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Abstracts
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Akutes dorsales Mittelhirnsyndrom mit nicht nur typischen okulomotorischen Störungen: ungewöhnliche Skew deviation bei links mesodienzephaler Blutung

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Beschreibung eines dorsalen Mittelhirnsyndromes bei einem 77jährigen Mann mit links mesodienzephaler hypertensiver Blutung. Neben typischen okulomotorischen Befunden (Blickpräferenz nach unten, Lidretraktion, vertikale Blickparese nach oben und unten, Retraktionsnystagmus, Konvergenzschwäche, vorübergehend auch Konvergenzspasmus und Pseudoabduzensparese) bestanden als ungewöhnliche Zeichen eine bilaterale Miose, die links im Verlauf in ein Horner-Syndrom überging, und eine konstante Skew deviation mit Hypertropie des *rechten* Auges. Mögliche Erklärungen werden diskutiert.

Spatial errors in Gerstmann's syndrome: a case report

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Background: Gerstmann's syndrome consists of left-right disorientation, finger agnosia, agraphia and dyscalculia. It is virtually never pure and usually accompanied by some degree of aphasia.

Case report: An 85-year-old ambidextrous man showed the tetrad of Gerstmann's syndrome one month after a left inferior parietal stroke.

Main neurobehavioural findings were:

– Errors in finger naming and identification of fingers named by the examiner as well as autotopagnosia.
– Peripheral dysgraphia with frequently ill-formed, rotated or inversed letters and number contrasting with their normal visual recognition even when rotated. Impaired mental imagery in tasks of verbal description and kinesthetic recognition of letters and numbers.
– Impaired number processing (decimal transfers) with preserved table calculation and normal cognitive estimation of quantities.
– Impaired visual and motor imagery when rotation or left-right discrimination were required (recognition of the right/left hand presented in different postures, matching of rotated objects, identification of towns on inversed maps).

He had no aphasia, apraxia, sensorial or visuo-perceptual deficits. Reasoning and semantic knowledge were normal.

Conclusion: The patient presents a pure Gerstmann's syndrome without any aphasic disturbance. The syndrome appears to be based on a severe impairment in mental generation and manipulation of visual images.

Dynamic Bielschowsky test

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Positive Bielschowsky head-tilt test (BHT) is the cardinal finding for diagnosing unilateral trochlear nerve palsy (uTNP): tilting the head towards the side of the paretic eye leads to an increase in vertical and torsional deviation between the two eyes due to the lacking contribution of the superior oblique muscle to ocular counterrolling in the intorsional direction. In clinical use, this test is performed in a static fashion, comparing the deviation upon tilting the head to each side. We determined the kinematics of eye rotations in the presence of uTNP by dynamically performing BHT. Patients with uTNP (n = 8) and healthy subjects (n = 11) were asked to fix upon targets on a Hess screen, while they were rotated about the roll axis on a motorised turntable (± 35°, 0.3 Hz). 3-D eye movements were recorded with dual search coils. In the patients, the ocular rotation axis, which is normally close to the line of sight, was

tilted in the nasal (15.4° ± 8.6 SD) and downward (12.2° ± 4.3 SD) direction, but remained stable during head roll. In 6 of 8 patients the angle between the rotation axes of the two eyes did not change significantly with changing gaze directions. We conclude that in uTNP the rotation axes of both eyes are relatively comitant during dynamic torsional vestibular stimulation, despite the pathological orientation of the axis of the paretic eye. – Supported by *Swiss National Science Foundation (32-51938.97 SCORE A / 31-63465.00)* and *Betty and David Koetser Foundation for Brain Research*.

Head impulse testing with electrooculography

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The Halmagyi-Curthoys head impulse test recorded with search coils (one coil on one eye, the other on the forehead) is a very sensitive method to quantify peripheral vestibular function during high-acceleration stimuli. This technique, however, bears some disadvantages: it is semi-invasive (may irritate the conjunctiva), costly, and requires technical support by an engineer. Thus, for a broader clinical use, we have developed a non-invasive, low-cost method to record the horizontal head impulse test based on electro-oculography (EOG). – Binocular horizontal DC-amplified EOG is recorded from two silver-chloride electrodes placed on the outer canthi of both eyes. A low-weight laser attached on a bite bar serves to indirectly measure the amplitude of each head thrust. The subject's head, firmly held with both hands from behind by the experimenter, is first positioned in such a way that the head-fixed laser dot projected on a tangent screen overlaps with a space-fixed LED in the centre of a screen. After the experimenter triggers the trial with a foot panel, the laser goes off, and the subject fixes upon the LED straight ahead. Then, the LED is turned off and the head thrust is applied in total darkness. After the impulse, the subject sequentially fixes upon the space-fixed LED and the head-fixed laser dot. The amplitude of this saccade corresponds to the amplitude of the head impulse. – Eye and light signals are digitised at 1000 Hz per channel with 12-bit resolution and stored on a computer hard disk. Offline data processing (written in MATLAB) compares the eye movements evoked by the head impulses with the eye movements between the two lights after the impulses. – EOG head impulse testing provides a non-invasive, low-cost method to assess bilateral peripheral vestibular function. – Supported by *Swiss National Science Foundation (32-51938.97 SCORE A / 31-63465.00)* and *Betty and David Koetser Foundation for Brain Research*.

Gravity-dependence of downbeat nystagmus

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Downbeat nystagmus (DBN) is a frequent ocular motor sign in patients with lesions of the vestibulocerebellum. The upward drift in downbeat nystagmus is a combination of a gaze-evoked drift, due to an impaired vertical neural integrator (gaze-holding network), and a velocity bias. Using a three-dimensional motorised turntable, we analysed the influence of gravity on these two mechanisms. Patients with cerebellar downbeat nystagmus (n = 6) and healthy subjects (n = 12) were placed in different whole-body positions along various planes. Ocular drift was monitored with scleral search coils. While there was no gravity-dependence of the vertical gaze-evoked drift, the vertical velocity bias consisted of two components: (1) a gravity-dependent (GD) component that sinusoidally modulated as a function of body position along the pitch plane, and (2) a gravity-independent (GI) component that was always directed upward. The combination of the gravity-dependent and gravity-independent component led to an overall drift that was minimal in supine position and maximal in prone position. Our results suggest that the intact vestibulo-cerebellum minimises an overacting otolith-ocular reflex elicited by pitch-tilt and cancels an upward ocular drift that is independent of gravity-modulated otolith signals. – Supported by *Swiss National Science Foundation (32-51938.97 SCORE A / 31-63465.00)* and *Betty and David Koetser Foundation for Brain Research*.

Chronic meningitis caused by varicella zoster infection

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A 48-year-old female presented with chronic headache, progressing over a period of more than six

months. The analysis of the cerebrospinal fluid (CSF) revealed a pleocytosis of 300/µl, predominantly consisting of lymphocytes. The diagnosis of a chronic meningitis was made. There were no signs of an associated encephalopathy neither in the EEG nor in the MR-images. In a further step, a genome-specific sequence of varicella zoster was detected in the cerebrospinal fluid. Furthermore, virus-specific oligoclonal banding was found by immunoblotting technique, indicating antibody activity against both varicella zoster and herpes simplex virus, with an oligoclonal response before treatment with aciclovir and an increase in the number of bands thereafter. Our observation suggests a varicella zoster infection with coactivation of an immunological response against herpes simplex virus, making definite assessment of the causative agent difficult.

Cerebrotendinous xanthomatosis: case report of a predominantly spinal form

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Cerebrotendinous xanthomatosis (CTX) is an autosomal recessive disease, due to a deficiency of sterol 27-hydroxylase, a key enzyme in the synthesis of primary bile acids. In cerebrotendinous xanthomatosis, deficiency of bile acid synthesis leads to the storage of cholestanol and cholesterol in many tissues, especially in the lens, tendons and the CNS. Typical disease onset consists of premature bilateral cataracts and diarrhoea in childhood, followed by progressive cerebellar and pyramidal signs, dementia, seizures and tendon xanthomas. MRI scan often reveals abnormality of the dentate nuclei and surrounding cerebellar white matter. Biochemical diagnosis is made on the basis of bile alcohol excretion levels in urine and serum cholestanol levels. Early diagnosis of the disease is particularly important, as patients may benefit a lot from therapy with chenodeoxycholic acid and simvastatin.

We will report on a 50-year-old woman with progressive spastic paraparesis since the age of about 25, neuropsychological deficits and a history of chronic diarrhoea. MRI demonstrated marked white matter abnormalities in the lateral and dorsal columns of the spinal cord. The sister of the patient, who was misdiagnosed of suffering from multiple sclerosis, presented with a very similar history of gait disturbance beginning at early adulthood. Biochemical analysis confirmed the diagnosis of CTX in both patients.