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ORIGINAL ARTICLE

Associations between ectodermal dysplasia, psychological distress and quality of life in a group of adults with oligodontia

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ABSTRACT

Objective: The aim of this study was to assess demographics, self-reported signs of ectodermal dysplasia (problems with hair, nails, skin and sweat glands), present teeth, previous dental treatment, psychological distress and QoL in individuals with oligodontia, and to explore the associations between these factors. We also aimed to compare the level of psychological distress and QoL between the study group and normative samples.

Methods: Forty-seven individuals with oligodontia registered at a resource centre in Norway were included in the study. The participants completed self-administered questionnaires on demographics, ED signs, dental treatments, psychological distress and QoL (overall, health-related- and oral health-related QoL). Relevant statistics (independent t-test, correlation analysis and hierarchical multiple regressions) were used.

Results: Thirty-five participants reported ED signs. Forty-one participants had tooth replacements (nine had removable dentures). Sixteen had ≤ 10 present teeth, 13 perceived dry mouth and seven were unemployed. Persons with ≤ 10 present teeth had higher anxiety- and depression-scores than those having > 10 present teeth. Unemployment, dry mouth and removable dentures indicated poor health-related- and oral health related QoL. Compared to a normative sample; the study group had significantly poorer mental health (MH)-related QoL, mean (SD); (51.1(8.2) versus 46.8(9.3), $p < .001$) and more anxiety, mean (SD); (4.5(3.1) versus 6.7(3.6), $p < .001$). ED signs and treatments were most important for psychological distress and MH related QoL (MCS), whereas demographic parameters were most important for the other QoL measures.

Conclusions: The psychological burden of oligodontia is significant, emphasizing the importance of a holistic approach by caregivers.

Introduction

Oligodontia is defined as the congenital absence of six or more teeth (excluding third molars) and accompanies several genetic syndromes [1,2]. Our understanding of the genetics underlying tooth agenesis has progressed greatly in recent years, with approximately 80 genes found to be related to tooth agenesis [3]. However, OMIM lists 111 different syndromes associated with oligodontia.

Ectodermal dysplasias (EDs) are the most common diagnoses associated with oligodontia [2]. Not all genes causing oligodontia or ED have been identified, and diagnosis is still often based on clinical signs. EDs comprise a large heterogeneous group of genetic disorders with developmental abnormalities in two or more of the following ectodermal structures: teeth, skin, sweat glands, nails and hair. The salivary glands are ectodermal in origin, and reduced salivary secretion has been demonstrated in both ED and isolated oligodontia [4,5]. In these conditions, it is also common for

present teeth to be peg-shaped. Reduced salivary secretion and missing and malformed teeth can negatively affect chewing, swallowing, speech and appearance in individuals with oligodontia or ED [6]. In the most severely affected individuals, the face has an edentulous appearance, possibly leading to psychological disturbances [1].

Missing teeth and perceived dry mouth are associated with decreased quality of life (QoL) [7–10]. Adolescents with ED have poorer oral health-related QoL (OHRQoL) due to functional limitations compared to children, and young females with ED have more emotional problems than males [11]. In a study of 14 adults, ED symptoms and findings affected, to some extent, health-related QoL (HRQoL) without high emotional or social consequences [12]. Previously, in a comparative study of three different adult groups with various orofacial conditions, our research group reported Treacher Collins syndrome, cherubism and oligodontia/ED. The study focused on psychological distress and QoL. The group with oligodontia/ED had a significantly higher level of

anxiety and worse mental HRQoL than the other two groups [13].

The aim of this study was to assess demographics, self-reported signs of ED (problems with hair, nails, skin and sweat glands), present teeth, previous dental treatment, psychological distress and QoL in individuals with oligodontia, and to explore the associations between these factors. We also aimed to compare the level of psychological distress and QoL between the study group and normative samples.

Methods

Study participants

All 75 individuals with oligodontia who were registered at a National Resource Center for Oral Health in Rare Disorders (TAKO-centre) and seen after 1999, were contacted during 2013 and invited to the study. The TAKO-centre is a nationwide resource center receiving patients from all parts of Norway. The inclusion criteria were having been examined at the TAKO-centre after 1999, age ≥16 years at time of inclusion, and the absence of ≥6 teeth (excluding third molars). The presence of ED included Rieger syndrome and incontinentia pigmenti (IP). Only a few of the participants had been genetically tested.

All registered individuals fulfilling the inclusion criteria received a written request asking them to complete self-administered questionnaires on demographics, signs of ED and other symptoms associated with EDs, including present teeth (≤10 teeth/>10 teeth), previous dental treatment, psychological distress and QoL (overall-, health-related- and OHRQoL).

Individuals who did not reply after several weeks were re-contacted by either telephone or a new written invitation. Forty-seven individuals (63%) (24 females, 23 males) ranging in age from 16 to 82 years old responded and comprised the final study group. It was by coincidence that the number of males to females was almost equal. The participants were not selected by a convenience sampling technique, but consisted of the entire population of patients with oligodontia registered at the TAKO-centre.

Self-reported demographics, ED signs, ED-related health problems and treatments

A self-administered questionnaire was used to collect information on demographic variables, including gender, civil status and age, as well as information on employment (student, full time job or part time job). All respondents were asked if they had ED signs, such as missing teeth, dry skin/eczema, reduced sweat function, nail and hair defects or related health problems, like perception of dry mouth, hearing loss, eye dryness, nasal congestion/foul smell, chronic airway disease, problems with digestion, chewing or speech. Additionally, they were asked about previous and current dental treatments, such as surgical, orthodontic and prosthodontic treatments. This questionnaire was designed for the study.

Psychological distress

The Hospital Anxiety and Depression scale (HADS) was used to measure psychological distress. The HADS is a self-administered questionnaire measuring levels of distress [14] that comprises 14 items with two subscales, one for anxiety (HADS-A) and one for depression (HADS-D). Originally, the HADS was developed to identify cases with anxiety or depression in need of treatment among patients in non-psychiatric hospitals [15]. Each subscale has seven items rated on a four-point Likert-style scale from 0 (not present) to 3 (maximally present), with sum scores rated from 0 to 21. A sum ≥8 on each subscale was used as a cut-off to identify potential cases of anxiety and depression disorders that may require further clinical examination [14,16]. The psychometric properties of the Norwegian version of HADS have been demonstrated to be excellent [17].

Quality of life

Oligodontia is a complex condition challenging many aspects of life. It was therefore useful to reveal both overall-, health related- and disease specific health related QoL. Overall QoL (OQoL) is perceived as the subjective experience of QoL [18], and the HRQoL (physical and mental) is based on general health (GH) and may be defined as the 'objective' level of the experience in comparison with normative sample, while the disease-related QoL often is related with specific disease/diagnosis. Accordingly, three different scales of QoL were used.

Cantril's Self-Anchoring Ladder (CL) is a self-administered questionnaire assessing OQoL with only one question, 'How is your life?' Respondents are asked to rate their present experiences with life on a scale anchored by their own identified values between 0 (worst possible QoL) and 10 (best possible QoL) [19]. The CL has been shown to have high validity and reliability [20].

The Short-Form Health Survey (SF-36) was used to measure HRQoL. The SF-36 is a multipurpose, generic questionnaire with 36 items rated on eight scales [21], with two summary measures, the Physical Component Scale (PCS) and Mental Component Scale (MCS). The PCS consists of physical functioning (PF), role limitation owing to physical health problems (RP), bodily pain (BP), which addresses the degree of pain, and GH. The MCS consists of vitality (VT), social functioning (SF), role limitations due to emotional problems (RE) and mental health (MH).

Information from the SF-36 was transformed to a scale of 0 (worst) to 100 (best). Based on transformations, both the PCS and MCS have a mean of 50 and standard deviation of 10 in the Norwegian population, as well as many others [21–23]. Scores <40 on both PCS and MCS are defined as caseness, meaning that the participants have poor QoL requiring treatment or help to improve the QoL. The SF-36 has been extensively validated [24], and its reliability is well-documented in several countries, including Norway [23].

To investigate OHRQoL, a short form of the original OHIP-49 questionnaire developed by Slade and Spencer [25] was used. OHIP-14 is a self-reported assessment of the impact of oral health on daily life that consists of 14 items organized into seven dimensions: functional limitations, physical pain, psychological discomfort, physical disability, psychological disability, social disability and handicap [26]. The study

participants record experiences with different aspects of oral health-related situations during the past year. Response options for each item are: 0 = never, 1 = rarely, 2 = sometimes, 3 = quite often, 4 = often. OHIP-14 has been shown to have internal consistency, good construction, high stability, and discriminant validity, and the Norwegian translation used in this survey has been shown to have an acceptable reliability and validity [27]. The total score for OHIP-14 is calculated as a sum score of 0–56. A high score indicates poor OHRQoL.

Normative samples

Three different studies were used as the source for normative data; the Survey of Level of Living[1] for SF-36-data, the Nord-Trøndelag Health (HUNT3) Survey[2] for HADS data, and Dahl's study [28] for the OHIP-14 data.

In the survey of level of living carried out by Statistics Norway in 2002, the SF-36 was answered by 3648 survey participants. The HUNT Study is one of the largest health studies ever performed in Norway and includes a unique database of personal and family medical histories collected during three intensive studies. 50,805 individuals completed the HADS in the HUNT3 survey. From these studies, we were able to randomly draw and match data. As the number of participants was very different in the two studies, 10 controls were drawn for each patient for the SF-36 data and 75 controls were drawn for the HADS data. The data were first divided according to gender, then according to age and lastly the 10 versus 75 controls were drawn giving close to identical mean ages in the study and control groups. Thus, a total of 240 females, 51% and 230 males, 49% were used as controls for the SF-36 data, and 3525 (1800 females, 51% and 1725 males, 49%) individuals were used as controls for the HADS data. For the OHIP data, the gender distribution among the 2150 respondents by chance was identical to ours and also had a comparable age frame.

Data analysis

Descriptive statistics, chi-square, independent t-test and correlation analysis between independent and dependent variables (i.e. Pearson's product–moment correlation coefficient), as well as hierarchical linear multiple regression analysis were used.

Parametric tests were used when comparing means between subgroups in the study group and between the study group and normative samples to explore psychological distress and QoL. Due to the small N of subgroups, i.e. when comparing psychological distress and QoL in subgroups, non-parametric statistics (Mann–Whitney U-tests) was employed as well, without different results. The data are therefore presented according to parametric tests.

Hierarchical multiple regressions were conducted to examine possible associations and modifying effects between independent variables and QoL and psychological distress (dependent variables). The variables were entered in steps in a predetermined order. Step 1 comprised demographic variables and step 2 comprised clinical signs/removable denture. In regression analysis, one parameter per 10 persons studied can be included, thus five parameters that significantly correlated with dependent variables were included. Present teeth (having ≤10 teeth) correlated with HADS only; therefore this was one of the clinical variables in the HADS regression. On the other hand, the perception of dry mouth was only correlated with the dependent variables of QoL and was therefore chosen as one of the three clinical variables in all the QoL regressions. Due to relatively small sample, the adjusted R square statistics, which correct the R square value, provide a better estimation of the population value.

The internal consistencies of the HADS, SF-36 and OHIP-14 were examined using Cronbach's coefficient α. Values >0.60 were considered acceptable, >0.70 good, and >0.90 excellent.

Data were analysed using SPSS (Statistical Package for Social Sciences version SPSS Statistics 22; SPSS Inc., Chicago, IL). A significance level of $p<.05$ was considered significant.

Ethical considerations

The Regional Committee for Medical and Health Research Ethics (2012/2003) approved the study. Participants provided written consent confirming their willingness to be included in the study. The participants were informed that they were free to withdraw from the study at any time.

Results

The demographics, ED signs, ED related health-problems, having ≤10 teeth versus having >10 teeth, and dental treatments reported by the study participants are presented in Table 1.

Thirty-five participants reported ED signs (skin, sweat glands, nails and hair). The 12 remaining participants reported oligodontia only. Additionally, the study group frequently reported to perceive dry mouth, hearing loss, eye dryness, nasal congestion/foul smell, chronic airway disease problems, and problems with digestion, chewing and speech. Sixteen of the participants reported that they had 10 present teeth or less. Most of the participants ($n=41$) had tooth replacements, such as removable dentures ($n=9$), bridges ($n=34$) and implants ($n=25$), and several had combinations of these. Three quarters of the participants had undergone orthodontic treatment ($n=31$) or were presently receiving orthodontic treatment ($n=5$) (Table 1). Nine of the 47 participants stated that they did not need orthodontic treatment. Thirty-three of those who were receiving or had undergone orthodontic treatment had received prosthodontic treatment.

Psychological distress

Compared to the normative sample, the participants in the present study had higher levels of and increased prevalence of anxiety (Table 2). No differences in the levels of anxiety and depression were found between men and women in the study group. Differences between subgroups according to

Table 1. Demographics, ectodermal signs or problems, and dental treatments of the study sample.

	Oligodontia N = 47
Demographics	
Mean age (SD) range, years	29.6 (14.9) 16–82
	n (%)
Gender	
Female	24 (51)
Male	23 (49)
Employed (student, full time job or part time job)	
Yes	40 (85)
No	7 (15)
ED signs and ED-related health problems	
Number of present teeth	
≤10	16 (34)
>10	31 (66)
Dry skin/eczema	
Yes	28 (60)
No	19 40)
Reduced sweat function	
Yes	10 (21)
No	37 (79)
Nail defect	
Yes	17 (36)
No	30 (64)
Hair defect	
Yes	21 (45)
No	26 (55)
Perceived dry mouth	
Yes	13 (28)
No	34 (72)
Hearing loss	
Yes	2 (4)
No	45 (96)
Eyes (dryness)	
Yes	6 (13)
No	41 (87)
Nose (nasal congestion/foul smell)	
Yes	5 (11)
No	42 (89)
Chronic airway disease	
Yes	8 (17)
No	39 (83)
Digestion problems	
Yes	9 (19)
No	38 (81)
Chewing problems	
Yes	3 (6)
No	44 (94)
Speech problems	
Yes	1 (2)
No	46 (98)
Dental treatments	
Orthodontics	
Yes	36 (77)
No	11 (23)
Removable denture	
Yes	9 (19)
No	36 (77)
2 missing	
Bridges	
Yes	34 (72)
No	12 (26)
1 missing	
Implants	
Yes	25 (53)
No	22 (47)

other demographic variables, perceived dry mouth, ≤10 teeth/>10 teeth and dental treatments are detailed in Table 3. Participants with 10 teeth or less showed higher levels of anxiety and depression versus those with more teeth. In addition, those with removable dentures had a higher level of depression compared to participants without removable dentures. Cronbach's alpha was 0.70 for the HADS-A score and 0.75 for the HADS-D score, respectively, which is considered good.

Overall quality of life

Significant differences in OQoL were found between those with and those without removable dentures. In addition, unemployed participants had poorer OQoL than those working or studying (Table 3).

Health-related quality of life

Role limitations owing to physical health problems (RP), role limitations due to emotional problems (RE) and MCS scores were significantly poorer in the study group compared to the normative sample (Table 2).

As shown in Table 3, the older age-group scored poorer on Physical Function, Role limitation owing to physical health problems (Role Physical), and the PCS, and those who were employed versus the unemployed reported better HRQoL on all scales. Only the SF-36 VT score differed between females and males.

Perceived dry mouth and removable dentures resulted in poorer HRQoL scores on several scales, including the PCS, as well as the MCS for those with removable dentures (Table 3). Cronbach's alpha for SF-36 scores was excellent, ranging from 0.80 to 0.92.

Oral health-related quality of life

Compared to the normative sample of Norwegian adults, the study group had significantly poorer OHIP-14 sum score (Table 2). Those who were unemployed, had perceived dry mouth, and wore removable dentures had the poorest OHRQoL (Table 3). Regarding subscores (data not shown), participants who were unemployed had increased functional limitations and more physical pain, psychological discomfort, physical disability, social disability and handicap. Similarly, those wearing removable dentures had increased functional limitations, physical pain, physical disability, social disability and handicap. In addition, participants with perceived dry mouth had more psychological and social disability related to oral health. Cronbach's alpha for the OHIP-14 sum score was excellent (0.94).

Hierarchical multiple regression analyses

Psychological distress
Age and present teeth made unique contributions to HADS-A ($\beta = 0.296$, p .05 and -0.513, $p = .001$, respectively). The change in HADS increased significantly indicating that ED signs, present teeth and treatments were of higher importance, and explained more of the variance in anxiety than the demographic variables.

For HADS-D, age made a unique contribution ($\beta = 0.350$, $p = .016$), as well as removable dentures and the number of present teeth ($\beta = 0.364$, $p = .006$ versus $\beta = -.473$, $p = .001$).

Table 2. Differences in psychological distress and health-related quality of life (QoL) across the study sample and age- and gender-adjusted normative samples among Norwegian adults.

		Oligodontia N = 47	Normative sample N = 3525[a]	p Value
Psychological distress	HADS-A	6.67 (3.56)	4.52 (3.14)	<.001
	HADS-A case, n (%)	22 (47)	681 (17)	<.001
	HADS-D	3.16 (3.20)	2.67 (2.60)	NS
	HADS-D case, n (%)	5 (11)	230 (7)	NS
			N = 470[b]	
Health-related QoL (SF-36)	Physical functioning	92.87 (12.67)	93.65 (12.50)	NS
	Role physical[d]	86.30 (23.69)	86.45 (29.86)	NS
	Bodily pain	78.57 (27.48)	79.12 (23.54)	NS
	General health	74.57 (24.15)	78.28 (18.83)	NS
	Vitality	60.46 (24.56)	60.05 (18.91)	NS
	Social functioning	80.85 (25.51)	88.93 (18.86)	.007
	Role emotional[d]	66.73 (18.36)	88.58 (27.61)	<.001
	Mental health	76.59 (18.17)	79.91 (14.17)	NS
	Physical Component Score (PCS)	53.41 (7.75)	52.40 (8.14)	NS
	PCS case, n (%)[e]	4 (9)	46 (10)	NS
	Mental Component Score (MCS)	46.83 (9.29)	51.12 (8.24)	<.001
	MCS case, n (%)[e]	9 (19)	45 (10)	<.05
			N = 2438[c]	
Oral health-related QoL	OHIP-14	8.50 (9.62)	4.1 (6.2)	<.01

NS: not significant.
Data are given as mean (SD) unless otherwise noted.
[a]The normative sample is from the Nord-Trøndelag Health (HUNT3) Study.
[b]The normative sample is from the Survey of Level of Living 2002 – Cross-sectional Theme: Health.
[c]OHIP-14 mean scores were compared to a normative Norwegian adult sample, N = 2438 [28].
[d]*Role Physical*= Role limitation owing to physical health problems and *Role emotional*= role limitations due to emotional problems.
[e]Score <40 on both the PCS and MCS is defined as cases, meaning that the participants have poor QoL requiring treatment or help to improve the QoL.

Table 3. Differences in psychological distress and quality of life (QoL) across demographic variables, ED signs and dental treatments for the study group (N = 47).

	Age		Employment/unemployment		Gender		Perceived dry mouth	
	16–29 years 34	30–82 years 13	Employment 40	Unemployment 7	Female 24	Male 23	Yes 13	No 34
n	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)
Psychological distress								
HADS-A	6.2 (3.6)	7.9 (3.3)	6.3 (3.6)	8.9 (2.5)	7.5 (3.7)	5.8 (3.3)	7.2 (4.2)	6.5 (3.4)
HADS-D	2.7 (3.0)	4.4 (3.5)	2.9 (3.3)	4.9 (1.8)	3.3 (3.6)	3.0 (2.8)	4.4 (2.6)	2.7 (3.3)
QoL variables								
Overall QoL								
Cantril ladder	7.6 (1.8)	6.8 (2.4)	7.8 (1.8)	5.1 (1.5) ***	7.4 (1.8)	7.4 (2.2)	6.5 (2.3)	7.7 (1.8)
Health-related QoL								
SF-36								
Physical functioning	96.5 (6.2)	83.5 (19.4)***	96.3 (6.3)	73.6 (21.5)***	91.7 (15.1)	94.1 (9.7)	87.7 (14.9)	94.9 (11.3)
Role physical	91.9 (14.1)	71.6 (35.9)**	93.0 (12.9)	48.2 (35.1)***	86.2 (27.0)	86.4 (21.6)	69.2 (35.0)	92.8 (13.3)***
Bodily pain	82.7 (24.3)	67.8 (33.1)	83.6 (24.2)	49.9 (29.3)**	79.1 (27.2)	78.2 (28.4)	66.3 (29.7)	83.3 (25.5)
General health	77.2 (23.2)	67.7 (26.3)	78.0 (21.7)	55.0 (30.0)*	74.6 (26.7)	74.5 (21.8)	56.0 (28.2)	81.7 (18.3)***
Vitality	64.6 (21.5)	49.7 (29.5)	63.6 (24.5)	42.4 (16.9)*	53.5 (25.9)	67.7 (21.3)*	52.1 (26.6)	63.7 (23.4)
Social functioning	83.1 (23.4)	75.0 (30.6)	84.1 (24.2)	62.5 (27.0)*	81.8 (21.8)	79.9 (29.4)	73.1 (26.9)	83.8 (24.7)
Role emotional	68.7 (17.8)	61.5 (19.5)	69.4 (17.2)	51.4 (18.7)*	66.0 (17.8)	67.5 (19.3)	55.9 (23.3)	70.9 (14.4)*
Mental health	77.6 (18.4)	73.8 (18.9)	79.1 (17.6)	62.3 (15.7)*	75.5 (16.6)	77.7 (20.0)	67.7 (19.2)	80.0 (16.8)*
Physical Component Score (PCS)	55.4 (5.5)	48.4 (10.4)**	55.4 (5.12)	42.0 (10.6)***	53.3 (9.00)	53.5 (6.5)	48.4 (9.4)	55.4 (6.2)**
Mental Component Score (MCS)	47.2 (9.5)	45.9 (9.1)	47.5 (9.4)	42.7 (7.8)	45.9 (7.76)	47.8 (10.8)	44.2 (10.6)	47.8 (8.7)
Oral health-related QoL								
OHIP-14								
Sum score	7.0 (7.4)	12.4 (13.4)	6.7 (7.3)	19.0 (14.7)***	9.9 (10.9)	7.0 (8.0)	12.8 (12.1)	6.8 (8.1)*

	Number of present teeth		Orthodontic treatment		Removable denture		Implants	
	≤10 16	>10 31	Yes 36	No 11	Yes 9	No 36	Yes 25	No 22
n	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)	Mean (SD)
Psychological distress								
HADS-A	8.2 (3.4)	5.9 (3.4)*	6.5 (3.9)	7.4 (2.4)	7.8 (36)	6.3 (3.6)	6.6 (3.8)	6.6 (3.3)
HADS-D	4.7 (3.2)	2.4 (3.0)*	3.1 (3.4)	3.7 (2.4)	5.9 (3.1)	2.6 (3.0)**	3.4 (3.3)	2.9 (3.2)
QoL variables								
Overall QoL								
Cantril ladder	7,0 (1.7)	7.6 (2.1)	7.4 (2.0)	7.2 (2.2)	6.0 (2.6)	7.64 (1.7)*	7.0 (2.2)	7.9 (1.7)

(continued)

Health-related QoL
SF-36

Physical functioning	95.3 (6.7)	91.6 (15.0)	95.1 (8.7)	85.5 (19.9)*	86.7 (18.5)	94.4 (10.8)	92.6 (13.5)	93.2 (11.9)
Role physical	91.8 (11.8)	83.5 (27.7)	91.3 (15.0)	69.9 (37.5)**	66.0 (27.8)	91.5 (20.3)**	85.0 (23.5)	87.8 (24.4)
Bodily pain	81.3 (20.3)	77.2 (30.8)	83.0 (24.0)	65.5 (34.9)	56.8 (28.0)	84.8 (23.9)**	75.8 (29.1)	81.7 (25.8)
General health	72.8 (22.1)	75.5 (25.4)	76.9 (21.5)	67.0 (31.3)	60.7 (29.6)	78.4 (21.8)*	73.9 (23.9)	75.3 (25.0)
Vitality	57.8 (25.2)	61.8 (25.6)	60.6 (25.0)	60.2 (24.1)	43.9 (28.8)	64.2 (21.3)*	59.2 (23.2)	61.9 (26.5)
Social functioning	82.8 (15.1)	79.8 (29.7)	81.6 (26.5)	78.4 (23.1)	52.8 (31.7)	88.2 (18.2)***	73.5 (30.3)	89.2 (15.6)*
Role emotional	64.4 (17.8)	68.0 (18.8)	68.1 (17.6)	62.4 (21.1)	51.9 (22.6)	80.4 (14.4)**	65.9 (21.2)	67.7 (15.0)
Mental health	71.0 (18.6)	79.5 (17.5)	75.7 (18.8)	79.6 (16.3)	60.4 (22.9)	80.4 (14.4)**	72.6 (20.0)	81.1 (15.1)
Physical Component score (PCS)	55.4 (5.2)	52.4 (8.7)	55.2 (5.4)	47.5 (11.2)**	48.2 (7.5)	54.9 (7.3)*	53.3 (7.52)	53.5 (8.2)
Mental Component Score (MCS)	44.5 (9.0)	48.0 (9.4)	46.1 (9.9)	49.2 (6.9)	38.6 (12.4)	48.8 (7.2)**	44.9 (10.3)	49.1 (7.6)

Oral health-related QoL
OHIP-14

Sum score	8.00 (9.1)	8.8 (10.0)	7.6 (8.5)	11.3 (12.7)	17.4 (11.0)	6.3 (8.2)**	8.6 (10.4)	8.4 (9.0)

Data are presented as mean (SD) unless otherwise noted.
*$p < .05$.
**$p < .01$.
***$p < .001$ when comparing the subgroups according to age <29/>30; present teeth $\leq$10/>10; employment/unemployed; female/male; perceived dry mouth, orthodontic treatment, removable denture and implants yes/no.

A significant change in the explained variance of HADS-D from step 1 (6%) to step 2 (36%) was found, showing demographics as less important for depression in this group (Table 4).

Quality of life

Only employment made a unique contribution to CL ($\beta = -0.349$, $p = .036$), with demographics explaining 19% of the explained variance (Table 4).

For PCS, step 1 explained 42% of the variance, with a significant increase to 55% in the explained variance in step 2. Employment and a greater number of ED characteristics made the strongest unique contributions to the variance ($\beta = -0.443$, $p < .001$ versus $\beta = -0.248$, $p = .025$, respectively).

For MCS, wearing removable dentures made the strongest contribution ($\beta = -0.406$, $p = .007$), with a significant change from 2% to 17% of the explained variance between steps 1 and 2 (Table 4).

Unemployment and wearing removable dentures were significantly associated with OHIP-14 ($\beta = 0.306$, $p < .05$ versus $\beta = 0.352$, $p = .01$, respectively). For OHIP-14, the explained variance changed significantly from 18% to 29% between steps 1 to 2 (Table 4).

Discussion

The main findings of the present study were that participants affected by oligodontia had more anxiety, poorer mental HRQoL including reduced SF and role limitations due to emotional problems, and poorer OHRQoL than a normative sample. Those with less than 10 present teeth had higher level of anxiety and depression than those with more teeth present. Also wearing removable dentures gave higher level of depression. Removable dentures reduced all levels of QoL, while SF was reduced when having implants. Orthodontic treatment, experience of dry mouth and age reduced physical health related QoL including PF and role limitations due to physical problems. Clinical variables explained more of the variance of psychological distress, both anxiety and depression and MH related QoL compared to demographic variables, while it was opposite for all the other QoL measures.

Psychological distress

Higher levels of anxiety in the study group compared to the normative sample were reported. Almost half of the participants had anxiety scores above the level requiring further clinical examination. Health authorities should pay attention to this important finding. The clinical variables explained 29% of the variance of anxiety in the current sample of individuals. Absence of teeth is one of these variables. An explanation for this finding may be that missing many teeth and a compromised facial profile may lead to reduced oral function and psychological distress [29]. Wearing a removable denture was also one of the variables that counted for the explained variance of depression. Having removable dentures was correlated with reduced SF, which in turn is plausible to have an impact on psychological distress. Unfortunately, other parameters that could explain the high level of anxiety were not identified, possibly due to the design of the questionnaires. However, some of the differences between the study group and the normative sample may be related to aspects that can cause worry and uncertainty about the future. Individuals with oligodontia may require advanced long-term interdisciplinary medical and dental treatment in order to improve oral function and appearance, and delays in completing dental treatment may cause considerable frustration [30]. It is common for individuals with oligodontia to wait for many years before undergoing their final rehabilitation, as growth must often be completed before final treatment can begin, possibly explaining some of the variance in anxiety.

Individuals with 10 present teeth or less and those using removable dentures had also significantly higher levels of depression than individuals without dentures and those with more present teeth. Former studies have shown that tooth

Table 4. Psychological distress and quality of life according to demographic and clinical signs.

				B	Beta	p	Adjusted R^2
Psychological distress	HADS-A Anxiety						
	Constant			2.970			
		Step 1	Age	0.0751.842	0.296	.051	.074
			Employment (yes/no)		0.187	.212	
		Step 2	ED signs (yes/no)	1.921	0.238	.080	.286**
			Present teeth (<10/>10)	−3.825	−0.513	.001	
			Removable dentures (yes/no)	0.764	0.086	.520	
	HADS-D Depression						
	Constant			0.542			
		Step 1	Age	0.079	0.350	.016	.060
			Employment (yes/no)	0.425	0.048	.733	
		Step 2	ED signs (yes/no)	0.425	0.062	.626	.361***
			Present teeth (<10/>10)	−3.168	−0.473	.001	
			Removable dentures (yes/no)	2.917	0.364	.006	
QUALITY OF LIFE	Cantrils Ladder (CL) OQoL						
	Constant			10.386			
		Step 1	Age	−0.011	−0.081	.604	.190
			Employment (yes/no)	−1.904	−0.349	.036	
		Step 2	ED signs (yes/no)	−0.043	−0.010	.947	.197
			Removable dentures (yes/no)	−1.066	−0.216	.132	
			Perceived dry mouth (yes/no)	0.491	0.113	.449	
	SF-36/PCS HRQoL						
	Constant			70.095			
		Step 1	Age	−0.087	−0.161	.174	.415
			Employment (yes/no)	−9.357	−0.443	.001	
		Step 2	ED signs (yes/no)	−4.301	−0.248	.025	.554**
			Removable dentures (yes/no)	−3.274	−0.171	.110	
			Perceived dry mouth (yes/no)	3.536	0.209	.064	
	SF-36/MCS HRQoL						
	Constant			53.299			
		Step 1	Age	−0.152	−0.236	.143	.023
			Employment (yes/no)	1.054	0.042	.800	
		Step 2	ED signs (yes/no)	3.366	0.162	.270	.174*
			Removable dentures (yes/no)	−9.306	−0.406	.007	
			Perceived dry mouth (yes/no)	3.100	0.153	.311	
	OHIP-14 OHRQoL						
	Constant			−6.127			
		Step 1	Age	0.041	0.061	.681	.181
			Employment (yes/no)	8.174	0.306	.050	
		Step 2	ED signs (yes/no)	2.277	0.104	.443	.291*
			Removable dentures (yes/no)	8.506	0.352	.011	
			Perceived dry mouth (yes/no)	−2.352	−0.110	.431	

B: unstandardized coefficient; Beta: standardized coefficient; p: p value; R^2 square: (coefficient of determination) (measures the proportion of the variation in the dependent variable explained by the independent variables); adjusted R^2 square: (adjusts the statistic based on the number of independent variables in the model).
*$p < .05$.
**$p < .01$.
***$p < .001$.

loss may lead to low self-esteem and depression [31,32]. Our studies are in line with these. The level of depression observed in our study varied from mild mood disorders, mainly including loss of pleasure response, to caseness.

Quality of life

Those with self-reported signs of ED reported poorer HRQoL than those with isolated oligodontia. Dysfunctions, such as a lack of sweat capacity, may prevent physical activities and coping in warm conditions [33] and can explain our findings. In addition, individuals with oligodontia or self-reported ED reported other problems mainly related to dryness like dry eyes, nasal congestion/foul smell, hearing loss, chronic airway disease, and problems with digestion, chewing or speech.

The MH-related scores were poorer in the current study group than in the normative sample. In particular, SF and MH scores were affected, which contradict the findings of Pavlis et al. [12], who concluded that there were few emotional and social consequences. Nine of 47 individuals (19%) in our study were considered to have mental HRQoL (MCS) caseness and in need of further clinical examinations to determine if treatment is necessary. Wearing a removable denture and being unemployed were strongly associated with reduced QoL on all levels. However, the results showed that clinical variables explained most of the variance of MCS. This finding is partly in line with former research. A national

population-based study in Germany of adults 16–79 years of age showed that wearing a denture is a stronger factor for impaired OHRQoL than demographic variables (i.e. age, gender, education, years of schooling, and residential status) [34] while studies with participants between 20 and 70 years of age, found that the significance of tooth loss was ranked as being equal to the dissolution of marriage, retirement and problems with other relatives [35,36]. Missing natural teeth may lead to lower self-esteem, lower social status, and impaired function and decreased QoL, as underscored by Saintrain and de Souza [7], and can be an explanation for our findings.

Dry mouth may lead to difficulties in chewing and speaking, as well as mucosal infections and caries. In individuals with ED, the salivary glands may be affected, and in a study of orofacial functions among patients with ED, dryness of the mouth was a frequently reported finding (45%) [6]. Other studies have demonstrated that dry mouth affects both HRQoL and OHRQoL [7–9] negatively. Thus, dry mouth may partly explain the poorer OHRQoL in our study group compared to a normative Norwegian sample.

Unemployed participants had increased functional limitations, as well as more physical pain, physical disability and handicap. These findings partly confirm the findings of Kohli et al. [11], who emphasized functional problems due to OHRQoL. Our study also showed reduced physical HRQoL (PCS) due to perceived dry mouth and removable dentures. These findings could be seen as being partly in line with the above finding [11].

Strengths and limitations

All invited participants were registered and examined at a national centre, which provided an opportunity for a wide range of participants from all parts of Norway to participate. However, as patients were recruited from a specialist centre, selection bias in terms of recruitment of more severely affected individuals cannot be disregarded. Even though travel expenses are covered by insurance it may be psychologically or physically challenging for some individuals living far from the centre to travel, possibly adding to a selection bias in this study.

Established, validated, self-administered questionnaires that were easy to understand and complete were used. All instruments showed a good or excellent value for Cronbach's alpha confirming their reliability in this particular study.

The recruitment of 63% of the invited group yielded an adequate sample. We did not collect the normative data, but a strength was that we had access to normative quality assured data from several studies, and that we could draw and match age- and gender adjusted data from the Survey of Level of Living and HUNT studies for comparison with our own sample. Both parametric and non-parametric tests were used when comparing means/medians between the study group and normative samples and between subgroups. Only independent variables significant correlated with the

dependent variables were included in the hierarchical regression analysis.

A limitation of our study is that it was a cross-sectional study, and relatively few study participants included. However, we found it reassuring that the results were similar when using non-parametric and parametric test. Even if our results revealed important knowledge about various aspects of psychological distress and QoL related to oligodontia, we could not determine whether these parameters change over time. Although many types of oligodontia and ED can be genetically tested, most of the participants had a diagnosis based on clinical findings and family history. Finally, we only included individuals who lacked six or more teeth; therefore, our results cannot be extrapolated to persons with other combinations of ectodermal symptoms.

The study was based on self-reporting, and we cannot totally disregard recall-bias.

Conclusions

Taken together, the results show that the physiological burden of oligodontia is significant. The clinical variables were strongly associated with psychological distress. Half of the participants were bothered by anxiety and reduced mental HRQoL to such an extent that they may be in need of evaluation, guidance or treatment. These aspects were particularly pronounced in the unemployed, those with less than 10 present teeth, those wearing removable dentures, and those experiencing problems with dry mouth. Additionally, having signs of ED, in addition to tooth agenesis, may add to the reduced HRQoL. Qualitative studies may explore further reasons of psychological distress and reduced QoL in individuals with oligodontia. This study emphasizes the importance of a holistic approach by caregivers.

Notes

1. The 'normative sample' in this study is from the 'Survey of Level of Living 2002 – Cross-sectional Theme: Health.' Statistics Norway (SSB) is responsible for collecting the data, and the survey is paid for by SSB, the Norwegian Institute of Public Health, and the Department of Public Health and General Practice. Data are arranged and the right to use given in an anonymous form by the Norwegian Social Science Data Services (NSD). The SSB, the Norwegian Institute of Public Health, the Department of Public Health and General Practice, and NSD are not responsible for analyses of or interpretations of the data in this study.
2. The Nord-Trøndelag Health (HUNT) Study is a collaboration between HUNT Research Center (Faculty of Medicine, Norwegian University of Science and Technology NTNU), Nord-Trøndelag County Council, Central Norway Health Authority, and the Norwegian Institute of Public Health.

Disclosure statement

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