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Quality of Life Outcomes in Primary Caregivers of Children with Esophageal Atresia

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Objective To investigate the quality of life (QoL) impact on primary caregivers of children with esophageal atresia.
Study design We used a prospective cohort study design, inviting primary caregivers of children with esophageal atresia to complete the following questionnaires: Parent Experience of Child Illness (PECI), Patient-Reported Outcomes Measurement Information System (PROMIS) Anxiety, PROMIS Depression, 12-Item Short Form Survey (SF-12), and Pediatric Quality of Life Inventory (PedsQL). The PECI, PROMIS Anxiety and Depression, and SF-12 assessed caregiver QoL, and the PedsQL assessed patient QoL. Patients with Gross type E esophageal atresia served as controls.
Results The primary caregivers of 100 patients (64 males, 36 females; median age, 4.6 years; range, 3.5 months to 19.0 years) completed questionnaires. The majority (76 of 100) of patients had Gross type C esophageal atresia. A VACTERL (vertebral anomalies, anorectal malformation, cardiac anomalies, tracheoesophageal fistula, renal anomalies, limb anomalies) association was found in 30, ≥1 esophageal dilatation was performed in 57, and fundoplication was performed in 11/100. When stratified by esophageal atresia types, significant differences were found in 2 PECI subscales (unresolved sorrow/anger, $P = .02$; uncertainty, $P = .02$), in PROMIS Anxiety ($P = .02$), and in SF-12 mental health ($P = .02$) and mental component summary scores ($P = .02$). No significant differences were found for VACTERL association, nor esophageal dilatation. Requirement for fundoplication resulted in lower SF-12 general health score, and lower PedsQL social and physical functioning scores.
Conclusions We have demonstrated that caring for a child with esophageal atresia and a previous requirement for fundoplication impacts caregiver QoL. *(J Pediatr 2021;■:1-7).*

Esophageal atresia is a significant congenital anomaly of the esophagus that affects one in 2600 newborns.[1] Despite successful repairs in infancy, up to 85% of patients with esophageal atresia have ongoing esophageal dysmotility extending into adulthood. This is likely due to a combination of congenitally impaired neural innervation and narrowing at the surgical anastomosis. Subsequently, a reduction in esophageal wall compliance may result in poor food bolus transport, possibly manifesting as difficulty swallowing, esophageal stricture formation, food bolus obstruction, recurrent aspiration events with resultant pneumonia, and/or acute life-threatening respiratory arrest.[2-4] Many of these patients also develop a significant esophageal dilatation requirement.[5] All of these may impact the quality of life (QoL) of the patients and their families.

The literature in adults and children suggests that patients with esophageal atresia have a long-term QoL comparable to that of the general population.[6] However, patients do suffer morbidity, particularly symptoms of dysphagia.[7]

PECI	Parent Experience of Child Illness
PedsQL	Pediatric Quality of Life Inventory
PROMIS	Patient-Reported Outcomes Measurement Information System
QoL	Quality of life
RCH	The Royal Children's Hospital
SF-12	12-Item Short Form Survey
TEF	Tracheoesophageal fistula
VACTERL	Vertebral anomalies, anorectal malformation, cardiac anomalies, tracheoesophageal fistula, renal anomalies, limb anomalies

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The current literature is limited in terms of patient age groups, with the majority of the studies conducted in older children (≥8 years).[8-10] In a study that included patients as young as age 2 years, no comparison with the general population was made.[11]

The published literature also does not describe the impact of esophageal atresia on primary caregivers and their resultant QoL. In the present study, we assessed QoL outcomes in the primary caregivers of patients with esophageal atresia to identify areas of concern for patients and their families, as well as risk factors for poorer QoL outcomes.

Methods

We used a prospective cohort study design, inviting the primary caregivers of children with esophageal atresia to complete validated questionnaires. Patients were identified from the Nate Myers Oesophageal Atresia Database at The Royal Children's Hospital, Melbourne (RCH), a prospective database of patients with esophageal atresia including data collected from 1948 onward. All patients aged ≤19 years with esophageal atresia listed in the database (all esophageal atresia types) were eligible for inclusion. The most common variant of esophageal atresia is associated with a distal tracheoesophageal fistula (TEF), an abnormal congenital communication between the trachea and the esophagus. The Gross classification scheme was used to define the types of esophageal atresia: type A, esophageal atresia without TEF; type B, esophageal atresia with proximal TEF; type C, esophageal atresia with distal TEF; type D, esophageal atresia with both proximal and distal TEF; and type E, TEF without esophageal atresia (**Figure 1**; available at www.jpeds.com). Type E served as a proxy "control" group, as this variant has an esophagus in continuity from birth and typically does not have the same issues of dysmotility as the other types of esophageal atresia. An association with VACTERL (vertebral anomalies, anorectal malformation, cardiac anomalies, tracheoesophageal fistula, renal anomalies, limb anomalies) was defined as 2 or more other associations besides esophageal atresia/TEF.[12]

Eligible patients were identified, and their primary caregivers were recruited to complete 5 questionnaires. This study forms part of a larger prospective study focused on esophageal motility outcomes in patients with esophageal atresia. A cohort size of 100 primary caregivers was determined, based on the ability to recruit families during the pilot motility study. The primary caregivers of patients with esophageal atresia completed the questionnaires over the phone or in person if they were in attendance for an appointment at RCH. The validated questionnaires used were the Parent Experience of Child Illness (PECI),[13] Patient-Reported Outcomes Measurement Information System (PROMIS) Anxiety and PROMIS Depression,[14] 12-Item Short Form Survey (SF-12),[15] and Pediatric Quality of Life Inventory (PedsQL).[16] These questionnaires explore health-related QoL outcomes, functional outcomes, social functioning, caregiver well-being, and impact of illness on the family. Short and simple validated questionnaires were used to reduce the risk of bias.

Specifically, the PECI examines parental adjustment related to caring for a child with chronic illness, measured over 25 items. It comprises of 4 subscales: guilt and worry, unresolved sorrow and anger, uncertainty, and emotional resources. The original study reporting PECI scores focused on pediatric patients with brain tumors and will be compared alongside our findings.[13] The PROMIS Anxiety short form assesses the pure domain of anxiety, measured over 7 items. The PROMIS Depression short form assesses the pure domain of depression, measured over 8 items. The SF-12 is a multipurpose generic measure of health status measured over 12 items. The PedsQL is a brief measure of health-related QoL in children and young people including a psychosocial health summary score, a physical health summary score, and a total scale score. The PedsQL is measured over 36 items for infants aged 1-12 months, 45 items for infants aged 13-24 months, 21 items for toddlers aged 2-4 years, and 23 items for children aged ≥5 years. The study was approved by the RCH Human Research Ethics Committee (HREC 35089).

Statistical analyses was conducted using Stata 16.1 (StataCorp). To compare outcomes between esophageal atresia types and other risk factors, exact nonparametric analyses were used to circumnavigate smaller group sizes and nonnormal outcomes. The Kruskal–Wallis test was used to explore differences between esophageal atresia types, and the Mann–Whitney *U* test was used to compare 2 groups on other risk factors. Median and IQR were presented for group comparisons. Spearman correlation coefficients (with bootstrapped 95% CIs; 1000 repetitions) examined the relationships between age and questionnaire outcomes. However, where relevant, the mean and IQR are also included to allow for comparisons with previously published data. Statistical significance was defined as $P < .05$. Missing data were excluded from the analysis.

Results

The primary caregivers of the 100 children with esophageal atresia (64 males, 36 females; median age, 4.6 years; range, 3.5 months to 19.0 years) completed all questionnaires. The questionnaires generally took 15-20 minutes to complete. The primary caregivers included 86 mothers, 13 fathers, and 1 grandmother. Demographic data of the patients are summarized in **Table I** (available at www.jpeds.com). The majority (76 of 100) of the children had Gross type C esophageal atresia. A VACTERL association was found in 30 of the 100 children, ≥1 esophageal dilatation was performed in 57, and fundoplication was performed in 11. In terms of respiratory morbidity, 45 patients had tracheomalacia, 8 required continuous positive airway pressure or airway clearance therapy, 3 required a tracheostomy, 2 required home oxygen therapy, and 1 required aortopexy.

Parent Experience of Child Illness (Assessing Primary Caregivers)

The overall median scores were as follows: guilt and worry, 1.5 (IQR, 0.9-2.0); emotional resources, 3.2 (IQR, 2.8-3.4); unresolved sorrow and anger, 1.1 (IQR, 0.6-1.6); and uncertainty, 1.2 (IQR, 0.6-2.0). For comparison with previous publications, the overall mean scores were as follows: guilt and worry, 1.5 ± 0.8; emotional resources, 3.1 ± 0.5; unresolved sorrow and anger, 1.2 ± 0.7; and uncertainty, 1.3 ± 0.8. A comparison with the originally reported PECI scores that focused on pediatric brain tumors is presented in Table II (available at www.jpeds.com).[13] Our primary caregiver group had lower (better) scores for guilt and worry, unresolved sorrow and anger, and uncertainty compared with the brain tumor cohort, as well as higher (better) emotional resources scores.

When stratified by esophageal atresia type, a significant difference was found in 2 PECI subscales: unresolved sorrow and anger and uncertainty (both *P* = .02) (Table III). Bonferroni-adjusted post hoc examination of medians suggested that the primary caregivers of children with type A esophageal atresia had higher scores (worse) compared with the primary caregivers of children with type E esophageal atresia (proxy controls) for the PECI subscales of unresolved sorrow and anger and of uncertainty. The primary caregivers of children with type C esophageal atresia also had higher scores (worse) compared with the primary caregivers of children with type E esophageal atresia (proxy controls) for the PECI subscale of uncertainty. No significant differences were found when comparing for VACTERL association (Table IV; available at www.jpeds.com), esophageal dilatation (Table V; available at www.jpeds.com), previous fundoplication (Table VI), or age of the child (Table VII).

Patient-Reported Outcomes Measurement Information System: Anxiety (Assessing Primary Caregivers)

The overall median PROMIS Anxiety T score was 51.6 (IQR, 42.9-58.5). This is interpreted as "none to slight" levels of anxiety (<55).

When stratified by esophageal atresia type, a significant difference was found in PROMIS Anxiety (*P* = .02) (Table III). Bonferroni-adjusted post hoc examination of medians suggested that the primary caregivers of children with type A esophageal atresia and type C esophageal atresia had higher scores (worse) compared with the primary caregivers of children with type E esophageal atresia (proxy controls) for PROMIS Anxiety. No

Table III. Nonparametric comparison (Kruskal–Wallis) of outcomes by esophageal atresia type: PECI, PROMIS, and SF-12 for primary caregivers, PedsQL for patients

	Type A			Type B			Type C			Type E			
Esophageal atresia type	**n**	**Median**	**(IQR)**	**n**	**Median**	**(IQR)**	**n**	**Median**	**(IQR)**	**n**	**Median**	**(IQR)**	***P***
PECI guilt and worry	12	1.4	(1.4-1.9)	4	1.5	(0.9-1.9)	76	1.7	(1.0-2.0)	8	0.9	(0.7-1.0)	.14
PECI emotional resources	12	3.2	(3.0-3.3)	4	3.4	(2.3-4.0)	76	3.1	(2.8-3.4)	8	3.7	(2.6-3.9)	.60
PECI unresolved sorry and anger	**12**	**1.2**^{e}	**(1.0-1.6)**	**4**	**0.8**	**(0.4-1.8)**	**76**	**1.3**	**(0.6-1.7)**	**8**	**0.5** ^{a}	**(0.4-0.6)**	**.02**
PECI uncertainty	**12**	**1.4**^{e}	**(0.8-2.1)**	**4**	**1.7**	**(0.7-2.5)**	**76**	**1.2**^{e}	**(0.8-2.0)**	**8**	**0.6** ^{a,c}	**(0.3-0.7)**	**.02**
PROMIS anxiety t-score	**12**	**51.6**^{e}	**(48.4-60.8)**	**4**	**54.8**	**(45.3-61.5)**	**76**	**52.5**^{e}	**(42.8-58.8)**	**8**	**36.3** ^{a,c}	**(36.3-46.5)**	**.02**
PROMIS depression t-score	12	45.3	(37.1-50.2)	4	46.0	(41.5-54.3)	76	49.4	(37.1-54.5)	8	37.1	(37.1-47.2)	.32
SF-12 Physical functioning	12	100.0	(100.0-100.0)	4	100.0	(87.5-100.0)	76	100.0	(100.0-100.0)	8	100.0	(100.0-100.0)	.99
SF-12 Role-physical	12	100.0	(87.5-100.0)	4	100.0	(75.0-100.0)	76	100.0	(75.0-100.0)	8	100.0	(87.5-100.0)	.99
SF-12 Bodily pain	12	100.0	(87.5-100.0)	4	87.5	(62.5-100.0)	75	100.0	(75.0-100.0)	8	100.0	(62.5-100.0)	.74
SF-12 General health	12	85.0	(60.0-85.0)	4	85.0	(55.5-85.0)	76	85.0	(60.0-85.0)	8	85.0	(60.0-85.0)	.99
SF-12 Energy/fatigue	12	75.0	(50.0-75.0)	2	50.0	(50.0-50.0)	64	75.0	(50.0-75.0)	7	75.0	(75.0-75.0)	.27
SF-12 Social functioning	12	100.0	(87.5-100.0)	4	62.5	(50.0-87.5)	76	100.0	(75.0-100.0)	8	100.0	(100.0-100.0)	.42
SF-12 Role-emotional	12	100.0	(87.5-100.0)	4	56.3	(50.0-81.3)	76	87.5	(75.0-100.0)	8	100.0	(81.3-100.0)	.25
SF-12 Mental health	**12**	**87.5**^{b}	**(87.5-88.6)**	**4**	**56.3** ^{a,c,e}	**(31.3-64.5)**	**76**	**75.0**^{b}	**(52.9-87.5)**	**8**	**87.5**^{b}	**(64.5-93.8)**	**.02**
SF-12 Physical component summary	12	56.4	(51.9-57.1)	4	55.4	(51.7-63.8)	76	56.2	(52.8-58.6)	8	56.0	(51.8-57.4)	.89
SF-12 Mental component summary	**12**	**56.5**^{b}	**(54.1-58.5)**	**4**	**39.9** ^{a,c,e}	**(31.1-46.7)**	**76**	**52.4**^{b}	**(43.6-57.3)**	**8**	**57.5**^{b}	**(49.1-59.7)**	**.02**
PedsQL physical functioning score (infants)	1	75.0	(75.0-75.0)	1	100.0	(100.0-100.0)	23	75.0	(70.8-94.4)	0			.38
PedsQL physical symptoms score (infants)	1	45.0	(45.0-45.0)	1	77.5	(77.5-77.5)	23	82.5	(67.5-87.5)	0			.25
PedsQL physical score (2 y+)	11	90.6	(46.9-93.8)	3	46.9	(40.6-100.0)	53	93.8	(78.1-100.0)	8	95.3	(89.1-100.0)	.13
PedsQL emotional score	12	77.5	(62.5-82.5)	4	53.8	(42.5-76.3)	76	70.0	(55.0-90.0)	8	72.5	(67.5-87.5)	.55
PedsQL social score	12	80.0	(60.0-92.5)	4	92.5	(72.5-100.0)	76	90.0	(67.5-100.0)	8	90.0	(80.0-100.0)	.43
PedsQL school score (2 y+)	10	67.5	(50.0-83.3)	2	72.5	(45.0-100.0)	49	75.0	(55.0-91.7)	8	75.0	(65.8-92.5)	.73
PedsQL cognitive score (infants)	1	50.0	(50.0-50.0)	1	100.0	(100.0-100.0)	23	72.2	(50.0-91.7)	0			.27
PedsQL psychosocial score	12	73.3	(53.3-84.2)	4	67.9	(54.2-87.1)	76	76.7	(61.9-88.5)	8	80.8	(74.2-88.3)	.63
PedsQL physical score	12	84.4	(51.6-93.8)	4	66.4	(43.8-93.0)	76	89.1	(75.0-96.9)	8	95.3	(89.1-100.0)	.10
PedsQL total score	12	70.7	(54.6-88.5)	4	66.7	(50.4-89.5)	76	81.2	(66.5-91.1)	8	86.3	(76.1-92.4)	.28

Different superscript alphabets indicate the significant findings. Letter "e", indicates the the finding is significant for comparison with Type E, and letter "a", indicates the finding is significant for comparison with Type A.
Bold type are significant findings (*P*<.05).

Table VI. Nonparametric comparison (Mann–Whitney *U* test) of outcomes between requirement/no requirement for fundoplication surgery: PECI, PROMIS, and SF-12 for primary caregivers, PedsQL for patients

	Fundoplication surgery not required			Fundoplication surgery required			
Outcomes	**N**	**Median**	**(IQR)**	**n**	**Median**	**(IQR)**	***P***
PECI guilt and worry	89	1.5	(0.9-2.1)	11	1.8	(1.3-1.9)	.42
PECI emotional resources	89	3.2	(2.8-3.4)	11	3.0	(2.6-3.4)	.78
PECI unresolved sorry and anger	89	1.1	(0.6-1.6)	11	1.4	(1.0-1.9)	.12
PECI uncertainty	89	1.2	(0.6-2.0)	11	1.6	(1.0-2.2)	.33
PROMIS anxiety T score	89	50.9	(42.8-58.7)	11	58.8	(51.0-62.3)	.10
PROMIS depression T score	89	46.7	(37.1-54.3)	11	46.8	(44.5-50.2)	.99
SF-12 physical functioning	89	100.0	(100.0-100.0)	11	100.0	(100.0-100.0)	.91
SF-12 role-physical	89	100.0	(87.5-100.0)	11	100.0	(75.0-100.0)	.45
SF-12 bodily pain	88	100.0	(75.0-100.0)	11	75.0	(75.0-100.0)	.21
SF-12 general health	**89**	**85.0**	**(60.0-85.0)**	**11**	**60.0**	**(25.0-85.0)**	**.03**
SF-12 energy/fatigue	78	75.0	(50.0-75.0)	7	75.0	(50.0-75.0)	.73
SF-12 social functioning	89	100.0	(75.0-100.0)	11	100.0	(50.0-100.0)	.58
SF-12 role-emotional	89	100.0	(75.0-100.0)	11	75.0	(62.5-100.0)	.17
SF-12 mental health	89	87.5	(62.5-87.5)	11	62.5	(45.0-75.0)	.12
SF-12 physical component summary	89	56.4	(53.9-58.4)	11	53.3	(47.3-59.7)	+++.24
SF-12 mental component summary	89	53.4	(44.1-57.4)	11	48.4	(39.0-52.3)	.18
PedsQL physical functioning score (infants)	25	75.0	(70.8-94.4)	0			
PedsQL physical symptoms score (infants)	25	80.0	(67.5-87.5)	0			
PedsQL physical score (2 y+)	**64**	**93.8**	**(78.1-100.0)**	**11**	**78.1**	**(56.3-93.8)**	**.02**
PedsQL emotional score	89	70.0	(56.3-90.0)	11	60.0	(50.0-90.0)	.41
PedsQL social score	**89**	**90.0**	**(80.0-100.0)**	**11**	**60.0**	**(50.0-95.0)**	**.04**
PedsQL school score (2 y+)	58	75.0	(58.3-91.7)	11	60.0	(50.0-80.0)	.20
PedsQL cognitive score (infants)	25	72.2	(50.0-91.7)	0			
PedsQL psychosocial score	89	76.9	(63.8-88.3)	11	56.7	(50.0-88.3)	0.11
PedsQL physical score	89	90.6	(75.0-96.9)	11	78.1	(56.3-93.8)	0.08
PedsQL total score	89	81.5	(67.4-90.5)	11	59.8	(56.5-88.1)	0.06

Bold type indicates statistical significance.

Table VII. Nonparametric correlation between child age and outcomes: PECI, PROMIS, and SF-12 for primary caregivers and PedsQL for patients

	Age			
Outcomes	**n**	***r***	**(95% CI)**	***P***
PECI guilt and worry	100	−0.11	(−0.31 to 0.09)	.28
PECI emotional resources	100	0.17	(−0.04 to 0.35)	.09
PECI unresolved sorry and anger	100	−0.09	(−0.28 to 0.12)	.38
PECI uncertainty	100	0.08	(−0.10 to 0.27)	.41
PROMIS anxiety T score	100	−0.04	(−0.23 to 0.17)	.73
PROMIS depression T score	100	−0.12	(−0.32 to 0.08)	.23
SF-12 physical functioning	100	0.00	(−0.23 to 0.21)	.97
SF-12 role–physical	100	−0.09	(−0.30 to 0.12)	.36
SF-12 bodily pain	99	−0.16	(−0.34 to 0.05)	.13
SF-12 general health	100	−0.15	(−0.34 to 0.04)	.14
SF-12 energy/fatigue	85	0.10	(−0.13 to 0.32)	.36
SF-12 social functioning	100	−0.03	(−0.25 to 0.18)	.75
SF-12 role– emotional	100	−0.09	(−0.27 to 0.14)	.38
SF-12 mental health	100	−0.02	(−0.22 to 0.18)	.81
SF-12 physical component summary	100	−0.15	(−0.35 to 0.05)	.15
SF-12 mental component summary	100	0.00	(−0.21 to 0.20)	1.00
PedsQL physical functioning score (infants)	25	0.35	(−0.11 to 0.66)	.09
PedsQL physical symptoms score (infants)	25	0.26	(−0.19 to 0.60)	.21
PedsQL physical score (2 y+)	**75**	**−0.31**	**(−0.50 to −0.08)**	**.01**
PedsQL emotional score	100	0.07	(−0.13 to 0.26)	.51
PedsQL social score	100	−0.16	(−0.37 to 0.04)	.11
PedsQL school score (2 y+)	**69**	**−0.40**	**(−0.61 to −0.13)**	**.001**
PedsQL cognitive score (infants)	25	0.13	(−0.36 to 0.55)	.54
PedsQL psychosocial score	100	−0.04	(−0.25 to 0.18)	.68
PedsQL physical score	100	0.04	(−0.17 to 0.23)	.68
PedsQL total score	100	−0.05	(−0.24 to 0.17)	.61

Bold values indicate statistical significance.

significant differences were found when comparing for VACTERL association (Table IV), esophageal dilatation (Table V), previous fundoplication (Table VI), or age of the child (Table VII).

Patient-Reported Outcomes Measurement Information System: Depression (Assessing Primary Caregivers)

The overall median PROMIS Depression T score was 46.8 (IQR, 37.1-53.2). This is interpreted as a "none to slight" level of depression (<55).

No significant differences were found when comparing for esophageal atresia type (Table III), VACTERL association (Table IV), esophageal dilatation (Table V), previous fundoplication (Table VI), or age of the child (Table VII).

SF-12 (Assessing Primary Caregivers)

The median physical component summary score was 56.2 (IQR, 52.3-58.4), and the median mental component summary score was 52.8 (IQR, 44.1-57.4). This is interpreted as same or better than the general population (50 is average score or norm).

When stratified by esophageal atresia type, significant differences were found in the mental health score (*P* = .02) and mental component summary (*P* = .02) (Table III) for the primary caregivers of children with type B esophageal atresia, lower scores (worse) compared with those of children with other types of esophageal atresia. Previous fundoplication resulted in a lower general health score

(Table VI). No significant differences were found when comparing for VACTERL association (Table IV), esophageal dilatation (Table V), or age of the child (Table VII).

PedsQL Inventory (Assessing Patients)

The overall median total score for the entire patient cohort was 81.1 (IQR, 65.80-90.2). The overall median psychosocial score for patients was 76.7 (IQR, 60.9-88.3), and the overall median physical score for patients was 90.6 (IQR, 71.9-96.9). For comparison with previous publications, the overall mean total score for patients was 77.3 ± 16.0. The overall mean psychosocial score for patients was 74.0 ± 16.9, and the overall mean physical score for patients was 82.4 ± 18.4. These values are presented in Table VIII for comparison with the recommended cutoff scores that identify children with special health care needs (categorized as <8 years and ≥8 years).[17]

No significant differences were found when comparing for esophageal atresia type (Table III), VACTERL association (Table IV), or esophageal dilatation (Table V). Previous fundoplication resulted in a lower PedsQL social functioning score, as well as a lower physical functioning score in children aged ≥2 years (Table VI). When compared for age, significant correlations were found between age and physical and school functioning scores in children aged ≥2 years (Table VII and Figure 2 [available at www.jpeds.com]).

Discussion

The findings in this study are consistent with those reported by Witt et al in a cohort of 49 families recruited from 2 centers in Germany, in which parents of children with esophageal atresia were found to have lower mental health scores compared with the general population, representing an emotional burden that highlights the need for psychosocial support.[18] In another study examining posttraumatic stress reactions in parents of children with esophageal atresia, 59% had posttraumatic stress disorder, and 8% had severe anxiety.[19] Kumari et al reported that in mothers of children with esophageal atresia, 47% had moderate stress and relatively poor QoL in environmental and psychological domains, as assessed with the Parental Stress Scale and World Health Organization Quality of Life: Brief Version questionnaire.[20]

Table VIII. PedsQL scores of patients with esophageal atresia and recommended cutoff scores to identify children with special health care needs (patient scores)[17]

Patients	Esophageal atresia patients, mean (SD)	Recommended cutoff scores
All patients		
Total score	77.3 (16.0)	
Psychosocial score	74.0 (16.9)	
Physical score	82.4 (18.4)	
Children <8 y		83 (moderate, 79; major chronic conditions, 77)
Total score	79.1 (14.8)	
Children ≥8 y		78 (moderate, 76; major chronic conditions, 70)
Total score	67.4 (19.1)	

We have demonstrated that unresolved sorrow and anger, uncertainty, anxiety, and lower mental health scores were prominent in our cohort of primary caregivers. There are several possible explanations for these findings. In the case of unresolved sorrow and anger, this possibly could be related to the initial diagnosis of a newborn with esophageal atresia. This diagnosis is typically unexpected for parents, because the majority of children are not detected prenatally.[21] In the case of uncertainty, this possibly may be related to the morbidity associated with esophageal atresia and the fact that predicting the development of a stricture or dysmotility may be challenging at times, and may be associated with highly stressful choking episodes in the child.[5] Although the PECI is based on thoughts and feelings over the past month, the PROMIS Anxiety questionnaires are based on the past 7 days. As such, this could be affected by any acute event, which would have measured differently compared with years after a particular event. Similarly, mental health scores are related to these issues and are likely to be more prominent in caregivers of children with type A or type B esophageal atresia, who may have more significant symptoms. This is due to the longer gap between the 2 ends of the esophagus that needs to be repaired in these esophageal atresia types.

Furthermore, we have found differences in the QoL outcomes for the primary caregivers of children with different esophageal atresia types. As we have designated type E esophageal atresia as proxy controls, we are in fact demonstrating a difference in the QoL for the primary caregivers based on the presence of an atresia and anastomosis of the esophagus, given the fact that type E is an isolated TEF. We also found that having a VACTERL association did not have an impact on the QoL outcomes. These findings are consistent with those of Flieder et al, who previously demonstrated that differences in QoL in patients with esophageal atresia were determined by the type and severity of atresia, rather than on the age or presence of associated malformations.[22] However, Rozensztrauch et al found that the presence of associated malformations, specifically skeletal impairment, had an impact on family functioning.[23] Statistically significant correlations were found in the social, cognitive, and communication spheres using the PedsQL Family Impact Module. This difference from our findings is likely related to the different domains assessed by different questionnaires. The lack of impact of a VACTERL association may be explained by the fact that esophageal morbidity has a regular impact on mealtimes. Therefore, this should be at the forefront of caregivers' minds, especially with regard to choking risk. In comparison, the impact of other aspects of a VACTERL association may be less dramatic, despite the need for long-term care.

In terms of patient outcomes, we have found that PedsQL physical score for children aged ≥2 years, as well as social score, were significantly different for the need for fundoplication surgery as well as for age. Regarding the fundoplication requirement, this may be related to these patients' more severe reflux, which consequently impacts physical functioning. It also has an impact on missing school for medical appointments, which may impact school functioning. This certainly is just one of many plausible explanations, given the multifactorial contribution to patient outcomes, and the severity of reflux may be just one of many associations. The need for fundoplication also very well may be a surrogate for the severity of esophageal atresia morbidity in the patients, and the impact might not be related to the need for fundoplication alone. In addition, the older the child, the more likely this also may impact functioning as the comparisons with their peers become starker. As reported in the literature, physical scores also tend to be lower in patients with esophageal atresia.[24]

Notably, the PedsQL has been used to assess outcomes in children with other congenital conditions, such as congenital diaphragmatic hernia.[25,26] Morseberger et al found that older age was correlated with poor school function in these patients, not unlike our findings on the impact of age on scores.[25]

A significant strength of the present study is the larger number of primary caregivers of children with esophageal atresia recruited compared with previous studies. Moreover our cohort was recruited from a single pediatric center, minimizing any potential biases that could arise from differing parental experiences owing to varying institutional practices. In addition, our cohort includes a wide age range of patients (3.5 months to 19 years), which is more inclusive than the cohorts included in previous studies.

Our present findings are important, as they may be used to guide the counseling of the primary caregivers of children with esophageal atresia, including identifying and preventing mental health issues in their primary caregivers. It is important that psychosocial supports are provided for primary caregivers from the earliest stages to ameliorate the significant impacts on caregiver QoL.

We recommend considering the use of these questionnaires to assess caregiver QoL and identify needs for counseling. Over time, the early and timely identification of the impact on primary caregivers may lead to an improvement in QoL. In addition, there is scope for further studies investigating any change in questionnaire scores when interventions of psychosocial supports are instituted between administrations of questionnaires.

Most of the participants were mothers, which limited our ability to compare findings against other caregiver types, such as fathers. Our PedsQL questionnaires were also completed as parent-proxy responses, which also could have influenced the responses. This may underestimate or overestimate the impact of QoL. Older children (aged ≥13 years, of whom there were 7 in this cohort) were encouraged to provide responses with their primary caregiver completing the questionnaires, and younger children required their primary caregiver to complete the questionnaires on their behalf. Witt et al previously demonstrated that mothers' and fathers' responses tend to correspond with each other, but that mother–child and father–child agreement may show differences.[27] Similarly, in other pediatric studies, parent-proxy responses were used for younger children. Nevertheless, the recruitment of patients with this wide age range, is a strength of this study.

Although recruiting from a single center afforded a more consistent clinical practice, it also limits our findings in terms of generalizability. The impact on quality of life also may be influenced by other factors in the patients' and parents' healthcare experience, which may differ from the cohort at our institution. Therefore, our findings might not consider the impact of this on QoL.

In conclusion, this study shows that the presence of an atresia (and thus anastomosis) of the esophagus had a significant impact on long-term QoL of the primary caregivers of children with esophageal atresia, and that a previous requirement for fundoplication and age had significant impacts on the QoL of the affected children. ■

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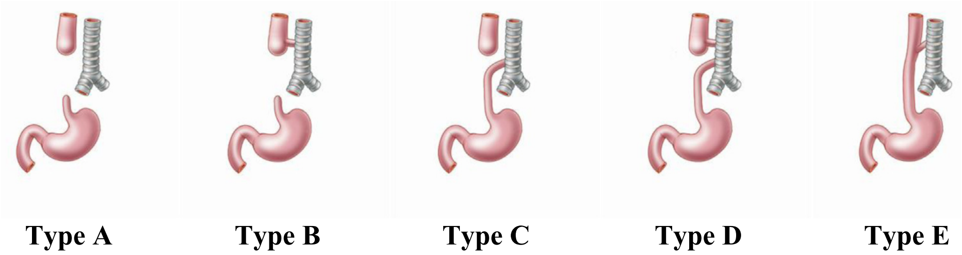

Figure 1. Types of esophageal atresia by the Gross classification scheme. (Illustrations © The Royal Children's Hospital, Melbourne. Reproduced with permission from The Royal Children's Hospital, Melbourne.)

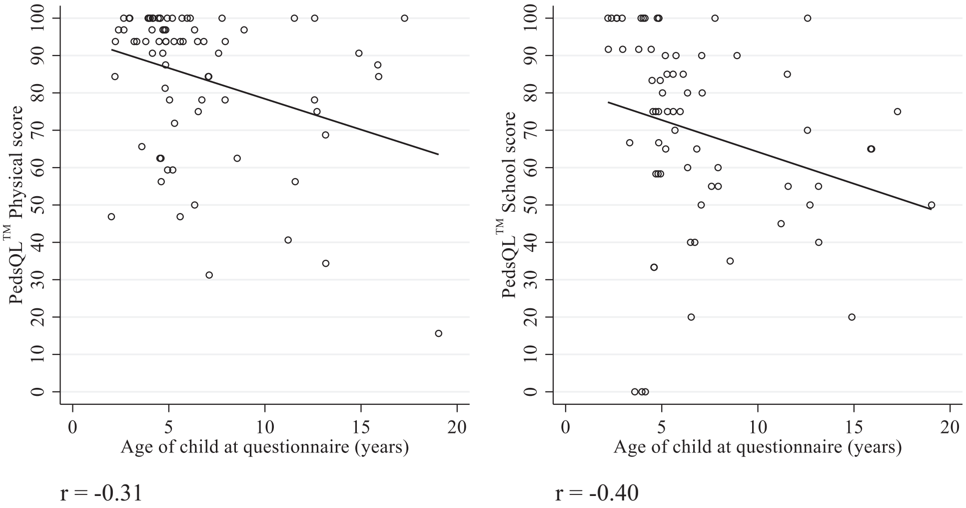

Figure 2. Illustration of statistically significant correlations for age and PedsQL scores (patient scores).

Table I. Patient demographics (n = 100)

Characteristics	Value
Male sex, n	64
Gestational age, wk, median (range)	$37^{4/7}$ ($30^{1/7}$-$41^{5/7}$)
Birth weight, kg, median (range)	2.9 (1.2-4.5)
Age at questionnaire, median (range)	4.6 y (3.5 mo-19.0 y)
Esophageal atresia type, n	
Type A	12
Type B	4
Type C	76
Type D	0
Type E	8, including 1 double-TEF
VACTERL, n	30
Requirement for ≥1 esophageal dilatation, n	57
Requirement for fundoplication surgery, n	11

Table II. PECI scores of patients with esophageal atresia and patients with brain tumor[13]

PECI subscales	Esophageal atresia: Median (IQR)	Esophageal atresia: Mean (SD)	Brain tumor, mean (SD)
Guilt and worry	1.5 (0.9-2.0)	1.5 (0.8)	1.7 (0.8)
Emotional resources	3.2 (2.8-3.4)	3.1 (0.5)	2.7 (0.7)
Unresolved sorrow and anger	1.1 (0.6-1.6)	1.2 (0.7)	1.5 (0.8)
Uncertainty	1.2 (0.6-2.0)	1.3 (0.8)	2.0 (0.9)

Scores are based on items ranging from 0, “never,” to 4, “always.” Therefore, lower scores are interpreted as better in the subscales of guilt and worry, unresolved sorrow and anger, and uncertainty; and higher scores are interpreted as better in the subscale of emotional resources.

Table IV. Nonparametric comparison (Mann–Whitney *U* test) of outcomes between presence/absence of VACTERL association in the patient: PECI, PROMIS, and SF-12 for primary caregivers and PedsQL for patients

	No VACTERL association			VACTERL association			
	n	Median	(IQR)	n	Median	(IQR)	*P*
PECI guilt and worry	70	1.4	(1.0-1.8)	30	1.8	(0.9-2.3)	.11
PECI emotional resources	70	3.2	(2.6-3.4)	30	3.2	(2.8-3.4)	.55
PECI unresolved sorry and anger	70	1.1	(0.6-1.6)	30	1.4	(0.8-1.9)	.19
PECI uncertainty	70	1.1	(0.6-2.0)	30	1.6	(0.8-2.2)	.09
PROMIS anxiety t-score	70	51.6	(47.0-58.8)	30	48.5	(36.3-59.4)	.47
PROMIS depression t-score	70	46.3	(37.1-52.9)	30	49.3	(37.1-54.3)	.94
SF-12 physical functioning	70	100.0	(100.0-100.0)	30	100.0	(100.0-100.0)	.59
SF-12 role–physical	70	100.0	(75.0-100.0)	30	100.0	(75.0-100.0)	.96
SF-12 bodily pain	69	100.0	(75.0-100.0)	30	100.0	(75.0-100.0)	.86
SF-12 general health	70	85.0	(60.0-85.0)	30	85.0	(60.0-85.0)	.45
SF-12 energy/fatigue	57	75.0	(50.0-75.0)	28	75.0	(50.0-75.0)	.20
SF-12 social functioning	70	100.0	(75.0-100.0)	30	100.0	(75.0-100.0)	.80
SF-12 role–emotional	70	93.8	(75.0-100.0)	30	93.8	(75.0-100.0)	.54
SF-12 mental health	70	75.0	(62.5-87.5)	30	81.3	(50.0-87.5)	.62
SF-12 physical component summary	70	56.1	(51.8-58.4)	30	56.4	(54.4-58.6)	.92
SF-12 mental component summary	70	52.8	(46.2-57.4)	30	52.8	(42.2-57.5)	.72
PedsQL physical functioning score (infants)	15	72.2	(70.8-100.0)	10	78.5	(75.0-94.4)	.52
PedsQL physical symptoms score (infants)	15	77.5	(65.0-85.0)	10	82.5	(72.5-87.5)	.33
PedsQL physical score (2 y+)	55	93.8	(78.1-96.9)	20	93.8	(75.0-100.0)	.36
PedsQL emotional score	70	65.8	(55.0-83.3)	30	72.9	(65.0-90.0)	.12
PedsQL social score	70	86.3	(70.0-100.0)	30	93.8	(65.0-100.0)	.57
PedsQL school score (2 y+)	53	70.0	(55.0-90.0)	16	77.5	(60.0-95.8)	.37
PedsQL cognitive score (infants)	15	72.2	(47.2-97.2)	10	71.9	(56.3-91.7)	.76
PedsQL psychosocial score	70	76.7	(58.3-84.6)	30	81.2	(63.8-90.4)	.32
PedsQL physical score	70	89.1	(68.8-96.9)	30	91.4	(75.0-100.0)	.45
PedsQL total score	70	79.4	(65.0-89.1)	30	86.5	(66.3-92.2)	.34

Table V. Nonparametric comparison (Mann-Whitney *U* test) of outcomes between requirement/no requirement for any esophageal dilatation: PECI, PROMIS, SF-12 for primary caregivers and PedsQL for patients

	Esophageal dilatation requirement			No esophageal dilatation requirement			
Outcomes	n	Median	(IQR)	n	Median	(IQR)	*P*
PECI guilt and worry	43	1.5	(0.9-2.0)	57	1.6	(1.1-2.1)	.39
PECI emotional resources	43	3.2	(2.8-3.6)	57	3.0	(2.8-3.4)	.43
PECI unresolved sorry and anger	43	0.8	(0.5-1.6)	57	1.3	(1.0-1.6)	.06
PECI uncertainty	43	1.0	(0.6-1.8)	57	1.4	(0.8-2.0)	.23
PROMIS anxiety T score	43	49.8	(42.1-57.6)	57	51.6	(47.3-60.1)	.30
PROMIS depression T score	43	48.0	(37.1-52.9)	57	46.7	(37.1-53.3)	.67
SF-12 physical functioning	43	100.0	(100.0-100.0)	57	100.0	(100.0-100.0)	.78
SF-12 role - physical	43	100.0	(75.0-100.0)	57	100.0	(75.0-100.0)	.93
SF-12 bodily pain	43	100.0	(100.0-100.0)	56	100.0	(75.0-100.0)	.45
SF-12 general health	43	85.0	(60.0-85.0)	57	85.0	(60.0-85.0)	.47
SF-12 energy/fatigue	36	75.0	(50.0-75.0)	49	75.0	(50.0-75.0)	.84
SF-12 social functioning	43	100.0	(100.0-100.0)	57	100.0	(75.0-100.0)	.30
SF-12 role - emotional	43	100.0	(75.0-100.0)	57	87.5	(75.0-100.0)	.16
SF-12 mental health	43	75.0	(62.5-87.5)	57	75.0	(50.0-87.5)	.77
SF-12 physical component summary	43	56.1	(53.9-58.6)	57	56.4	(51.4-58.4)	.95
SF-12 mental component summary	43	53.3	(47.5-57.4)	57	52.8	(43.1-57.4)	.49
PedsQL physical functioning score (infants)	16	75.0	(68.8-97.2)	9	77.8	(72.2-83.3)	.71
PedsQL physical symptoms score (infants)	16	76.3	(66.3-86.3)	9	82.5	(72.5-90.0)	.53
PedsQL physical score (2 y+)	27	93.8	(71.9-100.0)	48	93.8	(76.6-96.9)	.53
PedsQL emotional score	43	70.0	(56.3-90.0)	57	70.0	(55.0-90.0)	.71
PedsQL social score	43	93.8	(70.0-100.0)	57	90.0	(65.0-100.0)	.51
PedsQL school score (2 y+)	25	75.0	(65.0-90.0)	44	68.3	(50.0-90.8)	.26
PedsQL cognitive score (infants)	16	70.8	(57.3-100.0)	9	75.0	(47.2-83.3)	.45
PedsQL psychosocial score	43	78.8	(64.4-85.0)	57	76.7	(56.7-88.5)	.50
PedsQL physical score	43	90.6	(68.4-96.9)	57	87.5	(77.6-96.9)	.85
PedsQL total score	43	82.1	(66.7-90.2)	57	79.3	(64.1-90.2)	.70