Abnormalities of ocular motility in myotonic dystrophy

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Abbreviations: OKN = optokinetic nystagmus; SP = smooth pursuit; VOR-S = vestibulo-ocular reflex suppression

Anastasopoulos et al. (1996) reported a parallel degradation of smooth pursuit (SP) and vestibulo-ocular reflex suppression (VOR-S) indicating a central nervous system dysfunction in patients with myotonic dystrophy, who all had visual acuity better than 0.6. However, in our study on 13 myotonic dystrophy patients (Verhagen et al., 1992), we found SP impairment only in the two cases with the lowest visual acuity (0.3 and 0.5), but optokinetic nystagmus (OKN) responses were normal in all cases. We were therefore inclined to attribute the poor SP response in these latter cases to their poor visual acuity. Because OKN responses are less dependent on foveal vision than SP, we wrote that in view of the fact that most myotonic dystrophy patients have reduced vision due to cataract and retinal abnormalities (Miller, 1985), we would not be inclined to accept a deficit in visual following responses, unless it showed up in both the SP and the OKN responses (Verhagen et al., 1992).

Unfortunately OKN was not examined by Anastasopoulos et al. (1996). In fact, OKN responses have never been included in reports on myotonic dystrophy, as far as we know, except in ours (Verhagen et al., 1992) and in a case report by Emre and Henn (1985). In our opinion, the VOR-S findings add nothing substantial to the SP findings in the study by Anastasopoulos et al. (1996), as the visual target (a laser spot) was identical in both cases, as was, apparently, the visual background (Ganzfeld-like structure of the cylindrical screen or darkness?) which, if not identical, must have been at least so designed that background structures could not compete with SP of the moving target by eliciting OKN responses in the opposite direction. In other words, the method was such that the emphasis was very much, in a fairly similar way, on foveal vision in both cases. Therefore an additional finding of OKN impairment might have made it more easy to accept central dysfunction as the underlying cause. Studying VOR-S using the whole (structured) visual surround instead of a laser spot might be a suitable alternative.

Anastasopoulos et al. (1996) suggested that the accentuation in SP/VOR-S deficits at higher frequencies might be related to a similar frequency-dependent performance of parieto-occipital visual association areas. Frequency-dependent impairment of SP/VOR-S on the basis of suboptimal visual acuity (foveal function) might be a more trivial suggestion. We could not find any report on the dependence of SP gain on visual acuity in a 1966–96 Medline search, other than one example of impairment of accommodation degrading SP (Penetar et al., 1988), but it is common knowledge that SP deteriorates in persons with refraction errors when they are not using their glasses and, presumably, so does VOR-S. It could well be that such deterioration is more marked at the higher frequencies of stimulation.

References


We thank Verhagen and Huygen for their comments. These authors suggest that the deficit for SP and VOR-S, which we observed in our myotonic dystrophy patients (Anastasopoulos et al., 1996), could possibly be due to low visual acuity. However, visual acuity in all of our patients was good (>0.6) and clearly better than in the two patients in their study (Verhagen et al., 1993), who were the only ones showing a clear deficit in SP and visual acuity (0.3 and 0.5).

It should be mentioned, however, that the two patients in their study had, in addition to the SP and visual acuity deficit, convergence paralysis and divergent strabismus. While slowing of SP with low visual acuity appears not to be documented in the vast SP literature (at least we did not find an indication for it, either), it is known that abnormalities of SP are found in a considerable proportion of patients with strabismus. Therefore, we may have to look for reasons other than visual acuity, which could explain the discrepancy between the SP deficit in our patients and the negative findings in the vast majority of their patients.

Important differences concern the methods used to elicit SP and to analyse the responses. The authors applied only one stimulus, consisting of a spot moving in a circle of 10° radius at a horizontal/vertical peak velocity of 20°/s. Thus, the horizontal and vertical components of their stimulus had a frequency of ~0.32 Hz. We mentioned that the SP deficit in our patients was most pronounced at higher frequencies (i.e. at 0.4 and 0.8 Hz). Furthermore, the authors used conventional EOG, the spatial resolution of which is clearly less than that of the infrared device which we used. This point might be of relevance for detecting small catch-up saccades which compensate for SP slowing. Unfortunately, we do not learn from their article whether they separated saccades at all from the responses. Saccade separation in their study might have been more difficult with the strong low pass filtering (LP 30 Hz) and an apparently rather low (but not precisely specified) sampling rate. Such methodological differences might be the reason why our data are more comparable with the work of Bollen et al. (1992) who did observe an SP slowing, using methods for data acquisition and analysis similar to ours.

The authors appear to suggest that a deficit in visual-following responses in patients can only be proved if it shows up in both SP and OKN responses. We have not tested OKN in our patients, so that we cannot make a statement about their OKN performance. However, we would disagree with the authors' suggestion. It is well known that both OKN and SP are often impaired in parallel in patients with parietal lesions, for instance, but there are cases in which OKN is relatively spared compared with SP (Baloh et al., 1980; Chambers and Gresty, 1982; Heide et al., 1990). Furthermore, we find it difficult to accept the authors' finding of a normal OKN in their two patients with defective SP when considering the broad variation of their normative data (‘the lower 5% confidence limit of response velocity was 20°/s, with the stimulus velocity being 40 and 60°/s’).

Coming back to the question as to what extent SP may be affected by low visual acuity, it is our experience from clinical nystagmography that a moderate or medium impairment has little effect on SP. This experience is based, for example, on our observations in patients suffering from low vision of one eye due to optic neuritis; when comparing SP in the normal versus the impaired eye, we noticed hardly any difference. This observation is in line with the generally accepted notion that the internal (open loop) gain of the SP system is very high (Gresty and Hamalgyi, 1979; Leigh et al., 1982) and that central visual motion processing depends mainly on the so-called magnocellular visual route, which favors high temporal and low spatial properties of visual stimuli (see Merigan and Maunsell, 1993).

Finally, we address the authors' question about the background conditions in our SP and VOR-S tests. They were identical (darkness), as was the visual stimulus (light spot). We disagree with the authors' claim that our VOR-S test added nothing substantial to the SP findings; we hold that the parallel degradation of SP and VOR-S performance, which we observed in our myotonic dystrophy patients, clearly indicates a central rather than a muscular origin of the SP deficit.

References
The course of cortico-hypoglossal projections in the human brainstem: functional testing using transcranial magnetic stimulation

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Abbreviations: CMAP = compound muscle action potential; PES = peripheral electrical stimulation; TMS = transcranial magnetic stimulation

Urban et al. (1996) recently assessed the course of the cortico-hypoglossal projections in the human brainstem using transcranial magnetic stimulation (TMS) of the motor cortex. To this interesting study we want to add some important findings we have made with a similar technique, especially concerning the uncrossed central motor pathways.

A special bipolar surface electrode was used to record the compound muscle action potentials (CMAPs) from the lingual muscles after TMS of the motor cortex and peripheral electrical stimulation (PES) of the hypoglossal nerve medial to the angle of the jaw. With this technique, which was described previously in detail (Muellbacher et al., 1991, 1994; Urban et al., 1994), the assumed bihemispheric motor representation of the lingual muscles was confirmed. In their study, Urban et al. (1996) used a similar recording technique, but PES was only performed in subjects with a medullary lesion (Urban et al., 1996). However, for a reliable estimation of the pattern of crossed and uncrossed central innervation, not only TMS, but also PES with simultaneous recording from either side of the tongue should be performed in every subject.

TMS of one hemisphere produces CMAPs in the contralateral and in the ipsilateral lingual muscles. The ipsilateral CMAPs usually represent responses propagated via the uncrossed corticobulbar pathways, but they may partially be derived from a peripheral ‘contamination’. Interestingly, some individuals do show CMAPs in the contralateral half of the tongue after PES (Fig. 1), despite a careful positioning of the mouthpiece in the midline (Muellbacher et al., 1994; montage B). In our last series of 40 control subjects (80 sides), 23 presented contralateral CMAPs with a sharp initial negative deflection from the baseline with amplitudes up to 64% of the ipsilateral responses. The exact origin of these contralateral responses after PES is unclear, but a similar peripheral ‘cross-over’ of the CMAPs from one side to the other also seems possible after cortical stimulation. The ipsilaterial responses after TMS may therefore be derived from the contralateral lingual side, thus not exactly representing CMAPs propagated via the uncrossed cortico-hypoglossal projections.

In their study, Urban et al. (1996) analysed the ipsi- and contralateral responses after cortical stimulation in patients with different brainstem lesions. As a peripheral cross-over

![Fig. 1](image-url)  
**Fig. 1** Contralateral (upper traces) and ipsilateral (lower traces) compound muscle action potentials evoked by peripheral electrical stimulation of the hypoglossal nerve medial to the angle of the jaw in a control subject (two responses superimposed).