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Abstracts
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Freie Mitteilungen SNG

Session: Donnerstag, 22.11.2007, 16.30–17.30 Uhr

Implicit motor sequence learning in Parkinson's disease patients with Levodopa-induced dyskinesia

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Evidence suggests that secondary plastic changes occur in Parkinson's disease (PD) in addition to the primary neurodegenerative processes. These changes may lead, among other functional disorders, to Levodopa-induced dyskinesia (LID). Studies in humans and animals have in particular revealed disturbed synaptic plasticity in LID.

Our hypothesis is that abnormal neuronal plasticity in PD patients with LID leads to impairments in motor learning.

Six dyskinetic and 9 non-dyskinetic PD patients under Levodopa therapy and 15 age-matched healthy subjects performed an implicit sequence learning task, where they had to press keys in response to the turning-on of lamps. Unbeknownst to the subjects, the lamps turned on in 2 different specific sequences.

We found a significant general learning effect ($p < 0.05$) in healthy subjects as well as in PD patients without LID, but not in patients with LID. Additionally, a calculated interference parameter, measuring the interaction between the two sequences, was significantly greater in PD patients with LID than in healthy controls ($p < 0.05$).

We conclude that the concurrent learning of similar motor tasks is impaired in patients with LID whereas patients without LID perform very similar to healthy subjects. These results might be due to secondary plastic changes in PD patients with LID which cannot be compensated by the application of Levodopa.

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IVIg-dose increase in multifocal motor neuropathy – a prospective six-month trial

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Background: IVIg is the only treatment option in multifocal motor neuropathy (MMN) based on RCTs, but the optimal IVIg dose remains unclear.

Methods: Nine patients with MMN on IVIg for at least 1 year and persistent but stable paresis and CB were selected. Dose was increased from a baseline of 0.5 g/kg per month [0.1–1.1], given at variable intervals [4–12 weeks], to 1.2 g/kg per month, given over 3 consecutive days, planned for 6 cycles. If patients' motor function did not improve after 2 cycles, they entered step two: dose was increased to 2 g/kg per month, given over 5 consecutive days.

Results: Following step one, 6 patients improved, 2 patients entered step two, 1 patient withdrew due to absent efficacy. Side effects (6 patients) were: fatigue (5), headache (4), nausea (2), hypertension (2), vertigo (1), abdominal pain (1); one serious adverse event was: infection of i.v. line with bacteriaemia. Study was stopped in one patient due to multiple side effects.

Conclusions: IVIg-dose increase may lead to clear improvement of motor functions in patients with stable MMN on long-term IVIg therapy. Improvement was independent from baseline IVIg dose, disease duration, presence of atrophy, number of proven CB and presence of EMG signs of axonal injury. With this small sample of patients there was no clear correlation between amount of dose increase and degree of improvement. Higher doses of IVIg caused more side effects, however, transient and usually not severe.

Copeptin, the C-terminal part of the vasopressin pro-hormone to predict outcome in patients with stroke

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Background: Prognostic factors to predict outcome in stroke patients would be helpful to guide decisions on treatment and interventions. Vasopressin (AVP) levels were found to be elevated in patients with stroke; however, its measurement is cumbersome. Copeptin is more stable, released in an aequimolar ratio with AVP and can be assayed readily in plasma.

Methods: From a prospective observational study data of the first 49 patients with an ischaemic stroke were analysed. On admission severity of stroke was assessed with the National Institute of Health Stroke Scale (NIHSS). In all patients copeptin levels were determined with a sandwich immunoassay and compared with functional outcome after 3 months as assessed with the modified Ranking score (mRS).

Results: 20 (41%) patients were female and 29 (59%) male. The median age was 73 years. On admission the median NIHSS was 6 (1–46). NIHSS and copeptin levels correlated with the mRS at 3 months ($r = 0.59$ and 0.46; $p < 0.001$). Copeptin levels on admission were higher in patients with a worse outcome (mRS ≥ 3) compared to patients with a good outcome (mRS < 3, $p = 0.03$). For the outcome "death", copeptin had an area under the receiver operating curve (0.86, CI 0.73–0.94) within the range of the NIHSS (0.99; CI 0.85 to 0.99).

Conclusion: Copeptin levels measured on admission were associated with poor outcome after 3 months. Thus, our pilot data indicate

that copeptin is a novel predictor of outcome in patients with ischaemic stroke

Sleepiness is not always perceived prior to falling asleep in healthy sleep-deprived subjects and sleepy patients

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Objective: We prospectively evaluated the subjective awareness of sleepiness (SubS) prior to sleep onset during MWT in young healthy sleep-deprived subjects and in sleepy patients.

Method: 159 patients (mean age 39.8 years; 59 females) with sleepiness of various origin and 28 young healthy students (mean age 22.4 years; 13 females) after a whole night sleep deprivation underwent 4 MWTs. They received the instruction: "Indicate your earliest symptoms of sleepiness and try to stay awake as long as possible!" Overt sleep (OS) and microsleeps (MS) of at least 3-second duration were scored separately.

Results: Overall 17 of 28 healthy subjects (60.7%) and 64 of 159 patients (40.3%) presented either an MS- or an OS-fragment before indicating SubS at least in one of 4 MWT-trials. In both healthy subjects and patients females demonstrated a better awareness of SubS than male subjects.

Conclusion: Our unexpected finding is in sharp contrast to the general assumption that nobody can fall asleep without prior awareness of sleepiness while driving. If the results will be confirmed in larger series, far-reaching consequences will ensue. (1) The simple advice to sleepy subjects that they should not drive when sleepy would no longer be adequate. (2) Motor vehicle crashes due to microsleeps could no longer be judged as due to "reckless driving" in all cases. (3) Prevention strategies against sleepiness-induced motor vehicle crashes would have to include efforts to improve perception of SubS.

Freie Mitteilungen SNG

Session: Freitag, 23.11.2007, 14.45–15.15 Uhr

ERP correlates of word production before and after stroke in an aphasic patient

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We analysed the electrophysiological correlates of severe anomia in an aphasic patient (C.W.) in comparison to a control group and himself, since he had been recorded with the same task as a control subject one year before suffering a haemorrhagic stroke.

EEG was recorded during a picture-naming task with 128 channels covering the entire scalp one year before stroke (as a control subject) and at 3, 4 and 5 months post-stroke. Two other subjects from the control group were recorded again one year later with the same task in order to ensure that the observed changes are not due to intra-subject variability. No changes were observed in the follow-up recording of these two control subjects.

Behavioural results showed severe anomia in C.W. after stroke, stable across the three recording sessions. Waveform analysis and temporal segmentation were carried out on the patient's data in comparison to the control group and his own data before stroke. The analyses revealed normal amplitudes and topographic maps from 0 (picture on screen) to about 270 ms. Abnormal amplitudes and topographic maps appeared after 270 ms. The same abnormal topographic map lasting over 200 ms was identified across the three post-stroke recording periods. The stability of this abnormal pattern seems to indicate a fixation on a cortical activation leading to inefficient word retrieval.

Abnormal cortical network activation in amnesia

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Amnesic patients show reduced variation of evoked potential amplitudes over single electrodes between 300 and 600 ms when processing new, as opposed to familiar information. Whether this abnormality reflects reduced modulation of similar cortical networks or activation of different networks, is unknown. In this study we used electrical spatiotemporal mapping to explore cortical network activation during memory processing in a group of amnesic patients. High-density evoked potentials were recorded as 11 amnesic patients with various aetiologies and 12 control subjects performed a continuous recognition task composed of meaningful designs. Presentation of pictures induced precisely the same electrocortical map configurations in amnesics and controls during the first 200 ms. Beyond this period processing differed significantly. Between 200 and 350 ms amnesic patients expressed different topographical maps from controls. Maps specific to amnesic patients correlated with the visual P1 component. From 350 to 550 ms presentation of repeated items induced modulation of the same cortical maps as new items in controls. In amnesics, by contrast, it induced different electrocortical maps, indicating distinct cortical processing of new and old information. The study shows that cortical mechanisms underlying memory formation and re-activation in amnesia basically differ from normal memory processing.

Poster SNG

Session: Freitag, 23.11.2007, 13.30–14.00 Uhr

P 01 Gait parameters during walking with a four-point cane, simple cane and Nordic walking stick in patients with post-stroke hemiparesis

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Purpose: To examine the effects of 3 walking aids on hemiparetic patients' gait parameters; to assess patient satisfaction with each type of cane.

Relevance: Aids improve gait stability but their effect on hemiparetic patients' gait is rarely studied.

Participants: 21 patients, suffering a first stroke, with no prior walking difficulties, able to walk 10 metres holding a handrail, unfamiliar with any of the aids tested.

Method: After a clinical assessment patients were tested on a GAITRite® walkway system and with a 6-minute walking test over three consecutive days, each day using a different aid (in randomised order). Patients subsequently rated (VAS) each aid.

Analyses: Friedman/Wilcoxon test.

Results: Patients walked further ($p = 0.048$), faster ($p = 0.033$) and with less left/right step time differential ($p = 0.005$) when using a simple cane with ergonomic handgrip (SCEH) than with a four-point cane (FPC). They also walked significantly further ($p = 0.003$) and relatively faster when using the SCEH compared to a Nordic walking stick (NWS). Patients' found both the SCEH and the FPC to be significantly more helpful than the NWS ($p = 0.0004$; $p = 0.01$ respectively).

Conclusion: In Switzerland the SCEH, FPC and NWS are the most commonly used aids in hemiparetic patients' gait rehabilitation. We demonstrated the advantages of using a SCEH and showed that both patients' subjective preference and objective criteria should be considered when choosing a walking aid.

P 02 Complications neurologiques après chirurgie de by-pass gastrique pour obésité morbide

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Arrière-plan: Les traitements chirurgicaux de l'obésité peuvent mener à de graves complications neurologiques.

Cas cliniques: Nous rapportons l'histoire de 3 patients hospitalisés en janvier 2006 dans notre service de neuroréadaptation, à la suite de 3 complications après by-pass gastrique (BPG).

Madame S.G., 25 ans, bénéficie d'un BPG en novembre 2005. Elle développe une encé-

phalopathie de Gayet-Wernicke. On constate aussi une polyneuropathie axone. L'évolution est lentement favorable et une réinsertion professionnelle dans une activité adaptée est planifiée.

Madame S.M., 29 ans, a bénéficié d'un BPG en 2005. Elle développe un arr√™t cardio-respiratoire sur dysfonction ventriculaire gauche (FE 25%) avec encéphalopathie post-anoxique diffuse sévère avec syndrome de Balint. La patiente reste partiellement dépendante.

Monsieur F.J., 30 ans, a bénéficié d'un BPG en mai 2005. Il développe une neuropathie axonale sensitivomotrice sévère, reliée à des carences multiples (acute post-gastric reduction surgery). Une myélopathie sur carence en cuivre est aussi découverte. L'évolution est partiellement favorable, avec l'utilisation de moyens auxiliaires pour la marche.

Conclusion: Le BPG concerne des patients jeunes et peut conduire à des complications neurologiques dévastatrices. La plupart de ces troubles suivent des carences, il y a donc un potentiel de prévention. Un suivi régulier auprès de centres spécialisés est nécessaire.

P 03 Asymmetric POSTS associated with ipsilateral EEG abnormalities

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Background: Positive occipital sharp transients of sleep (POSTS) are considered a normal EEG variant. They appear as positive waves in posterior regions during non-REM sleep and are found in 50–80% of recordings. There are rare reports on pathological conditions associated with POSTS (syncope, headaches, behavioural disorders); however, no relationship with definite electrographic abnormalities has been described. We here present a series of consecutive patients with strictly unilateral POSTS, associated with ipsilateral electrographic abnormalities.

Patients' descriptions: Over 30 weeks 4 female and one male subject (aged 7–76) were identified with POSTS restricted to the right, representing 0.4% of performed EEGs (5/1130). Their recordings also showed ipsilateral abnormalities (sharp waves, rhythmic theta activity, postictal slowing). All patients were diagnosed with partial epileptic seizures or epilepsy (cryptogenic or symptomatic).

Conclusion: The fact that POSTS were clearly asymmetric and found only on the same side as the abnormalities raises the question whether they should always be considered physiological or could, rarely, be interpreted as a marker of underlying increased cortical excitability. The right-sided location in all patients might be a mere coincidence or suggest that the non-dominant hemisphere could be more likely to produce POSTS associated to abnormal EEG findings. Larger observations are needed to confirm our preliminary findings.

P 04 Hereditary spastic paraplegia, molecular and clinical correlation in Swiss and Chinese families

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An increasing number of mutations in genes causing different forms of hereditary spastic paraplegia (HSP) have been published in the last years. Here we report on our recent findings in two cohorts.

A large group of families with autosomal dominant pure HSP in central Switzerland has been followed up and we have found one single mutation leading to skipping of spastin exon 16. A large variability in the age at onset and in the course of the disease has been found. In a Chinese cohort we have found two families with a complex form of the disease inherited in an autosomal recessive way. The patients had a slowly progressive spastic paraparesis with mental deterioration and a thin corpus callosum. No mutation was found in the spastin gene, however, two new mutations were found in the spatacsin gene, both leading to a stop codon. Furthermore, diffusion tensor imaging was used to determine additional tract involvement. Significant mean diffusion (MD) increase and fractional anisotropy (FA) reduction were found in the thin corpus callosum. However, significant MD changes where also found in the thalamus and subcortical hemispheric white matter.

These results confirm the wide clinical and molecular spectrum found, and point at the importance of including HSP in the differential diagnosis of spasticity. Furthermore, molecular data now allow a good understanding of HSP pathophysiology and may also shed light on the mechanisms involved in spasticity due to more frequent causes.

P 05 Indirect pathway and early compensatory mechanisms in Parkinson's disease

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The main neuropathological feature of Parkinson's disease (PD) is a degeneration of dopaminergic cells in the substantia nigra (SN), leading to an imbalance between the direct and indirect pathways in the basal ganglia. Enkephalinergic/GABAergic neurons belong to the indirect pathway and an upregulation of striatal enkephalin (ENK) mRNA has been described in PD. Animal models of PD show a partial behavioural recovery, even after an almost complete dopamine depletion. This could be explained by the existence of early compensatory mechanisms. Here, we investigated the role of the indirect pathway in the early post-surgical phase.

We used the unilateral 6-hydroxydopamine (6-OHDA) rat model of PD and control animals. We measured the spontaneous locomotor activity, using the video-tracking system Ethovision (Noldus, NL), and performed in-situ-hybridisation histochemistry with DNA oligoprobes for ENK mRNA in the striatum.

We observed, in parkinsonian rats, that a high ipsilateral ENK mRNA expression correlated with a higher locomotor activity (lower bradykinesia). In contrast, this correlation was not found in the control group. These results are in contradiction with the classical model of basal ganglia function which proposes that an increase in indirect pathways activity leads to bradykinesia.

In conclusion, our results suggest that ENK increase in the indirect pathway may be linked with early compensatory mechanisms leading to partial behavioural recovery in Parkinson's disease.

P 06 Right ventricular arrhythmic cardiomyopathy with an autosomal dominant R355P DES gene mutation

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Objective: To describe a peculiar cardiac involvement in a Swiss family with an autosomal dominant DES mutation.

Methods: Our index patient, a 29-year-old man, was investigated for a progressive limb weakness of a 5-year duration. He was still hardly ambulant. His paternal grandmother walked with 2 canes since her fifties but died at 80 years of age. His father died at 42 years and his aunt died at 55 years of age from a dilated heart and had dysphagia.

Results: On clinical examination a severe four-limb proximodistal weakness was found, greater in the lower limbs. Serum CK level was elevated 10-fold. A right ventricular arrhythmic cardiopathy was found and on cardiac MRI evidence of fibrofatty degeneration in the left ventricular posterior wall was observed. The four family members had a muscle biopsy that revealed a dystrophic pattern and cyptoplasmic spheroid-like inclusions. A heterozygous point mutation c.1064G>C (p.R355P) in exon 6 of the desmin gene (DES) was found. The mutation causes an amino-acid exchange from arginine to proline and is located in the 2B subdomain of desmin, a hotspot for mutations.

Discussion/conclusion: In this family 2 points could be underlined: (1) there was a severe skeletal and rapidly progressive muscle weakness, (2) the type of cardiomyopathy of our index patient has not been described yet, although the R355P mutation of the DES gene has already been described.

P 07 Familial late-onset inclusion body myositis in the search of a gene

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We describe an inclusion-body-myositis (IBM) Swiss family with autosomal dominant inheritance, in whom no mutation in GNE and VCP genes was found.

Typically, hIBM is a recessively inherited rimmed-vacuole myopathy sparing the quadriceps and caused by mutations in the GNE gene. Sporadic IBM (sIBM) is an inflammatory myopathy; in a small number of families with late onset IBM is associated with Paget's disease and FT dementia due to mutations in the protein p97/VCP.

Methods: We studied the family members of a 75-year-old man, who was investigated for a late-onset progressive difficulty in climbing stairs; as the whole family had fallen out, sIBM was the initial diagnosis based on history, clinical examination and muscle biopsy. During the following months we saw 2 ladies with the same family name and found out they were sisters of our index patient. All 3 had a late and insidious onset.

Results: Clinical examination of these 3 still ambulant patients revealed the same proximo-distal muscle weakness, particularly of the hip flexors, leg extensors and adductors; finger flexors VI and V were slightly weak in the upper limbs. CK were normal and muscle MRI demonstrated a fatty involvement. Muscle biopsies demonstrated rimmed vacuoles and endomyseal lymphocytic inflammatory infiltrates. No mutation in the GNE or VCP genes was found. We conclude that further investigations would be worth performing to find out the causative gene defect in this familial IBM.

P 08 Bilateral paramedian artery stroke presenting as an amaurosis fugax

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Bilateral paramedian thalamic infarctions account for ⅓ of the paramedian infarcts and are associated with impaired consciousness, oculomotor and neuropsychological disturbances. A 44-year-old healthy woman suffered from a brief loss of vision of the right eye. Amaurosis fugax and an embolic origin were suspected. Neurological examination and 3-lead ECG in the acute stage were normal. CT of the brain was considered as normal. DWI-MRI showed bithalamic paramedian ischaemic lesions. Oculomotricity, bilateral visual acuity and comprehensive neuropsychological examinations were normal. MR angiography and neurosonology showed normal extra- and intracranial vessels. However, neurosonology and transoesophageal echocardiography revealed a large right-to-left cardiac shunt due to an atrial communication. 24-hour cardiac monitoring was normal but she underwent prior to an eventual closure of the cardiac defect an ambulatory R-test which revealed several paroxystic short-lasting passages into atrial fibrillation. She was anticoagulated and remained free of symptoms. The interest in this case is threefold: (1) bilateral paramedian infarction which usually presents with a devastating clinical picture may occur clinically silent; (2) amaurosis fugax which is usually associated with ipsilateral carotid disease may be the consequence of cardiac embolism, and (3) atrial fibrillation is never completely ruled out and consequently can prevent closure of a possibly asymptomatic right-to-left cardiac shunt.

P 09 Measurable balance impairment in minimally affected multiple sclerosis patients

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Background: Impaired balance is present in the majority of patients with multiple sclerosis (MS) throughout the disease course. However, neurological examination of mildly affected patients complaining of poor balance often does not show clinical signs of imbalance.

Aim: To evaluate postural stability in MS patients with and without subjective balance impairment.

Methods: MS patients without overt signs of imbalance or gait ataxia were examined. For clinical balance testing we used accepted clinical tests and for laboratory testing the SwayStar™ system.

Results: We examined 25 patients. Ten of them (40%) complained of impaired balance (group 1), but only 2 of them had pathological signs during clinical testing. In the group without symptoms of impaired balance (group 2) (n = 15) 2 patients had pathological signs during clinical testing. After excluding all patients with pathological clinical measurements, patients were assessed using the SwayStar™ system. Group 1 (n = 8, mean age 42.2 years, mean EDSS 2.2) performed significantly worse than group 2 (n = 13, mean age 38.9 years, mean EDSS 1.8) during laboratory testing.

Conclusion: Mildly affected MS patients with subjective symptoms of impaired balance and normal findings on clinical examination show a specific impairment of balance when examined with the SwayStar™ system. Clinical assessment of mildly affected MS patients complaining of impaired balance is inadequate. Our study shows that more refined tools are needed.

P 10 Amplitude asymmetry of the occipital EEG alpha background may be determined by structural features (cerebral torque), not by hemispheric dominance

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In routine EEG a moderate asymmetry of the normal occipital alpha background activity is often seen, usually with higher amplitudes on the right side. It is commonly assumed that the greater amplitudes are found on the non-dominant hemisphere, usually the right one in right-handers, although there is no convincing explanation for this. In most subjects a structural hemispheric asymmetry of the occipital lobe, which is termed "cerebral torque", can be found on neuroimaging. This asymmetry means that the occipital end of the interhemispheric fissure (occipital falx) points to the right side.

The occipital alpha activity is blocked by visual input and generated by the primary visual cortex. This calcarine area is not located at the surface of the convexity but mesial, adjacent to the interhemispheric fissure. Therefore the asymmetry of interhemispheric fissure would explain the amplitude asymmetry of alpha background activity.

In a few subjects, showing a reversed EEG asymmetry with an alpha-amplitude predominance on the left side, we also found a reversed structural asymmetry with the interhemispheric fissure pointing to the left side.

We conclude that the amplitude asymmetry of the occipital EEG alpha background may be determined by structural features (brain torque), not by hemispheric dominance.

P 11 Very late onset Friedreich's ataxia with disturbing head tremor and without spinal atrophy – a case report

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Friedreich's ataxia is the most common inherited recessive ataxia, caused by expansion of a GAA trinucleotid repeat in the frataxin gene. Atypical late onset forms with milder symptoms are linked with a smaller size of expanded trinucleotid sequences. Here we report a further case of a very late onset Friedreich's ataxia.

A female patient showed first symptoms of mild gait ataxia and head tremor at the age of 64. In the following years the head tremor worsened to be quite disturbing, whereas the gait disorder remained mild. At the age of 73, additional signs were dysarthrophonia and oculomotor abnormalities. The diagnosis of Friedreich's ataxia was confirmed by genetic testing with a short GAA expansion of about 95–105 repeats on the smaller allele and

about 360 on the longer allele. The spinal MRI showed no cervical cord atrophy.

In conclusion, our data show that Friedreich's ataxia should be considered in the differential diagnosis of cerebellar ataxia also in older patients.

P 12 Rehabilitation der Retentionsblase mittels intravesikaler Elektrostimulation (IVES) bei neurologischen Patienten

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Anhand einer retrospektiven Auswertung von 36 neurologischen Patienten mit Restharnblase im Jahr 2006 konnte gezeigt werden, dass IVES eine gut geeignete und sichere Methode ist, auch bei neurologischen Patienten eine Retentionsblase zu beseitigen und zu einem erheblichen Teil Dauerkatheter (DK) und intermittierende Einmalkatheterisierung (IEK) zu erübrigen.

P 13 An unusual late recovery from locked-in syndrome after basilar artery occlusion

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A patient with locked-in syndrome (LIS) is conscious but "closed" inside her or his body because of a pontic insult resulting in a quadriplegia and lower nerves' paralysis with anarthria. The classical picture of LIS includes preservation of vertical gaze and upper eyelid movement allowing communication through an eyelid opening-closing code. The most common aetiology of LIS is basilar artery occlusion. LIS has generally been associated with high mortality and poor recovery and is a challenge for the rehabilitation team. Casanova et al., 2003, report only 21% of significant motor recovery.

We report a 68-year-old man with complete locked-in syndrome after basilar artery occlusion. MRI confirmed destruction of pons. The patient has flaccid quadriplegia, lower nerves' paralysis with anarthria and swallowing incapacity requiring tracheotomy and gastrostomy. Intestinal paralysis requires discharge colostomy.

The recovery took time, was late, but despite the severity of the initial clinical state and the extent of the lesion was unusually good with, 17 months after the bilateral pontic infarct, verbal communication, swallowing recovery, bladder and bowel control, functional right upper limb and ability to walk.

P 14 Ictal bradycardia and asystole: an uncommon but life-threatening cause of syncope

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Although seizures often produce consciousness impairment, seizure-induced syncopes are extremely rare. We present two patients with ictal bradyarrhythmias elicited by seizures.

Patients' descriptions: A 46-year-old man with repetitive "drop attacks" for 5 years, preceded by prodromal stereotypical feelings of fear, was admitted following a new episode. The EEG captured a typical episode: a seizure arising in the left frontotemporal region, followed by asystole with transitory EEG attenuation. He underwent a pacemaker implantation and received valproate. After six months he only reported one prodromal episode.

A 75-year-old man had several unexplained syncopes for many years, heralded by ascending gastric discomfort. EEG disclosed a left frontotemporal seizure leading to a severe bradycardia (15/min), inducing a presyncopal state. He was started on valproate and a pacemaker was implanted; at 18 months he remained asymptomatic.

Comment: Ictal bradyarrhythmias represent a rare phenomenon (about 2% of partial seizures), but may be potentially lethal, as discussed in cases of sudden unexpected death in epilepsy (SUDEP). Seizures from the mesiotemporal region spreading contralaterally seem to induce a paroxysmal dysfunction of limbic autonomic centres. Investigation of syncope preceded by stereotypical prodromes should therefore include long-term EEG. The treatment is double-headed, including an antiepileptic drug without cardiac arrhythmic effects and pacemaker implantation.

P 15 Status epilepticus prognostic score: multicentric validation and application

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Background: It is unclear how the potential risk of an aggressive treatment of status epilepticus (SE) balances with outcome improvement. We have recently developed a prognostic score (Neurology 2006;66:1736–8), relying on age, previous seizures, seizure type and consciousness impairment. The aim of this study was to validate the score and to analyse its potential impact on treatment choice, particularly coma induction (CInd).

Methods: Consecutive adults with SE were prospectively studied in three university hospitals: in Switzerland (CHUV) and in Massachusetts (BWH and MGH). The score was calculated before outcome assessment (at discharge). Prediction for survival was assessed with the negative predictive value (NPV) and effect of CInd on survival using a propensity-based matching.

Results: 23% of 135 patients died; the score had a sensitivity of 94%, specificity of 58% and NPV of 97%. Return to baseline clinical conditions was higher for subjects with a favourable score ($p = 0.001$). CInd was more frequent at BWH ($p = 0.003$), but other variables were similar among the centres. Propensity-based estimation of CInd effect on survival was non-significant (95% CI: –0.335 to 0.111).

Conclusion: The SE clinical prognostic score reliably identifies patients who will survive and is simple to use in practice. Our observations also suggest that aggressive treatment (i.e. CInd) is not routinely warranted in patients with a favourable SE score, who will almost certainly survive their SE episode.

P 16 Reversed flow in the external carotid artery despite ipsilateral patent common carotid artery as diagnosed with colour-coded duplex flow imaging

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Occlusion of the common carotid artery (CCA) is generally associated with occlusion of the ipsilateral internal carotid artery (ICA) and external carotid artery (ECA). However, collateral circulation to the ECA may occasionally preserve patency of the ICA via retrograde perfusion through the bulb. We report a case of reversed flow in the ECA in the presence of a patent CCA as diagnosed with colour-coded duplex flow imaging (CDFI). An 85-year-old man presented with acute dysarthria associated to left hemiparesis. Cerebral CT scan and 3-lead ECG performed in the acute stage were normal but MRI showed an acute ischaemic lesion in the territory of the perforators of the superficial branches of the right middle cerebral artery. CT and MR angiography showed patent ICA and ECA but no CCA on the right side and an occlusion was suspected. At the same time CDFI demonstrated patency of the right CCA showing high-resistance flow pattern due to a large partially calcified atheromatous plaque arising from the carotid bifurcation into the proximal ICA where it determined a stenosis >70% NASCET. Surprisingly, flow in the ipsilateral ECA was reversed and supplying, through the stenosed carotid bifurcation, the ICA which was dampened. Ipsilateral ophthalmic artery was orthograde. This case demonstrates that collateralisation with reversal flow in the ECA can occur in a patent CCA when associated to a stenosis

of the bifurcation and that CDFI stays the method of choice to detect such haemodynamic conditions.

P 17 Faecal incontinence in Guillain-Barré syndrome (GBS): a case report

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Faecal incontinence is no usual symptom of Guillain-Barré syndrome (GBS). We report an 81-year-old woman presenting ascending flaccid tetraplegia, sensitive and respiratory impairment, cardiac arrhythmia and – most notably – urinary retention and anal incontinence 5 days after onset of severe diarrhoea.

Electroneuromyographic findings showed demyelinating lesions in the upper extremities, axonal lesions in the lower extremities. Cerebrospinal fluid (CSF) was normal on the fifth day. Anti-GM1 and anti-GD1b antibodies were elevated. MRI of the spinal cord was normal. The patient received intravenous immunoglobulins for 5 days, followed by 5 plasmapheresis sessions. Perineal exam revealed hypoaesthesia, areflexia and hypotonia. Cystomanometry showed hypoaesthesia and bladder hypocontractility, findings typical of peripheral bladder paralysis. Anal incontinence and urinary retention recovered by the third month. Walking capacity recovered within 5 months.

Whereas urinary dysfunction with favourable evolution has repeatedly been reported in connection with GBS, faecal incontinence is highly unusual. This case demonstrates that anal incontinence may be a feature of GBS.

P 18 Successful treatment of medically refractory spasticity in upper motor neuron predominant amyotrophic lateral sclerosis (ALS) with intrathecal baclofen – a case report

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Objective: To report on beneficial therapy with an intrathecal baclofen pump in an ALS patient.

Case report: A 66-year-old male was diagnosed with probable laboratory-supported ALS according to Airlie House criteria in September 04. Disease onset was bulbar with dysarthria, followed by increasing tetraspasticity predominantly in the lower limbs, associated with bilateral myoclonus of the gastrocnemius muscles. Baclofen orally up to 150 mg/d and tizanidin up to 24 mg/d were administered with no benefit. A 10-day treatment with intrathecal baclofen extern pump system started. Ability to walk 10 m with one crutch. Baclofen i.t. was started with 24 µg/d. Daily evaluation by Ashworth scale, timed 10 m walk and video.

Results: Around 43.5 µg/d baclofen, spasticity in leg flexors declined from AS IV to II, walking time / 10 m shortened from 37.5 to 20 seconds. After definitive pump implantation, dosages up to 50 µg/d. At discharge the patient was able to walk 70 m with rollator and could climb 10 steps on a stairway.

Conclusions: I.t. application of baclofen might be an option in ALS patients with predominantly upper motor neuron signs and unsatisfying orally medication. Mobility, activities of daily living and quality of life can improve in ALS under i.t. baclofen therapy, as shown here.

P 19 Enterale Langzeiternährung und Stoffwechselkontrolle bei Patienten mit zerebral bedingten Schluckstörungen und insulinpflichtigem Diabetes mellitus Typ 2 mit einer fettmodifizierten Spezialnahrung, reich an einfach ungesättigten Fettsäuren, Fischöl, Chrom und Antioxidantien

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Fragestellung: Ergebnisse von Stufe I einer zweistufig geplanten Studie (Design nach Bauer & Koehne) an Typ-2-Diabetikern mit zerebral bedingten Schluckstörungen gaben Hinweis auf eine bessere Diabetesbehandlung mit einer enteralen fettmodifizierten Spezialnahrung (Diben, Fresenius Kabi) verglichen mit einer Standardnahrung (Pohl et al., Eur J Clin Nutr 2005;59:1221–32). Stufe II der Studie wurde mit reduzierter Zahl an Primärvariablen und angepasster Patientenzahl durchgeführt.

Methoden: Randomisierte, doppelblinde, kontrollierte multizentrische Studie. Insulinbehandelte Typ-2-Diabetiker [HbA_{1c} ≥7,0% und/oder Nüchternblutzucker (NBZ) >120 mg/dl] mit Indikation zu einer Sondenernährung erhielten bis zu 84 Tage entweder 27 kcal/kg KG/d (max. 2025 kcal/d) der Spezialnahrung (Testgruppe T) oder eine isoenergetische, isonitrogene enterale Standardnahrung (Kontrollgruppe K). Der gesamte Insulinbedarf, Nüchternblutzucker und der Blutzuckerspiegel (BZ) am Nachmittag wurden täglich, HbA_{1c} und weitere Variablen an den Tagen 1, 28, 56 und 84 gemessen. Die Ergebnisse (Differenz zum Basiswert) sind als Effektgrössen des univariaten Wilcoxon-Mann-Whitney Tests (MW) dargestellt.

Ergebnisse: 105 Patienten wurden in Stufe II der Studie aufgenommen (53T/52K). 55 Patienten (33T/22K) nahmen bis Tag 84 an der Studie teil. Die Daten zeigten nach 84 Tagen Veränderungen (Medianwerte) des täglichen Insulinbedarfs von –8,0 vs. +2,0 U (MW = 0,8326; p <0,0001) und des NBZ von –39,0 vs. –12,1 mg/dl (MW = 0,8030; p <0,0001) (T vs. K). Die Effektgrössen zeigen eine grosse Überlegenheit der Testgruppe für beide Variablen und die Kombination der Wirksamkeitsparameter (Tage 56/84; MW = 0,7899; p <0,0001). Für HbA_{1c} an Tag 84 wurde für die Testgruppe eine relevante sowie für den BZ am Nachmittag der Tage 56 und 84 eine grosse Überlegenheit gezeigt. Die Anzahl der relevanten Hypoglykämien (NBZ <60 mg/dl) war 1 (Testgruppe) vs. 5 (Kontrollgruppe), ein Hinweis auf einen günstigen Effekt der Testnahrung.

Schlussfolgerung: Die Ergebnisse der formalen Kombination von Stufe I und II dieser Studie zeigen im Vergleich zur Standardnahrung einen statistisch signifikanten positiven Effekt der Spezialnahrung auf den täglichen Insulinbedarf und den Nüchternblutzucker. Bei Patienten mit zerebral bedingten Schluckstörungen und einem Typ-2-Diabetes wird mit dieser Spezialnahrung die Diabetesbehandlung verbessert. Die Nahrung erwies sich als gut verträglich.

P 20 Spinal cord syndrome and optic neuritis after FSME vaccination

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We describe a 41-year-old patient showing a reduced general condition with normal vital signs but an incomplete sensorimotor spinal cord syndrome at level Th 9/10 accompanied by urinary retention, spastic paralysis, gait disturbance and pyramidal tract signs. Three weeks earlier the first FSME (tick-borne encephalitis) vaccination had been administered.

Cerebrospinal fluid analysis revealed a lymphocytic pleocytosis with 84 cells/µl, elevated protein and lactate. Magnetic resonance imaging of the brain and spinal cord and routine blood examination were normal. Immunological investigation revealed increased FSME IgG titres and borderline values of FSME IgM. Electrophysiologic examination showed pathologic visual evoked potentials on both sides.

In the absence of viral or bacterial infections we supposed a paravaccinal myelitis due to FSME vaccination and started therapy with i.v. high-dose corticosteroids resulting in partial remission of the spinal cord syndrome except for a urinary retention. The patient presented here is one of the rare cases of paravaccinal myelitis and optic neuritis in adults after FSME vaccination.

P 21 Don't know was it means, don't know whether it exists, but I know it is in English (or French)

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Pure alexia is a reading deficit following occipitotemporal lesions. In the most severe cases the capacity to read isolated letters is lost. Most patients have residual implicit reading abilities, testimonied by the preserved capacities to make lexical and semantic decision tasks with quickly presented stimuli. Here we describe a patient with pure alexia who showed striking preservation of the ability to identify the language in which words were written while he performed at chance in tasks usually preserved in pure alexic patients. To our knowledge, this residual visual word processing ability in pure alexic patients has only been mentioned by Cohen and Dehaene (2000) in patient VOL with pure alexia.

Reading abilities were assessed by using several tasks including lexical, semantic, emotional decision and language decision on words, sentences and texts. The patient was able to decide the language (French or English) in which a shortly (400 ms) visually presented word was written, even if he made close to 93% errors in reading. Reaction times in reading were slow (mean reaction time, 2300 ms for correct answers). In lexical decision (400 ms) the patient performed above 90% when pseudowords were orthographically illegal; but 46% errors when the pseudowords were orthographically legal. When asked to make a semantic and an emotional decision on rapidly presented words, A. N. responded at chance level. In language decision on individual words he made 80% correct and his mean reaction time ranged from 950 to 2300 ms. Performance was neither influenced by word frequency nor by word length. Language recognition was influenced by orthographic regularities as A. N. performed 54% correct when words were orthographically legal French and English words. For sentences and texts, performance was not influenced by syntactic legality.

Together, these results indicate that A. N.'s residual ability in language decision was most likely due to a preserved ability to extract and recognise combinations of letters, specific to a given language.

Neurologist-in-training

The aim of this section is to prepare the neurologist-in-training for the FMH examination, to confront her or him with specific problems of everyday neurological practice and to give him or her updates on recent controversies in clinical neurology.

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Case vignette

The 54-year-old right-handed male patient originated from Malaysia but had been working for many years in Canada as a professor of technical sciences. Currently, he made a tour throughout Europe after a one-year sabbatical in Germany. At the evening before the admission his wife noticed that he could not memorise new information and repetitively asked the same questions. The neurological examination on the next morning revealed a reduced digit span (0), loss of anterograde memory and impairment of retrograde memory covering several years. There were no other neurological deficits, physical examination and routine laboratory work-up (including white and red cell count, CRP, TSH, liver and kidney parameters) revealed no abnormalities. The resident in the emergency room suspected a transient global amnesia (TGA) and proposed a clinical follow-up in 24 hours without further examinations.

Question 1

Which clinical finding indicates that this initial appraisal might not be correct?