

## The effect of lip strengthening exercises in children and adolescents with myotonic dystrophy type 1

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### ABSTRACT

**Objective:** Myotonic dystrophy type 1 (DM1) is a slowly progressive neuromuscular disease. Most children and adolescents with DM1 have weak lips and impaired lip function. The primary aim of the present study was to investigate if regular training with an oral screen could strengthen the lip muscles in children and adolescents with DM1. If lip strength improved, a secondary aim would be to see if this could have an immediate effect on lip functions such as lip mobility, eating and drinking ability, saliva control, and lip articulation.

**Methods:** Eight school aged children and adolescents (7–19 years) with DM1 were enrolled in an intervention study with a single group counterbalanced design. After three baseline measurements four children (Subgroup A) were randomly chosen to start 16 weeks therapy while the others (Subgroup B) acted as controls without therapy. After 16 weeks the subgroups changed roles. During treatment the participants exercised lip strength with an oral screen for 16 min, 5 days/week. Lip force, grip force (control variable), and lip articulation were followed-up every fourth week. At baseline, after treatment, and after maintenance, the assessment protocol was completed with measurements of lip mobility using 3D motion analysis and parental reports concerning eating ability and saliva control.

**Results:** Seven of eight participants improved maximal lip strength and endurance but only four showed significant change. Increased lip strength did not automatically lead to improved function. There was a wide intra-individual variation concerning speech and eating ability within and between assessments. The treatment programme could be carried out without major problems but the frequency and the effect of training were affected by recurrent infections in some.

**Conclusions:** Maximal lip force and lip force endurance can improve in school aged children and adolescents with DM1. Improved lip strength alone cannot be expected to have an effect on lip articulation, saliva control, or eating and drinking ability in this population. Lip strengthening exercises can be a complement but not a replacement for speech therapy and dysphagia treatment. A prefabricated oral screen is an easy to use tool suitable for strengthening lip exercises.

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### 1. Introduction

Myotonic dystrophy type 1 (DM1) is a multisystemic disease with muscle weakness and myotonia (delayed muscle relaxation) as cardinal symptoms. Other systems commonly affected are the heart, lungs, smooth muscle, endocrine regulation, and brain [1]. Developmental delay and learning disabilities of varying degrees, are common in the congenital and childhood forms of the disease

and the prevalence of neuropsychiatric disorders is higher in this group than in the general population [2,3]. DM1 has an autosomal dominant inheritance pattern and is caused by an expansion of a CTG-repeat sequence (trinucleotide expansion) on chromosome 19q13. In general, the earlier the symptoms occur, the more severe the clinical symptoms of the disease will be [4].

In a Swedish study [5] of 56 children and adolescents with DM1 all had impaired facial expression, 60% moderately or severely reduced intelligibility, 52% eating difficulties, and 37% drooling. Six individuals had no speech. Lip force was measured with a lip force meter (LF100). The majority had very weak lips compared to healthy controls. A threshold of 8 N for weak lips in children and

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adolescents older than 4 years was proposed. Thirty-five patients had been assessed with the same protocol 3–4 years earlier [6] which made it possible to study the progression of orofacial dysfunctions during this time span. It was found that the progressive weakening of the orofacial muscles often began before puberty and manifested as deteriorated facial expression, reduced intelligibility, and increased drooling. Orofacial functions improved during childhood in some patients.

There is no consensus regarding the effect of oral motor exercises on lip strength in individuals with DM1. Earlier studies have stated that physiotherapy can be recommended without an increased risk of damage to the muscles [7,8]. However, it still remains to be shown if physical exercises can improve strength and motor function and postpone the progression of the muscular atrophy in DM1 [7].

An oral screen has been used as a therapy tool for the purpose of strengthening the chain of muscles also known as the buccinator mechanism [9–12]. Anatomic research on the perioral muscles [13] has revealed that there is a *deep unit* composed of the buccinator muscle and the inner ring of the orbicularis oris, and a *superficial unit* built up by the depressor anguli oris, the zygomaticus, the risorius and the outer ring of orbicularis oris. The muscles included in each unit are densely interrelated. The perioral muscles are active in lip closure and lip rounding and thus involved in the production of bilabial and labiodental consonants and rounded vowels. Lip closing and lip control are also important for the oral phase of swallowing and saliva management [14].

The oral screen is placed inside the lips and in front of the teeth. There are prefabricated oral screens in hard or soft plastic material (Fig. 1) but they can also be custom made by dental technicians. The training procedure is not complicated. For active use, the patient should try to keep the oral screen inside the lips while someone is pulling the ring. Daily exercises are often recommended and each training session lasts just a few minutes.

The effect of exercises with an oral screen for orthodontic treatment in typically developing children has been studied. Owman-Moll and Ingervall [11] studied children with incompetent lip function and found a substantially increased lip force after 1 year of treatment. It was discussed if increased lip force would be of value for the stability of orthodontic treatment. Thuer and Ingervall [10] found in their study of children with protruding maxillary incisors, an increase in lip force but the pressure from the lips on the teeth was unaffected. Tallgren et al. [12] treated children with lip and/or tongue dysfunction with a custom made oral screen in order to study the effect on orofacial muscle activity and changes in craniofacial morphology. Their results indicated a

decrease in orofacial muscle activity during oral functions but no significant change in tooth position. Hägg et al. [15] found in a retrospective study that self-training with an oral screen could improve lip force and swallowing capacity in stroke patients with oropharyngeal dysphagia. It was concluded that the treatment results were more likely attributable to sensory motor stimulation and the plasticity of the central nervous system, than the training of the lip muscles per se.

Different instruments and techniques have been suggested for the measure of maximal lip force [15–18]. A number of video-computer interactive systems for objective evaluation of facial expressions have also been explored [19–23]. Facial expressions that are performed with maximal effort such as an open mouth smile and lip pucker have been found to be the most reproducible [22,24,25]. In a methodological study (unpublished data) comparing quantitative measurements and qualitative assessments of lip function, a strong relation was found between lip force, lip contraction, eating ability and saliva control. These results raised the question if improved lip strength could have a positive effect on lip functions. The primary aim of the present study was therefore to investigate if regular training with an oral screen could strengthen the lip muscles in children and adolescents with DM1. If lip strength improved, a secondary aim would be to see if this could have an immediate effect on lip functions such as lip mobility, eating and drinking ability, saliva control, and lip articulation.

## 2. Materials and methods

School aged children and adolescents with congenital or childhood DM1 from the region of Västra Götaland (approximately 2 million inhabitants) were invited via the paediatric neurologist or the local habilitation team to participate in an intervention study. A letter of invitation was sent out to 18 families. Eight families accepted the invitation. Another two individuals wanted to participate but after discussion with the parents it was decided that they were too weak and affected by the disease to take part in the study. Two more families explained that they were interested but participation was not possible for practical reasons. Six families did not answer the invitation for unknown reasons. No reminder letter was sent out. Of the eight participants enrolled in the study (Table 1), seven attended special schools, six due to learning disability and one because of autism.

Informed consent was obtained from all the participants and the study was approved by the Ethics Committee at the University of Gothenburg.

### 2.1. Study design

A one-group single-treatment counterbalanced design was used [26]. The participants came to the clinic once a week during 3 weeks for baseline measurements. The six first participants were randomised to either immediately start the 16 week exercise program (Subgroup A) or to act as a control during the following 16 weeks (Subgroup B). Two children were enrolled later in the study and had the opportunity to choose if they wanted to start treatment immediately or wait as the intervention interfered with the summer holidays. After the baseline was established, examinations were carried out every fourth week. A more extensive follow-up was carried out at the clinic after the treatment and maintenance periods. The examinations made in between could be done at school or in the patient's home. None of the participants were receiving concomitant speech therapy or oral motor treatment in addition to that provided by the study. The following examinations were included in the baseline and follow-up protocol in order to monitor the possible effect of therapy.



Fig. 1. Prefabricated oral screen used in the present study.

**Table 1**  
Subgroup affiliation, case number, age, sex (f, female and m, male) and diagnosis in eight children and adolescents with myotonic dystrophy type 1 participating in an intervention study.

Subgroup & no.	Age (years)	Sex	Form of DM1	Percentage correct production (%)		Drooling	Eating difficulties	Lip force (N)
				/b, p, m/	/f, v/			
A1	15	f	Childhood	52	90	No	Not really	4
A2	19	m	Congenital	100	100	No	Not really	4
A3	7	m	Congenital	35	95	Mild	Not at all	2
A4	19	f	Congenital	6	85	Mild	Not at all	6
B1	17	f	Congenital	100	100	No	Not at all	21
B2	15	m	Childhood	100	100	No	Somewhat	3
B3	11	m	Congenital	0	0	No	Not at all	6
B4	13	m	Childhood	100	100	Mild	Not really	18

The percentage correct production of bilabial (b, p, m) and labiodental (f, v) consonants, the degree of drooling and eating difficulties and the maximal lip force at first baseline is also presented.

### 2.1.1. Lip force

Maximal lip force and endurance were measured with a calibrated lip force meter (LF100, Detektor AB, Gothenburg, Sweden) [5,15]. The same type of oral screen that was used for the intervention (Ulmer model, Dentarum, Pforzheim, Germany) was connected to the measuring device. The patient was told to keep the oral screen inside the lips while the examiner pulled the handle with an increasing force until the oral screen was dropped. The best of three values obtained (Newton) was saved. Endurance of the lip muscles was evaluated by testing for how many seconds the patient could keep the oral screen inside the lips against a resistance equal to 50% of the achieved maximum lip force.

### 2.1.2. Grip force

Grip force was used as a control variable and was measured with a grip force meter (Grippit, Detektor AB, Gothenburg, Sweden) [27]. As with the lip force meter, the best of three values obtained (Newton) was saved.

### 2.1.3. Lip mobility

A 3D analysis of lip mobility was performed with a tracking system for mimic muscle evaluation (SmartEye Pro 3.7 - MME, SmartEye AB, Gothenburg, Sweden) [23] and two calibrated video cameras (Sony XC-HR50). The facial expressions used for analysis of lip mobility were open mouth smile and lip pucker. Certain landmarks on the face were marked with a mouse on chosen snapshots from the video recording. The position of the landmarks was then automatically tracked during voluntary movements. There was a built-in correction in the program for any head movements accompanying the facial movements. A computer generated log file registered the 3D position of the oral commissures.

### 2.1.4. Lip articulation

The treatment effect on lip articulation was tested with SVANTE—a Swedish Articulation and Nasality Test [28]. The complete test was performed three times, before, in the middle of, and after the study period. A shorter version including only words and sentences containing bilabial and labiodental speech sounds was carried out during the less extensive examinations. The articulation test was video recorded. The speech evaluation of the recordings made at first baseline, after treatment, and after maintenance (when the complete test was presented) was independently carried out by two speech language pathologists, the first author (LS) and one not involved in the project. When disagreement arose, they watched the video together and made a consensus decision. Compensatory strategies were described as: labiodental, labiolingual, dental, or other. Exact inter-judge percentage agreement compared point-by-point on single phonemes was 100% in four participants without impaired lip articulation and 95% in the group with impaired speech. The

consensus was considered so satisfactory that the rest of the video recordings could be evaluated by the original examiner alone.

### 2.1.5. Eating and saliva control

The parents filled out a questionnaire with scales for rating eating and drinking ability and saliva control before (three times), in the middle of, and after the study period. They answered on a 4-point scale if there were any difficulties with eating and drinking and could choose between: not at all, not really, somewhat, and very much. The parents were also asked to specify difficulties by answering some yes/no questions (Table 2). If the child had a drooling problem it was rated if the drooling was mild, moderate, severe, or very severe.

## 2.2. Training procedure

The method of intervention was training with an oral screen for 16 min, 5 days a week. The treatment programme was designed to be intense enough to be effective and yet not too tiring for a child or adolescent with DM1. The daily programme was divided into three training sessions. Session one and two both included 3 min of active training with the oral screen. A teacher, a parent, or the patient himself/herself pulled the oral screen for 5 s as much as possible without pulling it out of the mouth and then paused for 5 s. This action was repeated over a 3-min period. Session three included a 10 min passive use of the oral screen inside closed lips in order to experience closed mouth and nasal breathing. A sand glass or a timer was used as a reminder of time. A log book was used to keep record of the training. The exercises were preferably done at school during the day as individuals with DM1 are generally extremely tired in the morning and in the evening, but this was not always possible to arrange. Four participants carried out the exercises at school, three at home, while one child carried out two of the daily sessions at school and one at home.

## 2.3. Statistical analysis

The Wilcoxon Signed Ranks Test (related observations) was used in order to study if there was a significant difference in the

**Table 2**  
Questions specifying the child's eating and drinking ability.

Yes/No questions on eating and drinking ability	
Q1	Is it difficult to get food off spoon with lips?
Q2	Do food and liquids leak out of corners of mouth?
Q3	Does food remain in mouth after finished meal?
Q4	Do you swallow large pieces of food without chewing?
Q5	Is it difficult to chew—to grind food between the teeth?
Q6	Do you cough daily during meals?
Q7	Do you press the tongue forward when you swallow so that food comes out of the mouth?
Q8	Does it take long time to swallow bites of food?

change of maximal lip force after treatment compared to after maintenance or no treatment. Calculations were made with SPSS for Windows, version 15.0.

### 3. Results

#### 3.1. Lip force and grip force

The mean deviation of maximal lip force and maximal grip force across the three first baseline measures was determined across participants and was used as references for significant change. The mean deviation of maximal lip force during baseline was 3.2 N and the corresponding value for maximal grip force was 20 N.

#### 3.2. Case A1

##### 3.2.1. Maximal force

Maximal lip force improved significantly during treatment and returned to baseline after 16 weeks of no treatment (Fig. 2). Grip force varied in a similar pattern to the lip force but within or more close to baseline.

##### 3.2.2. Endurance

Lip force endurance improved during treatment and the improvement maintained. At baseline 3 a lip force of 3.5 N (50% of maximal lip force) could be sustained for 3 s, after treatment 4.5 N for 6 s, and after no treatment 3.5 N for 15 s.

#### 3.3. Case A2

##### 3.3.1. Maximal force

Case A2 improved maximal lip force during treatment and the result was kept above baseline after no treatment (Fig. 2). The change was significant. Maximal grip force decreased during the same time period.

##### 3.3.2. Endurance

There was no obvious change related to lip force endurance. At baseline 3, 2.5 N could be sustained for 14 s, after treatment 4.5 N for 5 s, and after no treatment 3.5 N for 6 s.

#### 3.4. Case A3

##### 3.4.1. Maximal force

Case A3 improved the maximal lip force from 2 N during baseline to 3 N after treatment and returned to the baseline results after no treatment (Fig. 2). The improvement was not considered to be significant. There was a peak performance of grip force at baseline 2, otherwise the grip force varied between 39 and 52 N.

##### 3.4.2. Endurance

Lip force endurance improved somewhat from baseline 3 where 1 N was sustained for 0 s, after treatment 1.5 N for 2 s, and after no treatment 1 N for 1 s.

#### 3.5. Case A4

##### 3.5.1. Maximal force

The results from the maximal lip force measurements fluctuated in case A4 (Fig. 2). Baseline varied between 2 and 6 N. There were two peak performances of 7 N during treatment but after treatment and after no treatment there was a decrease to baseline. Grip force measurement also fluctuated within baseline results except for a dip at assessment 5.

#### 3.5.2. Endurance

Lip force endurance improved. At baseline 3, 2.5 N was sustained for 3 s, after treatment 1.5 N for 34 s, and after no treatment 3 N for 6 s.

#### 3.6. Case B1

##### 3.6.1. Maximal force

Case B1 (Fig. 3) improved maximal lip force significantly after treatment. Grip force also increased above baseline measurements but in contrast to lip force the grip force had a decreasing trend during treatment.

##### 3.6.2. Endurance

There was an improvement in lip force endurance. At baseline, 10.5 N was sustained for 5 s, after maintenance 11 N for 7 s, and after treatment 14.5 N for 18 s.

##### 3.6.3. Maximal force

The lips were stronger during and after treatment compared to baseline measures but the difference did not reach the criteria for significant change (Fig. 3). Grip force was below baseline during both treatment and maintenance.

##### 3.6.4. Endurance

Lip force endurance showed the best result during maintenance. At baseline 3, 2 N was sustained for 3 s, after maintenance 2.5 N for 9 s, and after treatment 2.5 N for 3 s.

#### 3.7. Case B3

##### 3.7.1. Maximal force

Lip force increased significantly after treatment compared to baseline but not compared to maintenance (Fig. 3). Grip force varied approximately in the same pattern as lip force but the variations were within or close to baseline.

##### 3.7.2. Endurance

Lip force endurance improved. During maintenance 5.5 N was sustained for 1 s and after treatment the same lip force for 5 s.

#### 3.8. Case B4

##### 3.8.1. Maximal force

Both lip force and grip force varied within the baseline results during maintenance and treatment (Fig. 3). The peak performance of 18 N in lip force was already achieved at baseline 1. This result was not reached again until the end of treatment.

##### 3.8.2. Endurance

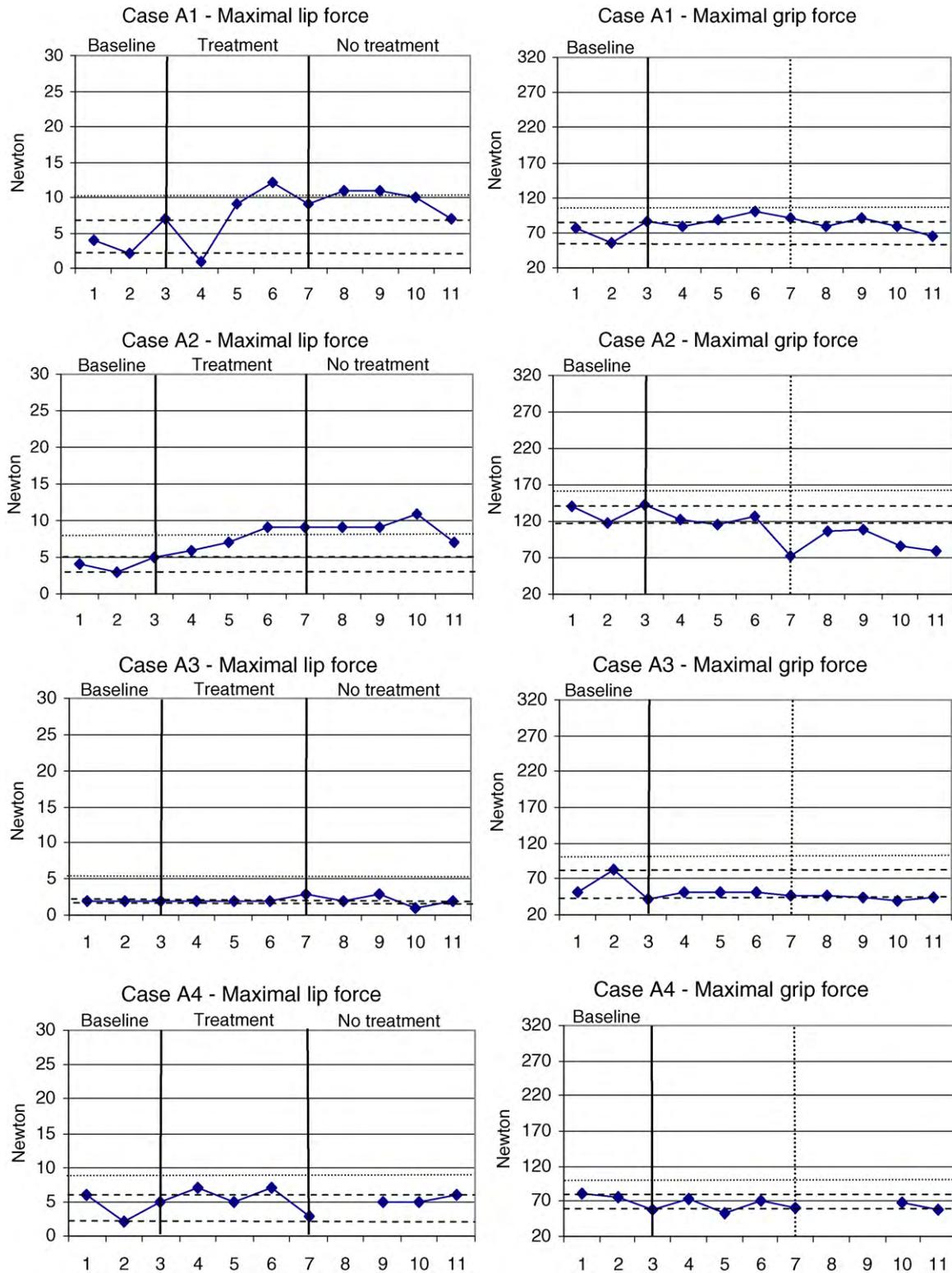
At baseline 1, 9 N was sustained for 0 s and after treatment for 3 s.

#### 3.9. Counterbalancing

In Subgroup A, treatment was carried out before the period of no treatment. Two individuals in this group had a significant improvement in maximal lip force after treatment compared to baseline (cases A1 and A2) and the improvement lasted for 12–16 weeks after treatment.

Two individuals in Subgroup B (cases B1 and B3) improved maximal lip force considerably already during maintenance. Only in case B1 the maximal lip force improved further after treatment.

Fifty percent of the participants reached the criteria for improved maximal lip force after treatment. However, when analysing the results on a group level there was no significant



**Fig. 2.** The results from lip force and grip force measurements in four individuals with myotonic dystrophy type 1 (Subgroup A) during baseline, treatment with an oral screen, and no treatment. The broken lines indicate the baseline area and the dotted line the proposed level for significant change.

difference between the changes in lip force obtained after treatment compared to those obtained after no treatment ( $z = -0.911, p = 0.362$ ).

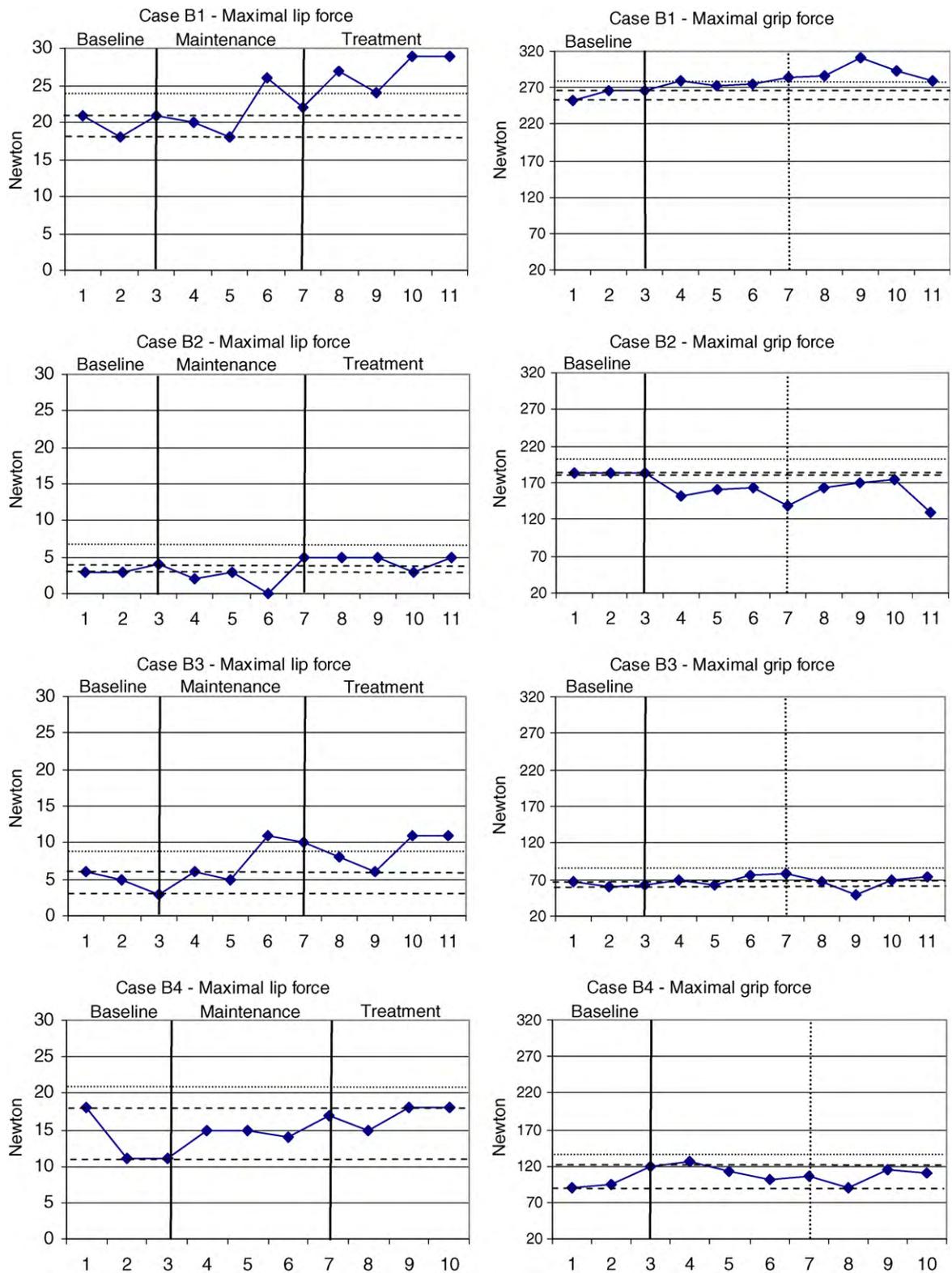
**3.10. Lip function**

The results from the evaluation of lip mobility, lip articulation, saliva control and eating and drinking ability are summarised in

Tables 3 and 4. It was analysed whether a significant change in maximal lip force after treatment could be related to changes in lip function (see below).

**3.10.1. Lip mobility**

The mean deviation of mouth width during baseline was 2.3 mm for lip pucker and 4 mm for open mouth smile. A change in mouth width of 5 mm or more was chosen as clinically relevant



**Fig. 3.** The results from lip force and grip force measurements in four individuals with myotonic dystrophy type 1 (Subgroup B) during baseline, maintenance, and treatment with an oral screen. The broken lines indicate the baseline area and the dotted line the proposed level for significant change.

criteria for significant change. Of those who got stronger lips after the intervention, one individual (case B1) had a significant decrease in mouth width in an open mouth smile after treatment and one after no treatment (case A2). No one had a significant change in mouth width in a lip pucker or a significant increase in mouth width in an open mouth smile.

### 3.11. Lip articulation

Four individuals had impaired lip articulation (cases A1, A3, A4, and B3). Impaired lip closure in bilabial consonants was in general compensated with labiodental or labiolingual articulation. Labiodental consonants were mostly compensated with interdental or

**Table 3**

The percentage correct production of bilabial and labiodental consonants and the mouth width in an open mouth smile and a lip pucker assessed in an intervention study of eight children and adolescents with myotonic dystrophy type 1.

Case	Percentage correct production (%)						Mouth width (mm)					
	Bilabials (b, p, m)			Labiodentals (f, v)			Smile			Lip pucker		
	I	II	III	I	II	III	I	II	III	I	II	III
A1	37	32	61	93	100	100	48	47	md	36	36	34
A2	100	100	100	100	100	100	44	40	38	32	32	31
A3	60	26	6	95	100	100	39	36	34	32	29	31
A4	10	17	16	80	75	70	44	43	42	37	36	39
B1	100	100	100	100	100	100	56	48	49	31	29	28
B2	100	100	100	100	100	100	50	50	46	30	32	30
B3	0	0	0	0	0	0	46	44	44	32	38	35
B4	100	100	100	100	100	100	63	62	62	38	39	42

I, baseline (mean value); II, after treatment; III, after maintenance/no treatment. md, missing data.

linguadental articulation. The intra-individual variation of the speech performance was high, especially in bilabial consonants. Four individuals had impaired lip articulation (cases A1, A3, A4, and B3) and two of them improved maximal lip force after the intervention (cases A1 and B3). In case A1, the percentage correct production of bilabial consonants increased after no treatment and of labiodental consonants after treatment. Case B3 never used labial articulation of consonants, neither before nor after treatment. Bilabial consonants were most often substituted with dentals and labiodentals with /h/.

### 3.12. Saliva control

According to the questionnaire, none of the participants who got significantly stronger lips after intervention had any problems with saliva control at baseline. Case A1 reported drooling (mild) for the first time after no treatment.

### 3.13. Eating and drinking

When the parents rated the degree of eating and drinking difficulties, all but one (B2) had no or very mild difficulties. The most common difficulties were related to lip function: Q1–Q3 (Table 2). The answers from ratings and yes/no questions varied considerably within baseline and between assessments. Case A1 had mild difficulties with eating and drinking at baseline. It was difficult to get food off spoon with lips (Table 2; Q1). She no longer complained of this symptom after treatment but the problem returned after no treatment. The other three participants who got significantly stronger lips after intervention reported no difficulties with eating and drinking, neither before nor after treatment.

## 4. Discussion

An intervention study was carried out in order to examine if school aged children with DM1 could improve lip strength through lip exercises with an oral screen. The results showed an increased maximal lip force compared to the baseline measurements in seven participants but only four showed a significant change after treatment. Some improvement was already achieved before treatment in three individuals which indicates that other variables than treatment influenced the performance of maximal lip force. The main explanation is considered to be that the frequent assessments improved the technique to keep the oral screen inside the lips against resistance. Hägg et al. (2008) investigated the reliability of the lip force meter that was used in the present study (LF100) and found that the intra-investigator reliability was excellent and the inter-investigator reliability was good or excellent [15].

DM1 is a multisystemic disease and the general condition could therefore be affected for various reasons such as excessive tiredness, gastrointestinal problems and recurrent infections [1]. Two individuals wanted to but could not participate in the study for these reasons. It was expected that variations in general health condition would interfere with the results and therefore the measurement of grip force was included as a control variable in the study protocol. In most cases, grip force and lip force followed the same curve but grip force variations were more close to baseline or below baseline. Case A1 had two viral infections during the treatment period which were followed by major decreases in lip force. Grip force was less affected. The reason for this could be that not only impaired health but also the intermission of treatment had an influence on the results. Case A4 had recurrent viral infections during treatment and maintenance and both maximal

**Table 4**

Days of treatment and the occurrence of eating and drinking difficulties and drooling reported by the parents to eight children and adolescents with myotonic dystrophy type 1 participating in an intervention study.

Case	Days of treatment	Eating/drinking difficulties <sup>a</sup>			Drooling <sup>b</sup>		
		I	II	III	I	II	III
A1	48	1	0	1	0	0	1
A2	75	0–1	0	0	0	0	0
A3	66	0	0	1	0–1	0	0
A4	No info	0–1	1	1	1	1	1
B1	85	0	0	0	0	0	0
B2	63	1–2	2	0	0	0	0
B3	54	0	0	0	0	0	0
B4	66	0–1	2	1	0–1	1	0

I, baseline (mean value); II, after treatment; III, after maintenance/no treatment.

<sup>a</sup> Eating difficulties: 0 = not at all, 1 = not really, 2 = somewhat, and 3 = very much.

<sup>b</sup> Drooling: 0 = no, 1 = mild, 2 = moderate, and 3 = severe.

lip force and grip force results seemed to follow her health condition. Case B4 had a sensitive mouth due to the eruption of teeth. The pain interfered with both treatment and assessment. Although, all participants had the same neuromuscular disease the group was very heterogeneous. The participants were of different age and had different cognitive and behavioural profiles. Some were more affected by the disease. The majority had very weak lip muscles while two adolescents were quite strong. The three participants who got significantly stronger lips after treatment were all motivated to follow the training recommendations and they always made an effort to improve records. The two youngest children did not fully understand why they were doing the exercises and were less motivated for training.

Another aspect of lip force is endurance. Endurance was defined as the length of time that an individual can sustain a given force [29,30]. As endurance was related to maximal performance it was not always possible to directly compare results from different assessments. Estimations were done from the given results which showed that lip force endurance can improve in children and adolescents with DM1. A more effective way of examining lip force endurance would probably have been to examine for how long time the individual could sustain the same force at each examination and not to change the force according to variations in maximal performance.

The flaccid dysarthria in individuals with DM1 is primarily caused by weak and hypotonic oral and velopharyngeal muscles and respiratory insufficiency [5,31] but the symptoms worsen when general health condition and alertness is affected. The experience from clinical practice is that parents of children with DM1 and patients with DM1 often confirm that tiredness negatively affects speech. The present study offered unique information concerning speech in children and adolescents with DM1 as lip articulation was tested so frequently during a time span of approximately 9 months. An interesting finding was that the production of bilabial consonants could vary so much from one assessment to the other and even during the same assessment. No single explanation for these variations was detected. One child was at one point obviously affected by a viral infection. The children with dysarthria and impaired lip articulation had cognitive deficits. They did not seem to be aware of their deviant articulation. Three of them were able to produce bilabials and labiodentals but despite this, they never tried to correct themselves during the assessments. An urgent subject for further research would be to study if speech therapy in combination with exercises for improved strength could improve lip articulation in individuals with DM1 and dysarthria.

Stronger lips were expected to result in a decrease in mouth width when performing a lip pucker [32] but this assumption was not confirmed in the present study. Mouth width in an open mouth smile was *not* expected to increase or decrease since the orbicularis oris muscle is not active in this expression and yet, this was the case in two individuals with improved lip strength. One possible explanation for the decrease in lip mobility could be myotonia in the orbicularis oris muscle secondary to the efforts to maximize the expression. The accuracy of the system for measuring facial distances that was used was investigated by Schimmel et al. (2010). They found it possible to measure geometrical distances with high precision and facial distances with good accuracy and precision [23].

In the questionnaires, no one reported severe difficulties with eating and drinking or with saliva control and the answers could not be related to changes in lip strength. Difficulties related to eating and drinking varied considerably between assessments concerning degree and type of difficulties. Intra-individual variations in eating and drinking ability in children and adolescents with DM1 are, like variations in speech production, thought to be related to general health and alertness.

The treatment programme used in the present study has been tested in clinical practice and found to be realistic to perform on a daily bases. The two youngest participants did not have own motivation to carry out the exercises but all the others were determined to follow the programme. In order to achieve gains in strength there should be an overload of the In musculature [33]. There was no way to completely monitor or verify whether appropriate resistance was applied during exercise. This could have had an impact on the results. Furthermore, it is not known if the chosen treatment programme was optimal. Perhaps, lip force endurance would have improved more if the intermittent pulling of the oral screen had been more than 5 s. Treatment can always be more intense and more frequent. It is of great importance however, that children with DM are not tired out as they need their restricted strength for participation in ordinary daily activities. Oral screens can also just be held in place, for instance during sleep. The reason for passive use could be to elicit nose breathing, to change oral habits such as thumb sucking, or to work as an orthodontic appliance balancing muscle forces affecting the position of the teeth.

A secondary aim of the study was to investigate if stronger lips could have an immediate effect on lip functions such as lip mobility, eating and drinking ability, saliva control, and lip articulation. Due to the fact that only 50% of the participants improved lip strength significantly after treatment, that there were major inter-individual variations concerning lip dysfunction and considerable intra-individual variations between assessments it was not possible to answer the question if stronger lips could improve lip function on the basis of the results from the present study. Therefore, if lip exercises are included in an intervention programme we recommend that they are used together with exercises that enhance the functional use of the achieved strength such as speech therapy and dysphagia treatment. Automation of new behaviours can probably not be expected without repetitive training. When the outcome of treatment is evaluated the intra-individual variations have to be taken into account. Further studies are warranted in order to answer questions related to oral motor treatment and speech therapy in children and adolescents with DM1.

## 5. Conclusions

Maximal lip force and lip force endurance can improve in school aged children and adolescents with DM1. Improved lip strength alone cannot be expected to have an effect on lip articulation, saliva control, or eating and drinking ability in this population. Intra-individual variations concerning speech and eating can be of major importance. A prefabricated oral screen is an easy to use tool suitable for strengthening lip exercises.

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