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Oral health status in a group of children and adolescents with myotonic dystrophy type 1 over a 4-year period

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Aim. The aim of this longitudinal study was to evaluate changes in oral health, orofacial function, and dental care in children with myotonic dystrophy type 1 (DM1) in comparison with a control group.

Methods. Thirty-six DM1 patients and 33 control patients out of originally 37 in each group were examined on two occasions about 4 years apart. Caries, plaque, and gingivitis were registered, mouth opening capacity assessed and the ability to cooperate in dental treatment estimated. Questionnaires concerning different aspects of oral health and care, symptoms of temporomandibular dysfunction (TMD), and dental trauma were also used.

Results. The DM1-patients, in particular the boys, had significantly more caries, plaque, and gingivitis than the control patients on both occasions and the increase in decayed missing or filled permanent teeth (DMFT) and surfaces (DMFS) was significantly larger. They received more dental care and had lower cooperation ability. Mouth opening capacity and increase of it was significantly lower and symptoms of TMD were significantly more frequent.

Conclusions. DM1 patients, as they grow older, have increasing amounts of plaque and risk of caries and gingivitis. They have more TMD problems. Behaviour management problems do not seem to decrease with age. Increased prophylactic care is essential for DM1 patients.

Introduction

Muscular weakness has been shown to have an impact on oral health in various ways. It affects craniofacial growth[1,2] as well as chewing capacity and oral self-cleaning ability[3–5]. Myotonic Dystrophy type 1 (DM1) is a hereditary neuromuscular disease with a most variable clinical picture. Progressive muscular waste and myotonia are the hallmarks of the disease, but almost every system in the body is affected in some way. Facial weakness is one of the earliest and most constant features[6].

The prevalence of DM1 worldwide is around 5–19/100000 but there are regions with a much higher prevalence, e.g., the Saguenay province in Quebec, Canada, and the northern parts of Sweden[6]. In a recent study from western Sweden, the prevalence in childhood was 5 per 100000[7]. The pattern of inheritance in DM1 is autosomal dominant and exhibits a phenomenon known as anticipation, which is defined as a progressively earlier onset of the disease with increasing severity in successive generations within a family. The genetic locus is an unstable CTG trinucleotide expansion on chromosome 19. The length of the expansion is roughly correlated to the onset and clinical severity of the disease. According to age at the onset of symptoms four types are discerned: mild with onset late in life and very slight symptoms, classical or adult with a median age at onset of around 20 years, childhood with onset before 10 years old and congenital with a severe and a mild subdivision according to

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the severity of symptoms at birth. The most serious forms are the congenital, severe or mild, and childhood onset types of the disease. In these types there is an obvious affect on CNS and learning and speech difficulties and neuropsychiatric disorders are common[6,8].

Reduced muscle strength and motor function in a group of children and adolescents with DM1 was shown by Kroksmark *et al.*[9] Oral motor dysfunction with impairment of lip motility, tongue motility, and lip force, most prominent in congenital DM1, and with males more affected than females, was reported in that same group of patients by Sjögreen *et al.*[10] Earlier studies of DM1 have shown an increased occurrence of plaque, gingivitis, and caries possibly due to a slower oral clearance, lower salivary secretion rate and lower motor coordination ability when compared with healthy control persons, both in adults[11,12], children and adolescents[13]. An increased frequency of temporomandibular dysfunction problems (TMD) both in adults, children and adolescents has been reported[6,13]. Behaviour management problems in the dental situation have also been shown in children and adolescents with DM1 and this was significantly correlated to the number of CTG-repeats[13].

Beside these cross-sectional studies[11–13], there are to our knowledge only some case reports on the cariological and periodontal situation in DM1 patients[14,15]. No prospective longitudinal study has been undertaken as far as we know. Since DM1 is a slowly progressing disease, the oral symptoms shown could possibly increase over time. The aim of this longitudinal study therefore is to assess the changes in oral health, orofacial function and dental care in growing children with this progressive neuromuscular disease. The findings may lead to improved preventive measures for children with DM1.

Material and methods

Subjects

Thirty-seven children with DM1, confirmed by DNA analysis of blood samples, were examined in 1999–2001 and about 3–4 years later they were invited to participate in a longitudinal examination. The inclusion criterion was a minimum of 40 CTG repeats with a range from 130 to 2300 repeats amongst the children. The patients were originally in the age 2.7–17.7 years and constituted a major part of diagnosed cases (37 out of 50) of DM1 in that age group at the time of the first examination in the counties of Västra Götaland and Skåne in the western and southern parts of Sweden. They were compared with a control group of age and gender matched children selected randomly from a public dental clinic in Gothenburg. In connection with the follow-up examination another 19 children with newly diagnosed DM1 were examined. Together with the original 37 patients they participated in a cross-sectional study of 56 children and adolescents with DM1[13]. Of the original 37 patients in the longitudinal study one boy with DM1 had moved out of the area and could not participate in the second examination. Four control patients could not be examined; one boy denied participation, one boy was not reached and two girls had moved from the area. Thus the subjects in this longitudinal study of oral health in children and adolescents with DM1 were 36 DM1 patients and 33 control patients. The time between the two examinations varied, with a mean of 44.5 (range 26–61) months in the DM1 group and a mean of 45.2 (range 35–51) months in the control group. Ten DM1 patients, eight boys and two girls, had congenital DM1 of the severe type with serious symptoms at birth. Thirteen, six boys and seven girls, had the mild congenital type with slight symptoms at birth. Thirteen, eight girls and five boys, had DM1 with childhood onset, which is between 1 and 10 years of age. Thirty of the DM1 patients went to special schools for children with learning disabilities and six went to mainstream schools. All control patients went to mainstream schools.

Methods

The study was performed as a part of a multidisciplinary study of children and adolescents with DM1 at Göteborg University, Sweden. The study was approved by the Human Ethics

Committees at the Medical Faculties at Göteborg and Lund Universities. Written informed consent was obtained from each participant or his or her parents.

The two examinations in the longitudinal study were carried out in a dental office with optimal lighting. The examinations were undertaken by one examiner (ME) and the same methods were used on both occasions. Clinical examination was made with probe and mirror to detect caries and restorations. Oral hygiene and gingivitis were recorded. Examination of oral function and temporomandibular dysfunction was also performed. Bite-wing radiographs and/or ortopantomograms were available or taken if indicated in 25 DM1 patients and in 27 control individuals at examination 1 and 28 DM1 patients and 30 control individuals at examination 2. It was not always possible to carry through the whole examination protocol on all children due to their lack of cooperation ability.

A standardised questionnaire commonly used in examination of orofacial manifestations in persons with rare disorders[16] was used. To this specific questions were added about dental care, oral hygiene habits, use of fluorides and temporomandibular function. The questionnaires were filled out by the parents.

Caries and restorations

Caries was recorded clinically according to criteria described by Koch[17]. On radiographs, proximal carious lesions within the enamel or just reaching the enamel–dentin border with no spread into the dentin were diagnosed as initial, and lesions with a clear spreading into the dentin as manifest. Manifest caries was recorded on all surfaces and the number of decayed, missing (due to caries) and filled permanent teeth (DMFT) and surfaces (DMFS) were calculated. Decayed, extracted (due to caries), and filled primary teeth (deft) and surfaces (defs) were recorded in the primary and mixed dentitions. Initial caries on permanent (DSi) and primary (dsi) teeth was also recorded at the examination. Molar and premolar teeth, extracted due to caries, were recorded as five surfaces.

In order to be able to make a true longitudinal comparison of caries in spite of different ages and different number of teeth, a key tooth at risk, the first permanent molar, was selected for examination. Comparison of caries index in this tooth was made when it was present at both examinations or missing due to caries. Teeth that erupted between examinations and were decayed at examination 2 also were included in the indexes. The same criteria for calculation of increment of caries between the two examinations were used for caries on occlusal, proximal, buccal, and lingual surfaces on first and second permanent molars.

A questionnaire regarding the intake frequency of 25 cariogenic food products according to the method used by Kristofferson *et al.*[18] was given to the participants in the study. The intake frequency of each food product was given a score according to whether they were consumed seldom = 0, every week = 2, daily = 4, two to three times a day = 8, more than three times a day = 16. An individual dietary score was calculated by adding the scores for all 25 products.

Oral hygiene and gingivitis

Plaque and gingivitis were recorded according to the visible plaque index (VPI) and the gingival bleeding index (GBI), that is the percentage of surfaces with visible plaque or bleeding on probing, respectively, out of the total number of surfaces examined[19].

Information on daily oral hygiene habits such as frequency of tooth brushing, use of toothpaste and other fluoride-containing products and hygiene aids, such as electric tooth brushes, was elicited from questionnaires and case histories.

Oral function and temporomandibular dysfunction

Maximal mouth opening as the most reliable sign of oral functional status[20] was recorded. Symptoms of temporomandibular dysfunction were elicited by a questionnaire on pain or fatigue in function. The presence of findings was recorded (symptoms = 1, no findings = 0).

There were also questions regarding oral functions such as mouth-breathing and duration of meals.

Dental care

In the questionnaires, there were questions regarding the frequency of visits to the dentist and if the child attended a specialist or public dental care clinic. Behaviour management problems during dental treatment, and the need for sedation or general anaesthesia to carry through the treatment, were assessed. An estimation of the ability to cooperate was made on a scale from zero (no problems) to ten (extreme difficulties) by the examiners and by the patient and/or parents. The patients and/or parents were also asked about satisfaction with the dental care received.

In the taking of case history, data on traumatic injuries to the teeth were recorded. To ascertain diagnosis only crown fractures and avulsions of permanent teeth, as most objective diagnoses, were included in the comparison between the groups of patients.

Statistical methods

Statgraphics plus version 3 software for Windows and the Statistical Package for the Social Sciences (SPSS), version 15 for Windows were used to analyse the data. Student's *t*-test for unpaired data, Fischer's exact test and one-way Anova test completed with Student-Newman–Keuls multiple test were used to compare the differences between the groups. Pearson's correlation was used to calculate correlations. $P < 0.05$ was considered statistically significant. Estimates of means and differences are given at 95% confidence interval if not otherwise stated. Multiple inference aspects were taken into account by not over-interpreting single weak significances.

Results

Caries and restorations

In this study the children and adolescents with DM1 had more caries than the individuals in the control group. There was a significant difference in manifest caries in permanent teeth, DMFS and DMFT, between the groups at both examinations and in the increase between the examinations (Table 1). In first permanent molars the differences in manifest caries, DMFS, were significant at both examinations and in initial caries on buccal surfaces, DSi b, at the second examination (Table 2). All other increases in indices between examinations during the study period were also larger for the DM1 group than for the control group although

Table 1. Mean value and SD for the caries indices DMFS and DMFT in permanent teeth at examination 1 and 2 and the difference between examinations in the DM1 and control group.

	DM1			Control			
Variable	***n***	**mean**	**SD**	***n***	**mean**	**SD**	***P*-value**
DMFS							
ex 1	31	3.9	6.9	29	0.6	1.5	0.014
ex 2	36	5.4	9.8	33	1.0	1.7	0.012
Diff	31	2.5	4.8	29	0.5	0.8	0.028
DMFT							
ex 1	31	2.6	4.2	29	0.6	1.4	0.012
ex 2	36	3.3	4.9	33	0.9	1.6	0.008
Diff	31	1.2	1.9	29	0.4	0.7	0.047

DMFS, decayed, missing and filled surfaces; DMFT, decayed, missing and filled teeth; Diff, difference between examination 1 and 2.
P-values concern the differences between the groups.

Table 2. Mean value and SD of DMFS, DMFS b and DSi b in first permanent molars at examination 1 and 2 and difference between examinations.

	DM1 (*n* = 29)		Control (*n* = 27)		
	mean	**SD**	**mean**	**SD**	***P*-value**
DMFS					
ex 1	2.4	3	0.3	0.8	0.001
ex 2	3.5	4	0.7	1.1	0.001
Diff	1.1	2.2	0.4	0.8	0.125
DMFS b					
ex 1	0.2	0.6	0	0	0.142
ex 2	0.4	0.9	0	0	0.051
Diff	0.2	0.5	0	0	0.061
DSi b					
ex 1	0.5	1.1	0.1	0.5	0.126
ex 2	0.6	1.2	0	0	0.017
Diff	0.1	1.2	−0.1	0.4	0.555

DMFS, decayed, missing and filled surfaces; DMFS b, decayed, missing and filled buccal surfaces; DSi b, buccal surfaces with initial caries; Diff, difference between examination 1 and 2.
P-values concern the differences between the groups.

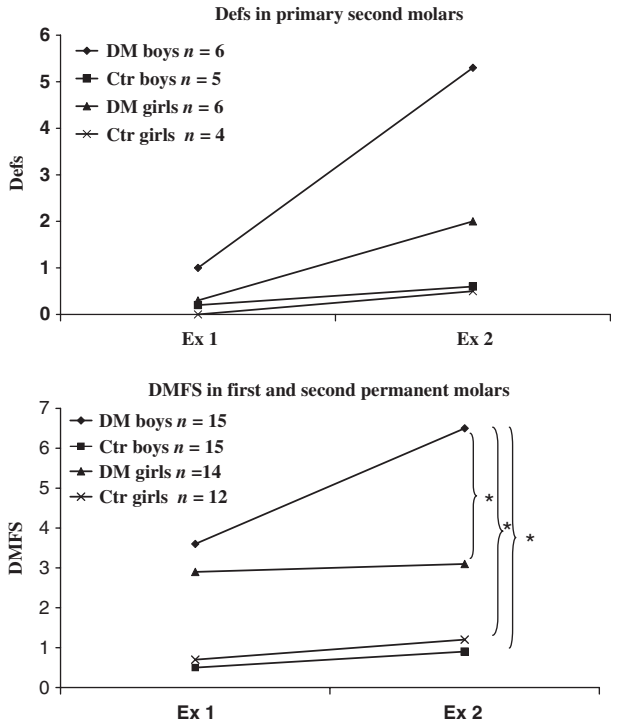

Fig. 1. Changes between examination 1 and 2 split for group and gender in decayed, extracted (due to caries) and filled surfaces (defs) in second primary molars and decayed, missing (due to caries) and filled surfaces (DMFS) in first and second permanent molars present at both examinations. Permanent molars that erupted during the time of the study and were decayed at examination 2 were also included. *Significant difference between groups ($P < 0.05$).

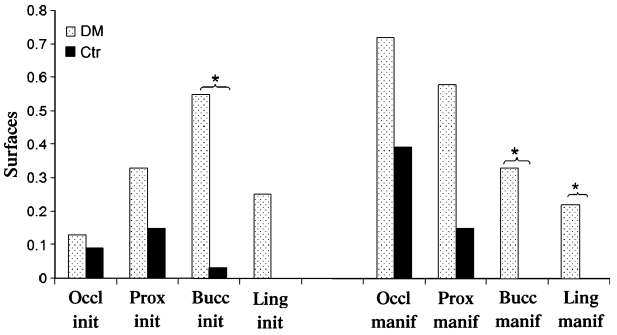

Fig. 2. Increase between examination 1 and 2 in caries on different surfaces of permanent molars present at both examinations in DM1 and control group. Surfaces of permanent molars that erupted during the time of the study and were decayed at examination 2 were also included. Occl, occlusal; prox, proximal; bucc, buccal; ling, lingual; init, initial caries; manif, manifest caries. *Significant difference between the groups ($P < 0.05$).

not significantly so. The increase in surfaces with manifest caries, defs and DMFS, in molars in the two groups split for gender is presented in Fig. 1. The increase in DMFS differs significantly ($P < 0.0005$) and boys stand for the major part of the increase in caries in the DM1 group, mean 2.9 ± 1.8 compared to DM1 girls 0.3 ± 0.5, control boys 0.4 ± 0.5, and control girls 0.5 ± 0.5.

The patients in the DM1 group had a significantly larger increase in both manifest and initial caries on buccal and lingual smooth surfaces of permanent molars (Fig. 2). Manifest caries in incisors was found in one DM1 patient at examination 1 and three at examination 2. Four patients in the DM1 group had permanent molars, seven in all, extracted because of caries. In the control group no patient had manifest caries in incisors or a permanent molar extracted due to caries.

Dietary score did not show any significant difference between the groups during this follow-up period, DM1 group 26.9 at examination 1, 27.8 at examination 2, control group 26.5 at examination 1, 24.2 at examination 2.

Oral hygiene and gingivitis

The DM1 patients, especially the boys, had more plaque and gingivitis than the children in the control group, and this was most evident on buccal surfaces. The differences between the groups were significant at both examinations (Fig. 3) e.g., visible plaque on buccal surfaces (VPIb) for DM1 51.5 ± 13.6%, control 9.3 ± 3.2% at examination 1 ($P < 0.000$) and DM1 69.1 ± 13.3%, control 16.4 ± 7.2% at examination 2.($P < 0.000$). When the groups were split for gender and for age up to ten and over 10 years old, VPIb was significant for boys all ages and for girls over 10 years of age at examination 1 and for all ages both girls and boys at examination 2.

Oral hygiene habits did not change in any remarkable way between examinations. According to the questionnaires the vast majority of children in both groups brushed their teeth two times a day. Twenty-five in the DM1 group and five in the control group were still helped with their tooth brushing at examination 2. All persons in both groups used fluoride-containing toothpaste and the use of other fluoride products was 2–3 times

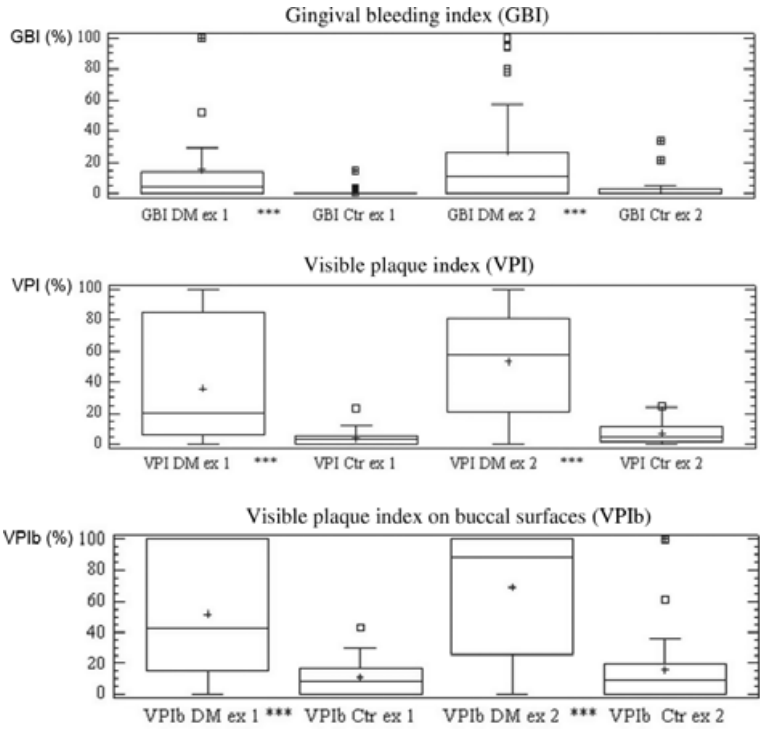

Fig. 3. Box and whisker plot of Gingival Bleeding Index (GBI), Visible Plaque Index (VPI), and Visible Plaque Index on buccal surfaces (VPIb) in DM1 group and control group at examination 1 and 2. The box extends from the lower quartile to the upper quartile, covering the centre half of each sample. The centre lines within each box show the location of the sample medians and the plus signs indicate the sample means. The whiskers extend from the box to the minimum and maximum values in each sample. Outside points (more than 1.5 times the interquartile range) are plotted separately as small squares and far outside points (more than 3.0 times the interquartile range) as small squares with plus signs through them. ***Significant differences between the groups ($P < 0.001$).

more frequent in the DM1 group than in the control group. The use of fluoride was more common in the DM1 group and the difference between DM1 group and control group was significant at examination 2 ($P = 0.019$). Devices to improve oral health, such as electric toothbrushes were also more frequent in the DM1 group than in the control group ($P = 0.048$ at examination 1 and $P = 0.003$ at examination 2).

Oral function and temporomandibular dysfunction

Symptoms of temporomandibular dysfunction were significantly more common in the DM1 group than in the control group at both examinations. Mean maximal mouth opening also differed significantly at both examinations (Table 3) and increased on the average more in the control group, 5.2 ± 1.8 mm, than in the DM1 group, 1.4 ± 2.3 mm, difference 3.8 ± 2.9, ($P = 0.009$) during the follow-up period. In ten DM1 patients over the age of ten it had even decreased at examination 2 compared to two patients in the control group.

Based on the questionnaires, a longer time for completing meals was reported by most children with DM1. At examination 1 the mean time was 23.2 ± 3.8 min in the DM1 group and 16.3 ± 1.7 min in the control group ($P = 0.001$). At examination 2 no significant change had taken place and the difference remained. The mean time was 24.1 ± 4.5 min in the DM1 group and 16.4 ± 2.8 min in the control group ($P = 0.005$). At examination 1, 53 per cent of the DM1 patients and 30 per cent of the control group said that they never used chewing gum ($P = 0.037$). This had increased to 70 per cent and 31 per cent respectively at examination 2 ($P = 0.002$). Mouth breathing was common in the DM1 group, 33 reported to sleep with their mouth open compared to four

Table 3. Reported symptoms and signs of temporomandibular dysfunction at examination 1 and 2. (*n* = number of subjects).

	Examination 1			Examination 2		
	DM1 (*n* = 36)	Control (*n* = 33)	*P*-value	DM1 (*n* = 36)	Control (*n* = 33)	*P*-value
Reported symptoms*						
Number of individuals presenting with symptoms	12	2	0.005	15	4	0.015
Clinical measuring						
Maximal opening of the mouth (mm)	41.0	48.3	0.000	42.4	53.5	0.000

*Pain or difficulty in maximal mouth opening and/or pain or fatigue when chewing.
P-values concern the differences between the groups.

in the control group at both examinations ($P = 0.000$ at both examinations).

Dental care

The frequency of visits to dental clinics was, according to the questionnaires, significantly higher in the DM1 group than in the control group at both examinations, 3.2 ± 1.3 compared to 1.1 ± 0.9, ($P = 0.002$), at examination 1, 2.8 ± 0.6 compared to 1.2 ± 0.2, ($P < 0.000$), at examination 2. Pedodontic and/or orthodontic specialist care was more common among the DM1 patients than the control patients, 16 compared to three at examination 1 ($P = 0.001$) and 21 compared to two at examination 2($P = 0.000$). The estimation of behaviour management problems did not change much between the examinations and the DM1 group had more problems than the control group (Table 4). The correlation between estimations at examination 1 and 2 were $r = 0.79$, for examiner and $r = 0.49$ for patient/parents indicating a somewhat larger variation in the estimations of the patients/parents than of the examiner. The need for general anaesthesia, carried out or deemed necessary for more extensive treatment, had increased somewhat in the DM1 group from 18 at examination 1 to 20 at examination 2. In the control group, there had been no need for general anaesthesia.

According to the questionnaires all patients and their families were satisfied with the dental care that the patients received, except for one DM1 patient at examination 2 whose father thought his son needed more time for adjusting to the dental situation and one control patient, also at examination 2, who wanted to be called to dental care more often.

Table 4. Behaviour management problems and patient cooperation problems on a scale from 0 = no problems to 10 = major problems. Dentist's and patient's/parents' estimation at examination 1 and 2.

	DM			Control			
Variable	*n*	mean	SD	*n*	mean	SD	*P*-value
BMP ex 1	31	3.9	3.2	33	0.2	0.5	0.000
Coop ex 1	32	3.9	3.5	30	0.8	1.6	0.000
BMP ex 2	35	3.7	3.1	33	0.2	0.3	0.000
Coop ex 2	35	3.9	3.2	30	0.9	1.1	0.000

BMP, behaviour management problems, dentists estimation.
Coop, patient cooperation problems, patient's/parents' estimation.
P-values concern the differences between the groups.

Dental trauma registered as the number of crown fractures was eight and avulsions one in the DM1 group. In the control group, there had been five crown fractures and one avulsion. The differences were not statistically significant.

Discussion

This longitudinal prospective cohort study showed that the children and adolescents with DM1 in it had abundant and increasing amounts of plaque and a continuous and increasing risk of caries and gingivitis. They also had more TMD problems and less cooperation ability in dental treatment than the children and adolescents in the control group.

The characteristic personality traits with mental retardation, neuropsychiatric problems and fatigability of the DM1 patients meant that it was not always possible to carry through the whole examination protocol even if the examination methods were simplified as much as possible. The fact that many of them could only tolerate extra-oral radiographs, which are not so good for caries diagnosis, might have led to under-scoring. Moreover, the questionnaires were sometimes answered by parents with classical DM1 who might have cognitive deficits affecting their ability to answer questions[21,22]. The overall problem with a study of this rare disease is to obtain a large enough number of patients. The number of patients in this study is small but still comprises a fair number (37 out of 50) of the known cases of young patients with the disease in the western and southern parts of Sweden at the time of the first study.

In spite of the fact that the DM1 patients, according to questionnaires and case histories, had as good habits concerning diet and oral hygiene as the healthy control patients and got more frequent dental care, they still had more caries, plaque, and gingivitis and the differences between the groups increased between the examinations. Boys with DM1 answered for the majority of the increase. This is in agreement with the results in a study by Crossner & Unell[23], but in contrast to data from the National Board of Health and Welfare[24] according to which there is no difference in caries prevalence between boys and girls in Sweden. The majority of the children with the severe congenital form of DM1 in this study were boys. However, severity of DM1 did not always coincide with high caries prevalence, which often affected the boys with the less serious types more. An explanation for this could be that the children most seriously affected by DM1 are the ones with least ability to manage by themselves. They get more care and could thus be less seriously affected by lifestyle diseases like caries. This is in accordance with studies on oral health in mentally retarded adults which have shown that individuals with mild mental retardation, living with their family or in their own apartments, show a higher caries incidence and prevalence compared to subjects with severe mental retardation living in institutions[25].

The correlation between oral motor function, oral clearance, plaque, and caries is well known[3–5]. In general, small children have slower oral clearance than adults[4]. The ability to maintain good oral hygiene is hampered by decreased muscle strength[26] and abundant plaque is a recognised risk factor in the caries process[27]. DM1 children in general show reduced muscle strength[9,10] and progressive weakening of the orofacial muscles[21]. Boys with DM also have significantly lower motoric ability both as to lip function and tongue motility[10]. This could be an explanation for the large amount of plaque and gingivitis found in the DM1 group compared to that of the healthy controls in this study. This was especially the case for the older boys. As long as the parents or care-takers took the responsibility for the oral hygiene procedures the situation was better. Similar results have been shown in a study of patients with Duchenne Muscular dystrophy where decreasing muscle function was associated with increased plaque amount leading to gingival inflammation[28]. It seems likely that the slower oral clearance and less pronounced self-cleaning ability demonstrated in adults with DM1[12] is present in young persons with DM1 as well.

A higher prevalence of temporomandibular dysfunction is documented in adults with DM1[6] and was also shown in a previous cross-sectional study of children and adolescents with DM1[13], although it was not a serious complaint. The symptoms of TMD increased in both DM1 and control groups between examination 1 and 2 in this longitudinal study. The maximal opening capacity of the mouth was lower in the DM1 group than in the control group at both examinations. In a longitudinal view, it has been shown that there are more TMJ-problems if the mouth opening capacity is lower than normal[29]. The most frequent symptom in the DM1 group was pain or fatigue when chewing. Eating and drinking difficulties also has been reported in the DM1 group[10]. This may explain the longer time for meals elicited

from the questionnaires. Longer time for completing meals has also been demonstrated in a group of adults with DM1[30].

Mental retardation and neuropsychiatric conditions such as attention deficit hyperactivity disorder (ADHD) are prevalent in congenital and early childhood DM1[6,8]. This is also the case for the children and adolescents with DM1 in this study among whom there is a high percentage of mental retardation and autistic spectrum disorders[31]. Fatigue and excessive day-time sleepiness is reported as well[31]. Higher caries prevalence and more behaviour management problems (BMP) have been demonstrated in a study of oral health, dental anxiety, and behaviour management problems in children with ADHD[32]. Adult patients with DM1 score high in the personality traits harm avoidance and fatigability[22]. High anxiety score also has been shown[33]. It may be reasonable to believe that children and adolescents with DM1 have these personality traits as well. The DM1 group in this study demonstrated less cooperation ability than the control group at both examinations. They were anxious and easily fatigued. Since they have become older during the time of the study it might be expected that the cooperation ability should improve but that was only marginally the case. The number of patients in need of general anaesthesia for their dental treatment increased. As general anaesthesia entails a greater risk for patients with DM1[6] this leads to more precautions and planning than general anaesthesia for healthy children. Prophylactic treatment and acclimatisation is essential to avoid the need for general anaesthesia both by preventing caries and gingivitis and allowing the patient to become accustomed to the dental clinic and the treatment there. This is even more important in view of the fact that past caries experience remains the most powerful known single predictor of future caries in the young permanent dentition[34,35]. The patients in the DM group were treated in specialist care significantly more often then the patients in the control group, probably largely on account of cooperation problems in the dental situation and the impact of the disease on the orofacial development. Cooperation between the public dental clinic close to where the patients live, for prophylactic care with short intervals, and the specialist dental clinic, for treatment planning and specialist care, ought to be optimal for the DM1 patients. Methods for plaque removal and fluoride treatment, that are easy to manage, are also desired for the patients' home-care.

There are several reports on increased frequency of tumbles and falls in DM1[36,37]. However, no significant difference as to dental trauma was shown between the DM1 group and the control group in this study. To evaluate this better, a larger patient/control group and a more specific investigation are probably needed.

The DM1 group in this study is small and heterogeneous in age since DM1 is a rare disease. Nevertheless, it is important that the influence of the disease on oral health is observed and paid attention to. The methods to examine the patients suffering from it ought to be simplified as far as possible since many of them have great problems with cooperation. From this study it may be concluded that the different traits of DM1 seem to combine to result in more pronounced deterioration over time of oral health in young patients with DM1 compared to control patients. They show increasing amounts of plaque and are at continuous and increasing risk of caries and gingivitis. They have more TMD problems. Behaviour management problems are common and do not seem to decrease with age. Intensified prophylactic care with methods that are easy to manage is of great importance for them.

What this paper adds

- The study shows that children and adolescents with myotonic dystrophy (DM) in this study have poor oral health that deteriorates over time.
- Children and adolescents with DM in this study have difficulties cooperating in dental treatment and this does not seem to improve with age.

Why this paper is important to paediatric dentists

- The results stress the importance for increased prophylactic care for children and adolescents with myotonic dystrophy (DM) from an early age.
- The results further add to the basis of knowledge for oral health care programmes for DM patients.

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