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Health-related quality of life of children treated for non-syndromic craniosynostosis

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ABSTRACT

Health-related quality of life (HRQoL) allows the acquisition of the subjective perspective of patients regarding their health and function; yet a very few studies have been evaluated HRQoL of patients treated for craniosynostosis (CS). In this retrospective, descriptive cohort study, school-aged children (7–16 years) treated for non-syndromic CS were assessed using the Pediatric Quality of Life Inventory (PedsQL) 4.0 Generic Core Scales. Seventy-three patients and their parents responded to the PedsQL (response rate: 80.2%). Patients generally estimated average HRQoL with no difference compared to the normal population sample. Further, no difference in HRQoL was found between treated sagittal (SS) or metopic synostosis. In the SS group, surgical methods involving spring-assisted surgery and pi-plasty were unrelated to HRQoL outcomes. Additionally, HRQoL was highly correlated with intelligence quotient (IQ, $r = 0.42$; $p = 0.0004$) and adaptive behavior skills (ABAS, $r = 0.57$; $p = 0.0001$). Furthermore, differences were observed in estimated physical function ($p = 0.002$) and school function ($p = 0.012$) between self- and proxy reports (*i.e.* parents estimated child HRQoL as higher than did the children). Children treated for CS have a generally average HRQoL, and neither CS type nor surgical method influenced HRQoL outcomes. Moreover, children and parents estimated HRQoL differently, suggesting the importance of using both self- and proxy reporting in patient-reported measures. HRQoL was strongly related to IQ and ABAS, indicating that the PedsQL can be used as a screening instrument to identify craniofacial patients in need of further psychological assessment.

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Introduction

Treatment outcomes for non-syndromic craniosynostosis (CS) have been studied from multiple perspectives, with numerous previous studies assessing cognitive, behavioral, and emotional function and the impact of surgical methods or timing on development [1–17]. However, little is known about the relationship between CS and health-related quality of life HRQoL, despite previous claims that QoL has been included as part of psychosocial assessment. In fact, few previous studies have used validated instruments for the purpose of measuring the HRQoL of treated CS patients [18–20]. Previous studies report a high QoL in adult patients [18], whereas in children treated for non-syndromic CS an elevated risk of lower HRQoL was revealed [19]. Additionally, children with untreated sagittal synostosis (SS) score low in terms of positive emotions [20].

In recent years, HRQoL has become increasingly clinically relevant as a tool for providing patient perspectives regarding their health; however, the relationship between health and HRQoL is not always consistent, as having a disease does not always negatively impact perceived QoL, and health does not always guarantee a better QoL [21].

The definition of HRQoL is multifaceted, with several definitions existing in the literature. The World Health Organization's definition of QoL is described as the perception by an individual of their position in life regarding their physical health, psychological state, level of independence, and social relationships [22]. HRQoL represents a multidimensional concept that also includes health status and its impact on well-being or QoL. The Pediatric Quality of Life Inventory (PedsQL) was developed to measure HRQoL associated with pediatric health care by James Varni in the late 1990s and covers important domains, including physical, psychological, and social function, with the recent addition of school function [23–25].

The aim of this study was to evaluate the HRQoL of children treated for non-syndromic CS and using the Swedish version of the validated PedsQL 4.0 Generic Core Scales. The rational for the study was to contribute to fill the gap in knowledge about HRQoL in children treated for non-syndromic CS.

Patients and methods

Patients and procedures

In this retrospective, descriptive cohort study, children (aged 7–16 years) treated for non-syndromic CS and living in the regions of Västra Götaland and Halland in Sweden were included in this study. The exclusion criteria were language barriers and secondary CS. The patients were initially informed of the study by letter, and after 1 week, received a telephone call with additional information

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about the study and invited to participate by scheduling a visit to the clinic. Background data were extracted from the Gothenburg Craniofacial Registry and previous studies of the same patient cohort [8,26].

Measurements using PedsQL 4.0 generic score scales (swedish version)

PedsQL is a modular questionnaire that measures the HRQoL of children ages 2 to 18 years. The questionnaire comprises 23 items measuring four dimensions of HRQoL: physical (8 items), emotional (5 items), social (5 items), and school function (5 items). PedsQL comprises both self- and proxy reports and has four age- and language-appropriate versions (2–4 years, 5–7 years, 8–12 years, and 13–18 years).

The questionnaire asks about difficulties with functions during the previous month. The response alternatives are presented on a 5-point scale: 0, never a problem; 1, almost never a problem; 2, sometimes a problem; 3, often a problem; and 4, almost always a problem. The response alternatives are converted to an overall scale (0–100 points) and reversed, so that higher points indicate better HRQoL.

The Swedish version of PedsQL has been translated and validated as demonstrating good psychometric properties [27]. Additionally, the instrument has been evaluated in large groups of Swedish school children ($n = 1455$) [28]. In this study, used both self- and proxy reports were used, and children and parents responded separately.

Statistical analysis

Comparison of background variables between two groups was conducted using Fisher's exact test for dichotomous variables, a Mann–Whitney *U* test for continuous variables, and a chi-squared test for unordered categorial variables. Comparisons with normative sample were analyzed with Fisher's non-parametric one-sample permutation test on the z-scores, (value-mean norm/SD norm). Comparison between self- *vs.* proxy reports were analyzed using Fisher's non-parametric permutation test for matched pairs. For the paired analysis between self- vs. proxy reports both effect size (mean difference/SD for the child) and standardized response mean, SRM, (mean difference/SD for the difference). Between-group comparisons were performed using Fisher's non-parametric permutation test for continuous variables. Adjustment for confounders was performed by analyses of covariance (ANCOVA). Calculation of 95% confidence intervals for mean differences was based on using Fisher's non-parametric permutation test between groups. The between-group effect size, ES, is defined as mean difference/pooled SD. The relationship between HRQoL (PedsQL), adaptive behavior skills (ABAS), and intelligence quotient (IQ) was analyzed by Spearman's correlation coefficient (r_s). All significance tests were two-sided, with a $p < 0.05$ considered significant. All statistical calculations were performed using SAS (v.9.4; SAS Institute, Cary, NC, USA).

Ethics

The study was approved by the Gothenburg Ethics Committee (no. 856-13) and conducted according to principles in the Declaration of Helsinki.

Results

Attrition analysis

Attrition analysis revealed no significant differences between the responding ($n = 73$) and non-responding ($n = 18$) groups regarding background variables, including gender, age at study, CS type, surgical method, age at surgery, and premature birth (Table 1).

Participants

A total of 73 patients (24 females and 49 males; response rate: 80.2%) and their parents participated in the study. The mean age

Table 1. Attrition analysis.

Variables	Non-participating ($n = 18$)	Participating ($n = 73$)	*P*
Sex			
Female	6 (33.3%)	24 (32.9%)	
Male	12 (66.7%)	49 (67.1%)	1.00
Age at study	11.7 (2.2)	11.0 (2.4)	0.23
	12 (8; 15)	11 (7; 15)	
	$n = 18$	$n = 73$	
Age at surgery (days)	277.1 (338.6)	237.0 (224.7)	0.74
	159.5 (94; 1523)	165 (84; 1484)	
	$n = 18$	$n = 73$	
Craniosynostosis			
Sagittal	9 (50.0%)	41 (56.2%)	
Metopic	6 (33.3%)	24 (32.9%)	
Unicoronal	1 (5.6%)	6 (8.2%)	
Bicoronal	1 (5.6%)	1 (1.4%)	
Lambdoid	1 (5.6%)	1 (1.4%)	0.64
Surgery method			
Pi-plasty	1 (5.6%)	17 (23.3%)	
Spring-assisted surgery	8 (44.4%)	23 (31.5%)	
Fronto-orbital reshaping with bone graft	6 (33.3%)	14 (19.2%)	
Fronto-orbital reshaping with spring	3 (16.7%)	18 (24.7%)	
Barrel-stave osteotomy	0 (0.0%)	1 (1.4%)	0.29
Born premature			
Yes	3 (17.6%)	5 (6.8%)	
No	14 (82.4%)	68 (93.2%)	0.34

For categorical variables, *n* (%) is presented.
For continuous variables, mean (SD) / median (min; max) / *n* is presented.
For comparison between groups, Fisher's Exact test (lowest one-sided.
p-value multiplied by 2) was used for dichotomous variables, a chi-squared test was used for non-ordered categorical variables, and the Mann–Whitney *U* test was used for continuous variables.

Table 2. Demographics and baseline characteristics.

Variables	Total ($n = 73$)
Sex	
Female	24 (32.9%)
Male	49 (67.1%)
Age at study	11.0 (2.4)
	11 (7; 15)
	$n = 73$
Craniosynostosis	
Sagittal	41 (56.2%)
Metopic	24 (32.9%)
Unicoronal	6 (8.2%)
Bicoronal	1 (1.4%)
Lambdoid	1 (1.4%)
Surgery method	
Pi-plasty	17 (23.3%)
Spring-assisted surgery	23 (31.5%)
Fronto-orbital reshaping with bone graft	14 (19.2%)
Fronto-orbital reshaping with spring	18 (24.7%)
Barrel-stave osteotomy	1 (1.4%)
Age at surgery (days)	237.0 (224.7)
	165 (84; 1484)
	$n = 73$
Involved parent	
Mother	36 (50.0%)
Father	12 (16.7%)
Both	24 (33.3%)
Maternal education level	
Primary school	2 (3.4%)
High school	21 (35.6%)
College/university	36 (61.0%)
Paternal education level	
Primary school	2 (5.7%)
High school	19 (54.3%)
College/university	14 (40.0%)
Born premature	
Yes	5 (6.8%)
No	68 (93.2%)
Did the child have other diseases/diagnosis?	
Yes	22 (30.6%)
No	50 (69.4%)
What kind of condition?	
Heart disease	4 (23.5%)
Neuropsychiatric diagnosis	5 (29.4%)
Asthma/allergy	4 (23.5%)
Migraine	3 (17.6%)
Metabolic disease	1 (5.9%)
Has your child been assessed by a psychologist?	
Yes	14 (19.2%)
No	59 (80.8%)
What did the psychological assessment show?	
Autism	4 (30.8%)
Problem with executive function	1 (7.7%)
ADHD	2 (15.4%)
Problem with attention	2 (15.4%)
Both autism and ADHD	2 (15.4%)
Development delay	2 (15.4%)
Is your child under medical treatment?	
Yes	8 (11.4%)
No	62 (88.6%)
Right- or left-handed?	
Right	62 (87.3%)
Left	9 (12.7%)
Wechsler full scale intelligence quotient	98.1 (14.6)
	100 (59; 135)
	$n = 72$
Wechsler verbal comprehension intelligence quotient	99.5 (14.6)
	100 (57; 130)
	$n = 72$
Wechsler perceptual reasoning intelligence quotient	104.5 (13.7)
	102 (69; 135)
	$n = 72$
Wechsler working memory intelligence quotient	94.9 (13.0)
	97 (56; 120)
	$n = 71$
Wechsler processing speed intelligence quotient	92.1 (14.4)
	91 (53; 126)
	$n = 71$
Adaptive behaviour skills full scale	94.4 (20.2)
	97.5 (47; 120)
	$n = 72$
Conceptual composite scale	93.9 (20.1)
	95.5 (40; 119)
	$n = 72$
Social composite scale	93.4 (19.6)
	97 (42; 118)
	$n = 72$
Practical composite scale	94.6 (19.8)
	97 (55; 120)
	$n = 72$

For categorical variables, n (%) is presented.
For continuous variables, mean (SD) / median (min; max) / n is presented.

at study was 11.0 ± 2.4 years (range: 7–15 years), and all patients were treated for non-syndromic CS [SS, $n = 41$; metopic (MS), $n = 24$; and other, $n = 8$]. Mean age at surgery was 237.0 days, and the mean IQ (98.1) and ABAS (94.4) were within average ranges (Table 2).

Health-related QoL of patients treated for CS relative to normative sample

The HRQoL of treated CS patients was estimated as generally average, with no differences found on a full-scale level relative to normative sample. At a detailed level, the SS group reported significantly lower school function ($p = 0.002$) and psychosocial function ($p = 0.031$) as compared with normative sample. The MS group reported an average HRQoL, with no significant difference relative to normative sample (Table 3).

The impact of CS type

Background data variables 'age at study' and 'working memory IQ' differed between the SS and MS groups, with this adjusted for prior to the analysis. No differences were found in HRQoL outcomes between the SS and MS groups (Table 4).

The impact of surgical method used to treat SS

Comparisons of SS treated with spring-assisted surgery and pi-plasty revealed no significant differences, indicating that the effect of the surgical method used to treat SS was unrelated to HRQoL outcomes (Table 5).

HRQoL correlates with IQ and ABAS

HRQoL significantly correlated with ABAS ($r = 0.57$; $p = 0.0001$) and IQ ($r = 0.42$; $p = 0.0004$) (Table 6).

Conformity of self- and proxy reports

The conformity of self- and proxy reports was strong in regard to emotional function; however, significant differences were found in estimated physical function ($p = 0.002$) and school function ($p = 0.012$), for which the parents estimated better functions than the children (Table 7).

Table 3. Child-rated PedsQL generic core scales relative to normative sample.

Variables	Craniosynostosis ($n = 73$) Mean (SD) Median (Min; Max) n	p (within group)	Sagittal ($n = 41$) Mean (SD) Median (Min; Max) n	P (within group)	Metopic ($n = 24$) Mean (SD) Median (Min; Max) n	p (within group)
HRQoL	−0.18 (1.14)	0.20	−0.26 (1.11)	0.14	−0.24 (1.26)	0.45
PedsQL Full Scale	−0.04 (−4.57; 1.24) $n = 67$		−0.09 (−3.49; 1.04) $n = 40$		−0.04 (−4.57; 1.24) $n = 21$	
Physical Function Scale	0.10 (0.91)	0.36	0.09 (0.87)	0.54	−0.02 (1.02)	0.94
	0.14 (−3.80; 1.07) $n = 69$		0.14 (−1.95; 1.07) $n = 40$		0.14 (−3.80; 1.07) $n = 23$	
Emotional Function Scale	−0.06 (1.00)	0.61	−0.10 (0.97)	0.51	−0.09 (1.04)	0.69
	0.09 (−3.00; 1.12) $n = 69$		0.26 (−3.00; 1.12) $n = 40$		0.09 (−3.00; 1.12) $n = 23$	
Social Function Scale	−0.27 (1.31)	0.082	−0.37 (1.35)	0.085	−0.30 (1.36)	0.34
	0.28 (−5.12; 0.70) $n = 69$		0.28 (−5.12; 0.70) $n = 40$		0.28 (−4.29; 0.70) $n = 23$	
School Function Scale	−0.51 (1.32)	0.0014	−0.68 (1.37)	0.0018	−0.43 (1.31)	0.14
	−0.34 (−4.05; 1.14) $n = 67$		−0.34 (−4.05; 1.14) $n = 40$		−0.34 (−4.05; 1.14) $n = 23$	
Psychosocial Function Scale	−0.33 (1.28)	0.035	−0.45 (1.28)	0.031	−0.32 (1.37)	0.32
	0.00 (−4.50; 1.20) $n = 67$		−0.23 (−4.35; 1.20) $n = 40$		−0.15 (−4.50; 1.20) $n = 23$	

For continuous variables, mean (SD) / median (min; max) / n is presented.
For comparison within groups, Fisher's non-parametric permutation test for matched pairs was used.
For each variable, the z-score was calculated as (PedsQL − normal mean) / normal SD.

Table 4. Difference in the mean of child- and parent-rated PedsQL generic core scales between treated sagittal and metopic CS patients.

Variables	Sagittal ($n = 41$)	Metopic ($n = 24$)	p	Adjusted p*	Mean difference with 95% CI[a]	**Effect size**[b]
HRQoL	85.4 (9.8)	85.8 (11.2)	0.90		−0.41 (−6.09; 5.05)	0.040
PedsQL Full Scale	86.4 (53.3; 98.4) $n = 39$	87.5 (52.2; 97.8) $n = 21$				
Physical Function Scale	89.5 (8.0)	87.8 (11.7)	0.53	0.82	1.72 (−3.37; 6.39)	0.181
	90.6 (64.1; 100) $n = 40$	89.1 (39.1; 98.4) $n = 23$				
Emotional Function Scale	81.7 (11.8)	83.4 (11.8)	0.61		−1.68 (−7.92; 4.42)	0.142
	81.3 (55; 100) $n = 40$	85 (52.5; 97.5) $n = 23$				
Social Function Scale	89.7 (12.0)	88.4 (14.1)	0.70		1.31 (−5.67; 7.69)	0.102
	92.5 (37.5; 100) $n = 39$	92.5 (42.5; 100) $n = 23$				
School Function Scale	77.6 (17.7)	83.0 (14.2)	0.24	0.76	−5.41 (−14.55; 3.39)	0.326
	81.3 (35; 100) $n = 40$	87.5 (47.5; 97.5) $n = 21$				
Psychosocial Function Scale	83.3 (11.9)	84.8 (12.5)	0.66		−1.53 (−8.12; 4.88)	0.127
	85.8 (47.5; 97.5) $n = 39$	86.7 (51.7; 98.3) $n = 21$				

For continuous variables, mean (SD) / median (min; max) / n is presented.
For comparison between groups, Fisher's non-parametric permutation.
test was used for continuous variables.
[a]Data represent the mean (95% CI). The CI for the difference between groups is based on Fisher's non-parametric.
permutation test.
[b]Data represent the difference in the mean / pooled SD.
* Adjustment of physical function for age at study and school function for WMIQ was performed by ANCOVA.
CI: confidence interval.

Discussion

In this study of children treated for non-syndromic CS, patients were asked and their parents to estimate HRQoL using the PedsQL 4.0 Generic Core Scales as a reliable and validated HRQoL-measurement tool specifically constructed for children in pediatric care. Measuring HRQoL with a patient-reported instrument allows access to information directly from the perspective of the patient, which remains a rare but coveted phenomenon in both research and clinical settings. This cohort of 73 patients treated for non-syndromic CS, revealed a generally average HRQoL, with no significant differences identified between those treated for CS and normative sample or between those treated for SS and MS. However, reports of psychosocial and school functions were lower in the SS group, although no differences in functions were found between the SS and MS groups according to the estimated HRQoL. Furthermore, the surgical method used for treatment (*i.e.* spring-assisted surgery and pi-plasty) in the SS group was unrelated to better or worse HRQoL outcomes.

There are few studies examining the subjective perception of the HRQoL of patients treated for non-syndromic CS, with only

Table 5. Difference in the mean of child- and parent-rated PedsQL generic core scales between pi-plasty and spring-assisted surgery used to treat patients with sagittal CS.

Variables	Pi-plasty ($n = 17$)	Spring-assisted surgery ($n = 23$)	p	Difference between groups[a]	Effect size[b]
HRQoL	84.9 (11.7)	85.6 (8.6)	0.83	−0.74 (−7.41; 5.98)	0.074
PedsQL Full Scale	88.3 (53.3; 97.8)	86.4 (67.4; 98.4)			
	$n = 16$	$n = 22$			
Physical Function Scale	89.2 (10.0)	90.1 (6.4)	0.75	−0.90 (−6.25; 4.49)	0.111
	89.1 (64.1; 100)	91.4 (76.6; 100)			
	$n = 17$	$n = 22$			
Emotional Function Scale	82.1 (13.6)	81.4 (10.8)	0.89	0.70 (−7.19; 8.75)	0.057
	82.5 (55; 100)	80 (57.5; 95)			
	$n = 17$	$n = 22$			
Social Function Scale	87.2 (15.0)	91.1 (9.4)	0.38	−3.95 (−11.79; 4.06)	0.328
	92.5 (37.5; 100)	92.5 (65; 100)			
	$n = 16$	$n = 22$			
School Function Scale	77.4 (18.0)	77.3 (18.2)	1.00	0.08 (−11.67; 12.19)	0.004
	80 (42.5; 95)	81.3 (35; 100)			
	$n = 17$	$n = 22$			
Psychosocial Function Scale	82.9 (13.2)	83.3 (11.4)	0.94	−0.34 (−8.42; 7.81)	0.028
	86.7 (47.5; 96.7)	84.2 (54.2; 97.5)			
	$n = 16$	$n = 22$			

For continuous variables, mean (SD) / median (min; max) / n is presented.
For comparison between groups, Fisher's non-parametric permutation.
test was used for continuous variables. .
[a]Data represent the mean (95% CI). The CI for the difference between groups is based on Fisher's non-parametric.
permutation test.
[b]Data represent the difference in the mean / pooled SD.
CI: confidence interval.

Table 6. Correlation analysis of the mean of child- and parent-rated PedsQL generic core scales with ABAS and IQ.

Variables	HRQoL PedsQL Full scale	Physical Function Scale	Emotional Function Scale	Social Function Scale	School Function Scale	Psychosocial Function Scale
Adaptive behaviour skills full scale	0.57	0.32	0.50	0.40	0.55	0.57
	<0.0001	0.0084	<0.0001	0.0008	<0.0001	<0.0001
	65	68	68	67	66	65
Conceptual composite scale	0.57	0.33	0.46	0.38	0.57	0.57
	<0.0001	0.0053	<0.0001	0.0013	<0.0001	<0.0001
	65	68	68	67	66	65
Social composite scale	0.52	0.38	0.52	0.35	0.44	0.52
	<0.0001	0.0015	<0.0001	0.0038	0.0002	<0.0001
	65	68	68	67	66	65
Practical composite scale	0.53	0.28	0.46	0.37	0.52	0.53
	<0.0001	0.022	<0.0001	0.0020	<0.0001	<0.0001
	65	68	68	67	66	65
Wechsler full scale intelligence quotient	0.42	0.16	0.28	0.34	0.47	0.42
	0.0004	0.19	0.022	0.0046	<0.0001	0.0004
	65	68	68	67	66	65
Wechsler verbal comprehension intelligence quotient	0.39	0.26	0.26	0.30	0.40	0.39
	0.0014	0.030	0.036	0.013	0.0009	0.0014
	65	68	68	67	66	65
Wechsler perceptual reasoning intelligence quotient	0.31	0.05	0.18	0.22	0.34	0.31
	0.013	0.66	0.13	0.069	0.0047	0.013
	65	68	68	67	66	65
Wechsler working memory intelligence quotient	0.20	0.06	0.14	0.16	0.29	0.20
	0.12	0.60	0.27	0.20	0.019	0.12
	65	68	68	67	66	65
Wechsler processing speed intelligence quotient	0.36	−0.01	0.19	0.33	0.43	0.36
	0.0030	0.93	0.12	0.0065	0.0003	0.0030
	65	68	68	67	66	65

For each variable, Spearman's correlation is presented with the corresponding p-value and the number of observations used.

one study focusing on children treated for CS and reporting results indicating a risk of low HRQoL [19]. At the same time, in a series of untreated SS patients, there was a tendency toward a low score in relation to positive emotions [20].

Numerous studies have focused on assessing neuropsychological and cognitive functions in order to reach a consensus regarding developmental impacts related to CS diagnosis and surgical treatment [1–4,6–15,17]. Previous studies indicate that non-syndromic CS patients are generally expected to exhibit average cognitive development [6,8,10,12,29]. The cohort of patients in this study showed average performance in terms of IQ and ABAS, which were extracted as background data from previous studies [8,26]. Notably, associations between HRQoL, IQ, and ABAS were significant, with moderate correlations. This is an important aspect to consider when measuring HRQoL, given that cognitive and adaptive abilities can affect HRQoL outcomes. Therefore, it is crucial to

Table 7. Change in PedsQL generic core scales from child rater to parent rater.

Variable	Child ($n = 73$) Mean (SD) Median (Min; Max) (95% CI for the mean), n	Parent ($n = 73$) Mean (SD) Median (Min; Max) (95% CI for the mean), n	Change from child to parent ($n = 73$) Mean (SD) Median (Min; Max) (95% CI for the mean), n SRM ES	p (within group)
HRQoL	84.3 (12.5)	88.0 (11.4)	3.61 (13.15)	0.031
PedsQL full scale	85.9 (35.9; 100) (81.3; 87.3) $n = 67$	90.8 (58.7; 100) (85.2; 90.7) $n = 68$	3.8 (−19.57; 33.7) (0.40; 6.84) $n = 66$ 0.27 0.29	
Physical function scale	87.0 (12.2) 87.5 (34.4; 100) (84.0; 89.8) $n = 69$	92.2 (10.9) 96.9 (43.8; 100) (89.6; 94.8) $n = 69$	5.25 (13.53) 3.13 (−21.88; 37.5) (2.00; 8.52) $n = 69$ 0.39 0.43	0.0022
Emotional function scale	82.8 (14.5) 85 (40; 100) (79.3; 86.2) $n = 69$	82.5 (15.4) 85 (40; 100) (78.9; 86.2) $n = 69$	−0.290 (18.921) 0 (−40; 60) (−4.833; 4.242) $n = 69$ −0.02 −0.02	0.94
Social function scale	88.3 (15.8) 95 (30; 100) (84.6; 92.1) $n = 69$	91.4 (13.2) 97.5 (35; 100) (88.2; 94.6) $n = 68$	2.87 (15.34) 0 (−30; 45) (−0.78; 6.56) $n = 68$ 0.19 0.18	0.14
School function scale	77.7 (17.8) 80 (30; 100) (73.4; 82.0) $n = 67$	83.0 (19.6) 90 (30; 100) (78.4; 87.7) $n = 69$	5.82 (18.14) 10 (−50; 60) (1.41; 10.30) $n = 67$ 0.32 0.33	0.012
Psychosocial function scale	83.0 (14.2) 86.7 (36.7; 100) (79.5; 86.4) $n = 67$	85.8 (13.7) 90 (46.7; 100) (82.5; 89.1) $n = 67$	2.60 (14.98) 3.33 (−33.33; 45) (−1.03; 6.26) $n = 66$ 0.17 0.18	0.17

For continuous variables, mean (SD) / median (min; max) / (95% CI using the inversion of Fisher's non-parametric permutation test) / n is presented.
For comparison within groups, Fisher's non-parametric permutation test for matched pairs was used.
Change was calculated, as follows: difference = parent rate − child rate.
SRM, standardized response mean = mean difference / SD of the difference.
ES, effect size = mean difference / SD for child.
CI: confidence interval.

control for these variables in order to measure the intended phenomenon. In a clinical setting, HRQoL could be used to screen patients in need of further psychological assessment [30].

Patient-reported outcomes can offer information regarding patient perspective through self- or proxy reports. In this study, both self- and proxy reports were used to assess HRQoL. Interestingly, there were differences in how children and parents estimated HRQoL, with parents inclined to report higher HRQoL than the children. Previous studies frequently used proxy reports, where parents were asked to assess different aspects of developmental questions regarding their child [5,31–34]. The results of this study indicate that it is important to also use self-reports before drawing conclusions about patient status. Even though the actual patients are young, primary information rather than information from their parents, may add value to caregivers.

Strengths and limitations

The primary strength of this study is its methodological approach using a validated, reliable measurement, and comparison with a normative sample to assess HRQoL. Additionally, methodological strength is the advantages of controlling for confounders through the use of comprehensive amounts of background data. Additionally, the high response rate (80.2%) and the attrition analysis minimized the risk of selection bias. However, there are also limitations, as a larger study group would have increased the probability of the assumptions from the results. Moreover, a limitation with this study is the use of normative data instead of controls. However, the normative data was based on a large sample of children of the same age and the same ethnical background.

Additionally, there were no un-operated cases included. Therefore, the fundamental question regarding the benefit of surgery itself remains unsolved.

Conclusions

In summary, these results concluded that children treated for non-syndromic CS have a generally average HRQoL, with neither CS type nor surgical method used in the SS group related to better or worse HRQoL outcomes. Moreover, measuring HRQoL in a clinical setting can be used as a screening method to detect patients in need of profound psychological assessment. Furthermore, using both self- and proxy reports is crucial, given that parents tend to overestimate the HRQoL of their child.

Disclosure statement

No potential conflict of interest was reported by the authors.

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