthy population. No significant differences in (HR)QoL were found between GPU and JI patients. Furthermore, postoperative morbidity is not associated with changes into (HR)QoL.

In this study we found gastrointestinal-related QoL (GIQLI) to be generally good: only one patient (JI) scored below the cut-off for symptomatic patients, no significant differences were found between the groups and the controls nor between the two groups. Recently, Hannon et al. analyzed gastrointestinal-related QoL using GIQLI in 32 patients after GPU. Eighteen of them had a GPU for LGEA while in 14 patients GPU was performed as rescue procedure after failed primary repair or

Table 9b
Relation between HRQoL measurements in patients aged 16 years and older (TAAQoL) and morbidity. Data are reported as p value. A p value <0.05 indicates a symptom associated with significant lower HRQoL measurement.

	Heartburn	Esophageal dilatation	Episodic dysphagia	Asthma-like symptoms	Recurrent pneumonia	Dyspnea on exertion	Recurrent cough	Re-operation
Cognitive functioning	0.1	0.3	0.6	0.8	0.2	0.6	0.1	0.2
Sleep	0.1	0.5	1	0.6	0.4	0.2	0.1	0.4
Pain	0.2	1	0.8	0.4	1	0.6	0.2	0.2
Social functioning	0.4	0.7	0.2	0.4	0.4	0.2	0.4	0.2
Daily activities	0.3	0.8	0.6	0.1	0.6	0.1	0.3	0.6
Sexuality	0.3	0.4	1	0.1	1	0.1	0.3	1
Vitality	0.3	0.2	1	0.6	0.2	0.4	0.3	0.2
Positive emotions	1	1	0.4	0.4	0.4	0.4	1	0.2
Depressive emotions	1	0.3	0.3	1	0.4	0.8	1	0.5
Aggressive emotions	0.6	0.3	0.4	0.1	0.1	0.4	0.6	0.4

colon interposition [25]. Results showed that the median gastrointestinal-related QoL according to GIQLI was 113, therefore above the cut-off point of symptomatic impairment (105), comparable to our findings. Dingemann et al. investigated gastrointestinal-related QoL in 27 patients who had an ER for complex/complicated esophageal atresia. GIQLI scores were found significantly worse when compared to the reference group [26]. A recent systematic review [27] reported significant worse GIQLI measurements for LGEA patients compared to the normal population. However, the majority of included patients underwent colon interposition as ER procedure. These results appear to be in contrast with our findings, however, differences in the surgical strategies make comparison complicated.

In our study, general QoL in children after ER appeared comparable to the healthy population. There was no difference in the general QoL in young adults compared to healthy controls. However, young adults scored significantly worse on the domain physical functioning. Despite the physical limitation, the general QoL seems normal in young adults.

HRQoL was comparable to population average for both children and young adults. Young adults perceive their social functioning better than controls but described more aggressive emotions compared to the population average. This appears to be in contrast with previous studies investigating social functioning of children with chronic illness [28,29] and it might reflect a shift in the coping mechanisms of patients after ER towards a higher emotional sensitivity. Dingemann et al. [26] analyzed also HRQoL (KIDSCREEN27). Conform to our findings, HRQoL was perceived as generally good and with regard to the domain physical well-being patients scored even better than controls. However, a correlation between long-term morbidity and HRQoL was not investigated in this series. We did not identify significant differences in (HR)QoL after the two surgical procedures. Patients after GPU reported HRQoL measurements higher than JI patients although not statistically significant ($p = 0.077$).

In this study the relationship between postoperative morbidity and (HR)QoL was analyzed. Gastrointestinal and respiratory parameters were not associated with significant differences in (HR)QoL measurements. This outcome might suggest that physical complaints in ER patients do not affect patients´ perception of well-being. This may be due to the fact that LGEA patients and their families have accepted this morbidity. Patients and their families might have developed efficient coping strategies in order to face the challenges of life after ER. Interestingly, it has been suggested that patients with congenital diseases might report even better QoL scores than children with acquired conditions, due to stronger coping strategies elaborated from early childhood [30–31]. Fifty-seven patients that had a primary correction of EA demonstrated indeed better QoL measurements compared to children with diabetes and asthma [32].

Patients after ER might seek stability by evolving their expectations and conceptions of themselves and their social role [33]. LGEA patients might have developed different internal standards for daily activities compared to peers. They might have elaborated different life values and might have re-conceptualized their physical limitations, leading to paradoxical satisfactory findings when responding to the present questionnaires. Family influences on patient's daily life have to be considered as well. Parents of chronically ill children tend to overprotect their children [34]. One might assume that this happens for patients after ER as well. Although this is comprehensible parental behavior, it might represent a limitation to develop children's social functioning during adolescence. Moreover, somatic morbidity may affect the development of their personal identity and consequently may lead to social marginalization during a time when self-esteem largely depends on the acceptance by peers. Therefore, physicians should encourage the family of patients after ER to promote and sustain the social contacts and autonomy of their children. However, even if we noticed a shift towards more emotional sensitivity during transition into adulthood, emotional development seems adequate, with outcomes such as vitality, social and cognitive functioning comparable to controls.

Limitations of this study include the small sample size that may lead to the lack of significant differences between the two groups.

The GIQLI questionnaire represents a valid tool for evaluation of disease-specific QoL in patients with gastrointestinal disorder however, it is not tailored for patients with EA. Dellenmark-Blom et al. [35] recently developed and validated a German and Swedish condition-

Table 10
Relation between morbidity and QoL measurements (WHOQoL). Data are reported as p value. A p value <0.05 indicates a symptom associated with significant lower QoL measurement.

	Physical function	Psychological function	Social relations	Environment
Heartburn	0.8	0.4	0.2	0.8
Esophageal dilatation	0.2	0.4	0.1	0.1
Dysphagia	0.3	1	0.1	0.9
Asthma-like symptoms	0.3	0.1	0.6	0.6
Recurrent pneumonia	0.8	1	1	0.7
Dyspnea on exertion	0.6	1	0.6	0.7
Recurrent cough	0.8	0.4	0.2	0.8
Re-operation	0.8	0.5	0.1	0.5

specific HRQoL tool for patients who had a primary correction of EA. When implementing this for children with LGEA and ER, it might represent a more appropriate instrument to investigate disease-specific QoL in our patients. To date however, this questionnaire has not yet been validated for the Dutch population.

6. Conclusion

With the current validated QoL questionnaires, most patients after ER with GPU and JI for LGEA have normal generic and disease specific QoL scores. Postoperative morbidity and surgical reintervention do not seem to influence (HR)QoL. The question remains if non condition-specific HRQoL tools are suitable for this specific patients group. Condition-specific HRQoL tools may provide more detailed information on HRQoL for all EA patients. We expect that these tools may provide a tailor-made support if necessary.

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