



Published in final edited form as:

Spec Care Dentist. 2011 May ; 31(3): 88–94. doi:10.1111/j.1754-4505.2011.00189.x.

Comparison of perceptions of oral health-related quality of life in adolescents affected with ectodermal dysplasias relative to caregivers

Richie Kohli, BDS, MS^{1,*}, Steven Levy, DDS, MPH², Colleen M. Kummet, MS³, Deborah V. Dawson, PhD, ScM³, and Clark M. Stanford, DDS, PhD³

¹ TMD and Orofacial Pain Division, University of Minnesota, Minneapolis, Minnesota

² Department of Preventive and Community Dentistry, University of Iowa, Iowa City, Iowa

³ Dows Institute for Dental Research, University of Iowa

Abstract

The objective of this study was to assess the perceived oral health-related quality of life (OHQoL) of adolescents affected with one of the ectodermal dysplasias (EDs).

Data were collected from 2003 to 2007 in a cross-sectional study of a convenience sample of individuals affected by ED (n=35) using the Child Perceptions Questionnaire (CPQ11–14) for children and the Parent-Caregiver Perceptions Questionnaire (P-CPQ) for their caregivers. The main findings of this study were that individuals who were affected with ED in the older age group (15–19 years old) perceived more functional problems than younger individuals (11–14 years old) (p=0.04). Females with ED (n= 13) perceived more emotional problems than males (n=22; p=0.01). Although caregivers tended to report slightly higher OHQoL scores (indicating worse OHQoL), no significant differences were observed between children's and parents' total OHQoL and individual domains' median scores (p>0.05). Thus, the perceptions of oral health and well-being may vary by age and gender for children who have ED. Caution is warranted concerning using parents as proxies for their children when assessing the child's OHQoL.

Keywords

ectodermal dysplasias; oral health-related quality of life; children; teenagers; parents; perceptions

Introduction

Ectodermal dysplasias (EDs) are a heterogeneous group of disorders characterized by deficiency of ectoderm- and mesoderm-derived tissues, including skin, hair, teeth, and sweat glands.¹ Inheritance patterns are variable, and the incidence of ED is estimated at about 7 in 10,000 births.² There are nearly 200 distinct disorders that comprise the EDs.¹ Hypohidrotic ED (HED or Christ-Siemens-Touraine syndrome) is the most common form, inherited in an X-linked pattern in most cases, although autosomal dominant and autosomal recessive forms have been identified.¹

*Corresponding author: kohli007@umn.edu.

Conflict of interest

There is no conflict of interest in this study.

Besides facing challenges of functional limitations (increased body temperature, hearing problems, chewing difficulties, abnormal speech, and skin and eye infections), children with oro-facial problems associated with EDs may be prone to emotional, social and behavioral challenges related to their self-image.³ This can have a negative impact on the quality of life of the children and/or their parents.⁴ Additionally, patients with special care needs, including EDs, often spend a greater amount of time, money, and other resources traveling long distances to regional, often university-based, clinical care centers to obtain multi-specialist care.⁵ As a result, obtaining dental care for patients with ED is often challenging due to access issues and high costs. Thus, they may be expected to have poor oral health-related quality of life (OHQoL). On the other hand, research³ also has shown that children with special needs, especially those with oro-facial conditions (cleft lip/palate, etc.), may be fairly well adjusted to cope with the environment, probably because of the presence of the condition since birth and team management of their condition. In general, adequate dental care helps to have healthy nutrition, proper speech, favorable self-esteem, and greater educational development in children, resulting from proper concentration during learning and playing activities. Thus, individuals with ED can also have good OHQoL even though they are affected with an oral condition associated with one of the variant forms of ED. Further, due to different ages of cognitive development, younger children may perceive OHQoL differently than older children.

The literature focuses primarily on comprehensive reviews and case studies of EDs and dental management. The evaluation of general quality of life and especially oral-related issues in children with EDs has been limited. Mehta *et al.*¹ (2007) performed a study to delineate the head and neck manifestations and quality of life in EDs. Using the SF-8⁶ quality of life instrument, they reported that although a high percentage of participants with EDs (n=75) had otolaryngologic and dental conditions, the reported quality of life (QoL) scores indicated high quality of life. The literature also reveals that OHQoL concerns in children and adolescents with EDs are rarely addressed.

Several child OHQoL instruments are available. The Child Perceptions Questionnaire (CPQ)⁷ is the most established of these and was used in the present study. The CPQ was developed to take into account the children's cognitive abilities and lifestyles for age ranges from 8 to 10 years (CPQ8–10) and 11 to 14 years (CPQ11–14).⁷ This instrument uses a reverse scale approach in which higher scores equate to lower perceived QOL. Preliminary studies have confirmed the validity and reliability of the CPQ11–14 in other countries, such as the United Kingdom,^{8,9} New Zealand,¹⁰ Saudi Arabia,¹¹ China,^{12,13} Australia,⁴ and Brazil.^{14,15} The incorporation of an independent parental/caregiver component has resulted in the use of this instrument in populations of children with special needs.¹⁶ This was intended to be applicable to children with a wide variety of oral and orofacial conditions, to conform to contemporary concepts of child health, and to accommodate developmental differences among children of different ages.⁴

Jokovic *et al.*⁷ studied 123 children aged 11 to 14 years in Canada, who were attending clinics for the treatment of dental diseases (primarily caries), orthodontic disorders, and orofacial conditions (primarily cleft lip and/or palate). They observed that the impact of children's oral and orofacial conditions on functional and psychosocial well-being was substantial. In Hong Kong, Wong *et al.*¹⁸ assessed OHQoL impact among patients with oligodontia and the association between OHQoL and the number of missing teeth. Among this sample of Chinese children (11 to 15 years old, n=25), the impact of OHQoL measured by the Child Perceptions Questionnaire (CPQ) was substantial. The most frequent congenitally missing tooth was the esthetically important maxillary lateral incisor, which may in part explain the high CPQ scores (poor OHQoL) in this population.

The extent to which parents/caregivers understand the effects of ill health on their children's lives remains unanswered. In a 2003 study, Jokovic *et al.*¹⁹ assessed the agreement between children and their caregivers. A total of 42 pairs of mothers and children aged 11 to 14 years with oral and orofacial conditions completed the parental (P-CPQ) and child (CPQ11–14) components of the Child Oral Health Quality of Life Questionnaire.¹⁹ Overall scores indicated substantial agreement between mother and child pairs (ICC=0.70). However, the emotional (ICC=0.52) and social well-being (ICC=0.58) subscales indicated only moderate agreement. The study concluded that, although mothers may be used as proxies for their children in some circumstances and for some purposes, the views of both should be obtained in order to fully represent child OHQoL.¹⁹

A review of the literature reveals that OHQoL concerns in children with ectodermal dysplasias have rarely been addressed. Further, OHQoL differences have been found to differ according to age,¹⁵ gender,¹⁰ and child/parent reporting.¹⁹ The present research study assessed OHQoL in children with EDs using a valid and reliable measure. It addresses the following questions:

- Do children affected with EDs from different age groups and genders perceive OHQoL differently?
- Do children with EDs and their caregivers hold different OHQoL perceptions of the affected individuals' OHQoL?

The overall long-term goal is to provide evidence-based information for parents, clinicians, researchers, and public health professionals in order to better understand how the syndromes potentially interfere with people's day-to-day lives.

Materials and methods

OHQoL data were collected from 2003 to 2007 in a cross-sectional study with a convenience sample of individuals affected with EDs and their caregivers attending an annual conference of the National Foundation for Ectodermal Dysplasias (NFED) in the United States. Data were collected using the Child and Parent-Caregiver Perceptions Questionnaires, CPQ11–14 and P-CPQ, respectively. Children aged 11 to 19 years completed the CPQ11–14 questionnaire, while the caregivers completed the P-CPQ. The study protocol was approved by the University of Iowa Institutional Review Board (IRB Number 200302025). Consent (caregivers and children aged 18 years and older) and assent (children aged less than 18 years) were obtained at the time of data collection. Responses to the CPQ and P-CPQ questions were based on a recall period of three months and were scored on a five-point Likert scale: “Never”=0; “Once or Twice”=1; “Sometimes”=2; “Often”=3; and “Every day or almost every day”=4. Thus, higher scores resulted in less favorable OHQoL. Since a large number of the individuals participated in 2003 (17 out of 35) and two questions concerning emotional problems inadvertently were not included in the P-CPQ used in 2003 due to its development phase, we excluded those two questions from the entire analysis. Excluded questions were: 1) worried that he/she is not as healthy as other people, and 2) concerned what other people think about their teeth/mouth. Thus, 29 questions (6 questions for the domain of oral symptoms, 7 for functional limitations, 6 for emotional problems, and 10 for social problems) were used for overall data analysis. Questions common to the CPQ11–14 and P-CPQ are shown in Table 1.¹⁹

Overall composite and domain-specific scores for each child and caregiver (composition of caregiver group consisted of mother, father, sister, grandmother, and others) were created by summing the response codes for the questions (including “missing” values which were imputed as median values of the known responses per domain before summation). These scores were calculated separately for each age group (i.e., 11–14 years vs. 15–19 years).

Descriptive statistics profiled the characteristics of children affected by ED and their parents. Wilcoxon rank-sum tests were used to compare total OHQoL and individual domain scores by age group and gender. Wilcoxon signed-rank tests were used to determine if the medians of the directional differences between children and their caregivers were significantly different from zero. Spearman correlation coefficients were computed to assess the associations between children's and caregivers' scores. The weighted kappa statistic was used to assess agreement between caregivers and children for every common question. Bowker's test for symmetry²¹ determined whether there was a systematic overrating or underrating on the question level of the child's OHQoL by caregivers relative to the child's response.

Results

Questionnaires were completed by 35 pairs of children affected with ectodermal dysplasias and their caregivers. Among the children completing the CPQ11–14 questionnaire, 40% (n=14) were 11 to 14 years of age, 60% (n=21) were 15 to 19 years old, 63% (n=22) were male, and 37% (n=13) were female (Table 2). Based on detailed information available on 33 children affected with EDs, 72.8% (n=24) of the children were affected with Hypohidrotic Ectodermal Dysplasias (HED), 6.1% (n=2) with Hay Wells Syndrome, 3.0% (n=1) with Ectrodactyly-Ectodermal Dysplasia Clefing Syndrome (EEC), 3.0% (n=1) with KID (Keratitis-Ichthyosis-Deafness Syndrome), and 3.0% (n=1) with an unknown type; 12.1% (n=4) were missing responses. Sixty-one percent (20 out of 33) of children reported having removable partial dentures and 30% (10 out of 33) had dental crowns. The age of first dental visit (n=32) and the number of times dentures were fixed/replaced (n=19) ranged from 0 to 84 months and 0 to 50 times, respectively. The majority of caregivers completing the P-CPQ questionnaire were mothers (74%) or fathers (14%). Other caregivers were grandmother (3%), sister (6%), and other (3%).

A significant difference ($p = 0.04$) was identified in reported functional limitations based on child's age group with median score of 6 in the younger age group and 11 in older children (Table 3). For other domains (oral symptoms, emotional problems, and social problems), score differences were not statistically significant ($p > 0.05$). Overall OHQoL scores of children ranged from 2 to 80, with a median score of 25 in the younger children and 30 in older children. These differences were not statistically significant ($p > 0.05$). Individual domain-specific median impact scores corresponding to younger vs. older age groups of children were 6 vs. 7 for oral symptoms, 5 vs. 6 for emotional problems, and 6 vs. 6 for social problems.

Females (n=13) reported higher median total and domain scores than males (n=22), with differences statistically significant for emotional problems (3.0 vs. 10.0, $p = 0.01$) (Table 4).

On average, caregivers reported slightly worse oral health-related quality of life of their children than did the children themselves (Table 5), as indicated by the median total scores of 34.0 versus 26.0, respectively, for 11-to 19-year-olds. However, the total OHQoL median directional difference (child scores minus caregiver scores) of 8.0 was not statistically significant. The caregivers' median scores were the same or somewhat higher than the children's scores for all domains for the older age group, younger age group, and for both age groups combined, but differences were not statistically significant ($p > 0.05$). For both the younger and older age groups and for both age groups combined, the median directional differences were either absent or small, 0 for oral symptoms, 0 for functional limitations, 2.0 for emotional problems, and 0 for social problems (Table 5).

In 11- to 19-year-olds (n=35), positive correlations were obtained between child and caregiver for total oral health quality of life scores ($r=0.43$, $p=0.01$) and also for their individual domain scores ($r=0.53$, $r=0.47$, $r=0.45$, and $r=0.29$, respectively, for oral symptoms, functional limitations, emotional problems, and social problems). These correlations were statistically significant for the total and for individual domain scores ($p<0.05$), except for the social domain.

For individual questions, the level of agreement (weighted kappa) between children and their parents ranged from poor to substantial (-0.21 to $+0.74$). Overall, there was statistically significant agreement ($p<0.05$) between children 11 to 19 years old and their caregivers on questions like pain ($\kappa=0.39$), chewing difficulty ($\kappa=0.34$), mouth sores ($\kappa=0.32$), slow chewing ($\kappa=0.31$), nervousness ($\kappa=0.30$), and sleep disturbance ($\kappa=0.29$). The agreement was statistically significant ($p<0.05$) between older age group children and their caregivers for five questions (i.e., pain, sleep disturbance, chewing difficulty, slow chewing, and not wanting to talk to other children), whereas for the younger age group, there was no statistically significant agreement on any question except for “food stuck in palate.” Bowker’s test for symmetry²¹ was not statistically significant for any question in either age group.

Discussion

The children’s total OHQoL and individual mean domain scores obtained in this study were generally higher (*relatively worse overall OHQoL*) for the older age group, but only the differences in the domain of “functional limitations” were statistically significant. One explanation can be that the families of the older age group have experienced functional problems over a longer time period than is the case for the younger age group, in contrast to emotional and social problems for which they may have adjusted with time. Similarly, the older age group may be more conscious of the speech difficulties, sleeping problems, and chewing difficulties and hence restricted diet because of more independent food choices, but presence of fewer teeth. On the other hand, a recent study by Barbosa *et al.*¹⁵ reported significant differences among 11- to 14-year-old Brazilian public school children in the oral symptoms domain score, with the 11-year-olds having the perception of more oral symptoms than the 14-year-olds. The authors explain these differences on the basis of age-related experiences in the study populations as during the mixed dentition period (8 to 12 years of age), children may encounter many problems related to natural processes, such as exfoliating primary teeth, dental eruption, or space due to unerupted permanent teeth, which simultaneously affect their OHQoL. Significant differences in the oral symptoms domain scores in that study, in contrast to ours, can be explained by differences in the sample size and characteristics of the study population.

In the present study, females (n=13) reported higher median total and domain scores than males (n=22), but the differences were only statistically significant for emotional problems (10.0 vs. 3.0) ($p=0.01$). This suggests females may be experiencing more severe emotional problems than males. Foster Page and Thomson¹⁰ also reported that mean emotional well-being domain scores were higher for girls than for boys (4.0 vs. 2.7, $p<0.05$). This may be explained by a perception that girls are culturally more sensitive and concerned about their health and appearance. Barbosa *et al.*¹⁵ also observed the tendency of girls to report higher average impacts on OHQoL than boys, however these differences were not statistically significant ($p>0.05$).

In this study, parents tended to report more impact on OHQoL than their children, which is in contrast to the study by Jokovic *et al.*⁷ which included 11- to 14-year-old children (n=42) with pedodontic, orthodontic and oro-facial problems and observed that children on average

reported more problems than did their parents. However, in our study, the differences in scores were not statistically significant ($p>0.05$), which can be due to the small sample size ($n=35$).

In our study, at the individual question level, there was fair to moderate agreement (Kappa=0.21–0.60) between children 11 to 19 years old and their parents on questions referring to pain, mouth sores, chewing difficulty, sleep disturbance, slow chewing, and nervousness ($p<0.05$), which can be related to the concreteness of these events.¹⁹ There was little evidence of agreement ($p>0.05$) on questions concerning emotional or social problems, except for fair agreement on the topics of “being nervous” (Kappa=0.30, $p=0.02$) and “not wanted to talk to other children” in the older age group (Kappa=0.46, $p=0.03$). This could be due to greater interaction and time spent with peers, rather than family, as the child grows older, which decreases the chances of parents having knowledge about their children’s social context.¹⁹ Thus, one should be cautious while using caregivers as proxies for their children, especially at the individual question level.

There are limitations on the generalizability of these findings. Since the study used a convenience sample, there can be errors arising from a low response rate, due to the fact that some types of people (well-educated and motivated) are more willing than others to take part in surveys. There is the possibility of bias because of the site of collection, Hawthorne effect, nature of the sample, and lack of information on variables like race, ethnicity, socioeconomic status, family structure, and other confounding factors. Because of the cross-sectional nature of the study, we were not able to follow up with the participants and thus capture age-related differences in OHQoL perceptions over time. These limitations restrict generalizing the data and, thus, the prevalence estimates and scores apply only to those who took part in the study at that time.

No attempt was made to adjust for multiple testing or interactions. Given the modest sample sizes, any reasonable adjustment of this type would render results non-significant. Rather, noting the novel aspect of this study, an exploratory and descriptive approach to these analyses was taken. An important goal of the evaluation of these novel data was to identify suggestive results for future confirmation, and to identify promising directions for future work in this little-studied cohort.

Age differences found in this study may be a result of the extension of the CPQ11–14 to an older adolescent population for which it has not been validated. Justification for this extension was to capture this vital OHQoL information in a small population of teenage children and the opportunity for comparison with younger adolescents with a similar disease. For this reason we have presented group-specific data; however, we note that we would not expect older individuals to have difficulty with comprehension.

Despite the limitations mentioned above, this study has strengths. These include the value of these data, the novelty of this research with respect to persons with EDs, and the fact that there were suggestive findings despite the modest sample sizes. Our study provides valuable information on two counts: direction of future investigations, and need for caution regarding the use of caregivers as proxies for their children. Both children’s and their parents’ perspectives regarding children’s OHQoL were assessed, including the perceptions of individuals affected by ED of different age groups and gender. This study may help clinicians to understand and incorporate other important aspects of treatment like parents’ expectations and consideration of perceived emotional and social problems instead of physical treatment only.

For future studies, the results of the present study can be used to help estimate the minimum sample size required for desired power. Additional research is recommended to replicate the

present study in the general population with ED, which could confirm and expand on our findings. A key aspect of this study was to use a calibrated QoL instrument in a patient population that has some component of an affected disorder. Comparison of OHQoL between individuals who are and are not affected by ED (including both unaffected family members and unrelated controls) would further illuminate the expected patterns of responses for OHQoL perception in these two groups. Before-and after-treatment studies can help to assess treatment effects in terms of overall well-being.

This study was not intended as a validation study, but rather to describe self-and caregiver-reported OHQoL in this adolescent population and for that reason we confined our analyses to the 29 questions common to both children and parents. Although the CPQ has been validated in various studies, this instrument lacks a dynamic range. This is because anything that is “statistically significant” may not be significant clinically and a standardized range for clinical significance has not been reported in previous studies. Establishing cut-off points for good or poor OHQoL could help in better understanding the clinical implications of these studies. Short versions of the CPQ questionnaires²⁰ have been validated in some studies. If this validation can be extended for use in multiple settings, the short versions can certainly prove helpful in saving time and also retaining the interest of the study subjects to complete the questionnaire.

Conclusions

The main findings of this study were that people with ED in the older age group (15 to 19 years old) perceived more functional problems than the younger age group (11 to 14 years old) and females who had ED perceived more emotional problems than males who had ED. Although caregivers tended to report poorer OHQoL for their children than did the children, no significant differences were found between children’s and parents’ total OHQoL and individual domain median scores. Thus, among children affected by ED, the understanding of oral health and well-being may vary by age and gender. One should be cautious while using caregivers as proxies for their children in assessing the child’s OHQoL. Because of the small sample size and other limitations, the present study lays a foundation for additional studies needed to pursue and confirm these indicative results.

Acknowledgments

Our heartiest thanks go to the late Dr. Jane Chalmers, for taking intense academic interest in this study, as well as providing valuable suggestions that improved the quality of this study. We are very grateful to Drs. Jokovic and Locker for allowing us to use the CPQ and P-CPQ instruments. We are also very thankful to Ms. Cynthia Asmussen for her assistance in data collection, Ms. Chris White for library support, and finally all the participants of the ED study.

Funding support

This study was made possible by Grant Number UL1RR024979 from the National Center for Research Resources (NCRR), a part of the National Institutes of Health (NIH). Its contents are solely the responsibility of the authors and do not necessarily represent the official views of the Clinical and Translational Science Award (CTSA) or NIH. Thanks to the National Foundation for Ectodermal Dysplasias (NFED) for providing funding support through the Research Sponsorship Program of the Foundation.

References

1. Mehta U, Brunworth J, Fete TJ, Sindwani R. Head and neck manifestations and quality of life of patients with ectodermal dysplasia. *Otolaryngol Head Neck Surg.* 2007; 136:843–7. [PubMed: 17478227]
2. Itin PH, Fistarol SK. Ectodermal dysplasias. *Am J Med Genet C Semin Med Genet.* 2004; 131C:45–51. [PubMed: 15468153]

3. Locker D, Jokovic A, Tompson B. Health-related quality of life of children aged 11 to 14 years with orofacial conditions. *Cleft Palate Craniofac J*. 2005; 42:260–6. [PubMed: 15865459]
4. Do LG, Spencer A. Oral health-related quality of life of children by dental caries and fluorosis experience. *J Public Health Dent*. 2007; 67:132–9. [PubMed: 17899897]
5. Stanford CM, Guckes A, Fete M, Srun S, Richter MK. Perceptions of outcomes of implant therapy in patients with ectodermal dysplasia syndromes. *Int J Prosthodont*. 2008; 21:195–200. [PubMed: 18548955]
6. Roberts B, Browne J, Ocaña KF, Oyok T, Sondorp E. The reliability and validity of the SF-8 with a conflict-affected population in northern Uganda. *Health Qual Life Outcomes*. 2008; 6:108. [PubMed: 19055716]
7. Jokovic A, Locker D, Stephens M, Kenny D, Tompson B, Guyatt G. Validity and reliability of a questionnaire for measuring child oral-health-related quality of life. *J Dent Res*. 2002; 81:459–63. [PubMed: 12161456]
8. Marshman Z, Rodd H, Stern M, et al. An evaluation of the Child Perceptions Questionnaire in the UK. *Community Dent Health*. 2005; 22:151–5. [PubMed: 16161878]
9. O'Brien C, Benson PE, Marshman Z. Evaluation of a quality of life measure for children with malocclusion. *J Orthod*. 2007; 34:185–93. [PubMed: 17761802]
10. Foster Page LA, Thomson WM. Malocclusion and uptake of orthodontic treatment in Taranaki 12–13-year-olds. *N Z Dent J*. 2005; 101:98–105. [PubMed: 16416747]
11. Brown A, Al-Khayal Z. Validity and reliability of the Arabic translation of the child oral-health-related quality of life questionnaire (CPQ11–14) in Saudi Arabia. *Int J Paediatr Dent*. 2006; 16:405–11. [PubMed: 17014538]
12. McGrath C, Pang HN, Lo EC, King NM, Hagg U, Samman N. Translation and evaluation of a Chinese version of the Child Oral Health-related Quality of Life measure. *Int J Paediatr Dent*. 2008; 18:267–74. [PubMed: 18554335]
13. Li XJ, Huang H, Lin T, Huang GM. Validation of a Chinese version of the child perception questionnaire. [Article in Chinese.]. *Hua Xi Kou Qiang Yi Xue Za Zhi*. 2008; 26:267–70. [PubMed: 18705508]
14. Goursand D, Paiva SM, Zarzar PM, et al. Cross-cultural adaptation of the Child Perceptions Questionnaire 11–14 (CPQ11–14) for the Brazilian Portuguese language. *Health Qual Life Outcomes*. 2008; 6:2. [PubMed: 18194552]
15. Barbosa T, Tureli MCM, Gavião MB. Validity and reliability of the Child Perceptions Questionnaires applied in Brazilian children. *BMC Oral Health*. 2009; 9:13. [PubMed: 19450254]
16. Cheretakis C, Locker D, Dror Y, Glogauer M. Oral health-related quality of life of children with neutropenia. *Spec Care Dentist*. 2007; 27:6–11. [PubMed: 17388223]
17. Coffield KD, Phillips C, Brady M, Roberts MW, Strauss RP, Wright JT. The psychosocial impact of developmental dental defects in people with hereditary amelogenesis imperfecta. *J Am Dent Assoc*. 2005; 136:620–30. [PubMed: 15966649]
18. Wong AT, McMillan AS, McGrath C. Oral health-related quality of life and severe hypodontia. *J Oral Rehabil*. 2006; 33:869–73. [PubMed: 17168928]
19. Jokovic A, Locker D, Stephens M, Guyatt G. Agreement between mothers and children aged 11–14 years in rating child oral health-related quality of life. *Community Dent Oral Epidemiol*. 2003; 31:335–43. [PubMed: 14667004]
20. Jokovic A, Locker D, Guyatt G. Short forms of the Child Perceptions Questionnaire for 11–14-year-old children (CPQ11–14): development and initial evaluation. *Health Qual Life Outcomes*. 2006; 4:4. [PubMed: 16423298]
21. Zar, JH. *Biostatistical Analysis*. 4. Upper Saddle River: Prentice Hall; 1999.

Table 1

Questions common to the CPQ11–14 and P-CPQ.

Category	Symptoms	
Oral Symptoms (6 items)	Pain	Bad breath
	Bleeding gums	Food stuck in palate
	Mouth sores	Food stuck in/between teeth
Functional Limitations (7 items)	Sleep disturbance	Slow chewing
	Chewing difficulty	Difficulty eating hot/cold foods
	Mouth breathing	
	Speech difficulty	Diet restrictions
Emotional Problems (6 items)	Upset	Worried about appearance
	Irritable/frustrated	Shy/embarrassed
	Nervous/afraid	Worried that he/she is different than other people
Social Problems (10 items)	Missed school	Been teased by children
	Low concentration in school	Left out by children
	Not wanted to speak/read out loud in class	Not wanted to spend time with children
	Not wanted to talk to children	Avoided school or leisure activities
	Avoided smiling/laughing when with children	Asked questions by other children

Table 2

Characteristics of individuals affected by Ectodermal Dysplasia.

Age of child	N=35	%
<i>11–14 years</i>	<i>N=14</i>	<i>40.0%</i>
11 yrs	5	35.7
12 yrs	2	14.3
13 yrs	4	28.6
14 yrs	3	21.4
<i>15–19 years</i>	<i>N=21</i>	<i>60.0%</i>
15 yrs	11	52.4
16 yrs	2	9.5
17 yrs	4	19.0
18 yrs	3	14.3
19 yrs	1	4.8
<i>Gender</i>	<i>N=35</i>	<i>%</i>
Boys	22	62.9
Girls	13	37.1
<i>Type of ED</i>	<i>N=33*</i>	<i>%</i>
Hypohidrotic ED (HED)	24	72.8
Hay Wells	2	6.1
Ectrodactyly-Ectodermal Dysplasia-Clefting Syndrome (EEC)	1	3.0
Keratitis-Ichthyosis-Deafness Syndrome (KID)	1	3.0
Unknown	1	3.0
Missing response	4	12.1
<i>Removable partial dentures present</i>	<i>N=33*</i>	<i>%</i>
Yes	20	60.6
No	13	39.4
<i>Dental crowns present</i>	<i>N=33*</i>	<i>%</i>
Yes	10	30.3
No	23	69.7
<i>Age of first dental visit</i>	<i>N=32*</i>	
	Range= 0–84 months	Median= 21 months
<i>Times dentures fixed/replaced</i>	<i>N=19*</i>	
	Range= 0–50 times	Median= 4 times
<i>Caregiver respondents</i>	<i>N=35</i>	<i>%</i>
Mother	26	74.2
Father	5	14.3
Sister	2	5.7
Grandmother	1	2.9

Age of child	N=35	%
Others	1	2.9

* Based on detailed information available for these numbers of individuals affected by ED only.

Table 3

CPQ overall and domain scores of children by age group.

Variable	No. of items used (possible range of scores)	11-14 years and 15-19 years age group combined scores (n=35)		11-14 years age group scores (n=14)		15-19 years age group scores (n=21)		Wilcoxon Rank-Sum test p-value ^a
		Mean (s.d.), Median (min-max)	Mean (s.d.), Median (min-max)	Mean (s.d.), Median (min-max)	Mean (s.d.), Median (min-max)			
Total OHQoL	29 (0-116)	31.6 (19.0), 26 (2-80)	25.1 (13.8), 25 (2-46)	35.9 (21.0), 30 (12-80)	0.17			
Oral Symptoms	6 (0-24)	6.8 (3.3), 7 (0-14)	5.9 (3.8), 6 (0-14)	7.3 (2.9), 7 (2-13)	0.26			
Functional Limitations	7 (0-28)	9.4 (5.0), 9 (0-22)	7.3 (4.5), 6 (0-16)	10.8 (4.9), 11 (3-22)	0.04 ^b			
Emotional Problems	6 (0-24)	7.5 (6.6), 6 (0-24)	5.4 (4.0), 5 (0-12)	9.0 (7.6), 6 (0-24)	0.25			
Social Problems	10 (0-40)	7.9 (7.3), 6 (0-29)	6.6 (4.8), 6 (0-15)	8.9 (8.6), 6 (0-29)	0.73			

^a Significance probability (p-value) associated with the Wilcoxon rank-sum test of the null hypothesis that the distribution of scores is the same for each age group (older vs. younger).

^b Significance probability $\alpha < 0.05$.

Table 4

CPQ overall and domain scores of children by gender.

Total and domain scores	Males (n=22)	Females (n=13)	Wilcoxon rank-sum test
	Mean scores (s.d.), Median scores (min-max)	Mean scores (s.d.), Median scores (min-max)	p-value ^a
Total OHQoL	27.0(16.4), 23 (2–70)	39.4 (21.1), 34 (12–80)	0.06
Oral Symptoms	6.1 (2.7), 6.5 (0–10)	7.8 (4.0), 8.0 (2–14)	0.36
Functional Limitations	9.0 (5.0), 8.5 (0–20)	9.9 (5.3), 9.0 (3–22)	0.70
Emotional Problems	5.5 (5.7), 3.0 (0–23)	10.9 (6.8), 10.0 (2–24)	0.01 ^b
Social Problems	6.3 (5.8), 5.0 (0–20)	10.7 (9.0), 8.0 (1–29)	0.14

^aSignificance probability (p-value) associated with the Wilcoxon Rank Sum test of the null hypothesis that the distribution of scores is the same for both genders.

^bSignificance probability, $\alpha < 0.05$.

Table 5

Mean and median directional differences between overall and subscale/domain P-CPQ and CPQ scores in 11-to 19-year-olds (n=35).

Variable	No. of common items(possible range)	Child Scores		Caregiver Scores		Directional Differences ^d (child-caregiver)		p ^b -value
		Mean (s.d.), Median (min-max)	Mean (s.d.), Median (min-max)	Mean (s.d.)	Median	Mean (s.d.)	Median	
Total OHQoL	29 (0–116)	31.6 (19.0), 26 (2–80)	35.0 (16.8), 34 (6–66)	-3.4 (19.8)	-8.0	-3.4 (19.8)	-8.0	0.35
Oral Symptoms	6 (0–24)	6.8 (3.3), 7 (0–14)	7.9 (4.3), 7 (0–19)	-1.1 (3.9)	0	-1.1 (3.9)	0	0.21
Functional Limitations	7 (0–28)	9.4 (5.0), 9 (0–22)	9.8 (5.5), 9 (0–22)	-0.4 (5.8)	0	-0.4 (5.8)	0	0.63
Emotional Problems	6 (0–24)	7.5 (6.6), 6 (0–24)	8.4 (5.1), 8 (0–20)	-0.9 (6.2)	-2.0	-0.9 (6.2)	-2.0	0.37
Social Problems	10 (0–40)	7.9 (7.3), 6 (0–29)	8.9 (7.7), 6 (0–26)	-1 (9.6)	0	-1 (9.6)	0	0.65

^a Difference between child and caregiver scores accounting for the direction of differences (indicator of bias).

^b Significance probability associated with the test of the null hypothesis that the median change between the time points indicated is equal to zero, based upon the Wilcoxon signed-rank procedure.