Eye movements in myotonic dystrophy

An electrooculographic study

Gunter K. von Noorden, H. Stanley Thompson, and Maurice W. Van Allen

Fixation and pursuit movements, movements of regard, of convergence and divergence, and ocular reaction time were recorded electrooculographically in 10 patients with myotonic dystrophy. With the exception of pursuit movements, normal responses were present in 9 out of 10 patients. All of the patients demonstrated a selective and pronounced impairment of pursuit movements which were elicited by a horizontally and sinusoidally moving target. Pursuit movements which are normally smooth were entirely replaced by irregular saccades and gross movements of regard, in both directions of gaze, and irrespective of the target velocity. This selective disturbance of smooth eye movements cannot be explained on the basis of dystrophic changes, or of a myotonic reaction of the extraocular muscles, but is interpreted as evidence for an involvement of the central nervous system in myotonic dystrophy. This view is supported by the associated endocrine and mental changes which occur commonly in this disease, and by previously reported selective disturbances of pursuit movements in association with certain brain lesions.

Myotonic dystrophy is a progressive hereditary disease which is characterized by atrophy and weakness of muscles of the face, neck, and extremities. A striking clinical feature is the myotonic reaction, i.e., an abnormally delayed relaxation after muscular contraction. Other symptoms indicating a generalized disorder include frontal baldness, testicular and ovarian atrophy, and other endocrine disturbances.1,2 The most commonly described eye findings are cataracts. A review of the literature summarized by Davidson3 showed that the extraocular muscles are mentioned only rarely in connection with the disease. Lagophthalmos due to weakness of the orbicularis muscle and ptosis resulting from levator paresis have been described. Other reports mention squint and diplopia, especially when attempting to move the eyes suddenly, and limitation of the eye movements in extreme positions of gaze.2

In view of the scantiness of available information on the participation of the extraocular muscles in myotonic dystrophy, a study of the eye movements in patients with this disease seemed indicated. Electrooculography provides a convenient method to register different types of eye movement under various stimulus conditions. In this study the movements of the eyes during...
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fixation on a stationary target, during convergence and divergence, in response to a peripherally appearing visual stimulus (movements of regard), and in response to a sinusoidally moving target (pursuit movements) were registered electrooculographically in 10 patients with myotonic dystrophy.

Material and methods

Ten patients with myotonic dystrophy underwent a comprehensive neurological and ophthalmologic examination. The information obtained from this survey which included pupillographic studies in these and in additional patients with myotonic dystrophy will be presented in a forthcoming publication. This study deals exclusively with the findings pertaining to the ocular motility.

Ages, best corrected visual acuity, and the results of a complete ophthalmologic evaluation are summarized in Table I. Although some limitation of ductions and versions in the extreme lateral position of gaze were noted in four instances, no frank pareses of any of the extraocular muscles were present. All patients could move their eyes without effort at least 25 degrees to the right or left.

If visual acuity could be improved with glasses, the correction was worn during all experimental procedures. Reduced visual acuity was usually the result of incipient cataracts; in one instance (Case 7) the patient was practically monocular due to an old injury of one eye.

At the beginning of each experiment, the subject was seated comfortably 2 m. in front of a projection screen. Head movements were eliminated by a combined head and chin rest. Silver cup electrodes were fastened to the skin close to the lateral canthus and on the glabella, equidistant from both medial canthi. Except during convergence and divergence, the eye movements were recorded simultaneously (bitemporal leads) on one channel of a Schwarzer electrooculograph. Details regarding the electrooculographic recordings of horizontal eye movements have been described in earlier publications. The following types of eye movements were recorded.

**Fixation movements.** Eye movements were registered during fixation on stationary projected spots of red light in the center of the screen, and 15 degrees to the right and to the left of the patient. The size of the fixation target could be varied between 10 and 30 mm. according to the visual acuity of the patient, to assure good visibil-

<table>
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<th>Sex</th>
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<th>Versions</th>
<th>Convergence insufficiency</th>
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</tr>
<tr>
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<td>Limited in all directions</td>
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</table>
Movements of regard. A fixation light was projected at 15 degrees to the left of the center of the screen and remained during the entire experiment. A second light could be flashed on and off at 15 degrees to the right of the center. The size of the lights could be varied according to the visual acuity of the patient. The subject was instructed to fixate the light at his left, and to move his eyes only to fixate the target at his right when the latter was flashed on. The movements were elicited at random intervals to eliminate an anticipative reaction. Stimulus alternation was recorded on the second channel of the electrooculograph (Fig. 1).

Ocular reaction time. The ocular reaction time is defined as the time elapsing between presentation of a peripherally appearing visual stimulus and the resulting eye movement, which is performed to center the object of interest on the macula. The reaction time was obtained by measuring the time interval between the presentation of the stimulus recorded on the electrooculograph and the resulting movement of regard (Fig. 1). Data were obtained from 9 patients.

Convergence and divergence. A fixation light was projected into the center of the screen. A flashlight bulb mounted on an adjustable stand was placed in the median plane 15 cm. in front of the patient and in the same horizontal plane as the distant fixation light. The subject was instructed to converge on the near light as soon as the distant light would disappear, and to resume fixation of the latter when it became visible again. The movements of the right and left eye during convergence and relative divergence were recorded independently on the first (temporonasal lead) and second (nasotemporal lead) channels of the electrooculograph. The stimulus for convergence (disappearance of the distant light) and relative divergence (reappearance of the distant light) were registered on the third channel (Fig. 2). Thus it was not only possible to record the eye movements during convergence and relative divergence, but also to measure the elapsed time between stimulus presentation and the beginning of the oculomotor response. Since 6 patients did not summon sufficient effort for repeated voluntary convergence, this response could be measured only in the remaining 4 patients. Even these patients tired easily and, therefore, not more than 15 convergence and divergence eye movements were elicited.

Fig. 1. Movements of regard between 15 degrees to the left and 15 degrees to the right in a normal subject. Time signal (above), 1 second. Upper recording, EOG (bitemporal lead); lower recording, stimulus registration; RT, reaction time.

Fig. 2. Convergence and divergence movements in a normal subject. O.D., temporonasal lead; O.S., nasotemporal lead; lower recording stimulus registration. For details see text.

Fig. 3. Fixation movements (Case 3). Steady fixation of targets located 15 degrees to the right, in the center, and 15 degrees to the left was present in all patients with myotonic dystrophy.
Pursuit movements. Pursuit movements are smooth eye movements in response to a moving fixation target. The patient was instructed to follow with his eyes a spot of light which moved horizontally and sinusoidally back and forth at eye level. The stimulus was produced by a projector attached to a motor. The velocity of the movement was variable between 0.28 and 0.9 c.p.s. The excursions measured 15 degrees to each side. Technical details of the projection arrangement have been described in an earlier study. The movements of both eyes were recorded on the first channel of the electrooculograph. The actual motion of the target could be registered on the second channel. At the beginning of the experiment the projector was set in motion with the lowest speed obtainable (0.28 c.p.s.). The eye movements were recorded and the target speed was slowly increased until complete disintegration of the sinusoid pursuit movement was noted.

Results

Fixation movements. Fixation of targets located in front and 15 degrees to each side of the patient was steady in all instances. The recordings did not differ from those obtained from normal subjects. A recording demonstrating steady fixation (Case 3) is depicted in Fig. 3.

Movements of regard. Anomalies of the eye movements in response to a peripheral visual stimulus, were not present, except in one patient (Case 1). Movements of regard were performed swiftly and accurately, fixation was well maintained during the interval until execution of the next movement (Fig. 4).

Case 1, however, demonstrates anomalies, inasmuch as each movement was performed in 2 to 3 steps (saccades), and a quick and accurate one-step movement was only rarely performed. The saccadic decomposition was present when looking in either direction (Fig. 5).

Ocular reaction time. In order to create a basis for comparison, the ocular reaction time was determined in 3 normal subjects. Arithmetic means were calculated from 35 measurements in each subject, and the results are summarized and compared with

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*The reaction times quoted in the text and in Tables II and III represent relative rather than absolute values. Since submission of this paper, it was established that technical factors caused a constant delay of 0.15 seconds between the electrooculographic registration of the stimulus and its actual presentation to the subject. Thus, to arrive at absolute reaction times, 0.15 seconds will have to be subtracted from all data quoted in this study. The constancy of this delay was equal for all subjects and, therefore, the interpretation of our results remains unchanged.

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Fig. 4. Normal movements of regard in myotonic dystrophy. a, Case 2; b, Case 5; c, Case 7. For pursuit movements in the same patients, see Figs. 7 and 8.

Fig. 5. Movements of regard (Case 1). Abnormally prolonged reaction (RT) and stepwise performance of a normally one-step and swift response. Compare with Fig. 1.
Table II. Arithmetic means of ocular reaction time in normal subjects and patients with myotonic dystrophy

<table>
<thead>
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<th>No.</th>
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<th>Patients with myotonic dystrophy Reaction time (s)</th>
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<td>360</td>
</tr>
<tr>
<td>10</td>
<td>380</td>
<td></td>
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</table>

Table III. Arithmetic means of reaction time for convergence and divergence in normal subjects and patients with myotonic dystrophy

<table>
<thead>
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<th>No.</th>
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<th>Patients with myotonic dystrophy</th>
</tr>
</thead>
<tbody>
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<td></td>
<td>Reaction time</td>
<td>Reaction time</td>
</tr>
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<td>1 830 1,170</td>
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<td>2 330 430</td>
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<tr>
<td>4</td>
<td>320 330</td>
<td>8 320 380</td>
</tr>
</tbody>
</table>

Convergence and divergence. The delay between stimulus presentation and the beginning of convergence or divergence was determined by 30 measurements in 4 normal subjects. The arithmetic means are compared with those obtained from 4 patients with myotonic dystrophy (Table III). This comparison fails to reveal any significant difference between the two groups except in Case 1, where a pronounced prolongation of the reaction time was present. In this instance an average of 1.17 seconds elapsed before the eyes moved from convergence into a relatively divergent position (longest average time measured in a normal subject: 0.45 seconds), and the eyes began to converge from a relatively divergent position after a delay of 0.83 seconds (longest average time measured in a normal subject: 0.39 seconds).

Pursuit movements. A pronounced interference with the performance of smooth pursuit movements was demonstrated in all patients. For purposes of comparison, pursuit movements at various target speeds were registered in a healthy subject (Fig. 6). The physiology of pursuit movements in normal subjects has been described in an earlier publication. The recording demonstrates that smooth pursuit movements are performed, providing the target speed does not exceed 0.5 c.p.s. The recording also demonstrates that ideally smooth eye movements do not occur when following a moving target at slower speed, and that small saccadic eye movements in both directions of gaze are performed even in normal subjects. With increasing velocity of the target, the sine-shaped curve begins to disintegrate. The eyes begin to lose the target temporarily and compensate for the positional error by saccadic movements. The beginning of the sine curve distortion (critical frequency) occurs between 0.5

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*A slight tendency toward a longer delay between stimulus presentation and divergent movements was also observed in the remaining 3 patients and in 3 of the normal subjects. However, additional measurements will have to be obtained to ascertain whether any statistically significant difference exists between the reaction times of convergence and divergence in normal subjects.*
and 0.8 c.p.s. in normal subjects. Decomposition of the sine curve becomes complete at frequencies from 0.8 to 1.3 c.p.s. when pursuit movements are entirely substituted by saccadic movements of regard.

None of our patients demonstrated normal pursuit movements at any speed of the target. In the majority of instances the smooth pursuit movements were replaced by jerky deviation, saccades (cogwheel movements), or movements of regard. This indicated that the eyes were unable to follow the target smoothly and continuously, and were able to seize the latter only momentarily and at the points of reversal.

The sine curve was distorted even when the target was moving very slowly. Target speeds which would still elicit smooth pursuit movements in healthy subjects produced jerky eye movements in these patients (Fig. 7). A recording through varying target speeds which may serve as a representative sample for the entire group demonstrates the pathologic alteration of pursuit movements (Fig. 8).

An unusual phenomenon was observed in Case 1. After presentation of the horizontally moving target, futile attempts to move the eyes were made for as long as 10 to 12 seconds. Only after this delay did the eyes finally begin to move and the patient endeavor to follow the target (Fig. 9).

The pathologic manifestations of pursuit movements were equally well demonstrated during both monocular and binocular stimulation.

Discussion

The results of this study show a selective impairment of pursuit movements in both directions of gaze in patients with myo-
Pursuit movements are smooth and conjugate eye movements, which are elicited by the optokinetic effect of a moving visual stimulus. Normal visual acuity is not an essential prerequisite, since it has been shown that extreme blurring of the retinal image does not interfere with the smooth execution of pursuit movements. The velocity of the eye movement is directly related to the speed of the moving visual object. Low amplitude saccadic deviations are superimposed on the electrooculogram of smooth horizontal eye movements. These auxiliary movements are designed to compensate for differences between target position and eye position. Continuous fixation of the moving target is not the only mechanism involved. Additional factors participate in the regulation of pursuit movements. It has been demonstrated, for instance, that the eyes continue to move smoothly for some time after the cessation of the stimulus. It must be assumed from this and other observations that the performance of pursuit movements is influenced by cortical centers. Another essential factor in connection with the mechanism of pursuit movements is that of attention. Hofmann pointed out that pursuit movements are psychooptical reflexes which require the attention of the subject in addition to visual stimulation for normal functioning.

Pathologic changes of pursuit movements in both directions of gaze have to our knowledge never been described in connection with myotonic dystrophy. In fact, reports on any disturbance of pursuit movements which may be comparable to the changes observed in our patients have so far been extremely rare, and limited to subjects with brain stem affections and to those with strabismic amblyopia, the latter condition, however, always being associated with abnormalities of other types of eye movements.

Several possibilities have to be considered when attempting to interpret the nature of the interference in patients with myotonic dystrophy. The disease is characterized by a wasting of skeletal muscles and by an abnormally delayed relaxation...
of contracted muscles or muscle groups. Is it possible that dystrophic changes in the eye muscles interfere with smooth eye movements?

It has been suggested that limitation of ocular movement which is occasionally found in the lateral positions of gaze, as well as weakness of the orbicularis and levator muscles in patients with myotonic dystrophy, are caused by dystrophic changes in the extraocular muscles. However, the excursions of the eyes in our study did not exceed 15 degrees to either side during the performance of pursuit movements, and were thus well within the field of unimpaired motility, even in instances where lateroverision was restricted. This makes it unlikely that muscular weakness was responsible for the disturbance of pursuit movements. Moreover, other eye movements requiring high precision of function, such as movements of regard, convergence, and divergence were essentially normal, with the exception of Case 1, and should have been interfered with to an equal degree if muscle weakness was responsible for the abnormalities of pursuit movements. Is it possible that a myotonic reaction of the extraocular muscles is responsible for the abnormal conditions?

Davidson points out that “slight” eye movements usually do not elicit myotonia, whereas a prolonged motor effort such as strong closure of the eyes will be followed by a long delay in relaxation. He obtained electromyographic recordings from extraocular muscles in patients with the disease which yielded activity similar to that encountered in myotonic limb muscles. Paradoxically, these patients failed to present any anomalies of motility apparent to ordinary observation. Prolonged contraction of any of the extraocular muscles does not occur during the performance of pursuit movements. Alpern mentions that only slight differences of tension in the medial and lateral rectus muscles can be measured during pursuit movements. Thus, myotonic changes would be more likely to occur during movements of regard where steady fixation of a target in the lateral visual field and continued contraction of one yoked pair of horizontal rectus muscles are required, or during prolonged convergence. Abnormal movements of regard and prolonged reaction time were observed only in Case 1, where they may have been the result of a myotonic reaction of the extraocular muscles. However, the selective impairment of pursuit movements and the normalcy of movements of regard and ocular reaction time in 9 out of 10 of our patients cannot be easily explained on the basic of muscular or neuromuscular anomalies. A different interpretation must be sought.

Movements of regard and pursuit movements are related, inasmuch as they are both psychooptical reflexes and require attention for their normal function; but they differ in many ways. Accurate and precise movements of regard can be performed 2 to 3 weeks after birth, while smooth pursuit movements cannot be carried out until the fourth month of life. Movements of regard are extremely quick and predetermined in their path, whereas the velocity of pursuit movements depends on that of the moving target. A selective impairment of movements of regard in the presence of normal pursuit movements and the reverse situation have been observed and thus suggest the existence of two distinct and different anatomical pathways for each type of movement. On the basis of these considerations, it would seem reasonable to suggest that the selective impairment of pursuit movements in patients with myotonic dystrophy might be interpreted as evidence for involvement of the central nervous system in this disease.

The view that the central nervous system may be involved in myotonic dystrophy is not new. Endocrine and mental disturbances which occur commonly in this disease attest to the involvement of much more than muscle and neuromuscular mechanisms. A clue to the suspected cerebral mechanisms involved is supplied by reports on interference with the perfor-
formance of smooth pursuit movements in encephalitis, Parkinsonism, and cerebello-pontine angle tumors. It is of interest in this connection to note that Parkinsonism and myotonic dystrophy have been reported to occur as associated diseases. Further evidence for the possibility of central nervous system involvement in myotonic dystrophy has been furnished by autopsy reports describing pronounced cellular degeneration and destruction in the basal ganglia and hypothalamus.

The available data on central nervous system involvement in myotonic dystrophy do not justify final conclusions as to the pathologic basis for the impairment of eye movements. Nevertheless, the selective abnormality of pursuit movements demonstrated for the first time in this study is not only of diagnostic significance, but points strongly toward central nervous system involvement.

REFERENCES

Discussion

Dr. Mathew Alpern, Ann Arbor, Mich. Since the problem raised in this paper has clinical as well as theoretical implications for the physiology of eye movements, I hope I may be excused for resisting the temptation to give a flattering superficial discussion of the paper by Drs. von Noorden, Thompson, and Van Allen. This is difficult to do because the authors have succeeded in posing an important empirical question.

In the first place, we must examine carefully the tool by means of which the inferences under question were made. I have elsewhere discussed in some detail the artifacts and distortions in the objective recording of eye position by electro-oculography, and more recently Byford has shown how such nonlinearities may give a quite erroneous estimate of the behavior of the eyes during pursuit and saccadic eye movements. This criticism is important not only for an adequate appraisal of the results of this present study but also of those from the authors' Reference 8, which forms the physiologic background upon which the present experiments were designed.

To be specific: The reaction times for saccadic

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Eye movements (the term "movements of regard" is to be discouraged) in normal subjects were measured in the present instance to have a value which varied between 350 and 380 msecs., and if we are to agree with the authors' interpretation of the data in Table II (c.f., however, below) between 120 and 200 msecs. Thus the present data are at least 100 per cent too large! Clearly there must be an important artifact in the experimental procedure which is sufficiently large to mask conceivably a very significant difference in reaction times for saccadic eye movements in normal and dystrophic patients, which would go undetected in the present experiments. This same criticism also applies to the data of Table III.

In the second place, the degree of severity of the test that is applied to saccadic eye movements, which we are told are normal in dystrophic patients, and pursuit movements, which we are told are distinctly abnormal, is quite different; and the difference is very much biased to favor the authors' theoretical interpretation! Thus we are led to the belief that the presence of infrequent saccades on pursuit tracking of a sinusoidal target moving as fast as 15 degrees per second is distinctively abnormal, when normal records of Rashbass and Westheimer show such saccades even for targets moving slower than this, and indeed the authors' own Reference 8 shows a record from a "healthy subject" (Ref. 8, Fig. 3) that differs only in degree and not at all in kind from the records of some dystrophic subjects in the present article.

But this rather severe criterion of abnormality of the pursuit movements is not at all matched with a comparable criterion of abnormality when we examine saccadic eye movements which we are told are normal in these patients. Here the test is only the duration of the reaction time. One wonders if reaction time duration is a satisfactory test of normality for saccadic eye movements (and convergence), why it was not used for a test of normality for pursuit movements as well. Conversely, if one would like to have a more severe test for saccadic eye movements, would not the threshold spatial separation of targets to evoke a saccade be a profitable measurement? Normally this value is about 0.4 degrees, but if it were appreciably reduced in dystrophic patients, most of the authors' results could be explained.

In the third place, the present authors to the contrast that is established, is the reaction time for saccadic eye movements in dystrophic patients normal? I must confess that the evidence in this article suggests that it is not! If, as the authors tell us we must, we leave out the results from Case 1, as well as those from Case 3 (apparently not tested), the mean reaction time for the 8 remaining patients was 371.2 ± 6.1 msecs., which is considerably larger than that for the 3 normals of 360 msecs. Admittedly, there are not enough normal eyes to make a statistical statement, but surely the authors' failure to supply such information can only be regarded as incompleteness of the results. What are the normal limits? And what conceivable justification can there be for leaving out the results of Case 1?

The normal eye movements in tracking consist of superimposed movements of two independent systems. The smooth movement is a response to a movement of the image across the retina. It causes the target image to become stationary on the retina, but without the saccadic movement it cannot by itself bring the target image onto the fovea. This latter function is the task of the saccadic eye movement alone.

To evoke a pursuit eye movement, a velocity detection system is needed, and the velocity of the target and the velocity of the pursuit movement are linearly related in the normal eye, as long as the former is smaller than about 30 degrees per second.

If we can believe the evidence in the article, saccadic and smooth eye movements are differentially affected in myotonic dystrophy. This could be due to a defect in (a) the sensory receptor system (b) the motor system, or (c) the central nervous system. The authors are in support of the last of these possibilities, but I want to emphasize that their evidence excludes neither of the other two: namely, (1) a defect in the velocity detecting system in the retina without impairment in the position detection system, or (2) a loss in motor requirements of pursuit movements without loss of the motor requirements for small amplitude (30 degrees) saccadic eye movements because the latter are mechanically less difficult to perform. Indeed this last possibility would be supported by the data from Case 1, which by the authors' thesis remains unexplained.

Nevertheless, there is one compelling fact brought out by the data in the report which does lead me to wonder whether the authors may not after all be correct in the inference that myotonic dystrophy is an involvement of the central nervous system. The most curious feature of this extremely curious paper is that one fact is not even mentioned at all. This is the fact that the degradation of the pursuit eye movements in sinusoidal tracking tasks demonstrated here in dystrophic patients is similar to what Rashbass induced in normal observers by extremely small doses of barbiturates. This latter phenomenon is probably to be explained as an involvement of the central nervous system, and it seems to me that the similarity in the actions of these drugs and this disease merits careful detailed investigation. If the authors use
the present work as the point of departure for such an analysis of this question, we may very well be in their debt someday.

REFERENCES

Dr. von Noorden (closing). We are fully aware of certain limitations of the electrooculographic method which were mentioned by Dr. Alpern. However, in this study we are not dealing with minute changes which may escape electrooculographic registration, but with gross disturbances in patients with myotonic dystrophy. These changes can be clearly differentiated from recordings of normal subjects in this and earlier publications. There is no basis for the implication whereby the electrooculographic demonstration of the pronounced disturbance of pursuit movements in patients with myotonic dystrophy can be explained by artifacts, or erroneous interpretation of the electrooculogram.

The fact that ocular reaction times as measured by us in normals and patients with myotonic dystrophy differ from data published by Westheimer does not necessarily prove that our data are "100 per cent too large," nor do we claim that Westheimer's data are 100 per cent too small. It simply means that different results are to be expected when entirely different experimental methods are employed. We have not attempted to simulate Westheimer's experiments in this study, but have tried to establish whether or not a significant difference exists between reaction times of normals and of patients with myotonic dystrophy. Such a difference was not thought to be present with the exception of Case 1, which indeed presented many unusual features and thus could not be included in the group. We think it is quite possible that a true myotonic reaction of the extracocular muscles may explain the unique findings in this case. A similar case was reported by Verbiest.

Dr. Alpern wonders if the duration of reaction time is a satisfactory test of normalcy of saccadic eye movements. This claim was never made at any point in our paper. Movements of regard, or saccadic eye movements, as the discussant prefers to call them, were evaluated on the basis of swiftness and accuracy of the movement response to a light appearing in the periphery of the visual field. Samples of what we interpret as normal saccadic movements in patients with myotonic dystrophy will be demonstrated in the published paper.

Finally, we are grateful to the discussant for pointing out to us the work of Rashbass, who was able to produce degradation of pursuit movements in normal subjects with small doses of barbiturates. Since this drug acts on a cerebral level, this observation would indeed, as Dr. Alpern suggests, lend further support to our hypothesis.