Parkinsonism and Related Disorders 21 (2015) 561–566

Contents lists available at ScienceDirect

Parkinsonism and Related Disorders

journal homepage: www.elsevier.com/locate/parkreldis

Why are voluntary head movements in cervical dystonia slow?

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ARTICLE INFO

Article history:
Received 10 December 2014
Received in revised form
11 February 2015
Accepted 5 March 2015

Keywords:
Saccade
Cerebellum
Movement disorder
Bradykinesia
Dystonia
Head movement

ABSTRACT

Introduction: Rapid head movements associated with a change in fixation (head saccades) have been reported to be slow in cervical dystonia (CD). Such slowing is typically measured as an increase in time to complete a movement. The mechanisms responsible for this slowing are poorly understood.
Methods: We measured head saccades in 11 CD patients and 11 healthy subjects using a magnetic search coil technique.
Results: Head saccades in CD took longer to reach a desired target location. This longer duration was due to multiple pauses in the trajectory of the head movement. The head velocity of each segment of the (interrupted) head movement was appropriate for the desired total movement amplitude. The head velocity was, however, higher for the amplitude of the individual interrupted movements. These results suggest that brain programs the proper head movement amplitude, but the movement is interrupted by pathological pauses.
Conclusion: Voluntary head saccades have a longer duration in CD due to frequent pauses. The frequent pauses reflect pathological interruptions of normally programmed intended head movement.

1. Introduction

The dystonias are a group of disorders characterized by excessive muscle contractions leading to involuntary movements and abnormal postures [1]. Muscles of the neck are affected in cervical dystonia (CD) leading to abnormal twisting and turning of the head, often combined with jerky spasms or tremor-like movements [2]. In addition to these involuntary abnormal movements, several studies have shown that rapid voluntary redirections of the head from one target to another (head saccades) take longer than normal in CD [3–5].

Although these studies have been interpreted to mean that head saccades in CD are slow, they did not take into account an important relationship between head saccade amplitude and velocity, known as the main sequence [6]. Specifically, the velocity of a head saccade depends on the intended amplitude of the movement. There are three different explanations for the increased time to complete head saccades in CD (Fig. 1 and Table 1). The first involves abnormal central commands that encode head velocity. This hypothetical mechanism predicts that peak head velocity will be reduced for the desired amplitude of the head saccade in comparison to normal (red trace in Fig. 1B,F). The second possible reason is a failure of the brain to program the head movement with an appropriate amplitude. Thus, instead of one adequately large movement the brain might program multiple small sequential movements (Fig. 1C), resulting in frequent small pauses during the head saccade, increasing the time to reach the desired location. This mechanism predicts normal head velocities for the amplitudes of each segment of the movement (θn) (blue trace in Fig. 1F), but peak velocity would be smaller compared to the larger desired movement ($\Sigma\theta n$, blue trace in Fig. 1E). A third possible reason for slower head saccades is that a pathological process interrupts the normally programmed head movement, producing a series of concatenated movement bursts interspersed by brief pauses (Fig. 1D). Upon resumption of the movement, the head continues to obey the kinematic parameters of the originally programmed head saccade. Because the main sequence relationship predicts that larger amplitude movements have higher peak velocities than smaller ones, the third hypothesis predicts that the overall peak head velocity would be normally programmed for the desired total head movement; consequently, each smaller movement segment will

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http://dx.doi.org/10.1016/j.parkreldis.2015.03.005
1353-8020/

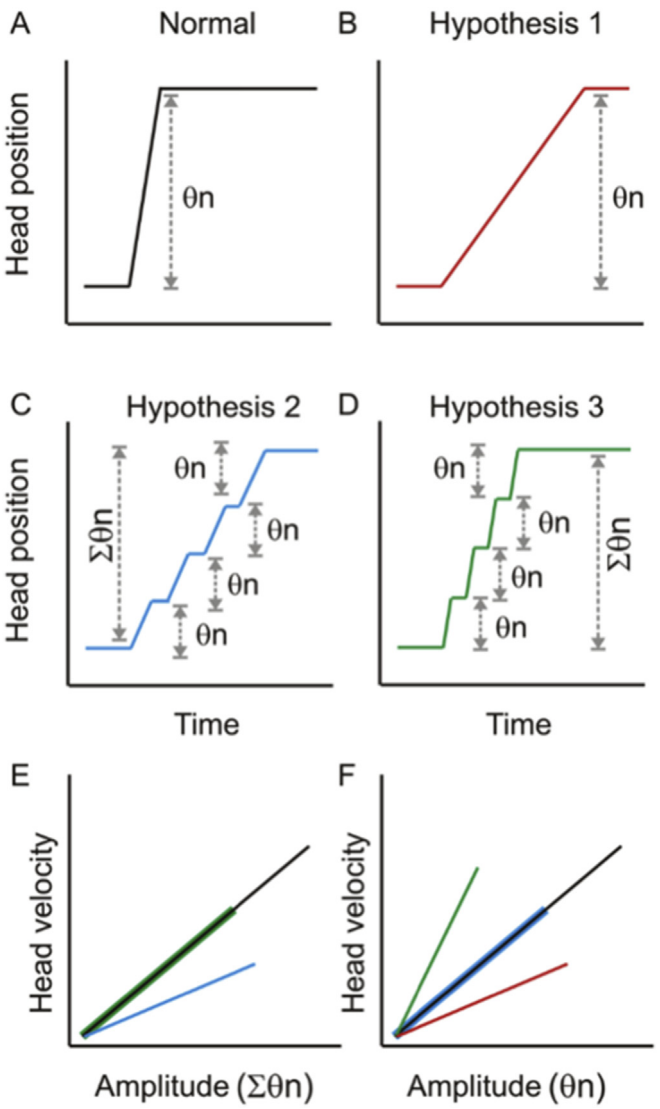

Fig. 1. Schematic illustration of three potential mechanisms for slowing of head saccades in CD. In panels A–D head position is shown on the y-axis while corresponding time is on the x-axis. (A) Normal subject, head position is depicted as black line. The subject makes one head saccade with amplitude θn. (B) Mechanism 1, head saccades are slow in CD. The peak head velocity is reduced leading to increased movement time for the same movement shown in panel A. (C) Mechanism 2, the brain programs multiple head movements each with small amplitude of head movement θn to accomplish desired head movement with amplitude ∑θn. (D) Mechanism 3, the brain programs appropriate amplitude ∑θn, but it is interrupted into smaller segments each with amplitude of θn. Panels E and F depict schematic representations of each mechanism in terms of peak head velocity to amplitude (main-sequence) relationship. Amplitude in panel E corresponds to the desired head movement, while the amplitude in panel F corresponds to the amplitudes of each interrupted segment. In mechanism 2, the brain programs smaller head movements, their velocities are appropriate for the segments with smaller amplitude θn, but they are too slow for desired (large amplitude) head movement ∑θn. The corresponding blue line in panel E depicts a smaller slope than the black line but the same slope as black line in panel F. In mechanism 3 (green line) the brain encodes head velocity that is appropriate for the desired head movement with the large amplitude but it is faster for the amplitude of small segments. Thus the corresponding line depicting the main sequence falls on the black line in panel E but it has a higher slope than black line in panel F. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

appear unusually fast in relation to its amplitude (green traces in Fig. 1E,F).

To discriminate among these mechanisms, we performed a quantitative dissection of head saccades in 11 patients with CD and compared head velocities for different movement amplitudes. The results suggest that slowing of head saccades is caused by pathological interruptions of an otherwise normally planned movement. Thus the terms “slowing” or “bradykinesia” are not entirely appropriate.

2. Methods

We evaluated 11 healthy subjects and 11 with isolated focal CD (Table 2). Patients with secondary dystonia, segmental or generalized dystonia, or features suggestive of a neurodegenerative disorder were excluded. The Johns Hopkins University Institutional Review Board approved the study; all subjects gave written informed consent before participating.

Head movements were recorded using a three-axis search coil (Skalar, Delft, The Netherlands) mounted on a head-fixed bar. Subjects sat in the magnetic coil frame such that the mid-sagittal plane coincided with the center of the frame. The trunk was restrained with a cushioned chest bar, hence allowing the head to move on a stationary torso. Horizontal head movements were defined as those around an earth vertical axis passing through the center of the coil frame (i.e., turning the chin to the right or left, or torticollis). Vertical head movements were those around an earth horizontal axis, passing through the center of the coil frame and parallel to the interaural line (i.e., flexion and extension of the head, or retrocollis-anterocollis). In this study we only quantified horizontal and vertical head movements. The angular position of the search coil with respect to the magnetic fields was digitized at 1000 Hz and the data were processed to compute the position of the head in three-dimensions [7].

Subjects wore a headband with an attached laser pointer. This setup used a head-fixed laser target to provide visual feedback about the spatial reference of the head position. Subjects were asked to aim their heads, using the head-fixed laser as a guide, at a light-emitting diode (LED) target located at 0°, and to turn the head as fast as possible (i.e., make a head saccade) toward LEDs located either to the right and left at 10°, 20°, and 30°, or up and down at 10° and 20°. Subjects made up to 2 head saccades to each eccentric target in either direction; hence up to 20 head saccades were recorded per subject. Three-dimensional head positions were further analyzed using Matlab® (The Mathworks™, Natick, MA). Mathematical derivations of angular head positions were used to compute head velocity and acceleration. Low-pass filtering and three-point averaging removed signal noise inherent to the mathematical derivations. Head saccades were interactively selected. Troughs in the velocity trace distinguished the breaks in the head saccades; those troughs reaching zero indicated a complete pause in the movement. A velocity criteria of 5°/s was used to determine the beginning of head saccade. Peak head velocity was defined as the maximum velocity achieved across the entire movement; when several local maxima were present in the velocity trace (i.e., when interruptions were present in the head saccade), this corresponded with the largest peak. We use a similar definition for peak acceleration. Statistical analysis was performed with the Matlab® statistics toolbox.

3. Results

3.1. Representative head movement traces

Fig. 2A shows an example of a 30°-amplitude horizontal head saccade in a healthy control. There was one uninterrupted movement lasting approximately 250 ms, with a single velocity peak (Fig. 2B) and a biphasic acceleration profile that had positive acceleration and negative deceleration peaks (Fig. 2C).

Fig. 2D depicts an example of a 30°-amplitude head saccade in a patient with CD, when moving in the same direction as their involuntary turning (pro-dystonic direction). The movement took 1088 ms; the longer duration was accompanied by frequent small pauses. The interruptions caused complete cessation of the movement as shown in the velocity trace where there are four peaks in horizontal head velocity and each peak is followed by a trough reaching zero (Fig. 2E). These interruptions were also seen in the acceleration trace (Fig. 2F).

Fig. 2G shows a 30°-amplitude head saccade in a patient with CD moving in a direction opposite to their involuntary turning (anti-dystonic). This movement spanned over 1323 ms, it again was associated with pauses seen in the head position (Fig. 2G), velocity (Fig. 2H), and acceleration traces (Fig. 2I). Results from this representative patient imply that the increased time required for head