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LETTER TO THE EDITOR

Contributions of visual and motor signals in cervical dystonia

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Sir,

We read with great interest the article by Shaikh *et al.* (2016) presenting a pathophysiological explanation of cervical dystonia. Cervical dystonia is a neurological disorder characterized by involuntary twisting and turning of the head in any of the three dimensions. The authors proposed that the abnormalities of head movements seen in the disease stem from a malfunctioning head neural integrator caused by either an intrinsic dysfunction or as a result of impaired cerebellar, basal ganglia, or peripheral feedback. Several studies hypothesized that the head neural integrator relies on feedback using visual information, neck proprioception, and input from the cerebellum (Chan-Palay, 1977; Noda *et al.*, 1990; Fukushima and Fukushima, 1992).

Starting from the hypothesis that visual feedback may play a crucial role in the functionality of the head neural integrator and, subsequently, in the pathophysiology of dystonia, we decided to analyse the straight-ahead preference in healthy human subjects and patients with cervical dystonia. Several studies had previously demonstrated a dysfunction of visuo-spatial perception in cervical dystonia (Anastasopoulos *et al.*, 1998; Müller *et al.*, 2005). In particular, it had been suggested that egocentric space representation was impaired in cervical dystonia and that visual straight-ahead perception was shifted towards the trunk compared to normal subjects whose visual straight-ahead were claimed to coincide with the head midsagittal plane in these publications (Anastasopoulos *et al.*, 1998; Müller *et al.*, 2005).

Our study was based on the observation that healthy human subjects gazing to the side had been reported to respond faster to objects appearing in the peripheral visual field when the objects were displayed at a location in the subjects' straight-ahead direction compared to objects displayed at an eccentric location to the subjects' position (Durand *et al.*, 2012). This effect was independent of the subjects' direction of gaze and could be found even though the objects formed similar images on the retina and had the same distance from the fovea. Recently, the described behavioural observations of a preferential processing for visual information coming from the straight-ahead direction could be confirmed by a functional MRI study in healthy subjects on stimulus-evoked blood oxygen level-dependent responses in V1 and V2 (Strappini *et al.*, 2015). The authors provided evidence that visual stimuli elicited an enhanced response when presented closer to the straight-ahead direction.

The straight-ahead preference for visual information coming from the peripheral visual field may be part of the head neural integrator system, however, the exact underlying

Table 1 Clinical features of the patients

Age	Gender	Dominant hand	Last injection of Botox (months)	Duration of CD (months)	Rotatocollis	Rotation angle (°)	BFM	TWSTRS	Medication
40	M	L	3	264	L	12.5	13	16	Baclofene Clonazepam
36	F	R	none	4	L	35	4	11	Codeine Levodopa Benserazide Naproxene
40	M	R	4	48	L	53	10	12	Venlafaxine Bisoprolol
61	F	R	7	348	L	31	2	6	Clonazepam
61	M	R	5	252	L	28.9	10	12	Bromazepam
75	M	R	3	216	L	40.7	8	11	Perindopril Clopidogrel Atorvastatine Lercanidipine
58	F	R	4	120	L	11	6.5	10	None
52	M	R	6	72	L	49.5	8	21	Clonazepam Trihexyphenidyl
64	M	R	5	240	L	25.5	4	8	Clonazepam
46	F	L	3	2	R	40	8	13	Levothyroxine
44	F	R	3	288	R	49.3	8	17	None
50	F	R	None	132	R	37.5	6	14	None
29	F	R	3	96	R	34.3	18	14	None
53	F	R	3	84	R	39	8	10	None
49	F	R	3	60	R	51	8	15	Ketoprofen
52	F	R	4	60	R	50.4	8	13	Thiocolchicoside
54	F	R	4	84	R	28.8	8	8	Propanolol Candesartan Escitalopram Clonazepam
48	F	R	6	336	R	28	8	15	None
46	F	R	3	60	R	32.1	10	9	Pramipexole

BFM = Burke-Fahn-Marsden Dystonia Rating Scale; CD = cervical dystonia; F = female; L = left; M = male; R = right; TWSTRS =Toronto Western Spasmodic Torticollis Rating Scale.

mechanism for this effect remains unclear. Possible explanations could be a top-down or attentional modulation of visual cortex processing that allows the subject to select behaviourally relevant information, or a bottom-up modulation by extra-retinal signals, such as proprioceptive information encoding the eye position. In their study with human subjects, Durand *et al.* used an attention-demanding test that required their participants to not only disengage their gaze but also their attention from the straight-ahead position (Durand *et al.*, 2012). As the results of this particular task showed the same preference for the straight-ahead perception of visual objects, the authors concluded that while an attentional explanation could not be excluded, the observed effect does not seem to require full attentional resources, and that there might be an automatic early integration of visual and postural inputs, which facilitate straight-ahead perception. This explanatory model would be in line with several studies reporting on the influence of gaze direction or proprioceptive eye position signals on visual cortex processing (Nieman *et al.*, 2002; Andersson *et al.*, 2007; Balslev *et al.*, 2012).

When adapting the paradigm of Durand *et al.* to an experimental setting (see Supplementary material for method) including healthy subjects and patients with cervical dystonia, we found that patients with cervical dystonia did not exhibit any preferential reaction for targets displayed closer to their head or their trunk axis at the population level (see Supplementary material for results). Our study may reveal a subtle perturbation of the visual information processing in cervical dystonia. It is unclear whether or not the described impairment of visual information processing in cervical dystonia is part of the cause or of the result of the disease's pathophysiology. However, our findings underline that other sensory inputs and cortical processes are affected in cervical dystonia and that their impairment may contribute to the hypothesized dysfunction of the head neural integrator. Additionally, they may broaden the spectrum of possible novel therapies mentioned by Shaikh *et al.* in that physical therapy using visual feedback techniques could be helpful in treating the disease (Harrison *et al.*, 1984).

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Supplementary material

Supplementary material is available at *Brain* online.

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