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We propose that the intial symptom of the very stereotyped simple partial seizure semiology of this patient originated from activation of the primary motor area. There is only one other case report in the literature mentioning an ear click occurring during epilepsia partialis continua [3]. Although we were not able to prove that the patient's ear clicks belong to the category of the objective clicking sounds [1], it is highly likely that this symptom is produced by an epileptic activation of the primary motor region rather than of Heschl's area in the superior temporal gyrus. In favour of this assumption is the location of the lesion as well as the sequence of the focal motor symptoms following the ear clicks.

Arroyo et al. [5] reported a patient with a perirolandic tumour in whom electrical cortical stimulation produced clicking sensations. The exact location of these electrodes could not be reconstructed retrospectively, i.e. the subdural electrodes could have been placed over the lower frontocentral or the superior temporal gyrus. It is the experience of others (Arroyo S, personal communication) as well as our own that electrical stimulation of the auditory area doesnot produce "clicking" but instead sounds, echoes, voices or more elaborated auditory hallucinations. The absence of ictal EEG changes indicates that the cortical area probably involved during the seizure is hidden in deeper parts of the brain or too small to produce visible changes in the surface EEG. This is not an unusual situation in simple partial seizures, in which the EEG is normal in about 50%–80% [6, 7]. The preservation of consciousness is also in accordance with the assumption of a rather small brain area being involved throughout the seizure.

In conclusion, the initial ictal symptom of rhythmic ear clicking can be explained by epileptic activation of the part of the motor cortex in which the palatal muscles that are capable of producing this sensation are represented. Ear clicks can be a manifestation of focal motor epileptic seizures.

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Quantitative assessment of speech in myotonic dystrophy

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Sirs: Myotonic dystrophy (MyD) is an autosomal dominant inherited multisystem disease which is characterized by progressive muscular weakness, atrophy and myotonia. Facial-bulbar muscle weakness is characteristic for adult onset MyD, which can result in a peripheral disorder of speech execution, in particular flaccid dysarthria [1]. The available clinical descriptions of speech in MyD patients [2, 3] are qualitative. Although the genetic basis for MyD has been found [4, 5], there remains the need for *quantitative* assessment of expression in order to detect early signs of the disease and to follow the progression over the years. In the present study we selected mildly affected, early-adult and adult onset MyD patients who had no intellectual impairment and no known neuropsychological dysfunction. We assessed quantitatively the extent to which their speech-motor function was affected, by administering a set of speech- and oral-motor tasks and measuring the performances acoustically.

Fifteen patients with MyD were asked to participate. The diagnosis of MyD was made by DNA linkage analysis in their families [5] and positive (myotonic) EMG and/or slit-lamp examination [2]. They were classified as adult or early-adult onset with a mild or moderate severity of neuromuscular disability (Muscular Disability Rating Scale score of 3 or less [6]) and normal hearing. They

were confirmed by neuropsychological assessment to have no intellectual impairment (mean IQ 107.6). The group consisted of eight female and seven male subjects with a mean age of 36.2 years (SD 8.6). The duration of the disease ranged from 0;3 years (0 years, 3 months) to 30;0 years with a mean of 11;0 years (SD 8;4). For each MyD subject, a control subject was selected, matched with respect to age, sex and educational level and with a history free of speech- or hearing-related problems.

In a brief interview at the beginning of the speech-motor assessment session, eight of the MyD patients reported speech initiation problems, which disappeared after "warming up". In three patients reduced intelligibility and increased speech exertion were reported as the first signs of fatigue. No effects of their psychological state, other than fatigue, were reported. The spontaneous speech of ten patients was judged as unremarkable by the speech pathologist. Five patients showed slight signs of imprecise articulation. Overall, the patients were perfectly intelligible and the speech signs were judged to be mild.

Patients and control subjects were administered a number of speech tasks. In the Maximum Sound Prolongation (MSP) task, the subjects were requested to sustain the speech sounds /a/, /z/, /s/, and /f/ for as long as possible. After instruction they were given three trials and the best performance was used for analysis. Performance on this task is of diagnostic significance for laryngeal problems [7] and for dysarthria [8,9]. The MSP score was calculated by averaging the durations of the longest /a/, /z/, /s/, and /f/ productions.

Table 1 Means and standard errors *(italics)* of maximum prolongations in seconds of /a/, /z/, /f/, and /s/ produced by the 15 MyD and 15 control subjects

	MyD		Controls	
/a/	21.3	*2.54*	26.5	*2.78*
/z/	19.7**	*2.69*	28.8	*3.54*
/s/	20.8**	*1.97*	32.4	*4.59*
/f/	14.5**	*2.73*	22.6	*5.44*
Overall	19.1**	*2.54*	27.6	*4.20*

* $P < 0.05$, ** $P < 0.01$ relative to controls

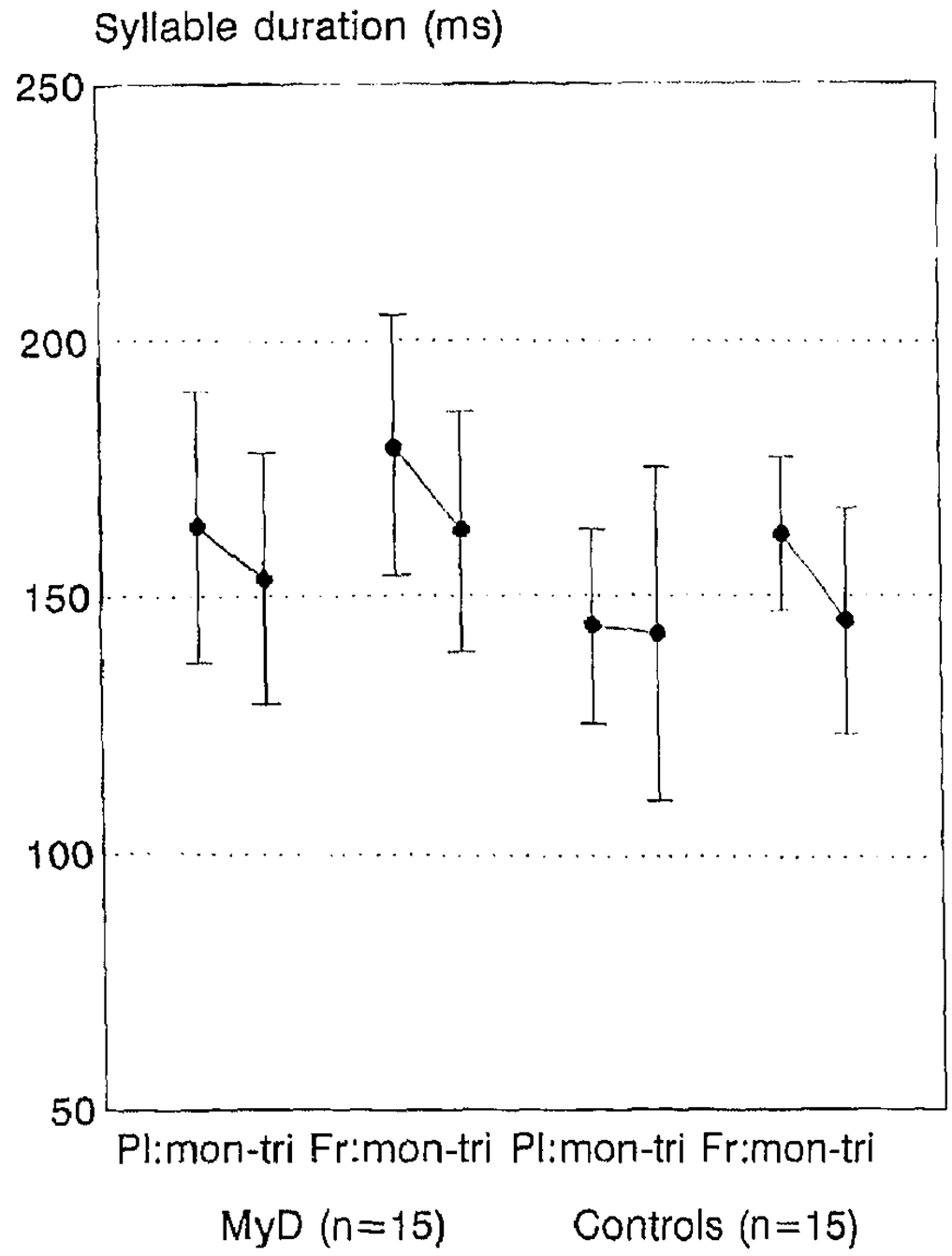

Fig. 1 Mean syllable durations in milliseconds (connected by drawn lines) and standard deviations of the mono- and trisyllabic plosive *(Pl)* and fricative *(Fr)* sequences for the MyD and control groups

The MyD group performed less well than the control group on the MSP task, as can be seen in Table 1. According to an analysis of variance this difference was significant ($F(1,28) = 4.20$, $P < 0.05$). Each group showed fairly equal durations for the sounds /a/, /z/, and /s/, but shorter durations for /f/ ($F(3,84) = 5.33$, $P < 0.01$). The interaction between group and sound was not significant ($F < 1.0$).

In the Maximum Repetition Rate (MRR) task, the subjects were asked to repeat as fast as possible the monosyllabic plosive sequences "papa..", "tata..", and "kaka.." and fricative sequences "fafa.. "," sasa..", and "xaxa..". Also, subjects repeated as fast as possible the multisyllabic sequences "pataka.." and "fasaxa..". The subjects were given several trials, and the best perfomance was selected for analysis. Poor performance on this task points to reduced motor speed or deviating stiffness [1, 8–11]. The MRR was measured in a semi-automatic way using the digitized signal [12].

The MyD group performed significantly ($F(1,28) = 5.29$, $P < 0.05$) less well than the control group on the MRR task for monosyllabic sequences, as can be seen in Fig. 1. The pattern of relative syllable durations (plosive sequences faster than fricative sequences) was identical for both groups. In order to disentangle the contribution of the duration of the consonant and the vowel, for each syllable a consonant ratio was calculated: the duration of the consonant (plosive or fricative) divided by the total duration of the syllable. The MyD subjects produced longer consonants (larger ratios) in the plosive sequences than in the fricative sequences (ratios 0.48 and 0.46, re-

spectively), in contrast to the control subjects (ratios 0.42 and 0.44); this interaction was significant ($F(1,27) = 4.23$, $P < 0.05$). The latter effect was strongest in the sequences with a labial place of articulation ("papa..", "fafa.."; $P < 0.05$).

The MyD subjects produced the multisyllabic sequences like the monosyllabic sequences, significantly more slowly than the control subjects ($F(1,28) = 5.58$, $P < 0.05$; see Fig. 1). More importantly, both the MyD and the control subjects produced the multisyllabic sequences slightly but significantly faster than the monosyllabic ones ($F(1,28) = 6.14$, $P < 0.05$). Thus, the relative durations of mono- and multisyllabic sequences was similar across groups.

To summarize: MyD patients showed overall poorer performance on the MSP and MRR tasks than the control subjects. The profile of scores was characteristic for flaccid dysarthria: shorter MSP, and slower MRR of both monosyllabic and multisyllabic sequences. This result shows that MSP and MRR can be used as quantitative measures of the integrity of speech-motor functions of MyD subjects.

Two further specifications of the dysarthria are in order. First, similar profiles were found for the MyD and control subjects, which suggests that the muscle weakness observed in the MyD subjects is not restricted to a particular articulatory organ. Second, the consonant ratios for the labial MRR sequences were the shortest for the control subjects and among the longest for the MyD subjects. This reflects problems with rapid alternating movement of the lips (and jaw). Furthermore, in contrast to the control subjects, the MyD subjects produced systematically higher consonant ratios in the plosive sequences than in the fricative sequences. In plosive sequences a complete occlusion of the vocal tract is made, which requires more muscle activity than in fricative sequences, where incomplete occlusion suffices [13]. The relatively high consonant ratios indicate overshoot rather than undershoot in the production of plosives by the MyD subjects. Taken together, the results suggest the effects of myotonia: the inability to relax in particular the lips, where the increase of the consonant ratio is most prominently present.

With the quantitative assessment procedure used in this study, one of the signs of MyD can be detected at an early stage. Moreover, an objective instrument is available to evaluate the progression of the disease, as well as the results of therapy that might be developed in the near future.

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