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HEALTH-RELATED QUALITY OF LIFE IN CHILDREN AND ADOLESCENTS WITH CEREBRAL PALSY

Z ZDRAVJEM POVEZANA KAKOVOST ŽIVLJENJA OTROK IN NAJSTNIKOV S CEREBRALNO PARALIZO

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ABSTRACT

Introduction. In a cross-sectional cohort study, health-related quality of life of Slovenian children and adolescents with cerebral palsy was examined, and factors associated with it have been identified.

Methods. Caregivers of 122 children and adolescents with cerebral palsy were addressed to fill out proxy versions of HRQoL questionnaires (DISABKIDS generic and cerebral palsy module). Children and adolescents without cognitive deficit were asked to fill out the self-report versions.

Results. Ninety-one families of 43 children (the mean age is 10 years, 6 months, SD 1.2; 26 males and 17 females) and 48 adolescents (the mean age is 14 years, SD 0.9; 23 males and 25 females) completed proxy-reports. Forty-eight individuals were able to self-report (26 children and 22 adolescents). Health-related quality of life was perceived as good. Self-reporting participants scored higher than their caregivers (mean score 75.6, SD 15.9 versus mean 72.3, SD 17.9; p=0.048). Adolescents scored lower than children in all domains (mean score 69.4, SD 19.4 versus mean 80.8, SD 10.0; p=0.01). Higher age (p<0.001), pain (p<0.001) and disturbed sleep (p=0.002) were strong predictors of worse health-related quality of life. Social Inclusion and Independence domains received the lowest scores.

Conclusions. Slovenian children and adolescents with cerebral palsy have a good health-related quality of life, with Social Inclusion and Independence being the weakest domains. Children reported higher scores than adolescents or their caretakers. Pain was the strongest predictor of poor health-related quality of life.

Keywords:
cerebral palsy, children, adolescents, quality of life, self-reports, proxy-reports

IZVLEČEK

Uvod. *V luči vse večjega trenda k celostnemu pristopu obravnave otrok s cerebralno paralizo se poleg dobrega poznavanja in vrednotenja otrokove oviranosti med glavna orodja, ki so v pomoč pri načrtovanju obravnav, uvrščajo vprašalniki za oceno z zdravjem povezane kakovosti življenja. Cilja raziskave sta bila pridobiti vpogled v z zdravjem povezano kakovost življenja pri skupini slovenskih otrok s cerebralno paralizo in najti morebitne povezave z njihovimi demografskimi in kliničnimi podatki.*

Metode. *V okviru presečne kohortne raziskave je bilo iz Slovenskega registra otrok s cerebralno paralizo naključno izbranih 122 družin. Skrbniki otrok so bili pozvani k sodelovanju z izpolnitvijo proxy različice vprašalnika o z zdravjem povezani kakovosti življenja. Otroci brez kognitivne okvare so bili naprošeni, naj izpolnijo različico vprašalnika, namenjeno samoocenjevanju.*

Rezultati. *Pri oceni z zdravjem povezane kakovosti življenja je sodelovalo 91 družin. Skrbniki 43 otrok in 48 najstnikov so izpolnili svoje različice vprašalnikov (proxy različica). Oseminštirideset otrok in najstnikov brez kognitivne okvare je samih izpolnilo vprašalnik (self različica). Ocenjena z zdravjem povezana kakovost življenja je bila dobra. Otroci so jo ocenili bolje kot najstniki (povprečno 80,8, SD 10,0 proti povprečno 69,4, SD 19,4; p=0,01). Preiskovanci so jo ocenili bolje kot njihovi skrbniki (povprečno 75,6, SD 15,9 proti povprečno 72,3, SD 17,9; p=0,048). Višja starost (p<0,001), prisotnost bolečine (p<0,001) in motnje spanja (p=0,002) so bili močni napovedni dejavniki za slabšo z zdravjem povezano kakovost življenja. Socialna vključenost in samostojnost sta bili najslabše ocenjeni domeni. Vprašalnik DISABKIDS se je izkazal za dobro orodje za oceno z zdravjem povezane kakovosti življenja otrok s cerebralno paralizo.*

Sklep. *Slovenski otroci s cerebralno paralizo ocenjujejo svojo z zdravjem povezano kakovost življenja kot dobro. Otroci jo ocenjujejo bolje kot najstniki ali njihovi skrbniki. Bolečina je najmočnejši napovedni dejavnik slabše z zdravjem povezane kakovosti življenja.*

Ključne besede:
cerebralna paraliza, otroci, najstniki, kakovost življenja

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1 INTRODUCTION

Cerebral palsy (CP) is a diverse condition with various levels of reduced motor function, often accompanied with cognitive deficit, epilepsy, vision or hearing impairment, orogastrointestinal malfunction and skeletal problems (1). All CP definitions share the fact that the injury to the immature brain results in a life-long disability (2-4). Current therapeutic interventions, focused on alleviating physical dysfunctions, can only offer limited relief and can cause additional pain (4, 5). Psychological problems and needs of CP patients are much less obvious and are usually poorly understood and tended by healthcare practitioners. Evidence shows that CP patients do not primarily search for physical improvement as much as they crave for social inclusion (5, 6).

QoL is a broad concept defined by the World Health Organization as "individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns" (7). Health-Related Quality of Life (HRQoL) represents the QoL in the view of an individual's health status (8). When managing chronic conditions, such as CP, it is an important marker of the efficacy of clinical interventions. It is, however, a complex, hard-to-define term and controversies exist about its detailed definition and appropriate measuring tools (8).

Studies that examined HRQoL of children and adolescents with CP showed scores similar to aged-matched general population with the exceptions in social participation and motor functioning (5, 6, 9, 10). In longitudinal studies, HRQoL in childhood correlated well with HRQoL in adolescence (5). Pain, parenting stress and psychological problems were identified as predictors of worse HRQoL (5, 10, 11). Although motor impairment influenced functioning and participation, it affected psychosocial wellbeing to a much lesser extent (9).

Not all existing HRQoL measuring tools actually measure HRQoL (a subjective perspective), but rather an objective interaction between body structure, function and participation (12). The weakness of many HRQoL measures is the lack of indicators that measure wellbeing (positive emotions and satisfaction about daily activities, relations and life overall), while being oriented towards asking how often patients feel sad and unsatisfied with their involvement in daily tasks (13).

We chose DISABKIDS questionnaires for their recognition as good HRQoL measuring tools (12). Generic and various disease specific modules enable a comparison between children with different chronic conditions (14, 15). A good correlation with KIDSCREEN measures (developed by the same group of professionals) offers an opportunity for comparing QoL of aged-matched general population. DISABKIDS questionnaires show good linkage with the International Classification of Functioning, Disability and Health (ICF) (16).

The objective of our study was to assess HRQoL of Slovenian paediatric patients with CP, identify possible underlying factors that are associated with it, and find potential differences in scoring between caregivers and children.

2 METHODS

2.1 Participants

At the time of data collection in July 2014, the Slovenian National Cerebral Palsy Registry (SRCP) included 371 children of all ages. They were registered through neurodevelopmental paediatricians in regional outpatient clinics covering around 90% of Slovenian paediatric CP population. Out of original 150 children aged 8-16 years, randomly selected from the SRCP, contact data for 122 families were available; 91 caregivers (of 43 children aged 6-12 years and 48 adolescents aged 13-16 years) were willing to cooperate. Children with tested IQ score >70 or attending regular school were considered cognitively able to self-report, and 48 of them (26 children and 22 adolescents) agreed to do so.

2.2 The Procedure

The caregivers of 122 children with CP were contacted by telephone at one or (in the case of first call non-responders) at two occasions by a single physician. Study aims were explained to them and caregivers were invited to participate. Questionnaires were sent to 103 caregivers who accepted the invitation, together with written instructions and informed consent form.

2.3 Measures

DISABKIDS instruments were used to assess HRQoL. DISABKIDS Chronic Generic Measure – long version (DCGM-37) – contains 3 domains (Mental, Social and Physical) that

are further divided in order to measure the following 6 dimensions: Independence, Emotion, Social inclusion, Social exclusion, Limitation, and Treatment. All items are Likert-scaled and transformed to a scale from 0 to 100, where higher scores indicate a better HRQoL. According to field testing, the instrument shows sound psychometric properties with satisfactory reliability, construct validity, convergent and discriminant validity (15). It is available in self-reported and proxy versions, and it was designed for children aged 8-16 years. DISABKIDS Cerebral Palsy Module (DCSM-CPM) and DISABKIDS Epilepsy Module (DCSM-EM) both address two dimensions, namely: Impact (10 items) and Communication (2 items) for DCSM-CPM, and Impact and Social (both 5 items) for DCSM-EM.
All six questionnaires (self-reported and proxy versions) underwent the translation and validation procedure with the guidance of European DISABKIDS Group and DISABKIDS manual, and were approved for the use in the study (17). Paediatric Quality of Life Inventory 4.0 (PedsQL) Generic Core Scales was used for comparison, as it has already been validated in the Slovenian language. Its concept is very similar to DCGM-37 and consists of 23 items encompassing: Physical functioning, Emotional functioning, Social functioning and School functioning (18).

The assessment bundle included: DCGM-37, DCSM-CPM, DCSM-EM (in cases of comorbid epilepsy) and PedsQL. In order to gain information about clinical usefulness of selected HRQoL measures, we added 5 additional questions asking for personal opinion about the questionnaires. All questionnaires were sent in proxy versions and, if applicable, in self-reported versions.

2.4 Statistical Analyses

Items from all questionnaires were scored according to DISABKIDS manual (17). Raw scores were converted into values on a 0-100 scale. A score was calculated if at least 80% of items were answered. Overall scores were calculated as a sum of all separate scores. Each score was expressed as mean and standard deviation (SD).

The effect of various factors on different dimensions and the final score was evaluated by independent samples t-test or one-way ANOVA, as the distribution of results was close to normal. Linear regression model was constructed to determine the most important factors influencing HRQoL. The differences were expressed as mean difference and 95% confidence interval (CI). A value of $p<0.05$ was considered statistically significant.

The partial and final scores of self-reports and proxy-reports were compared using Pearson's Product Moment Correlation coefficient (Pearson's r).

The partial and final scores of self-reports and proxy-reports were also compared to subscales and the total score of PedsQL measure, where Pearson's r was used again.

All the analyses were conducted with the SPSS (Statistical Package for Social Sciences Program) version 20.0.

3 RESULTS

Out of 122 parents of children with CP invited to partake in the study, 91 were willing to participate (the response rate was 75%). The main reason for refusal came from parents of children with very mild disability, as they considered their children healthy (not having cerebral palsy) and were concerned that the questionnaire would disturb them. In the initial sample, there were 60 families with cognitively intact children, among which 48 were willing to self-report (the response rate was 80%). The remaining 43 families with more severely affected children filled out only the proxy-reports. DISABKIDS final scores (transformed to a scale of 1-100) by different modules, items and demographic properties are presented in Table 1.

Table 1. DISABKIDS final scores (transformed to a scale of 1-100) by different modules, items and demographic properties.

SELF - REPORTS		ALL		CHILDREN		ADOLESCENTS		MALES		FEMALES	
Transformed scores (0-100; mean, SD)		valid items	score	valid items	score	valid items	score	valid items	score	valid items	score
DCGM-37-S	Independence	48	70.4,SD 20.1	26	75.6,SD 15.7	22	64.2,SD 23.1	23	69.4,SD 22.8	25	71.3,SD 17.6
	Physical	48	74.6,SD 18.9	26	80.3,SD 11.9	22	68.0,SD 23.5	23	74.5,SD 21.2	25	76.9,SD 12.9
	Emotion	48	85.3,SD 17.7	26	91.1,SD 9.8	22	78.6,SD 22.4	23	85.2,SD 20.5	25	71.5,SD 34.9
	Exclusion	48	80.2,SD 17.0	26	84.9,SD 10.5	22	74.6,SD 21.3	23	78.3,SD 17.7	25	69.5,SD 16.2
	Inclusion	48	65.7,SD 19.5	26	70.4,SD 16.9	22	60.2,SD 21.3	23	61.6,SD 22.2	25	82.0,SD 16.5
	Medication	14	78.6,SD 26.8	7	83.2,SD 12.4	7	74.0,SD 36.9	9	82.5,SD 22.6	5	85.4,SD 15.3
	General	48	75.6,SD 16.0	26	80.8,SD 10.0	22	69.4,SD 19.5	23	74.2,SD 19.0	25	74.8,SD 17.2
DCSM-CPM-S	Impact	44	76.1,SD 15.0	23	79.3,SD 10.4	21	72.6,SD 18.4	20	74.5,SD 18.3	24	77.5,SD 11.8
	Communication	44	92.3,SD 20.6	23	98.9,SD 3.6	21	85.1,SD 28.1	20	90.0,SD 25.2	24	94.3,SD 16.1
	Total	44	78.8,SD 14.7	23	82.6,SD 8.7	21	74.7,SD 18.7	20	77.1,SD 18.7	24	80.3,SD 10.6
PROXY - REPORTS		**ALL**		**CHILDREN**		**ADOLESCENTS**		**MALES**		**FEMALES**	
Transformed scores (0-100; mean, SD)		valid items	score	valid items	score	valid items	score	valid items	score	valid items	score
DCGM-37-S	Independence	83	58.1,SD 24.4	37	67.8,SD 20.8	46	50.3,SD 24.5	50	54.8,SD 24.6	33	63.1,SD 23.7
	Physical	83	66.2,SD 21.2	37	71.2,SD 17.1	46	62.1,SD 23.3	50	63.38,SD 22.5	33	70.4,SD 17.7
	Emotion	80	78.3,SD 20.4	36	82.7,SD 16.8	45	74.7,SD 22.5	47	77.9,SD 22.5	33	78.9,SD 17.3
	Exclusion	82	76.5,SD 19.9	36	82.5,SD 13.7	46	71.7,SD 22.8	49	73.6,SD 21.9	33	80.8,SD 15.8
	Inclusion	83	53.2,SD 24.5	36	65.2,SD 19.2	47	44.0,SD 24.3	49	49.9,SD 24.8	34	58.0,SD 23.7
	Medication	34	79.8,SD 21.0	15	79.8,SD 24.8	19	79.8,SD 18.3	23	78.7,SD 21.7	11	82.1,SD 20.5
	General	82	66.9,SD 18.2	36	75.1,SD 12.5	46	60.5,SD 19.5	49	64.8,SD 19.2	33	70.1,SD 16.3
DCSM-CPM-S	Impact	72	71.0,SD 17.3	33	74.5,SD 15.6	39	68.1,SD 18.3	20	72.1,SD 16.0	22	76.9,SD 11.1
	Communication	74	78.0,SD 29.2	33	87.9,SD 21.9	41	70.1,SD 32.0	20	88.1,SD 24.5	24	95.8,SD 12.6
	Total	74	72.6,SD 16.9	33	76.8,SD 13.6	41	69.2,SD 18.6	20	74.8,SD 14.9	24	80.9,SD 9.3

Abbreviations: DISABKIDS Chronic Generic Measure Self / Proxy (DCGM-37-S/-P); DISABKIDS Condition Specific Module - Cerebral Palsy Module Self / Proxy (DCSM-CPM-S/-P)

3.1 Sample Characteristics

The mean age of all participants with CP was 12 years and 4 months (SD 2.02); 43 (48%) were children (the mean age is 10 years, 6 months, SD 1.2) and 48 (53%) adolescents (the mean age is 14 years, SD 0.9). The sample consisted of 53 (58%) males and 38 (42%) females.

Among 48 self-reporting participants, there were 26 (54%) children (the mean age is 10 years 8 months, SD 1.1) and 22 (46%) adolescents (the mean age is 13 years, 11 months, SD 0.8). Gender distribution across the self-reported sample was balanced with 23 (48%) males and 25 (52%) females.

Compared to the data in the SRCP registry, the study sample shows similar gender, CP type and GMFCS distribution, and was therefore found to be representative of the population of Slovenian paediatric patients with CP (19).

All characteristics of children concerning their demographic parameters, level of impairment, comorbidities, therapeutic interventions and family social status are listed in Table 2 and Table 3.

Table 2. Demographic and health-related data of 91 participants.

	All participants - proxy reports			Self-reported participants		
	Children (8-12 y) N 43 (47.3%)	**Adolesc.** (13-17 y) N 48 (52.7%)	**All** N 91	**Children** (8-12 y) N 26 (54.2%)	**Adolesc.** (13-17 y) N 22 (45.8%)	**All** N 48
Age (mean)	10 y 6 mo, SD 1.2	14 y, SD 0.9	12 y 4 mo, SD 2.02	10 y 8 mo, SD 1.1	13 y 11mo, SD 0.8	12 y 2 mo, SD 1.9
Gender						
male	26 (23.3%)	27 (56.3%)	53 (58.2%)	13 (50.0%)	10 (45.5%)	23 (47.9%)
female	17 (39.5%)	21 (43.8%)	38 (41.8%)	13 (50.0%)	12 (54.5%)	25 (52.1%)
CP classification						
spastic	35 (81.4%) uni 14 (33%)	43 (89.6%) uni 11 (23%)	78 (85.7%) uni 25 (27%)	24 (92.3%) uni 13 (50%)	22 (100%) uni 7 (32%)	46 (95.8%) uni 20 (42%)
dyskinetic dystonia	7 (16.3%)	4 (8.3%)	11 (12.1%)	2 (7.6%)	/	2 (4.2%)
dys.choreoathetosis	/	1 (2.1%)	1 (1.1%)	/	/	/
ataxic	1 (2.3%)	/	1 (1.1%)	/	/	/
GMFCS						
I	18 (41.9%)	11 (22.9%)	29 (31.9%)	16 (61.5%)	8 (36.4%)	24 (50%)
II	10 (23.2%)	10 (20.8%)	20 (22.0%)	7 (26.9%)	7 (31.8%)	14 (29.2%)
III	3 (7.0%)	8 (16.7%)	11 (12.1%)	1 (3.8%)	4 (18.2%)	5 (10.4%)
IV	7 (16.3%)	8 (16.7%)	15 (16.5%)	2 (7.7%)	3 (13.6%)	5 (10.4%)
V	5 (11.6%)	11 (22.9%)	16 (17.6%)	/	/	/
IQ						
> 70	25 (58.1%)	21 (43.8%)	48 (52.7%)	26 (100%)	22 (100%)	48 (100%)
50-70	7 (16.3%)	10 (20.8%)	15 (16.5%)	/	/	/
20-50	7 (16.3%)	11 (22.9%)	18 (19.8%)	/	/	/
< 20	4 (9.3%)	6 (12.5%)	10 (11.0%)	/	/	/
Epilepsy	15 (34.9%)	17 (35.4%)	32 (35.2%)	3 (11.5%)	3 (13.6%)	6 (12.5%)
Speech disorder	19 (44.2%)	22 (45.8%)	41 (45.1%)	7 (26.9%)	6 (27.3%)	13 (27.1%)
Attention disorder	14 (32.6%)	11 (22.9%)	25 (27.5%)	10 (38.5%)	5 (22.7%)	15 (31.3%)
Visual impairment	15 (34.9%)	19 (39.6%)	34 (37.4%)	7 (26.9%)	9 (40.9%)	16 (33.3%)
- severe	2 (4.7%)	2 (4.2%)	4 (4.4%)	1 (3.8%)	/	1 (2.1%)
Hearing impairment	4 (9.3%)	3 (6.3%)	7 (7.7%)	1 (3.8%)	1 (4.5%)	2 (4.2%)
- severe	2 (4.7%)	1 (2.1%)	3 (3.3%)	1 (3.8%)	1 (4.5%)	2 (4.2%)
Reporting pain	7 (16.3%)	14 (29.2%)	21 (23.1%)	2 (7.7%)	5 (22.7%)	7 (14.6%)
Disrupted sleep	8 (18.6%)	12 (25.0%)	20 (22.0%)	2 (7.7%)	1 (4.5%)	3 (6.3%)
Gastrostomy	1 (2.3%)	/	1 (1.1%)	/	/	/
Reduced bone density	4 (9.3%)	5 (10.4%)	9 (10.0%)	/	/	/

Abbreviations: the number of cases (N); years (y); months (mo); unilateral (uni); Gross Motor Function Classification Scale (GMFCS); intelligence quotient (IQ)

Table 3. Implemented interventions and socio-economic data of 91 participants.

	All participants - proxy reports			Self-reported participants		
	Children (8-12 y) N 43 (47.3%)	**Adolesc.** (13-17 y) N 48 (52.7%)	**All** N 91	**Children** (8-12 y) N 26 (54.2%)	**Adolesc.** (13-17 y) N 22 (45.8%)	**All** N 48
Interventions:	3.9 per person (min 1, max 11)			/	/	/
Neurodevelopmental th.	91 (100%)			/	/	/
- age at onset	65 (71%) < 6 months, 77 (85%) <12 months			/ /	/ /	/ /
Physiotherapy	69 (75.8%)			/	/	/
Occupational therapy	48 (52.7%)			/	/	/
Speech-language therapy	31 (43.1%)			/	/	/
Special pedagogy	25 (27.5%)			/	/	/
Hypo therapy	20 (21.9%)			/	/	/
Hydrotherapy	17 (18.7%)			/	/	/
Psychological therapy	11 (12.1%)			/	/	/
Social pedagogy	9 (9.8%)			/	/	/
Complementary methods	7 (7.7%)			/	/	/
Orthopedic therapy	3 (3.3%)			/	/	/
Typhlopedagogy	2 (2.2%)			/	/	/
Surdopedagogy	2 (2.2%)			/	/	/

Education (stage):	no data	1	2	3	4	5	6	7	8	no data	1	2	3	4	5	6	7	8
- mother (N)	12	1	3	6	28	13	10	17	1	9	0	1	3	14	7	3	11	0
(%)	13	1	3	7	31	14	11	19	1	19	0	2	6	29	15	6	23	0
- father (N)	13	0	5	9	30	16	4	13	1	8	0	2	5	14	11	1	7	0
(%)	14	0	6	10	33	18	4	14	1	17	0	4	10	29	23	2	15	0

Unemployment:						
- mother	13 (30.2%)	20 (41.7%)	33 (36.3%)	7 (26.9%)	6 (27.3%)	13 (27.1%)
- mother (part time)	6 (14.0%)	2 (4.2%)	8 (8.8%)	2 (7.7%)	1 (4.5%)	3 (6.3%)
- father	4 (9.3%)	5 (10.4%)	9 (9.9%)	/	1 (4.5)	1 (2.1%)
- both	2 (4.7%)	3 (6.3%)	5 (5.5%)	/	1 (4.5%)	1 (2.1%)
Financial support						
- lost income substitute	8 (18.6%)	20 (41.7%)	28 (30.8%)	1 (3.8%)	5 (22.7%)	6 (12.5%)
- child care support	25 (58.1%)	36 (75.0%)	61 (67.0%)	10 (38.5%)	11 (50.0%)	21 (43.8%)
Child residence						
- home	41 (95.3%)	45 (93.8%)	86 (94.5%)	26 (100%)	22 (100%)	48 (100%)
- day care centre	1 (2.3%)	1 (2.1%)	2 (2.2%)	/	/	/
- 24h centre	1 (2.3%)	2 (4.2%)	3 (3.3%)	/	/	/
Schooling:						
- regular	25 (58.1%)	22 (45.8%)	47 (51.6%)	25 (96.2%)	22 (100%)	47 (97.9%)
- adjusted program	18 (41.9%)	26 (54.2%)	44 (48.4%)	1 (3.8%)	/	1 (2.1%)

Abbreviations: the number of cases (N); years (y)

3.2 Proxy-Reports

Caregivers rated their children's HRQoL as 'good' (DCGM-37 mean total transformed score 66.9, SD 18.2). The worse scored domains were Social Inclusion (mean 53.2, SD 24.5) and Independence (mean 58.1, SD 24.4). The highest rated were Emotion (mean 78.3, SD 20.4) and Medication (mean 79.8, SD 21.0). HRQoL of children was scored higher than HRQoL of adolescents (mean 75.1, SD 12.5 versus mean 60.5, SD 19.5; mean difference 14.64, 95% CI 7.21 - 22.07; p<0.001). The largest differences were for Independence (mean 67.79, SD 20.84 versus 50.34, SD 24.54; mean difference 17.45, 95% CI 7.36 - 27.54; p=0.001), Social Inclusion (mean 65.23, SD 19.17 versus 44.02, SD 24.34; mean difference 21.21, 95% CI 11.40 - 31.02; p<0.001) and Social Exclusion domains (mean 82.52, SD 13.65 versus 71.74, SD 22.76; mean difference 10.78, 95% CI 2.23 - 19.34; p=0.014). There were some missing data for each item, and 5 items had a missing data rate equal to 10% or more. In nine questionnaires, scoring was not possible due to too many missing values (>20% of unanswered items).

Seventy-nine DCSM-CPM completed proxy reports with a mean total transformed score 72.6, SD 16.9. The subscale Communication (mean 78.0, SD 29.2) was scored higher than Impact (mean 71.0, SD 17.3). Caregivers of adolescents scored lower (mean 69.2, SD 18.6) compared to caregivers of children (mean 76.8, SD 13.6), but the difference was borderline statistically important (p=0.050).

Nineteen DCSM-EM proxy reports were valid and gave the total transformed score of mean 89.3, SD 14.3.

3.3 Self-Reports

Among DCGM-37 self-reporters, HRQoL was perceived as 'good' (the mean total transformed score is 75.6, SD 15.9). The worse scored subscales were, as in proxy-reports, Social inclusion (mean 65.7, SD 19.5) and Independence (mean 70.4, SD 20.1). The highest score was given to the sub-scale Emotion (mean 85.3, SD 17.7). Children rated their HRQoL better than adolescents (mean 80.8, SD 10.0 versus mean 69.4, SD 19.4; mean difference 11.37, 95% CI 2.57 - 20.17; p=0.01). Between subscales, the biggest difference was seen in Emotion (mean 91.07, SD 9.76 versus mean 78.57, SD 22.43; mean difference 11.37, 95% CI 2.57 - 20.17, p=0.01), Physical (mean 80.29, SD 11.82 versus mean 67.99, SD 23.48; mean difference 12.30, 95% CI 1.74 - 22.85, p=0.02) and Social exclusion domains (mean 84.94, SD 10.54 versus mean 74.62, SD 21.28; mean difference 10.31, 95% CI 0.78 - 19.84; p=0.035). There were no missed items in the sample.

Twenty-three children and 21 adolescents filled out DCSM-CPM self-reports. The mean transformed total score of all 44 reports was 78.8, SD 14.7. The Communication domain scored higher (92.3, SD 20.6) than Impact (76.1, SD 15.0). No differences were observed among children and adolescents in DCSM-CPM reports (p=0.14).

Only 6 children and adolescents able to self-report had concomitant epilepsy. They scored their HRQoL through DCSM-EM as very good (80.4, SD 24.1).

3.4 Proxy and Self-Reports Comparison

The correlation between DCGM-37 proxy and self-reports was good (Pearson r=0.80 for total scores and r=0.59 - 0.80 for separate domains, where only Social Inclusion and Social Exclusion domains resulted in r<0.70). The absolute difference between proxy and self-reported scores was significant, self-reporting participants rating their HRQoL higher than caregivers (mean 75.6, SD 15.9 versus mean 72.3, SD 17.9; the mean difference 3.23, 95% CI 0.03 - 6.43; p=0.048).

The same was found for DCSM-CPM measure with the total transformed score r=0.76 and r=0.73 and r=0.81 for Impact and Communication subscales. There was no difference between proxy and self-reported DCSM-CPM scores (p=0.97).

3.5 PedsQL Reports

The mean total score of 78 PedsQL – proxy measures – was 61.5, SD 21.4, and of 41 PedsQL self-reported measures 75.6, SD 19.6. The correlation between DISABKIDS and PedsQL total transformed scores was very good (Pearson r=0.81 for self-reports and r=0.86 for proxy reports). Domains measuring similar concepts showed high correlation as well: r=0.70 for physical domains (self) and r=0.81 for physical domains (proxy), r=0.74 for emotional domains (self) and r=0.64 for emotional domains (proxy) and r=0.75 for social domains (self) and r=0.80 social domains (proxy).

3.6 Factors Influencing HRQoL

DCGM-37 proxy reports: lower age was found a strong single predictor of better HRQoL (mean 75.1, SD 12.5 versus mean 60.5, SD 19.5; the mean difference 14.64, 95% CI 7.21 - 22.07; p<0.001). There was a negative correlation between HRQoL and disease severity. GMFCS level alone, tested with one-way ANOVA, was negatively associated with HRQoL (p=0.023). So were pain (the mean difference 23.21, 95% CI 14.71 - 31.72; p<0.001), disturbed sleep (the mean difference 17.96, 95% CI 8.88 - 27.05; p=0.001) and cognitive abilities (the mean difference 13.38, 95% CI 5.84 - 20.93; p=0.001). However, the multivariable analysis of the same variables showed that the main factors reducing HRQoL were pain (p<0.001) and disturbed sleep (p=0.002), and not GMFCS itself (p=0.735). Adjusted r score for this model was 0.49

and the strongest standardised regression coefficient was found for pain -0.47, p<0.001. No correlation was found between the number of implemented therapeutic interventions and HRQoL, nor for comorbidities, such as epilepsy, and language, speech and attention disorders. There was also no HRQoL association with parents' education, employment status, financial support, child cognitive abilities and schooling type.

DCGM-37 self-reports: in the single variable analysis, besides higher age (the mean difference 11.37, 95% CI 2.57 - 20.17; p=0.012), the main factors reducing HRQoL were pain (the mean difference 18.57, 95% CI 6.03 - 31.11; p=0.005) and comorbidities (the mean difference 9.56, 95% CI 0.24 - 18.88; p=0.045). In the multivariable model using these variables, the adjusted r score was 0.27, and the strongest standardised regression coefficient was found for pain (-0.39, p=0.009), whereas age was only borderline significant (p=0.059).

DCSM-CPM proxy reports: in the multivariable model using variables that tested significant in univariate models (pain, disturbed sleep, GMFCS, cognitive impairment, speech impairment and epilepsy), pain was the only factor negatively influencing HRQoL (adjusted r score 0.34, standardized regression coefficient -0.30, p=0.009). DCSM-CPM self-reports: pain was the only significant single factor related to a lower perception of HRQoL (p=0.027).

3.7 Additional Questions

The caregivers needed, on average, 13.6 minutes to fill out DCGM-37 and DCSM-CPM proxy modules. Forty-six caregivers (54%) thought they were useful and 40 (47%) interesting. Eighty-three (87%) would potentially fill them out again, 50 (58%) would do that gladly.

The self-reported children and adolescents needed, on average, 14.5 minutes to fill out both questionnaires. Twenty-one (48%) of them found the questionnaires interesting and 16 (36%) useful. Only 3 participants (7%) considered the questionnaires stupid. The items made 2 participants (5%) feel uncomfortable, 12 (27%) felt a bit embarrassed, but were not bothered by them, and 30 (68%) were not bothered by them not at all. Forty-one (93%) individuals would potentially fill out the questionnaires again, 20 (45%) would do that gladly.

4 DISCUSSION

This is the first study that assessed HRQoL of Slovenian children and adolescents with CP. Overall, their self- and proxy reported HRQoL is good, which is similar to the findings in studies evaluating similar patient populations in other countries (5, 10, 11, 18).

Recently published data about HRQoL in children and adolescents with CP (SPARCLE I and II studies) convincingly show scores similar to the general age-matched population, with the exception of social support and peers domains (5). In these studies, HRQoL of children was a good predictor of HRQoL later in adolescence (5). In the present study, we compared HRQoL of children and adolescents in cross-sectional cohorts simultaneously and with the same HRQoL tool. We found significantly lower HRQoL scores in adolescents, as compared to those in children. QoL issues tend to change over time, as independence, relationships, sexuality and acceptance of disability increasingly gain importance in adolescence (20). This could potentially explain lower self- and proxy-perceived HRQoL scores in adolescents. However, our sample of self-reporting participants was small, and we did not include a control group of healthy children.

Pain is a well-recognised predictor of decreased participation and poor HRQoL (5, 10, 11, 21). In our study, it was related to lower scores in all groups. Disturbed sleep negatively associated with proxy-reported HRQoL, but it was a rare complaint with no impact in the self-reporting group. Our study was unable to show the impact of therapeutic interventions on HRQoL. This could be explained by a well-organized Slovenian neurodevelopmental network that enables all children with developmental delay to start a specific neurodevelopmental treatment at an early age, most of them within the first 6 months of life (19). Whereas most interventions aim to improve physical independence, most have a limited effect on HRQoL. It is therefore very important to design accessible interdisciplinary therapeutic approaches, which would better address HRQoL issues.

Children and adolescents rated their HRQoL higher than their caregivers in all domains, which is similar to the findings of other studies (11, 22). One reason for that could be that children focus on their abilities, as their disability has always been a part of their functioning, while caregivers tend to compare the abilities of their children to those of healthy children (23). Regardless of their level of disability, almost all children in our study resided at home, which indicates a high level of family engagement. It is possible that lower proxy scores reflect caregivers' psychological burden. It has been recognized that caregivers' well-being is significantly impaired, compared to matched adults from the general population (24). No proper supportive family-centred services or parent networks currently exist in Slovenia. Surprisingly, despite various unfavourable socio-economic factors, such as high maternal unemployment rate, none of them were significantly associated with HRQoL (20).

Consistent with other studies, there were limitations regarding Social Inclusion and Independence, which are, to some extent, expected due to the nature of the disease (5, 9). Whereas most interventions aim to improve physical independence, a lot more could be done in the wider society to improve social inclusion of children with CP.

This study has some limitations: The number of patients in subgroups was relatively small, there was no comparative sample of healthy children and adolescents, a non-personal approach was used, and a generic questionnaire was selected as a primary assessment tool.

5 CONCLUSIONS

This was the first study to assess HRQoL of children with CP in Slovenia. It is important to follow HRQoL of CP patients closely throughout their childhood and adolescence, and pay attention to the factors that might be negatively associated with it, such as pain. It is also important that therapeutic interventions are well-balanced and use integrated multidisciplinary approach to improve participation and social inclusion of individuals with CP.

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CONFLICTS OF INTEREST

The authors declare that no conflicts of interest exist.

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ETHICAL APPROVAL

The study was approved by the Slovenian National Ethics Committee, application number: 122/05/13. After receiving verbal and written information about the study, all caregivers signed written consent.

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