Movement Disorders
CLINICAL PRACTICE
CLINICAL VIGNETTES

Monocular Patching Attenuates Vertical Nystagmus in Wernicke's Encephalopathy via Release of Activity in Subcortical Visual Pathways

Christoph Helmchen, MD,[1,2,*] Björn Machner, MD,[1,2] Andreas Sprenger, PhD,[1,2,3] and David S. Zee, MD[4]

Downbeat nystagmus (DBN) is common in ataxia syndromes and usually related to cerebellar flocculus dysfunction. Persistent and disabling DBN commonly occurs in B1 deficient Wernicke's encephalopathy (WE) causing a functional cerebellar disorder.[1]

Here we describe a new, unusual phenomenon in a WE patient who has had severe vertical oscillopsia for years due to an incapacitating DBN: oscillopsia and his nystagmus is markedly attenuated when he covers one eye and views monocularly. We propose this is a new clinical sign of emerging activity in subcortical visual pathways.

The 54 year-old patient developed non-alcoholic WE from malnutrition after removal of a squamous cell carcinoma in the neck. We previously reported a salutary effect of alcohol on his nystagmus.[2] His thiamine levels were markedly reduced (9 μg/l, 24-99=normal). Following thiamine treatment the initial upbeating nystagmus changed to DBN. MRI showed high-signal intensities in paraventricular structures in the dorsal medulla, including the perihypoglossal complex (PHC) and the medullary paramedian tract neurons (PTN) that are related to normal gaze-holding. The patient consented to eye recordings using a video-based eye tracker (EyeLink II)[3] and the study was approved by the ethics committee of the University of Lübeck (20-498). DBN was 13.7°/s ± 5.2 (average of all gaze positions; increase on lateral and downward gaze) but there was a striking decrease of DBN in gaze straight ahead by monocular far viewing (Fig. 1, Video 1). Near viewing with convergence also attenuated his DBN, but remarkably, monocular viewing alone, even when looking at a distance, did the same. This curious effect recalled infantile latent nystagmus, in which monocular viewing releases a horizontal and vertical (usually upbeat) nystagmus, though in our patient paradoxically his vertical nystagmus lessened. The effect in infantile latent nystagmus has been attributed to residual activity in primitive, subcortical visual pathways that project, via the midbrain nuclei of the optic tract (NOT, horizontal) and the accessory optic nuclei (AON, vertical),[4] to lower brainstem structures that project to the cerebellum.[5] We speculate that monocular viewing in our patient unmasked an inherent tone asymmetry from the AON (which normally favors downward slow phases as in latent nystagmus) that opposed the pathologic upward bias of DBN. This hypothesis suggests that acquired pathology in adults may uncover activity in phylogenetically-old, subcortical visual pathways mediated by the accessory optic nuclei. The dynamic changes in the vestibular tone imbalance (upbeat to downbeat nystagmus) exerted by the affected brainstem structures in WE on the intact cerebellar flocculus in this non-alcoholic WE patient may be crucial for unmasking activity in the subcortical pathways. However, there are other neurodegenerative movement disorders apart from the cerebellar ataxias, for example progressive supranuclear palsy (PSP) and Multiple Systems Atrophy (MSA), that reveal different forms of vestibular tone imbalance and monocular patching may also uncover activity in these subcortical visual pathways that oppose pathological imbalances.

Clinically, this is important, as patients could wear an eye patch to abolish DBN. Our patient is watching movies on a TV screen with one eye covered. Prisms could also change viewing distance or to move the eyes or head where the nystagmus occurs less.

[1]*Department of Neurology, University Hospital Schleswig-Holstein, Lübeck, Germany;* [2]*Center of Brain, Behavior and Metabolism (CBBM), University of Lübeck, Lübeck, Germany;* [3]*Institute of Psychology II, University Lübeck, Lübeck, Germany;* [4]*Department of Neurology, Ophthalmology, Otolaryngology-Head and Neck Surgery, and Neuroscience, Division of Neuro-Visual and Vestibular Disorders, The Johns Hopkins Hospital, Baltimore, Maryland, USA*

***Correspondence to:** Prof. Christoph Helmchen, Department of Neurology, University of Lübeck, University Hospitals Schleswig Holstein, Campus Lübeck, Ratzeburger Allee 160, D-23538 Lübeck, Germany; E-mail: christoph.helmchen@neuro.uni-luebeck.de
Keywords: Wernicke's encephalopathy, monocular viewing, subcortical visual pathways.
Andreas Sprenger and David S. Zee contributed equally as co-last authors.
[The copyright line for this article was changed on 07 December 2021, after original online publication.]

Received 23 September 2021; revised 19 October 2021; accepted 24 October 2021.
Published online 4 December 2021 in Wiley Online Library (wileyonlinelibrary.com). DOI: 10.1002/mdc3.13380

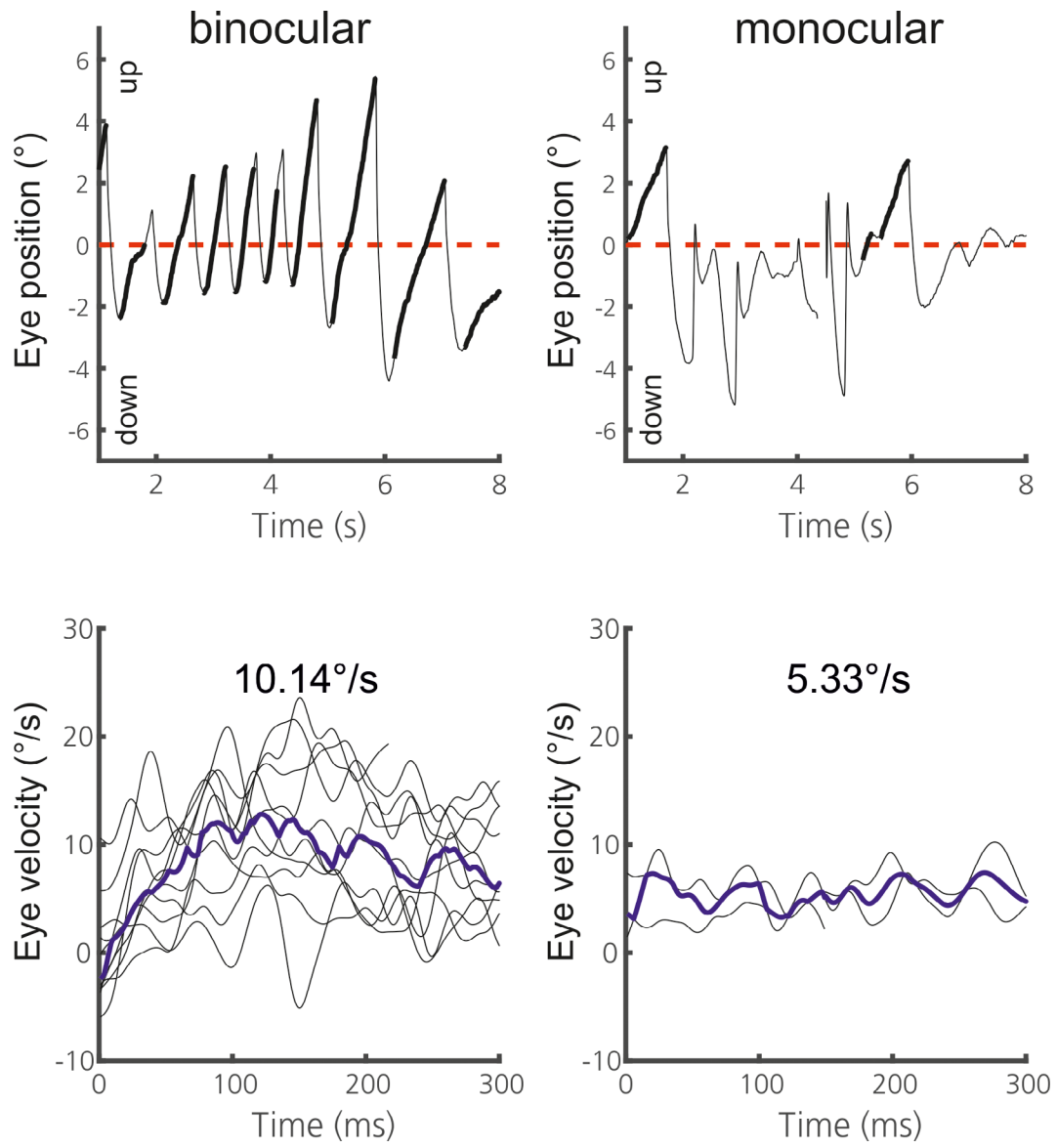


FIG 1. Modulation of DBN by monocular viewing conditions. Vertical eye position (upper panel) and eye velocity (lower panel) with DBN during binocular versus monocular far viewing. There is a marked (almost 50%) reduction of DBN during monocular far viewing with gaze in the straight-ahead position (1.2 m distance).

Acknowledgment

Open Access funding enabled and organized by Projekt DEAL.

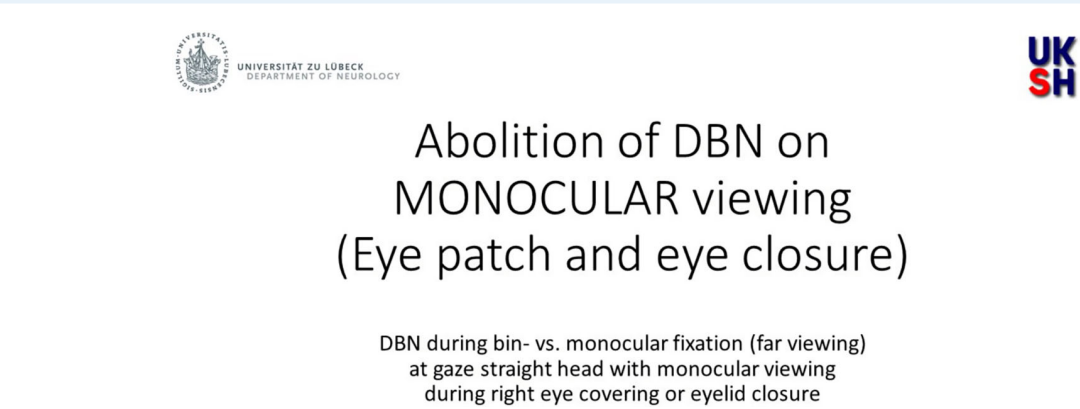
Author Roles

(1) Research project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript: A. Writing of the first draft, B. Review and Critique.

CH: 1A, 1B, 1C, 2A, 2B, 2C, 3A, 3B
BM: 1A, 3A, 3B
AS: 1A, 1B, 1C, 2A, 2B, 2C, 3A, 3B
DZ: 1A, 2C, 3A, 3B

Disclosures

Ethical Compliance Statement: The patient consented to eye recordings using a video-based eye tracker (EyeLink II) and video recordings and the study was approved by the ethics committee of the University of Lübeck (20-498). We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.



Video 1. DBN abolition by monocular viewing: DBN during bin- versus monocular fixation (far viewing) at gaze straight head with monocular viewing during right eye covering or eyelid closure.
Video content can be viewed at https://onlinelibrary.wiley.com/doi/10.1002/mdc3.13380

Funding Sources and Conflicts of Interest: The authors declare no conflict of interest relevant to this work and no additional disclosures to report.

Financial Disclosures for the Previous 12 Months: C.H., B. M., A.S., and D.Z. report no competing interests. ■

References

1. Kattah JC, McClelland C, Zee DS. Vertical nystagmus in Wernicke's encephalopathy: pathogenesis and role of central processing of information from the otoliths. *J Neurol* 2019;266(Suppl 1):139–145.
2. Helmchen C, Machner B, von der Gablentz J, Sprenger A, Zee DS. Downbeat nystagmus is abolished by alcohol in non-alcoholic Wernicke's Encephalopathy. *Neurol Clin Pract* 2021. https://doi.org/10.1212/CPJ.0000000000001138
3. Sprenger A, Hanssen H, Hagedorn I, et al. Eye movement deficits in X-linked dystonia-parkinsonism are related to striatal degeneration. *Parkinsonism Relat Disord* 2019;61:170–178.
4. Brodsky MC, Tusa RJ. Latent nystagmus: vestibular nystagmus with a twist. *Arch Ophthalmol* 2004;122(2):202–209.
5. Giolli RA, Blanks RH, Lui F. The accessory optic system: basic organization with an update on connectivity, neurochemistry, and function. *Prog Brain Res* 2006;151:407–440.