Neuro-ophthalmology 0165-8107/94/US$ 3.50

Neuro-ophthalmology – 1994, Vol. 14, No. 2, pp. 79-84

Buren (The Netherlands) 1994

Accepted 28 January 1994

Torsional rebound nystagmus in a patient with type I Chiari malformation

D. Straumann
E. Müller

Neurology Department, University Hospital, CH-8091, Zürich, Switzerland

Abstract In a 41-year old female patient with a type I Chiari malformation, the authors observed the following symmetrical oculomotor phenomenon: (1) With gaze straight ahead, eye position was rather stable with some drift about the torsional axis. (2) With lateral gaze, nystagmus developed with extorsional quick phases of the abducting, and intorsional quick phases of the adducting eye. (3) After returning to gaze straight ahead, there was rebound nystagmus with a torsional component in the opposite direction. The failure to keep the torsional eye position component constant independent of the actual direction of gaze and the preceding gaze displacement implies that the oculomotor output in this patient is violating Listing's law. Based on magnetic resonance imaging of the head, the authors hypothesize that important neural structures implementing Listing's law are localized in the caudal cerebellum or lower brainstem.

Key words Eye movements; three-dimensional; Listing's law; cerebellum; brainstem

Correspondence to:
D. Straumann, M.D., Neurology Department, University Hospital, CH-8091 Zürich, Switzerland, Phone: 41 1 255 55 00, Fax: 41 1 255 45 07

Introduction In Chiari malformation, which involves structures in the lower brainstem and the caudal cerebellum, eye movement disorders are often prominent.[1,2] These include rebound nystagmus,[3] which, in general, is considered to be a typical cerebellar sign,[4] possibly caused by a floccular or paraflocular lesion.[5-7] Rebound nystagmus is diagnosed if a horizontal gaze-evoked nystagmus persists and changes to opposite direction when the eyes are moved back to gaze straight ahead. Here, we describe a patient with a type I Chiari malformation in which we have found rebound nystagmus in the torsional direction evoked by eccentric horizontal gaze.

Case report At age 31, an Italian woman started to experience increasing weakness and pain of the distal left arm. Eight years later, the same symptoms developed on the right side. Examination revealed muscle wasting and reduced strength of the small hand muscles on both sides, and decreased sensibility on the ulnar side of the left forearm. Tendon reflexes were brisk for the lower and weak for the upper extremities. Sensibility for pain and temperature on the left side of the body was reduced, sparing

the head. Cranial nerves were found to be normal. Magnetic resonance imaging of the cervical and thoracic spine disclosed a syringomyelia with a central cavity extending from C2 to the upper thoracic region (diameter 1-3 mm).

At age 41, the patient again came to the hospital because of repeated occurrence of blurred vision, but no oscillopsia. Such episodes usually lasted several minutes and could sometimes be terminated by inclination of the head. Besides a rebound nystagmus in the torsional direction (see *Results*), clinical evaluation did not show significant changes compared with the records of two years ago. The patient was further evaluated in the vestibulo-oculomotor laboratory.

Methods After clinical neuro-ophthalmological examinations, partly documented by video recording, oculomotor abnormalities were measured with the three-dimensional search coil technique.[8] We used a 'Type 3000 Eye Position Meter (Skalar Instruments, Delft, The Netherlands). Digitized data from a dual search coil mounted on the right eye, sampled with 833 Hz, were stored on the hard disk of a computer. Because of small torsional drift of the eyes with gaze straight ahead (zero position), we could not determine a zero torsional eye position. As an estimate of torsional gain, we took values obtained by an *in vitro* calibration of the dual search coil on a gimbal system, performed prior to patient testing.

Tests were completed by direct current electro-oculography (EOG). The patient was seated on a vestibular chair, which was surrounded by an optokinetic drum. Chair and drum were independently driven by servo-controlled torque motors. In this set-up we tested vestibulo-ocular reflex, optokinetic nystagmus and smooth pursuit eye movements. Subsequent neuroradiological studies included magnetic resonance imaging of the head.

Results

CLINICAL OCULOMOTOR EXAMINATION With gaze straight ahead, fixation was stable with no spontaneous nystagmus. There was some slight drift of the eyes about the naso-occipital axis with a changing positive or negative torsional direction. Occasionally, one could observe quick torsional resetting of the eyes. When gaze was moved to the right or left, conjugate torsional nystagmus started immediately. Torsional quick phases were positive when gaze aimed to the right, and negative with gaze to the other side.* We did not notice any attenuation of torsional nystagmus or a change of nystagmus direction with prolonged eccentric gaze. However, there were repeated saccadic intrusions in the direction of primary position, and the patient reported increasing blurred vision, but no oscillopsia. With gaze back in zero position, torsional nystagmus changed direction and continued for more than a minute when the eyes stayed in this position. Torsional gaze-evoked and rebound nystagmus was not elicited with vertical eye displacement.

* Positive torsion is extorsion of the right eye and intorsion of the left eye.

THREE-DIMENSIONAL SEARCH COIL RECORDING Fig. 1 shows a representative portion of the data. The patient had to fixate a dot on a tangent screen at a distance of 164 cm. After initial fixation of the zero position, targets were consecutively given at 10, 20 and 30 deg to the right (as indicated by the dashed line); then the patient had to direct her gaze to zero position

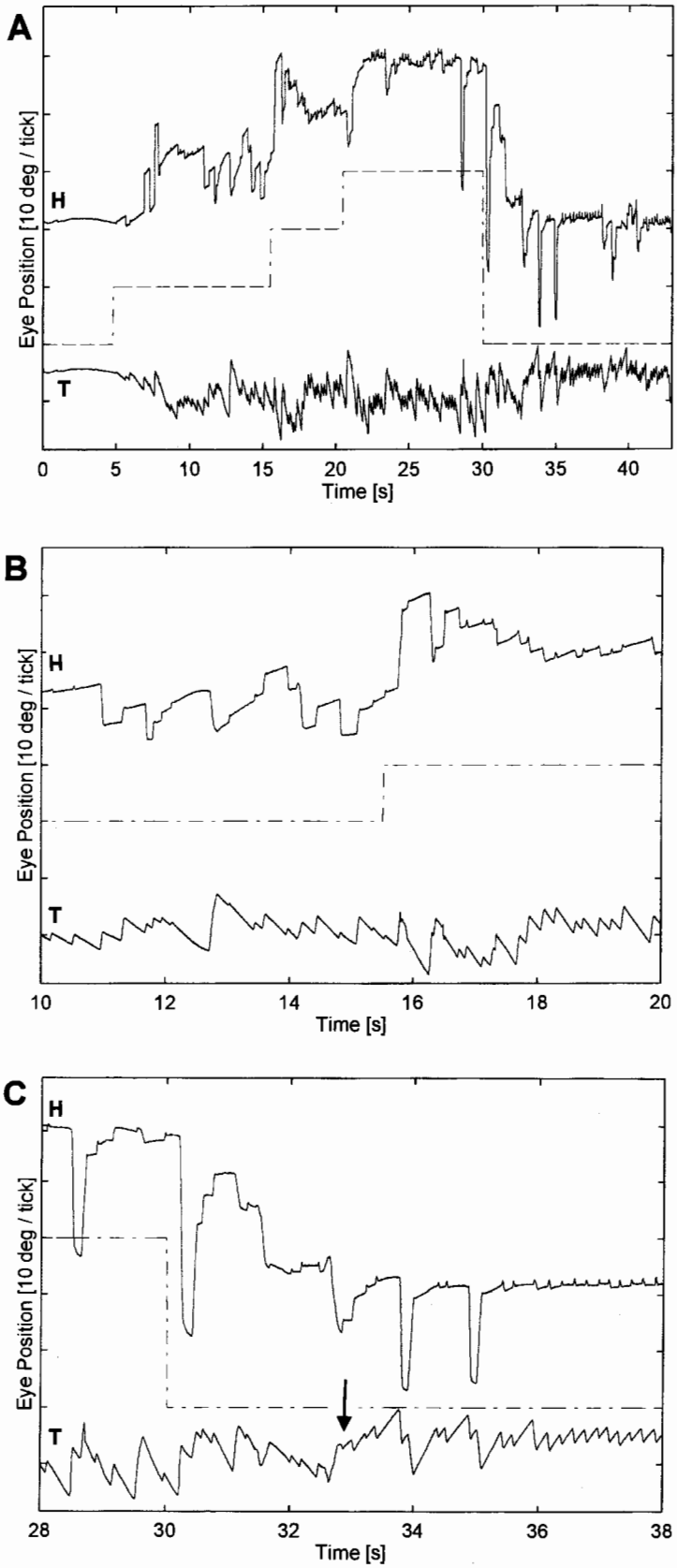

Fig. 1. Three-dimensional search coil recording in the patient. Right eye measured. H: horizontal eye position. T: torsional eye positon. Dashed line: target position. (A) Entire trial. (B) Section 10 s after start of trial with fixations at 10 and 20 deg to the right. (C) Section 28 s after start of trial with fixations at 30 deg to right and straight ahead. Arrow marks change of nystagmus direction.

again (Fig. 1A). With gaze shifted to horizontal eccentricities, positive torsional nystagmus (T) begins instantly. Probably due to this constant movement activity in the torsional direction, the horizontal eye position (H) is unstable, and there are repeated centripetal saccadic intrusions. Torsional nystagmus is intensified with increasing horizontal gaze eccentricity, as shown in a section of the trial (Fig. 1B). The patient needs about 2 s to recenter gaze from 30 to 0 deg (Fig. 1C). Hereafter, torsional nystagmus quickly changes its direction (arrow). The same trial, repeated with fixations to the left side (not shown), qualitatively yielded the same results.

ELECTRONYSTAGMOGRAPHY The vestibulo-ocular reflex to both sides showed a rather high gain of 0.9 with a time constant of about 10 s (measured after velocity steps of the turntable by 80 deg/s with 100 deg/s/s). A fast rise of eye velocity after the beginning of constant horizontal optokinetic stimulation was absent, and the optokinetic gain was very low for high drum velocities. Horizontal pursuit eye movements were saccadic to both sides with additional interruptions by torsional nystagmus. Visual suppression of horizontal vestibular nystagmus was impaired in both directions.

MAGNETIC RESONANCE IMAGING Fig. 2 shows a nearly midsagittal T1-weighted section of the brainstem and cerebellum: The caudal cerebellum reaches into the foramen magnum, and the slim lower medulla oblongata totally fills the subarachnoidal space at this level. Pons and midbrain show no abnormal configuration or signal intensity. In other sections (not shown), there was no supratentorial pathology except for a minimal internal hydrocephalus without transependymal secretion of liquor.

Discussion This 41-year old female patient with a type I Chiari malformation showed a torsional rebound nystagmus, evoked by horizontal gaze to

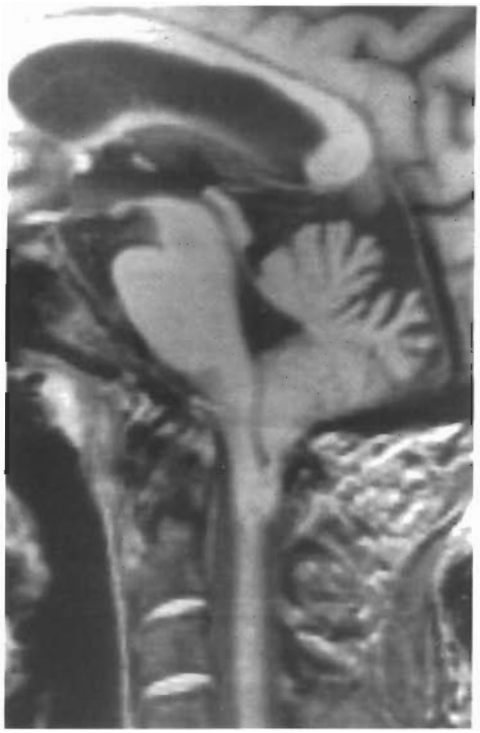

Fig. 2. Magnetic resonance imaging of the head. Section with enlargement of infratentorial structures.

both sides. As in other patients with pathological torsional nystagmus, *e.g.*, many patients with Wallenberg's syndrome, our patient did not complain of torsional oscillopsia but rather of blurred vision, which was the reason why she consulted her physician.

Occurrence of torsional nystagmus implies a violation of Listing's law. This law states that all axes about which the eye rotates from a reference position to any other physiological position normally lie in plane (Listing's plane), provided the head is stationary.[9-14] Hence, without vestibular input there are only two degrees of freedom for eye movements – horizontal and vertical – while the torsional degree of freedom is kept constant. It is generally assumed that Listing's law has neural, not mechanical origins, since vestibular stimulation leads to eye movements with three degrees of freedom, and therefore trajectories are not restricted to Listing's plane.[15] The anatomical localization of the neural structure implementing Listing's law is not yet known.

Theoretically, pathologies which induce eye movements out of Listing's plane are not necessarily situated inside the network that encodes Listing's law. For example lesions localized in the canal or otolith pathways of the peripheral or central vestibular system can produce spontaneous torsional nystagmus even if the 'neural Listing network' is still intact. The same argument applies for lesions of the eye plant or the motoneurons, where possible violations of Listing's law occur downstream of its neural implementation.

In the patient presented, we have reason to believe that the Listing network itself is affected: In normals, this network keeps the torsional eye position constant, which is – due to the non-commutativity of three-dimensional rotations – not a trivial task.[16] The instability of torsional eye position and the dependence of torsional nystagmus direction on actual and previous gaze direction is a clear indication that the patient's nervous system lacks the ability to compute synergistic activation of ocular motoneurons for eye movements in Listing's plane.

Based on magnetic resonance imaging of the head in the patient, we speculate that important structures involved in the implementation of Listing's law lie in the caudal cerebellum or lower brainstem. This hypothesis is supported by preliminary findings in rhesus monkeys, which indicate that bilateral ablation of the cerebellar nodulus and ventral uvula can lead to oscillations of the torsional eye position component.[17] Further experimental studies in animals and careful clinical evaluation of patients with pathologies in torsional eye movements should clarify the exact anatomical localization of the neural network which computes eye movements in Listing's plane.

References

1 Leigh RJ, Zee DS. The neurology of eye movements. Philadelphia: FA Davis, 1991; 425-426.

2 Saez RJ, Burton MO, Yanagihara T. Experience with Arnold-Chiari malformation. J Neurosurg 1976; 45:416-422.

3 Lennerstrand G, Gallo JE. Neuro-ophthalmological evaluation of patients with myelomeningocele and Chiari malformations. Dev Med Child Neurol 1990; 32:415-422.

4 Hood JD, Kayan A, Leech J. Rebound nystagmus. Brain 1973; 96:507-526.

5 Bondar RL, Sharpe JA, Lewis AJ. Rebound nystagmus in olivocerebellar atrophy: A clinico-pathological correlation. Ann Neurol 1984; 15:474-477.

6 Yamazaki A, Zee DS. Rebound nystagmus: EOG analysis of a case with a floccular tumor. Br J Ophthalmol 1979; 63:782-786.

7 Zee DS, Yamazaki A, Butler PH, Gucer G. Effects of ablation of flocculus and paraflocculus on eye movements in primate. J Neurophysiol 1981; 46:878-899.

8 Robinson DA. A method of measuring eye movements using a search coil in a magnetic field. IEEE Trans

Biomed Eng 1963; 10:137-145.
9 Tweed D, Fetter M, Andreadaki S, Koenig E, Dichgans J. Three-dimensional properties of human pursuit eye movements. Vision Res 1992; 32: 1225-1238
10 Haslwanter T, Straumann D, Hepp K, Hess BJM, Henn V. Smooth pursuit eye movements obey Listing's law in the monkey. Exp Brain Res 1991; 87:470-472.
11 Helmholtz H. Handbuch der Physiologischen Optik. Leipzig: Voss, 1867.
12 Hepp K. On Listing's law. Commun Math Phys 1990; 132:285-292.
13 Straumann D, Haslwanter T, Hepp-Reymond MC, Hepp K. Listing's law for eye, head and arm movements and their synergistic control. Exp Brain Res 1991; 86:209-215.
14 Tweed D, Vilis T. Geometric relations of eye position and velocity vectors during saccades. Vision Res 1990; 30:111-127.
15 Crawford JD, Vilis T. Axes of eye rotation and Listing's law during rotations of the head. J Neurophysiol 1991; 65:407-423.
16 Tweed D, Vilis T. Implications of rotational kinematics for the oculomotor system in three dimensions. J Neurophysiol 1987; 58:832-849.
17 Angelaki DE, Hess BJM. Vestibulo-ocular control by the cerebellar nodulo-uvular lobulus. In: Delgado-Garcia JM, Godaux E, Vidal PP (editors). Information Processing Underlying Gaze Control. Oxford: Pergamon, in press.