



ELSEVIER



REVIEW

Quality of life of children treated for cleft lip and/or palate: A systematic review[☆]

Anne F. Klassen^{a,*}, Elena Tsangaris^b, Christopher R. Forrest^c,
Karen W.Y. Wong^c, Andrea L. Pusic^d, Stefan J. Cano^e, Iqra Syed^a,
Meghna Dua^f, Sonika Kainth^f, Jessica Johnson^a, Tim Goodacre^g

^a Department of Pediatrics, McMaster University, Room HSC 3N27, 1200 Main Street West, Hamilton, Ontario L8S 4J9, Canada

^b Department of Clinical Epidemiology and Biostatistics, McMaster University, Hamilton, Ontario L8N 3Z5, Canada

^c Division of Plastic Surgery, Hospital for Sick Children, 555 University Ave, Toronto, Ontario M5G 1X8, Canada

^d Plastic and Reconstructive Surgery, Memorial Sloan–Kettering Cancer Center, Room MRI-1007, 1275 York Ave, New York, NY 10065, USA

^e Clinical Neurology Research Group, Peninsula College of Medicine and Dentistry, Room N13 ITTC, Tamar Science Park, Davy Road, Plymouth, Devon PL6 8BX, UK

^f Department of Health Sciences, McMaster University, Hamilton, Ontario L8N 3Z5, Canada

^g Spires Cleft Center, Oxford Radcliffe Children's Hospital, Oxford OX3 9DU, UK

Received 10 August 2011; accepted 6 November 2011

KEYWORDS

Quality of life;
Systematic review;
Cleft lip;
Cleft palate;
Children;
Pediatrics

Summary *Objective:* Cleft lip and/or palate (CLP) is the most common congenital craniofacial anomaly. As a first step toward developing a quality of life (QOL) questionnaire for CLP patients, our team conducted a systematic literature review to identify studies that measured child- or proxy-reported outcomes of CLP.

Design: PUBMED, CINAHL, EMBASE and PsycINFO were searched from their inception to July 2010 to identify studies that measured health-related concepts in CLP patients. Abstract and title screening was performed by two screeners. Full texts of all potentially relevant papers were obtained and examined by two reviewers. We identified publications that measured health concepts and categorized them to form a preliminary conceptual framework of CLP QOL issues.

Results: A total of 4594 publications were identified. Twenty-six studies met our inclusion criteria. Research involved CLP patients living in nine countries with sample sizes ranging from

[☆] *Meeting/conference presentations:* Findings have been presented at the following: Canadian Society for Epidemiology and Biostatistics National Student Conference in Montreal, Quebec, Nov 2011 and the International Society of Quality of Life Research conference in Denver, Colorado, Oct 2011.

* Corresponding author. Tel.: +1 905 521 2100x73775; fax: +1 905 521 4981.

E-mail address: aklass@mcmaster.ca (A.F. Klassen).

23 to 661. Health concepts were measured using 29 different questionnaires. No patient-reported outcome (PRO) instrument measuring QOL concerns of CLP patients currently exists. CLP-specific health concepts measured to date were categorized into a preliminary conceptual QOL framework with the following categories: physical, psychological and social health.

Conclusions: Our review has helped to identify areas of health that have been well researched using either a patient or proxy patient-reported outcome instrument (e.g., self-concept; behavior) and areas where more research is required.

© 2011 British Association of Plastic, Reconstructive and Aesthetic Surgeons. Published by Elsevier Ltd. All rights reserved.

Introduction

Cleft lip and/or palate (CLP) is the most common congenital craniofacial abnormality; occurring in approximately 1:1000 live births.¹ Children born with CLP may be affected by a combination of facial difference, swallowing and speech disorders, and various disturbances of dentition and growth. The course of treatment is complex and spans childhood, adolescence and frequently adulthood. Various methods of surgical care for CLP patients currently exist (e.g. cleft lip/palate repair, alveolar bone grafting, orthognathic surgery etc). Although much of the primary management of patients is surgically driven, the overall goal of treatment is to achieve psychological and social well-being for the individual and his or her family.

The complex interaction between individuals is intimately affected by appearance and 'visible differences' and involves communication, perception of self, and others' perceptions of oneself. Clefting that involves the lip and nose imposes evident facial difference; therefore, as a consequence, cleft related facial difference could be expected to have a profound impact on social interactions and quality of life (QOL) of patients. To adequately measure the QOL of CLP patients, a scientifically sound and clinically meaningful patient-reported outcome (PRO) instrument, that specifically addresses the unique issues of CLP patients, is required. PRO instruments, generic and/or disease-specific, are questionnaires that quantify QOL and/or other significant outcome variables (e.g. satisfaction, symptoms, function) from the patient's perspective.² Generic PRO instruments are ones developed specifically for use with any patient group and therefore allow direct comparison across disease groups, or with healthy children. Disease- or condition-specific PRO instruments address problems specific to a single disease or treatment group. These latter instruments include content that is more relevant to a given patient group, and are therefore better at detecting measuring change in health.² The lack of a PRO tool specific to children with orofacial clefts was identified as an important research gap in the January 2006 workshop entitled "Prioritizing a Research Agenda for Orofacial Clefts" held by the National Center on Birth Defects and Developmental Disabilities at the Centers for Disease Control.³ In a systematic review of literature published up to May 2007, our team was not able to identify any PRO instrument designed specifically for CLP patients.⁴

Developing a PRO instrument is a multi-phase undertaking that involves a literature review, in-depth

qualitative interviewing with the population of interest and input from experts working with the desired patient population.^{5,6} These three sources of information can provide a comprehensive understanding of important issues for patients and can inform the development of concepts, scales and items needed to measure patients' health concerns. That there is currently no comprehensive PRO instrument measuring QOL for CLP patients might well reflect the complexity involved in developing such an instrument – there are a number of quite distinct domains of QOL involved (e.g., appearance, speech, facial growth, psychosocial interaction). In addition, the relative importance of how these various domains might change over the period of a child's development to adulthood is unknown. A robust PRO instrument for the CLP population would need to be one that could address the domains of importance and be cognizant of any changes in aspects of QOL as the child matures.

As a first step toward developing a PRO instrument covering QOL issues for CLP patients, our team conducted a systematic literature review to identify all studies where either the child or parent had completed a PRO instrument that was designed to measure some aspect of child health. Our aims were three-fold as follows: (1) to identify all health concepts important to CLP patients; (2) to identify any PRO questionnaires developed specifically to measure QOL concerns of CLP patients; and (3) to identify important determinants of QOL for CLP patients.

Methods

Our review was guided by the PRISMA statement.⁷ PUBMED, CINAHL, EMBASE and PsycINFO were searched from the inception of each database to July 2010. A medical librarian developed the search strategy after consultation with the research team who developed a comprehensive list of topics for searching (details about the search strategy are available from authors). The search was limited to English language articles and articles focused primarily on children (birth to 21 years of age). In each database, a similar search strategy was used.

Abstract and title screening was performed by two screeners who worked independently. A third reviewer resolved any discrepancies of opinion. The full text of all potentially relevant papers were obtained and examined independently by two reviewers using the following inclusion criteria to identify relevant articles: (1) the sample included cleft lip and/or palate patients (if the study

included children with other health conditions, results for the CLP sample had to be presented separately); (2) the study sample included children (sample upper age limit was 21 years); and (3) the study described the use of a child- or parent-completed questionnaire measuring specific health constructs and compared findings for the CLP sample with population norms, or a comparison or control group.

Studies that used an ad hoc instrument (i.e., one without published evidence of a development or validation process) or a modified version of a questionnaire that was not itself validated were excluded.

Findings from studies that used an instrument that combined domain scores for different aspects of health (e.g., QOL summary score that combine scores across physical, psychological and/or social health domains) were not extracted given that it is unclear what summary scores actually measure. For the subset of studies that measured a determinant of some aspect of the child's QOL, the study had to present a *p*-value for a statistical test of the relationships between the concept and determinant.

Citations for included articles were examined to identify additional articles. One reviewer extracted study results from each eligible paper, and a second reviewer checked data extraction. In order to categorize the health concepts, we examined each questionnaire identified by our review in order to determine the constructs being measured. Health concepts were organized into a conceptual framework of QOL outcomes studied to date in CLP patients.

Information extracted for each determinant included the name of the determinant, the direction of the relationship with the health-related concept, and whether or

not the relationship between the determinant and the health concept was statistically significant (i.e., *p*-value for the relationship was less than 0.05). As the purpose of this review was to identify the evidence base (or lack thereof) to guide future pediatric plastic surgery research, we did not exclude research on the basis of poor methodological quality, nor did we assess the quality of each study.

Results

A total of 4594 publications were identified by our search strategy (see Figure 1). We found 26 articles that met our inclusion criteria. A citation review of included publications did not add any additional publications.

Characteristics for each publication included in our review appear in Table 1. Research involved CLP patients living in 9 countries. The sample sizes ranged from 23 to 661. QOL concepts were measured using 29 different questionnaires (see Table 2). The most frequently measured construct was that of self-esteem or self-concept, which was measured in 15 studies^{9,11,12,16,17,19–22,24,25,29,31–33} using 10 different questionnaires. Behavior, also frequently assessed, was measured in 13 studies^{8,10,17,18,23,24,26–28,30–33} using 8 different questionnaires. Three generic PRO instruments were used. Our search did not identify a PRO questionnaire developed specifically to address the health concerns of CLP patients.

A total of 18 studies^{8–13,16–19,21,23–26,29,31,33} measured some aspect of patient QOL compared with population norms or comparison groups. The main health concepts

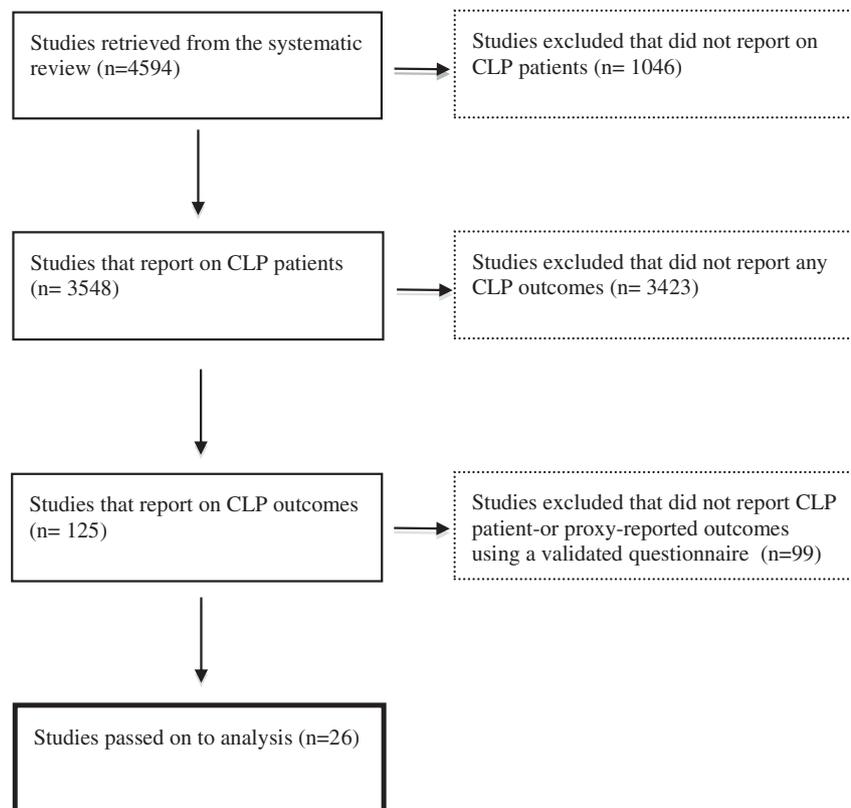


Figure 1 Flow chart of studies included and excluded.

Table 1 Study characteristics.

Author	Country	Type of cleft (%)	Gender (%)	Age range	Sample size	Type of comparison sample	Determinants
Berger ⁸	England	CLP (49); CP (37); CL (14)	Male (51) Female (49)	11 to 16	145	Normative data	^a Age, gender, cleft type
Boes ⁹	USA	CLP (50); CP (23); CL (27)	Male (100)	7 to 12	30	Comparison group	Brain structure
Brand ¹⁰	Switzerland	CLP (100)	Male (72) Female (28)	6 to 16	32	Comparison group	Not applicable
Broder ¹¹	USA	CLP (33); CP(35); CL (33)	Male (50) Female (50)	6 to 9	40	Comparison group	^a Cleft type
Cheung ¹²	Hong Kong	CLP (100)	Male (49) Female (51)	10 to 16	94	Comparison group	Not applicable
Damiano ¹³	USA	CLP (48); CP(24); CL (28)	Male (57) Female (43)	2 to 12	104	Normative data	Age, cleft type, ^a parent marital status, gender, ^a people in household, ^a household income
Feragen ¹⁴	Norway	CLP & CLA (63); CP (37)	Male (61) Female (39)	9 to 12	268	Not applicable	^a Comorbidity, cleft type, gender
Feragen ¹⁵	Norway	CLP (47); CP(35); CLA (18)	Not clear	10 & 16	661	Not applicable	Comorbidity, ^a gender, ^a cleft type
Gussy ¹⁶	Australia	CLP (100)	Male (61) Female (39)	12 to 17	23	Normative data	Not applicable
Hunt ¹⁷	Ireland	CLP & or CL (100)	Male (65) Female (35)	8 to 21	160	Comparison group	Not applicable
Hunt ¹⁸	Ireland	CLP (100)	Male (66) Female (34)	8 to 18	129	Comparison group	Not applicable
Kapp-Simon ¹⁹	USA	CLP (62); CP(30); CL(8)	Male (58) Female (42)	5 to 9	50	Comparison group	Cleft type, gender
Kramer ²⁰	Germany	CLP (33); CP(31); CL (37)	Male (59) Female (41)	5 to 7	147	Not applicable	^a Breast feeding, ^a parents' profession, ^a age of father, ^a siblings, ^a number of non-cleft operations, ^a financial status, cleft type
Kramer ²¹	Germany	CLP (42); CP(29); CL (29)	Male (61) Female (39)	8 to 12	132	Comparison Group	^a Cleft type, ^a gender
Leonard ²²	USA	CLP (70); CL (2); CP (29)	Male (64) Female (36)	8 to 18	105	Not applicable	^a Gender, age
Millard ²³	USA	CP(29); CLP (71)	Male (54) Female (46)	8 to 17	65	Normative data	^a Cleft type
Murray ²⁴	UK	CL & or CLP (100)	Male (62) Female (38)	Mean age 7.7	93	Comparison group	Cleft type
Persson ²⁵	Sweden	CLP (38); CL(29); CP(29); Unknown (4)	Male (49) Female (51)	17 to 20	55	Comparison group	Cleft type
Richman ²⁶	USA	CLP (100)	Not reported	14 to 17	36	Normative data	Not applicable
Richman ²⁷	USA	CLP (57); CP (43)	Male (52) Female (48)	6 to 12	65	Not applicable	Cleft type, ^a gender

Richman ²⁸	USA	CL (48); CLP (52)	Male (57) Female (43)	4 to 12	44	Not applicable	Cleft type, ^a gender
Sagheri ²⁹	Germany	CLP (47); CL (10); CP (42)	Male (52.5) Female (47.5)	4 to 7	59	Normative data	Cleft type
Schneiderman ³⁰	USA	CL/CLP (100)	Not reported	Not reported	58	Not applicable	^a Age, ^a gender
Stifer ³¹	USA	CLP (32); CL/CP (68)	Male (44) Female (56)	8 to 15	34	Comparison group	Not applicable
Starr ³²	USA	CLP (57); CP (43)	Male (65) Female (35)	13 to 18	94	Not applicable	Cleft type, age, ^a gender
Warschausky ³³	USA	CLP (74); CP (11); CL (7); CLA (7)	Not reported	5 to 18	27	Population norms	

Notes: CLP: Cleft lip and palate; CP: Cleft palate only; CL: Cleft lip only; CLA: Cleft lip alveolus.
^a statistically significant determinants for each study.

studied to date were categorized into a conceptual framework covering physical, psychological and social health (see Figure 2).

Eighteen publications^{8,9,11,13–15,19–25,27–30,32} measured 14 different determinants, ten of which were only measured in one study. The most commonly studied determinants included cleft type, child gender and child age. Table 3 shows each determinant along with the number of publications where it was included as a factor and total number of analyses where it was examined along with the number of statistically significant relationships with physical, psychological and social health of patients. Below we summarize the key health concepts and main determinants of QOL for each outcome.

Physical health

Six publications^{10,13,20,21,29,33} measured an aspect of physical health including physical function, pain, communication and general health of CLP patients. These publications show that scores for CLP patients did not differ from that of normative samples on any of the measures except for communicative competencies. More specifically, CLP patients reported more communication problems on two subscales (social environment and proactive behaviors) of the Participation in Everyday Life Communication Questionnaire (PIELCQ) than did an age and gender matched control group.¹⁰

Determinants of physical health were reported in four publications.^{13,20,21,29} One study measured QOL using the PedsQL 4.0 and reported better physical health scores in CLP patients in households with a higher income, a higher number of people and where the parents were married.¹³ Another study using the KINDL showed higher physical well-being scores for patients with older fathers.²⁰ When studied, gender was significant in one study, with boys reporting better physical function than girls.²¹ No significant difference by gender was found in the other study.¹³ Age¹³ and cleft type^{13,20,21,29} were not significant determinants in relation to physical health.

Psychological health

Psychological health was examined in 25 publications.^{8–12,14–33} Constructs studied under this health domain included the following: self-concept or self-esteem, behavior, satisfaction with appearance, psychological function and cognitive function.

Self-concept and self-esteem

Findings about self-concept and self-esteem were reported in 15 publications.^{9,11,12,16,17,19–22,24,25,29,31–33} Thirteen publications^{9,11,12,16,17,19,21,22,24,25,29,31,33} reported findings that compared CLP patients with population norms and/or a comparison sample. In most publications,^{9,16,19,21,22,24,25,29,31} but not all,^{11,12,19} the scores for CLP patients were either equivalent to or better than scores for control groups and/or normative data.

Ten publications^{9,11,19–22,24,25,29,32} examined a determinant of self-concept or self-esteem with cleft type (i.e.,

Table 2 Patient- and proxy-reported outcome instruments.

Name of questionnaire	Constructs measured	No. of studies
Behavior Problem Checklist ^{26–28,30}	Behavior	4
Child Behavior Checklist ^{18,24,31}	Behavior	3
Childhood Experience Questionnaire ^{8,14,15}	Social life experience	3
Children's Depression Inventory or Beck Depression Inventory ¹⁷	Depression	1
Children's Health and Illness Profile ³¹	Health and well-being	1
Child Health Questionnaire version PF28 ³³	Health-related quality of life	1
Comprehensive Assessment for Symptoms and History (CASH) Subscales ⁹	Social function	1
Culture-Free Self-Esteem Inventory ¹²	Self-esteem	1
Harter Self-Perception Profile for Children/Adolescents ³¹	Self-perception	1
Harter Social Support Scale for Children and Adolescents ³¹	Social support and positive regard from others	1
KINDL or KINDL-Kiddy ^{20,21,28}	Quality of life	3
Missouri Children's Behavior Checklist ³²	Behavior	1
Participation in Everyday Life Communication Questionnaire ¹⁰	Communicative competencies	1
Pediatric Behavior Scale ²³	Depression/anxiety, conduct and cognition	1
Pediatric Quality of Life Inventory ¹³	Health-related quality of life	1
Personality Inventory for Children Subscales ¹⁴	Psychological adjustment	1
Pictorial Scale of Perceived Competence and Social Acceptance for Young Children ²⁴	Self-concept/self-esteem	1
Piers-Harris Self-Concept Scale ²²	Self-concept	1
Primary Self-Concept Inventory ^{11,19}	Self-concept	2
Reynolds Child Depression Scale or Reynolds Adolescent Depression Scale ²³	Depression	1
Revised Children's Manifest Anxiety Scale ²³	Anxiety	1
Rosenberg Self-Esteem Scale or Self-Esteem Index ^{17,32}	Self-esteem	2
Satisfaction with Appearance Questionnaire ^{8,15}	Satisfaction with appearance	2
Self-Description Questionnaire I ⁹ or II ¹⁶	Peer relations and self-concept	2
Social Skills Rating System Questionnaire ³¹	Behavior and personality	1
State Trait Anxiety Inventory for Children or State Trait Anxiety Inventory ¹⁷	State and trait anxiety	1
Strengths and Difficulties Questionnaire ^{8,10}	Behavior, peer relations, emotional symptoms	2
Tennessee Self Concept Scale ²⁵	Self-concept	1
Youth Self-Report or Young Adult Self-Report ¹⁷	Behavior	1

CL, CP and/or CLP)^{11,19–21,24,25,29,32} the most frequently studied factor. In most publications where cleft type was examined^{19,20,24,25,29,32} scores for patients were not related with cleft type. In contrast, one study¹¹ used the Primary Self-concept Inventory and showed that patients with CLP had lower self-concept on two subscales (social and intellectual self-concept) as well as the total score compared with patients with CL or CP only. Gender as a determinant was studied in relation to self-concept or self-esteem in four publications.^{19,21,22,32} Female patients reported lower scores on two of eight subscales of the Piers-Harris Self-Concept Scale compared with male patients,²² but in another study, female patients reported higher self-esteem on the KINDL than did males.²¹ In the other two publications, gender was not related with self-concept scores.^{19,32} Age was considered in two studies.^{22,32} While no relationship with age was found in one publication,³² the other

publication reported that older boys had higher scores for self-concept than younger boys, and older girls had lower self-concept scores than younger girls.²²

Behavior

Findings about behavior were reported in 13 publications.^{8,10,17,18,23,24,26–28,30–33} Five publications^{8,10,23,24,33} reported no difference in behavior scores when CLP patients were compared with control groups or normative data. Four publications^{17,18,26,31} reported a significant difference in some aspect of behavior.

Determinants of behavior were reported in six publications.^{23,24,27,28,30,32} Five publications reported that cleft type was not related with behavior scores.^{23,24,27,28,32} Age was investigated as a determinant in two publications^{30,32} and one reported more behavior problems in younger children compared with older children.³² Three studies^{27,30,32}

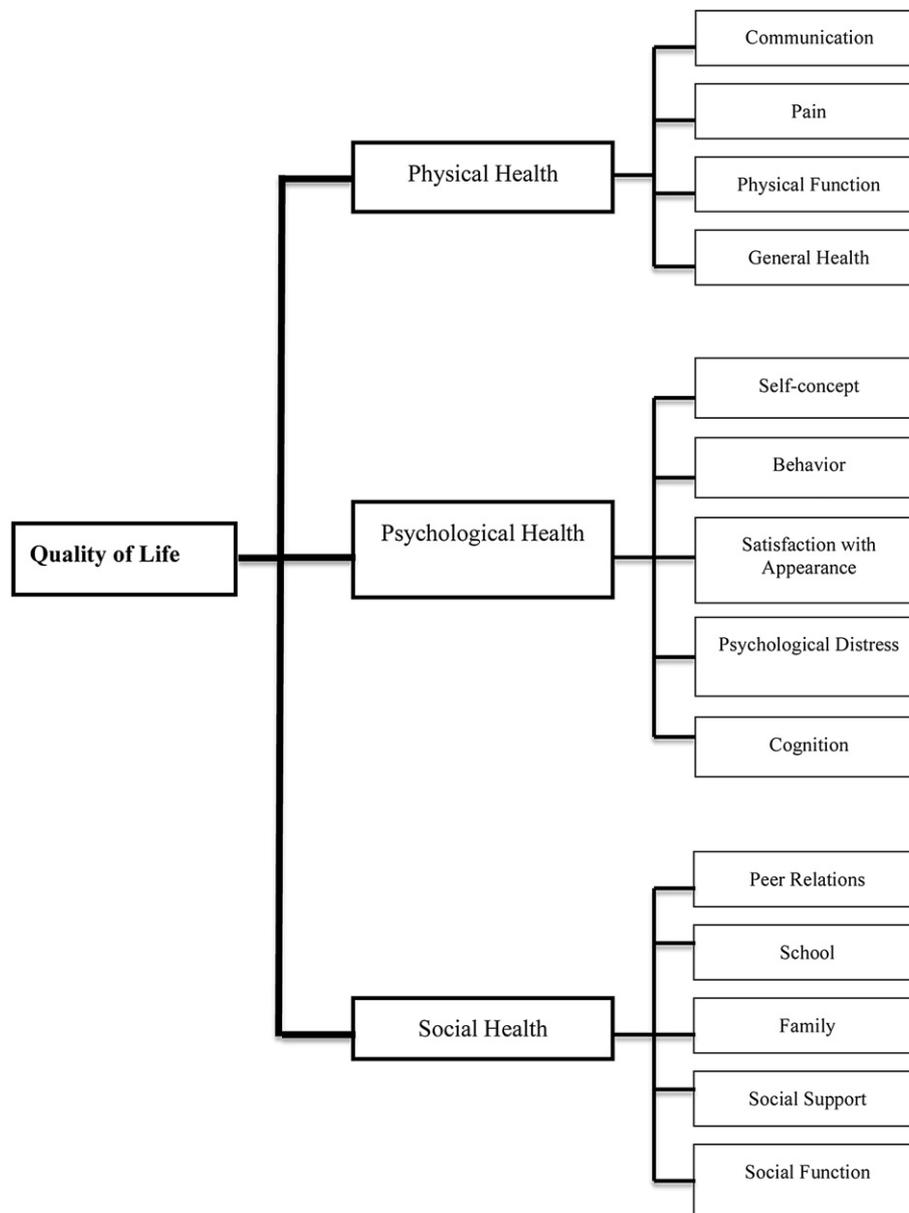


Figure 2 Conceptual framework of CLP QOL issues.

investigated gender and reported more behavior problems for boys than girls.

Appearance

Appearance was measured in two studies using a standardized questionnaire called the Satisfaction with Appearance Questionnaire.^{8,15} Compared with population norms, in one publication CLP patients were more satisfied with their facial appearance with the exception of their lips.⁸ In the other study¹⁵ determinants of satisfaction with appearance were studied in relation to age and gender, and the authors found that teenaged girls with non-visible clefts were more satisfied with their appearance than girls with visible clefts (no difference was found by cleft type for younger female patients or younger and teenaged male patients). The authors also reported that teenage girls were less satisfied with their appearance than were teenage boys.¹⁵

Psychological distress

Psychological distress was measured in 9 publications.^{8,10,14,17,20,21,23,29,33} One study reported that there were significantly more depressive symptoms for CLP patients on the Child Depression Inventory compared with a control group.¹⁷ Two studies^{14,15} investigated comorbidities as a determinant. One study reported that CLP patients with comorbid health problems had a mean score for psychological distress in the clinical range on the Personality Inventory for Children.¹⁴ In a third study, while the sample of teenagers with CLP did not differ from normative data on the Strengths and Difficulties Questionnaire, their mothers scored twice as many of them above the cutoff scores for adjustment difficulties compared with normative data.⁸

Determinants of emotional distress were investigated in 5 publications.^{14,20,21,23,29} Cleft type was studied three

Table 3 Determinants of QOL in research with CLP patients.

Determinant	Total number of analyses conducted for each determinant	Construct where relationship with determinant was statistically significant
CLP Variables		
Cleft type ^{8,11,13–15,19–21,23–25,27–29,32}	33	Psychological, ^{11,15,23} Social, ^{15,21}
Child Variables		
Gender ^{8,13–15,19,21,22,27,30,32}	18	Psychological ^{15,21,22,27,28,30,32} Social, ^{15,21} Physical ²¹
Age ^{8,13,22,30,32}	9	Psychological, ^{22,30} Social ⁸
Comorbidity ^{14,15}	4	Psych ¹⁴
Extent of breast feeding ²⁰	1	Social ²⁰
Number of non-cleft operations ²⁰	1	Physical ²⁰
Brain structure ⁹	3	Social ⁹
Family Variables		
Age of father ²⁰	2	Physical, ²⁰ Social ²⁰
Household income ¹³	3	Physical ¹³
Higher number of people in household ¹³	3	Physical ¹³
Parents married ¹³	3	Physical ¹³
Professional activity of parents ²⁰	1	Psychological ²⁰
Sibling present ²⁰	1	Psychological ²⁰

times^{20,23,29} with one publication reporting some differences in symptoms of anxiety and depressive by cleft type.²³ A study that used the KINDL PRO instrument reported that psychological health was better in female patients compared with males.²¹ A separate study found that psychological distress scores were higher in CLP patients on the Personality Inventory for Children for CLP patients with a comorbidity (e.g., ADHD, autism, dyslexia) publications.¹⁴

Cognitive function

One study by Millard et al. (2001) looked at cognitive attributes and found that the CLP sample had higher mean scores suggesting more problems in this area compared with normative data.²³

Social health

Ten publications^{8–10,14,15,20,21,29,31,33} reported findings on variables that were categorized under the heading of social health. Social health included the following concepts: social function, peer relations, school function, family function and social support.

Social function

Social function was measured with scales that assessed variables such as social experience, social adjustment and social competence. Three publications^{8,9,31} reported findings for CLP compared with population norms or a control group and provide mixed results. One publication³¹ found that CLP patients did not differ from a control group on subscales of the Child Behavior Checklist Social Competence Scale and Social Skills Rating System Questionnaire. In another publication, CLP patients reported better social function in terms of a more positive social environment compared with normative data.⁸ In contrast, social adjustment measured using the Social Adjustment Scale

(SAS) of the Comprehensive Assessment of Symptoms and History (CASH) was lower in a sample of CLP patients compared with a control group.⁹

Four studies examined a determinant of social function.^{8,9,14,15} In terms of gender, two studies^{8,14} found gender and social function were not related, while another reported that teenage girls reported more peer harassment than teenage boys.¹⁵ In terms of cleft type, two studies found this to not be a determinant of social function,^{8,14} while one study found that sixteen year-old girls with non-visible clefts reported less peer harassment than those with visible clefts.¹⁵

Peer relations

Six studies reported on peer relations.^{8–10,20,21,29} Two studies found no difference in peer relations using the Strength and Difficulties Questionnaire when CLP patients were compared with a control group or population norms.^{8,10} Another study, using the Self-Description Questionnaire-I, reported lower scores for the CLP patients compared with a healthy volunteer sample.⁹ Two publications that used a PRO instrument measuring QOL^{21,29} reported that CLP patients had similar scores for peer relations when compared with population norms.

Cleft type^{20,21,29} and gender²¹ were not significant factors in studies that examined these variables in relation to peer relations, although in one study that used the KINDL, girls in the CP only subgroup reported better scores for peer relations compared with boys with CP only.²¹

School function

One study examined school functioning and did not find a difference between CLP patients and population norms.²⁹ This same study found no relationship between cleft type and school function.²⁹

Family function

Four publications examined some aspect of family function including family relations, cohesion, and activities.^{20,21,29,33} CLP patients did not differ on outcomes relating to family when compared with population norms.^{29,33} Cleft type was studied in relation to family function but while no relationship was detected in two studies,^{20,29} it was an important factor in another study.²¹ Gender differences were found in relation to family function scores, with girls reporting better scores than boys.²¹

Social support

Social support was assessed using the Harter Social Support Scale which measures support received from parents, teachers, close friends, and classmates. This publication reported that there was no difference in scores when CLP patients were compared with a control group.³¹

Discussion

This systematic review synthesized evidence about QOL concepts from 26 studies of children with CLP. No PRO instrument measuring QOL concerns of CLP patients currently exists. However, using findings from studies using a range of questionnaires completed by the child and/or parent, we identified a number of health concerns of CLP patients and have developed a preliminary conceptual framework of QOL. Furthermore, our review has identified areas of QOL that have been well researched (physical health, self-concept/self-esteem, behavior, psychological distress and peer relations) and areas where more research is required (e.g., satisfaction with appearance, cognitive function, family function, social function, social support, and school function). We also identified a limited number of determinants of QOL that have received research attention to date (e.g., child age, gender, cleft type).

Conventional methods of outcome assessment, such as the reporting of complications data or photo analysis, represent the healthcare provider perspective. While these data remain important, they are no longer sufficient when considered alone. A more comprehensive approach involves capturing the patient perspective using questionnaires to measure important PROs. Our systematic review focused specifically on the identification of health concerns of CLP patients from the perspective of the patient and/or the patient's parent. Our focus was on PRO data to the exclusion of any other type of outcome data. We recognize that a number of important health outcomes associated with cleft care (e.g., hearing, speech, dental) are missing from our framework as research that captures the patient's point of view for such outcomes is currently lacking.

While our review confirmed that there are currently no PRO instruments developed to capture the specific concerns of CLP patients, there are a number of PRO instruments developed, or adapted for use in children with various dental, oral and craniofacial conditions. These instruments include the following: Youth Quality of Life Instrument—Craniofacial Surgery (YQOL-CS),³⁴ Youth Quality of Life Instrument—Facial Differences (YQOL-FD),³⁵ Child Oral Health Quality of Life Questionnaire (COHQOL),³⁶ Child Oral Health Impact Profile (COHIP),³⁷ Pediatric Voice

Outcome Survey (PVOS),³⁸ and the Pediatric Voice-Related Quality-of-Life (PVRQOL) survey.³⁹ The development process and psychometric properties of these instruments has been reviewed by our team and we reported that only the YQOL-FD team followed international guidelines for health outcomes instrument development and that this instrument alone could be recommended for use.⁴ However, the YQOL-FD was developed to measure QOL issue in adolescents with a broad range of craniofacial conditions and as such was not developed to cover the specific concerns of the CLP population. In a descriptive study using the YQOL-FD, the developers described how stigma experiences were frequently reported by youth with facial differences.⁴⁰ In addition, using the generic QOL measure (Youth Quality of Life Research Version – Revised) in a study of 56 adolescents aged 11–18 years with facial differences (including 24 CLP patients), the authors found that youth with facial differences reported lower overall QOL scores compared with a sample of 116 healthy peers.⁴¹

To enhance the QOL of children with CLP, it is essential to identify the most important determinants of QOL. Our review was used to identify factors (child age, gender, cleft type) studied in relation to health outcomes. Lach et al. (2006) have developed a conceptual framework to account for determinants of QOL in children with epilepsy, which they describe as including treatment, child, family and community variables, as these determinants form part of the broader context within which a child lives and has an impact on QOL.⁴² This conceptual framework has since been used to organize findings about determinants of QOL for children with cancer and childhood cancer survivors.⁴³ Future research with CLP patients could examine some of the unexplored variables described in the Lach et al framework as these may help to explain the different patterns of QOL of children with CLP. In addition, Yazdy et al. suggested a range of factors specific to orofacial cleft patients that could influence the QOL and represent areas for future research (e.g., access to an interdisciplinary team of specialists, the timing and type of surgery, family out-of-pocket costs and effect on caregivers).³

Our systematic review identified that while there is a lack of a CLP-specific PRO questionnaire for CLP patients, 28 different questionnaires have been used to measure outcomes from the perspective of the patient and/or parent, with the majority of questionnaires used in only one study. The content (items and scales) of these 28 questionnaires vary substantially indicating that there are very different ideas about what constitutes the most important components of health for this patient population as well as very little consensus on which questionnaires should be used to measure patient outcomes. Importantly, the use of so many different questionnaires with CLP patients makes it difficult to compare findings across studies. A standardized approach to QOL measurement, where everyone uses only the most scientifically sound and clinically meaningful PRO instrument, is needed and would lead to a better understanding of the health impact of CLP on children of different ages.

In order to develop a PRO instrument for CLP patients, in addition to a literature review, in-depth qualitative interviewing with the patient population and expert input from healthcare professionals that work with the patient

population is needed in order to identify patients' health concerns.^{5,6} A PRO instrument for CLP patients needs to measure the distinct domains of QOL of importance to patients (e.g., appearance, speech, facial growth, psychosocial interaction) and to do this in a way that captures differences in aspects of QOL as patients develops and matures. Our team has commenced with collecting this information and is currently developing a PRO instrument for CLP patients.⁴⁴

Conflict of interest/funding

None.

Acknowledgments

Anne Klassen is a recipient of Canadian Institute of Health Research career award. Elena Tsangaris is a recipient of a McMaster Child Health Research Institute scholar award.

References

- Dixon MJ, Marazita ML, Beaty TH, et al. Cleft lip and palate: understanding genetic and environmental influences. *Nat Rev Genet* 2011;**12**:167–78.
- Pusic AL, Lemaine V, Klassen AF, et al. Patient-reported outcome measures in plastic surgery: use and interpretation in evidence based medicine. *Plast Reconstr Surg* 2011;**127**:1361–7.
- Yazdy MM, Honein MA, Rasmussen SA, et al. Priorities for future public health research in orofacial clefts. *Cleft Palate Craniofac J* 2007;**44**:351–7.
- Klassen A, Pusic A, Skarsgard E. Clinical research in pediatric plastic surgery and systematic review of quality of life questionnaires. *Clin Plast Surg* 2008;**35**:251–67.
- Cano S, Klassen A, Pusic A. The science behind quality-of-life measurement: a primer for plastic surgeons. *Plast Reconstr Surg* 2009;**123**:98E–106E.
- U.S. Food and Drug Administration. *Patient reported outcome measures: use in medical product development to support labeling claims*. Available at: <http://www.fda.gov/downloads/Drugs/GuidanceComplianceRegulatoryInformation/Guidances/ucm071975.pdf>; 2009.
- Liberati A, Altman DG, Tetzlaff J, et al. The PRISMA statement for reporting systematic reviews and meta-analyses of studies that evaluate health care interventions: explanation and elaboration. *PLoS Med* 2009;**6**(7):e1000100.
- Berger Z, Dalton L. Coping with a cleft: psychosocial adjustment of adolescents with a cleft lip and palate and their parents. *Cleft Palate Craniofac J* 2009;**46**:436–43.
- Boes AD, Murko V, Wood JL, et al. Social function in boys with cleft lip and palate: relationship to ventral frontal cortex morphology. *Behav Brain Res* 2007;**181**:224–31.
- Brand S, Blechschmidt A, Müller A, et al. Psychosocial functioning and sleep patterns in children and adolescents with cleft lip and palate (CLP) compared with healthy controls. *Cleft Palate Craniofac J* 2009;**46**:124–35.
- Broder H, Strauss RP. Self-concept of early primary school age children with visible or invisible defects. *Cleft Palate J* 1989;**26**:114–7.
- Cheung LK, Loh JSP, Ho SMY. Psychological profile of Chinese with cleft lip and palate deformities. *Cleft Palate Craniofac J* 2007;**44**:79–86.
- Damiano PC, Tyler MC, Romitti PA, et al. Health-related quality of life among preadolescent children with oral clefts: the mother's perspective. *Pediatrics* 2007;**120**:e283–90.
- Feragen KB, Borge AI, Rumsey N. Social experience in 10-year-old children born with a cleft: exploring psychosocial resilience. *Cleft Palate Craniofac J* 2009;**46**:65–74.
- Feragen KB, Borge AI. Peer harassment and satisfaction with appearance in children with and without a facial difference. *Body Image* 2010;**7**:97–105.
- Gussy M, Kilpatrick N. The self-concept of adolescents with cleft lip and palate: a pilot study using a multi-dimensional/hierarchical measurement instrument. *Int J Paediatr Dent* 2006;**16**:335–41.
- Hunt O, Burden D, Hepper P, et al. Self-reports of psychosocial functioning among children and young adults with cleft lip and palate. *Cleft Palate Craniofac J* 2006;**43**:598–605.
- Hunt O, Burden D, Hepper P, et al. Parent reports of the psychosocial functioning of children with cleft lip and/or palate. *Cleft Palate Craniofac J* 2007;**44**:304–11.
- Kapp-Simon K. Self-concept of primary-school-age children with cleft lip, cleft palate, or both. *Cleft Palate J* 1986;**23**(1):24–7.
- Kramer FJ, Gruber R, Fialka F, et al. Quality of life and family functioning in children with non-syndromic orofacial clefts at preschool ages. *J Craniofac Surg* 2008;**19**:580–7.
- Kramer FJ, Gruber R, Fialka F, et al. Quality of life in school-age children with orofacial clefts and their families. *J Craniofac Surg* 2009;**20**:2061–6.
- Leonard BJ, Brust JD, Abrahams G, et al. Self-concept of children and adolescents with cleft lip and/or palate. *Cleft Palate Craniofac J* 1991;**28**:347–53.
- Millard T, Richman LC. Different cleft conditions, facial appearance, and speech: relationship to psychological variables. *Cleft Palate Craniofac J* 2001;**38**:68–75.
- Murray L, Arteche A, Bingley C, et al. The effect of cleft lip on socio-emotional functioning in school-aged children. *J Child Psychol Psychiatr* 2010;**51**:94–103.
- Persson M, Aniansson G, Becker M, et al. Self-concept and introversion in adolescents with cleft lip and palate. *Scand J Plast Reconstr Surg Hand* 2002;**36**:24–7.
- Richman LC, Holmes CS, Eliason MJ. Adolescents with cleft lip and palate: self-perceptions of appearance and behavior related to personality adjustment. *Cleft Palate J* 1985;**22**:93–6.
- Richman LC. Facial and speech relationships to behavior of children with clefts across three age levels. *Cleft Palate Craniofac J* 1997;**34**:390–5.
- Richman LC, Millard T. Brief report: cleft lip and palate: longitudinal behavior and relationships of cleft conditions to behavior and achievement. *J Pediatr Psychol* 1997;**22**:487–94.
- Sagheri D, Ravens-Sieberer U, Braumann B, et al. An evaluation of Health-Related Quality of Life (HRQoL) in a group of 4–7 year-old children with cleft lip and palate. *J Orofac Orthop* 2009;**70**:274–84.
- Schneiderman CR, Auer KE. The behavior of the child with cleft lip and palate as perceived by parents and teachers. *Cleft Palate J* 1984;**21**:224–8.
- Slifer KJ, Beck M, Amari A, et al. Self-concept and satisfaction with physical appearance in youth with and without oral clefts. *Child Health Care* 2003;**32**:81–101.
- Starr P. Cleft type, age, and sex differences in teen-agers' ratings of their own behavior, self-esteem, and attitude toward clefting. *Rehabil Lit* 1984;177–9.
- Warschawsky S, Kay JB, Buchman S, et al. Health-related quality of life in children with craniofacial anomalies. *Plast Reconstr Surg* 2002;**110**:409–14.

34. Edwards TC, Patrick DL, Topolski TD, et al. Approaches to craniofacial-specific quality of life assessment in adolescents. *Cleft Palate Craniofac J* 2005;42:19–24.
35. Patrick DL, Topolski TD, Edwards TD, et al. Measuring the quality of life of youth with facial differences. *Cleft Palate Craniofac J* 2007;44:538–47.
36. Jokovic A, Locker D, Guyatt G. Short forms of the Child Perceptions Questionnaire for 11–14-year-old children (CPQ 11–14): development and initial evaluation. *Health Qual Life Outcomes* 2006;4:4.
37. Broder HL, Wilson-Genderson M. Reliability and convergent and discriminant validity of the child oral health impact profile (COHIP Child's version). *Comm Dent Oral Epidemiol* 2007;35(Suppl. 1):20–30.
38. Hartnick CJ. Validation of a pediatric voice quality-of-life instrument. *Arch Otolaryngol Head Neck Surg* 2002;128:919–22.
39. Boseley ME, Cunningham MJ, Volk MS, et al. Validation of the pediatric voice-related quality-of-life survey. *Arch Otolaryngol Head Neck Surg* 2006;132:717–20.
40. Strauss RP, Ramsey BL, Edwards PC, et al. Stigma experiences in youth with facial differences: a multi-site study of adolescents and their mothers. *Corthod Craniofac Res* 2007;10:96–103.
41. Topolski TD, Edwards TC, Patrick DL. Quality of life: how do adolescents with facial differences compare with other adolescents? *Cleft Palate Craniofac J* 2005;42:25–32.
42. Lach LM, Ronen GM, Rosenbaum PL, et al. Health-related quality of life in youth with epilepsy: theoretical model for clinicians and researchers. Part 1: the role of epilepsy and co-morbidity. *Qual Life Res* 2006;15:1161–71.
43. Klassen AF, Anthony SJ, Khan A, et al. Identifying determinants of quality of life of children with cancer and childhood cancer survivors: a systematic review. *Support Care Cancer* 2011;19:1275–87.
44. Klassen A, Goodacre T, Wong K, et al. Understanding quality of life of cleft-lip and/or palate patients. 18th Annual International Society of Quality of Life Research, Denver USA, Oct 26–29 2011.