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Warming up improves speech production in patients with adult onset myotonic dystrophy

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Abstract

This investigation was conducted to study whether warming up decreases myotonia (muscle stiffness) during speech production or causes adverse effects due to fatigue or exhaustion caused by intensive speech activity in patients with adult onset myotonic dystrophy. Thirty patients with adult onset myotonic dystrophy (MD) and ten healthy controls were examined, using a protocol that requires subjects to speak continuously for at least 10 min. In MD patients, warming up led to an increase in speech rate and a decrease in speech variability without causing signs of fatigue or exhaustion as a result of prolonged and intensive use of the speech musculature. No significant changes were found in the controls. After warming up, MD patients achieved a habitual speech rate in reading and reciting similar to that of healthy controls.

Learning outcomes: As a result of this activity the reader will learn that

1. In contrast to most neuromuscular disorders, speech production in patients with adult onset myotonic dystrophy improves by activity.
2. Myotonia in speech musculature in patients with adult onset myotonic dystrophy can be reduced by instructing them to warm up their muscles by repetitive movements.
3. Warming up is a valuable intervention because it improves the velocity and fluency of speech production without aggravating the signs of flaccid dysarthria.

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1. Introduction

Myotonic dystrophy type I (MD), Steinert’s disease, which is caused by a mutation on chromosome 19, is an autosomal dominantly inherited multisystemic disorder that gives rise to internal, cardioligic, and ophthalmologic abnormalities (Harper, van Engelen, Eymard, & Wilcox, 2004). Neuromuscular characteristics are progressive muscle weakness (in speech flaccid dysarthria) and myotonia. Myotonia, i.e. muscle stiffness, is due to a slowed muscle relaxation caused by diminished chloride conductance of the muscle membrane.

MD can be divided into four types based on the onset, clinical features, and number of CTG repeats (cytosine, thymine, guanine) in DNA (Bruggen, 1994; Brunner, 1993; Koch, Grimm, Harley, & Harper, 1991). Congenital MD is the most severe form, with at birth severe hypotonia, insufficient respiration and impaired swallowing. Juvenile onset MD (1–12 years) is characterized by mental retardation, muscle weakness, multisystem pathology, and rapid disease progression (Harper et al., 2004; Hashimoto et al., 1995; Huber et al., 1989). Oral and maxillofacial manifestations, such as a tent-shaped mouth, have also been described (Mercier, Bennani, Ferri, & Piot, 1995; Peñarrocha, Bagan, Vilchez, Millian, & Fernandez, 1990). In contrast, adult onset MD (12–50 years) is mildly progressive with variable multisystem abnormalities but with predominant muscle symptoms and myotonia. Late onset MD (>50 years) is characterized by fewer symptoms, such as cataract, myotonia, and frontal balding.

In this study, we focused on flaccid dysarthria and myotonia in motor speech execution among patients with adult onset MD. Muscle weakness results in flaccid dysarthria (Darley, Aronson, & Brown, 1975; McNeil, 1997; Ramig, Scherer, Titze, & Ringel, 1988; Salomonson, Kawamoto, & Wilson, 1988; Weinberg, Bosma, Shanks, & DeMeyer, 1968), whereas clinical practice shows that myotonia impedes starting articulation, reduces speech rate and evokes disfluencies. There have been few studies of the effect of myotonia on speech production. Clinical practice indicates that patients cope poorly with myotonia in speech production, because they experience its onset as unpredictable and thus unmanageable, than with the more constant influence of muscle weakness. Most patients experience problems caused by myotonia at the moment they start a conversation, some others also experience problems during a conversation. Myotonia appears to affect the time, manner, and place of articulation so that some patients have more problems with bilabial sounds, and others with alveolar or velar sounds. Plosive sounds cause more difficulties than fricative sounds, and talking loudly and shouting seem to provoke myotonia (Maassen et al., 1995).

Our first aim was to determine the occurrence of symptoms of myotonia, and in particular whether symptoms of myotonia disappear after warming up. Studies on muscles in arms and hands have shown that myotonia of the hands is present at the start of activities but diminishes after repeated muscle contractions, an effect known as warming up (Cooper, Stokes, & Edwards, 1988). Our second aim was to determine whether symptoms of flaccid dysarthria increase after long, intensive speech activity, as a result of fatigue or exhaustion caused by warming up. The reason for examining this is that even if warming up is effective, it is not valuable if it leads to fatigue or exhaustion.
2. Methods

Consecutive outpatients with DNA-proven myotonic dystrophy of adult onset (12–50 years) were asked to participate in this study (Brunner, 1993; Brunner et al., 1989). Sixteen men and fourteen women, aged 18–68 years (mean 40.4 years; S.D. = 12.6 years) and with a disease duration of 1–31 years (mean 12.3 years; S.D. = 8.2 years), participated, as did 10 healthy controls (4 men and 6 women) aged 23–58 years (mean 34.4 years; S.D. = 12.6 years).

In order to demonstrate fatigue or exhaustion and warming up effects, we designed a protocol to ensure there was continuous speech production. Because there was no opportunity to rest between the various tasks, the protocol was explained before the examination and only brief instructions were given during the examination. All subjects completed the tasks three times and produced at least 10 min of continuous speech.

2.1. Speech protocol

- Maximum Repetition Rate tasks (3×).
- Reading a text aloud for 3 min (1× at customary speech rate).
- Read same text aloud (2× as quickly as possible).
- Maximum Sound Prolongation tasks (3×).
- Reciting the months of the year (1× at customary speech rate).
- Reciting the months of the year (2× as quickly as possible).
- Maximum Repetition Rate tasks (3×).

The first and last tasks of the protocol were identical, namely, the Maximum Repetition Rate task (MRR task) (Kent, 1994; Kent, Kent, & Rosenbek, 1987; Thoonen, Maassen, Gabreëls, & Schreuder, 1996; Wit, Maassen, Gabreëls, Thoonen, & de Swart, 1994), to enable comparison of speech at the beginning and end of the investigation. Patients were asked to produce monosyllabic sequences [papapa...] [tatata...] and [kakaka...] and tri-syllabic sequences [pataka...] as fast and for as long as possible. In between these tasks, the subjects were asked to read text out aloud for 3 min, three times. The first time subjects were asked to use their customary speech rate, and the second and third times to read the text as fast as possible. This task was followed by the Maximum Sound Prolongation task, in which subjects were asked to phonate the vowel ‘ah’ [a:] and the consonants ‘s’ [s:] and ‘f’ [f:] for as long as possible. This task is not influenced by myotonia, but because duration is an independent variable this task was administered to collect information about the presence of dysarthria, i.e. to differentiate between MD patients and healthy controls. Subjects were then asked to recite the months of the year, the first time at their habitual speech rate, and the second and third times as fast as possible.

All speech tasks were recorded using a Marantz CP 230 cassette recorder, a Sony ECM stereo microphone, and TDK high position SA 60 tape, while patients were sitting on a comfortable chair. The mouth to microphone distance was set at 30 cm for all measurements.

The recordings of the three MRR and MSP tasks were digitized at a sampling frequency of 10 kHz by an AD/DA converter (Kay 4300B). For the MRR task the repetition rate, intensity, and variability in speech rate were measured. The fastest MRR task and the longest MSP task were selected for analysis. Because many subjects produced a higher
intensity at the beginning than at the end of each task, it was not possible to insert an intensity baseline as needed by the software (Kay—multi speech program) to detect the different syllables automatically. Therefore, tags were inserted by hand for each syllable at burst onset. The last syllable in each series was excluded from the analysis. On basis of the tags, syllable duration was measured. All data were analyzed and judged by two speech-language pathologists on base of consensus. This procedure is described by Thoonen et al. (1996) who found an interjudge reliability of .88 or higher.

Duration was the only variable measured for the MSP task. The speech rate for the reading tasks and for reciting the months of the year is expressed in the number of syllables per minute.

All data were analyzed with SPSS version 12.0.1. The difference between the MRR values at the beginning and at the end of the examination was determined by using analysis of variance tests with repeated measures design (MANOVA). Paired-samples t-tests and one-way ANOVA were used to calculate the differences within and between MD patients and controls for the other tasks. If the Levine test of equality of error variances was significant, the non-parametric Mann–Whitney test was used; this was the case for the MSP tasks.

3. Results

In the MSP tasks the duration of phonation was significantly shorter in the MD patients than in the healthy controls (see Table 1). For [aː]: ($z = -2.811; p = 0.005$) and for [fː]: ($z = -4.685; p = 0.000$). When reciting the months of the year at their own speed, the patients and the controls had a similar speech rate ($z = -0.391; p = 0.696$); however, when instructed to recite the names of the months as quickly as possible, the controls were able to increase their speech rate significantly more than the MD patients could ($z = -3.764; p = 0.000$).

3.1. MRR tasks

Both the patients and the controls produced significantly fewer [ka] syllables than [pa] and [ta] syllables regardless of whether the test was performed at the beginning or end of the investigation. The trisyllabic sequences were, as has been found in healthy speakers (Wit, Maassen, Gabreëls, & Thoonen, 1993), produced significantly faster than the monosyllabic sequences (see Table 2).

The MD patients produced significantly fewer syllables in monosyllabic sequences at the beginning of the investigation than at the end of the investigation ($F_{(1,29)} = 11.995$;

<table>
<thead>
<tr>
<th>Measurement</th>
<th>MD patients</th>
<th>Healthy controls</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean</td>
<td>S.D.</td>
</tr>
<tr>
<td>MSP—[aː] in seconds</td>
<td>14.56</td>
<td>8.15</td>
</tr>
<tr>
<td>MSP—[fː] in seconds</td>
<td>7.21</td>
<td>6.16</td>
</tr>
</tbody>
</table>
In MD patients the maximum repetition rate increased slightly at the end of the examination for [pa] and significantly for [ta], [ka] and [pataka]. Thus at the end of the investigation, only the maximum repetition rate of [pa] was still significantly lower in the MD patients than in the controls ($F_{(1,38)} = 12.682; p = 0.001$) (see Figs. 1 and 2).

For the trisyllabic sequences [pataka] no significant differences between the MD patients and the controls were found at the beginning of the investigation ($F_{(1,38)} = 2.574; p = 0.117$). Furthermore, both groups could significantly increase the maximum repetition rate for these sequences (MD: $F_{(1,29)} = 8.724, p = 0.006$; controls: $F_{(1,9)} = 10.402, p = 0.010$), and so there were no differences in trisyllabic sequence production at the end of the investigation between the patients and the controls ($F_{(1,38)} = 2.442; p = 0.126$) (see Fig. 1).

### 3.2. Variability in repetition rate of MRR tasks

The variability in the rate of repetition of monosyllabic sequences was greater in the first test than in the last test in the patients ($F_{(1,29)} = 66.944; p = 0.000$), which was not the case in the controls ($F_{(1,9)} = 0.181; p = 0.681$) (see Table 2; Figs. 3 and 4). In 29 patients the variability decreased at the end of the examination and in only 1 patient the variability did not show any changes.

#### Table 2

Results of MD patients and healthy controls on MRR tasks at the beginning and at the end of the examination

<table>
<thead>
<tr>
<th>Measurement</th>
<th>MD patients Beginning</th>
<th>MD patients End</th>
<th>Healthy controls Beginning</th>
<th>Healthy controls End</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean  S.D.</td>
<td>Mean  S.D.</td>
<td>Mean  S.D.</td>
<td>Mean  S.D.</td>
</tr>
<tr>
<td>MRR—[pa] in syllables/seconds</td>
<td>6.28 1.11</td>
<td>6.35 1.04</td>
<td>7.41 0.66</td>
<td>7.68 0.92</td>
</tr>
<tr>
<td>MRR—[ta] in syllables/seconds</td>
<td>6.34 1.31</td>
<td>6.59 1.15</td>
<td>7.46 1.07</td>
<td>7.16 0.94</td>
</tr>
<tr>
<td>MRR—[ka] in syllables/seconds</td>
<td>5.41 1.29</td>
<td>5.98 0.94</td>
<td>6.30 1.00</td>
<td>6.36 0.86</td>
</tr>
<tr>
<td>MRR—[pataka] in syllables/seconds</td>
<td>7.03 1.41</td>
<td>7.68 1.28</td>
<td>7.80 0.87</td>
<td>8.40 1.25</td>
</tr>
<tr>
<td>Variability MRR</td>
<td>0.19 0.04</td>
<td>0.15 0.02</td>
<td>0.10 0.03</td>
<td>0.11 0.03</td>
</tr>
</tbody>
</table>

$p = 0.002$), whereas this difference was not seen in the controls ($F_{(1,9)} = 0.007; p = 0.937$).

In MD patients the maximum repetition rate increased slightly at the end of the examination for [pa] and significantly for [ta], [ka] and [pataka]. Thus at the end of the investigation, only the maximum repetition rate of [pa] was still significantly lower in the MD patients than in the controls ($F_{(1,38)} = 12.682; p = 0.001$) (see Figs. 1 and 2).

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Fig. 2. Increase in the repetition rate at the end of the investigation as an effect of warming up in MD patients; healthy controls did not show any changes. (♦), MD patients and (▲), controls.

Fig. 3. Scatter plot showing individual variability in performance of the monosyllabic MRR task by MD patients and healthy controls at the beginning and end of the investigation. Variability decreased after warming up in MD patients but not in healthy controls. (●), MD patients and (▲), controls.
3.3. Reading task

When instructed to read at their normal speed, the patients and the controls produced the same number of syllables per minute \( (F_{(1,34)} = 0.753; p = 0.392) \). When instructed to read as quickly as possible, the controls were able to increase their speech rate more than the MD patients, resulting in a significantly faster speech rate in the controls \( (F_{(1,34)} = 4.180; p = 0.049) \) (see Table 3). The mean speech rate of MD patients was the same for the first and last 100 syllables of text when they read at maximum speed.

3.4. Reciting task

When instructed to recite the months of the year at their natural speed, the patients and the controls did so at the same speed \( (F_{(1,38)} = 0.453; p = 0.505) \). However, when asked to perform the task as quickly as possible, the controls were able to increase their speech rate

<table>
<thead>
<tr>
<th>Measurement</th>
<th>MD patients</th>
<th>Healthy controls</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reciting (normal) in number of words/minute</td>
<td>293</td>
<td>309</td>
</tr>
<tr>
<td>Reciting (fast) in number of words/minute</td>
<td>392</td>
<td>514</td>
</tr>
<tr>
<td>Reading (normal) in number of words/minute</td>
<td>242</td>
<td>255</td>
</tr>
<tr>
<td>Reading (fast) in number of words/minute</td>
<td>344</td>
<td>398</td>
</tr>
</tbody>
</table>

Fig. 4. Variability in maximum repetition rate at the beginning and end of the investigation. At the end of the investigation, variability had decreased in MD patients but not in healthy controls. (●), MD patients and (▲), controls.
more than the MD patients, resulting in a significantly faster speech rate in controls ($F_{(1,38)} = 22.927$; $p = 0.000$) (see Table 3).

3.5. Intensity of MRR task

The mean intensity (in dB) of speech in the MRR task was not significantly different at the end of the protocol, then at the beginning of the experiment in either the subjects with myotonia or the control subjects. The greatest changes in intensity were seen in the controls (+1.25 dB for [pataka] and −1.25 dB for [ka]).

4. Discussion

We found that the influence of myotonia is significantly greater at the onset of a speaking episode but decreases during the production of continuous speech as a result of warming up. Warming up in general increased the number of syllables per second that could be produced and decreased the variability in speech rate, meaning that patients achieve both a higher speech rate and a more regular performance. After warming up, the speech rate for [pa] sequences did not increase significantly whereas for [ka] sequences the greatest increase of speech rate was found. A possible explanation is that for [pa] sequences the activity and muscle tension in lip and jaw muscles is too small to provoke great disturbances caused by myotonia, whereas the production of the [ka] sequences requires more muscle activity and especially tension, resulting in a greater influence of the myotonia.

The increase of speech rate was not the same for all patients and all sequences of the MRR tasks. In one patient no increase of speech rate was seen after warming up, in four patients only in one MRR task and in two patients in two MRR tasks. From six of these seven patients still the variability of speech production decreased, i.e. fluency improved. In the other 23 patients both speech rate improved in three or all MRR tasks and variability decreased. Despite the restricted results on the MRR tasks in some patients, the mean effect of warming up on speech rate was significant. This is consistent with our clinical experience that most patients with myotonia experience problems when starting to speak and have less difficulty once they are speaking.

For the minority of patients who experience unpredictable variation in occurrence of problems in everyday speech, we have two possible explanations. First, it is possible that patients’ speech production in some daily conversations is, due to turn taking, too short or not intensive enough to cause the warming up effect, and secondly, the moments of rest may be too long to allow the effect of warming up to be maintained.

After warming up, the speed at which the MD patients read aloud and recited words was not significantly slower than that of the controls. The MD patients also read the first 100 and last 100 syllables of a text at the same speed, which means that the effect of warming up was optimal at the beginning and lasted for at least 3 min, and that patients did not show signs of fatigue or exhaustion. Moreover, the intensity with which they spoke, which is an indicator of fatigue, did not change after they read the text aloud for at least 10 min. This indicates that the continuous activity needed to create the warming up effect does not lead to exhaustion or fatigue.
Even though the performance of the MD patients improved in terms of an increase in speech rate, decreased variability, and absence of fatigue and exhaustion after warming up, they still showed signs of flaccid dysarthria, as evidenced by their poor maximum performance on the sound prolongation task, by the higher variability of the repetition rate on the MRR tasks, and by the reduced maximum speech rate in reading and reciting tasks.

5. Conclusion

The speech of patients with adult onset MD shows signs of both flaccid dysarthria and myotonia. Weakness seems to be a less disturbing feature than myotonia, because after warming up the natural speed at which patients recited and read aloud was similar to that of healthy controls. Controls perform significantly better than patients only when participants are asked to function at their maximum.

Our results suggest that patients with adult onset MD can best be helped by speech-language pathologists if they focus on myotonia first. Just as with other motor skills, warming up affects speech production, leading to an increase in speech rate and a decrease in myotonia and variability without there being evidence of adverse effects, i.e. signs of exhaustion or fatigue as a result of long and intensive use of the speech musculature. Thus, warming up is valuable for patients with adult onset MD because it reduces or eliminates the influence of myotonia on speech muscles and results in a habitual speech production that is comparable with that of healthy controls.

Acknowledgements

We thank L. Kromhout and J. van de Kerkhof of the Department of Speech and Language of the Radboud University Nijmegen for their help with data analysis.

Appendix A. Continuing education

Study Questions and Answers

1. Disfluency in speech production in MD patients is seen mostly
   A. After intensive speech tasks.
   B. After rest.
   C. After long conversations.
   D. After taking a deep breath.

2. Myotonic dystrophy is more severe and progressive when
   A. The symptoms occur after the 12th year.
   B. MD can be proven by DNA diagnostics.
   C. The facial muscles are involved.
   D. Patients show characteristics of MD directly after birth.
3. Patients with adult onset MD do not have
   A. Dysarthria.
   B. Dysphagia.
   C. Muscle weakness.
   D. Cognitive disorders.
4. Myotonia can be reduced best by
   A. Massage.
   B. Warmth.
   C. Repetitive movements.
   D. Rest.
5. After warming up MD patients achieve a similar speech rate as controls
   A. In all MRR-tasks.
   B. In habitual reading.
   C. In fast reciting.
   D. A + B + C.

Key: 1B; 2D; 3D; 4C; 5B

References


