Ocular saccades in Duane's syndrome

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SUMMARY Since Hering's law might be affected when paradoxical innervation exists, 10 patients with Duane's syndrome were studied. Eye movements were measured by binocular recording of electro-oculogram. Our study shows clearly that the sound eye's motility is also affected. When adduction of the affected eye is present, Hering's law holds, whereas when adduction is limited this law cannot be demonstrated.

Duane's syndrome is considered to be a most controversial anomaly. In most cases the involved lateral rectus muscle is unable to abduct, while during adduction it contracts, hence the synonymous connotation of paradoxical innervation. Clinical diagnosis of Duane's syndrome is based on the congenital deficiency of horizontal eye movement and narrowing of the lid fissure on attempted adduction of the affected eye (Walsh and Hoyt, 1969). Electromyographic (EMG) measurements confirmed that the most common pathogenesis is paradoxical innervation of the lateral rectus muscle (Breinin, 1957; Blodi *et al.*, 1964; Sato, 1960; Reny and Brichet, 1972; Huber, 1974). However, the inability to abduct the affected eye might be due in part to restriction by stiff medial tissues. This can be tested by force generation tests (Scott, 1971, 1975). An additional test to study Duane's syndrome was suggested by Metz *et al.* (1975), who compared the adduction and abduction velocities of the involved eye. They showed that adduction saccadic velocity is reduced in all types of Duane's syndrome and abduction velocity is reduced only in types 1 and 3. It was suggested that the reduced saccadic velocity on adduction is due to the co-contraction. The co-contraction of medial and lateral rectus muscles of the same eye on attempted adduction is an exception to Sherrington's law. According to Hering's law the anomalous innervation might be reflected in the other eye. In an attempt to evaluate these laws as they apply to Duane's syndrome we have studied eye movements in patients when the fixating eye was the sound eye, the affected eye, or both eyes simultaneously.

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Patient and method

A total of 10 patients, 3 to 62 years of age, having Duane's syndrome (Table 1) underwent complete eye examination as well as traction test. Eye movements were measured by binocular recording of electro-oculogram (EOG). Electrodes (Ag-AgCl) were placed at the outer canthi of the 2 eyes and the resulting signal led through a DC amplifier (26A2, Tektronix) to a tape recorder (3960A, 4-channel FM, Hewlett Packard), on which eye movements were recorded. The patient was seated, and his head was held still in a firm chin rest facing a series of small lights arranged 10, 20, and 30° left and right of a central fixation point. When a light was switched on a pulse was also generated,

Table 1 *List of patients by age, sex, the affected eye, deviation in primary position, and Duane's syndrome type*

Patient	Age (years)	Sex	Affected eye	Deviation in primary position	Duane's syndrome type
1	3	F	Left	Orthophoria	1
2	8	F	Left	Orthophoria	1
3	9	F	Left	Esotropia	1
4	12	F	Left	Exotropia	2
5	13	F	Right	Exotropia	3
6	13	F	Left	Esotropia	1
7	18	F	Left	Esotropia	1
8	19	F	Right	Orthophoria	1
9	44	M	Right	Orthophoria	1
10	63	M	Right	Esotropia	1

its amplitude being dependent on the light which was switched on. This voltage was recorded and served to calibrate the eye movement recorded for a fixed subtending angle. A time index was recorded on a separate channel.

The patient was asked to fixate on a spot of light in the centre. When a new spot of light appeared 30° to the right, the patient was asked to fixate at it. This procedure was repeated several times for all the arranged spots of light when both eyes were open and when the left and subsequently the right eye were occluded. The baseline drift of the recorded EOG, if it appeared, was corrected during the experiment. The recorded data were reproduced, photographed with a movie camera (PC-2A, Nihon-Kohden), and analysed.

Results

In order to evaluate the speed of eye movements when fixation was with one or both eyes we measured the relationship between amplitude and duration of the saccades as illustrated on top of Fig. 1. Six of the patients had similar amplitude-duration relationships as illustrated at bottom of the figure. These eye movements were divided into 3 groups as shown for 1 patient with Duane's syndrome in the left eye (G_I , G_{II} , G_{III} in Fig. 1). The duration of the responses for 10, 20, and 30° target movements was measured for a total of 180 measurements and their standard deviation for each corresponding amplitude was plotted. For each group a line was drawn through the mean. The grouping itself is shown in Table 2, where the eye movement is shown to be affected by the type of fixation, by the direction of movement, and whether the recording is from the sound or the affected eye. The type of fixation was either with both eyes, the sound eye, or the affected one. The direction of movement was abduction or adduction from midline or back to midline. The details of types of fixation, the direction of movement, and the recorded eye are given in Table 2. This grouping purposely disregards the differences of amplitude-duration relationship between nasal and temporal saccades.

In contrast to these 6 patients in the other 4 patients the adduction of the affected eye was slower. In these patients the adduction of that eye was limited to various degrees in all types of fixation (both eyes, the sound eye, or the affected one). When the affected eye was fixating and abducting (from midline temporally), past pointing and staircase saccades were recorded from the other eye. The former is illustrated in Fig. 2 (lower right). In this figure the upper trace of each quadrant represents the EOG of the right eye (RE) and the lower

trace of the left eye (LE). All the traces are eye movement responses to a target movement of 30° from the midline. In the left column the amplitude-duration relationship of the RE is normal whether both eyes are open (left upper quadrant), the RE fixating (centre), or the LE fixating (lower quadrant). The affected LE performed limited adduction in gaze to right.

For 6 of the patients the eye movement traces in the right column of Fig. 2 show that the sound eye (RE) had normal adduction when both eyes were open (right upper quadrant) or the RE fixating (centre), but when the affected LE was fixating the covered RE past pointed the target as illustrated in

Fig. 1 The definition of amplitude-duration relationship of a saccade is illustrated in the upper part. In the lower part the 3 regression lines represent the mean saccadic movement velocity response to stimulation of 10° , 20° , and 30° target movement. Vertical lines represent standard deviation for a total of 180 measurements. For details see Table 2

Table 2 *The grouping of eye movements as illustrated by the linear regression lines in Fig. 1*

Group no.	Recorded eye	Type and direction of eye movement	Fixing eye(s)
G _I	Sound eye	Abduction	Nasal to medial
			Both
	Affected eye	Adduction	Medial to temporal
			Sound, affected
G _{II}	Sound eye	Abduction	Temporal to medial
			Both, sound
	Affected eye	Adduction	Medial to nasal
			Affected
G _{III}	Sound eye	Abduction	Nasal to medial
			Both, sound
	Affected eye	Adduction	Temporal to medial
			Affected

the lower right quadrant. The responses in the other 4 patients were different (Fig. 3). In this figure examples were recorded from a patient with Duane's syndrome in his right eye. On gazing to the right, when the affected RE was fixating, the covered LE performed staircase saccade (left central quadrant). Nystagmoid movements of about 2 to 3° were discovered in the affected eye of 5 of the patients. Only in 1 of these patients did the amplitude and rate of the nystagmoid movements decrease when fixation was with the affected eye.

EMG was performed in 5 patients and showed the paradoxical innervation of the lateral rectus muscle of the affected eye on attempted adduction. In all patients the traction test was positive. The abduction of the affected eye was limited by the contracted medial rectus muscle.

Discussion and conclusions

The major finding in this study is that all eye movements in Duane's syndrome can be divided into 2 groups. In 1, as found in 6 of the patients studied, the sound eye and the affected eye have similar amplitude-duration relationship with 3 subdivisions as illustrated in Fig. 1. In each subdivision for any gaze both eyes move with the same speed. For example, trace G_I in Fig. 1 illustrates gaze to the right, i.e., abduction of the sound eye (RE) and adduction of the affected one (LE) as indicated in Table 2. Thus, even in Duane's syndrome, where paradoxical innervation exists, the amplitude-duration relationship of the sound and the affected eyes for the same gaze suggests that the yoke and antagonist muscles work in co-ordination. In this group of 6 patients the affected eye had normal amplitude-duration relationship in adduction up to 30° in spite of the reciprocal contraction of the antagonist lateral rectus muscle. The second group of 4 patients had slower adduction of the affected eye.

Thus, adduction in all patients with Duane's syndrome in this group seems to fall into a

Fig. 2 A patient with left Duane's syndrome. In each quadrant all upper traces are from the right eye and lower traces from the left eye. All eye movements start from straight ahead gaze. In the right lower quadrant the occluded RE performed a past-pointing movement, while the affected fixating LE is attempting to abduct. BE = both eyes; RE = right eye; LE = left eye

Fig. 3 A patient with right Duane's syndrome. In each quadrant all upper traces are from the right eye and lower traces from the left eye. All eye movements start from straight ahead gaze. In the centre of the left column a staircase saccade is illustrated. The affected RE is attempting to abduct, while the covered LE performs multiple small saccades. BE = Both eyes; RE = right eye; LE = left eye

generalised pattern of eye movement amplitude-duration relationship. The abnormality of the cross-innervation in different patients can be of different severities. The co-contraction of the lateral rectus occurs at various angles of adduction of the affected eye. While for a particular patient this slower eye movement can reach a similar performance, as shown in G_{III} of Fig. 1 for another patient, this adduction can be anywhere between G_I and G_{III} (Fig. 1). This might explain the pathogenesis of types 2 and 3 of Duane's syndrome where adduction is limited to various degrees of eye movements. In such patients the co-contraction of the lateral rectus will contribute significantly for smaller adduction movements. Recently in a study of Duane's syndrome Metz *et al.* (1975) showed that in all their patients the adduction velocity was slower. Our study showed that 4 patients had indeed slower saccades on adduction, but 6 patients had eye movements with normal velocity.

Four out of 10 patients performed staircase saccades with the covered eye as a response to a target movement in the other eye. The intersaccade interval was above 200 ms, suggesting that the correction was based on vision for correcting eye position. The eye, however, was covered, and no retinal error in this eye could be detected. This is interesting, since this error did not come from the uncovered eye, which was the affected eye and therefore unable to abduct. A possible explanation could

be that the multiple saccades were the result of error in eye movements, whereas the feedback is based on continuous internal comparison between the target, as first recorded in that mechanism, and the sequential eye movement responses. For such comparison to take place during the staircase saccade no vision is necessary.

Our study shows clearly that the sound eye in Duane's retraction syndrome is also affected. Its saccadic movements were influenced in accordance with the fixating eye and the direction of the movement. In 6 patients with normal adduction of the affected eye Hering's law held, whereas in 4 patients where the adduction was limited it could not be shown.

This study was supported by a special grant from S.R. & P.N. Foundation.

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