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1 Scope for EEG post-processing – S.E. Petranek, V. Krajca, I. Patakova and J. Faber (Prague)

Some examples are presented of EEG data compression using a self-learning neuronal network. Even with saving the table of coefficients, such data can be compressed at a ratio of 1:3 without any substantial information loss, saving two-thirds of the recording medium. The time loss is practically nil – the data are compressed while being stored from the hard disk. The method can find good use in large laboratories where, considering the thousands of recordings annually, the financial effect is well worth the effort. Examples are also given of processing 13 min long full records (including 4 min hyperventilation, post-HV state, twice with eyes open and closed). The need is stressed to perform brain mapping as relative maps, each time with both basic reference electrodes (A1 + A2) and/or AVG, with the following as special cases: one map in the alpha band with a maximum up to the region of electrodes C3 and C4, for which the authors use the term 'MU type alpha' (typical of children's graphs), and the other with a maximum near Cz in the theta band referred to as 'sleep-type theta' (in adults with frequent NREM 1). Besides the conventional norm, no other typical picture was found in healthy controls. Finally, the authors present whole-record brain mapping processed with Laplace transformation in the form of absolute and relative maps. Despite their relatively low resistance to artifacts, the maps are of particular use to EEG beginners.

2 Coherence in clinical practice – S.E. Petranek, V. Krajca and I. Patakova (Prague)

The authors routinely apply two types of coherence to most EEG records, each of 13 min duration with 4 min hyperventilation, 3 min after HV, and twice with eyes open and closed, using the 10–20 system of electrodes free from major electrode artifacts. Interhemispheral coherences are calculated symmetrically relative to central electrodes Fz, Cz, Pz with the resulting values shown as brain maps in the usual bands. The resulting information corresponds to visual estimation of medial synchrony and lateral asynchrony. However, since there is no phase component, these maps are useful for determining the degree of maturation of children and adolescents under longitudinal follow-up. In adult patients, the uses are limited by the duration of vigilance and/or sleep variations in the course of recording. Local coherences are calculated from the same graphs between couples of electrodes arrayed longitudinally and transversely. The result is also placed in between the electrodes and processed as brain maps. Normal graphs show no major changes in any of the bands. However, local coherences show a sharp decline in different pathological conditions. Both types of coherence are noted for speed of calculation and ease of evaluation because of the length of the recording under study; all the reader can see are relationship disorders almost constantly present, independent of momentary correlations (coherence). The record requires meticulous description as the results of coherences are merely complementary information suggesting a reassessment of the graph, and an indication for detailed morphological scrutiny.

3 EEG biofeedback in minimal cerebral palsy – J. Faber, M. Pilarova, Z. Vuckova, F. Böhmova and L. Dobosova (Prague)

Children with minimal cerebral palsy (MCP) ($n = 30$, 24 boys, 6 girls, aged 7–14 years) had EEG, neurological, psychological and haematological examination performed before and after treatment with EEG biofeedback (EBF) in 20–30 sessions with sensorimotor rhythm (SMR) enhancement over C3 or C4. Half of the group live in conflict-ridden homes, 8 had asphyxia at birth, 3 had concussion, 7 suffered from frequent infection, 16 had the dys-dys-syndrome, and 5 showed genetic involvement, e.g. parental alcoholism, drug abuse and epilepsy. Neurological tests were topically negative in 24 children, while 6 had pyramidal manifestations of irritation and cerebellar syndrome; after EBF those findings showed no change. All the children had ADHD or ADD. Psychological tests showed above-average IQ in 14 children and average IQ in 16 children. EBF helped to improve IQ in 18 children, including all girls. Eleven children remained unimproved, and one showed deterioration. EEG was diffusely or episodically, less so even focally (5 ×), abnormal in 26 children, slightly abnormal in two children, and near normal in two children. According to optical description, post-EBF EEG showed improvement in 7 cases, no change in 18 cases, and deterioration in 5 cases. However, spectral analysis found improvement in those 18 children who improved in most of the psychological tests. Summed up, our 30 CMP children can be rated as a group with somatic, cerebral, psychic as well as social abnormalities. Two-thirds of them showed improvement after EBF, a treatment which in itself belongs somewhere between psychotherapy and biological therapy, however, without any side reactions.

4 EEG spectrum and consciousness – J. Faber, M. Pilarova, Z. Vuckova, S. Petranek and V. Krajca (Prague)

Thirty persons (10 controls, 12 epileptics and 8 dements) had EEG recorded at rest, reacting to sound and while listening to pure tones or chords from Smetana's symphonic poem Vyšehrad. Reaction time was found to depend on the EEG spectrum change calculated by means of FFT (Fast Fourier Transformation): the longer the time, the higher the delta and the lower the alpha activities. Between lucid wakefulness and sleep onset is a period of relaxation with a high alpha and relatively longer reactions time (500–800 ms). In terms of reaction time lengthening, it was irrelevant if delta activity was increased due to hyperventilation, sleep or subclinical epileptic discharges. During the perception of tone, FFT revealed changes in the alpha and delta bands different from those in the perception of chords, and different again from those in episodes of rest in normal and epileptic persons. Dements revealed no FFT differences while listening to tones or chords; the only differences were seen in the resting

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section of the spectrum. One and the same stimulus elicited a stereotypical FFT response. Different stimuli elicited different FFT responses in healthy and epileptic persons. The dements exhibited different responses to identical stimuli, and a relatively stereotypical response of low variability to different stimuli. FFT appears to be able to represent the information content of the EEG curve and, indirectly, also micro-EEG which reverberates between the thalamus and the cortex in the form of impulsation neuronal activity. This interneuronal impulsation coding is impaired in dements with cortical atrophy, displaying, on the one hand, increased variability in perceiving identical stimuli, and, on the other hand, insufficient differentiation and, consequently, increased stereotypy in the perception of different stimuli.

5 Topographical differences in neonatal EEG – a quantitative analysis – K. Paul, V. Krajca, J. Melichar and S. Petranek (Prague)

EEG activity in full-term neonates is often described as 'uniformly distributed'. Questioning this, the authors examined 21 full-term healthy neonates under standard conditions using a digital set to register the infants' EEG (Fp1,2, C3,4, O1,2, T3,4), respiration, ECG, EOG and EMG for a period of 90 min. Five minute samples of EEG activity in restful and active sleep were then passed on for automatic analysis based on adaptive segmentation. The activity in each lead was divided into 3 amplitude classes and numerically described using 13 parameters in terms of: performance in 5 frequency bands, proportional representation in the amplitude classes, stability and variability, and subsequently analyzed from bipolar and reference leads. Statistical analysis revealed significant difference in EEG activity taken from frontal, occipital, central and temporal regions, with low-amplitude activity predominating in the frontal and temporal regions, and high-voltage and/or medium-voltage activities prominent in central and occipital parts. In the alpha and theta-2 bands, maximum output was noted in the temporal area, and in theta-1 and delta-2 in the central region. Maximum variability was found over the posterior quadrants, in contrast to a rather stable activity over the anterior quadrants and temporal areas. Very little difference was noted between the homologous regions of the left and right hemispheres. Hence, rather than being 'uniformly distributed' neonatal EEG was found to be topographically differentiated.

6 Semiological seizure classification – P. Marusic and M. Tomasek (Prague)

Semiological seizure classification is based on a distinction between different types of seizure exclusively in terms of an ictal pattern of behavior or on the patient's subjective experiencing. Unlike the 1981 ILAE international classification of seizures, this classification works regardless of EEG or other clinical data. The ILAE classification takes for granted the presence of an unambiguous correlation between clinical ictal manifestations and interictal or ictal EEG findings; however, detailed analysis showed this presumption to be incorrect in many cases. Seizure semiology can be analyzed entirely independently and, together with further clinical information (history, EEG, methods of visualization, objective neurological findings), can then be correlated for the purpose of defining the epileptic syndrome. The SSC has been in use for a number of years at some epileptological centers, particularly in assessments of video EEG monitoring, but also in routine outpatient practice. The authors who present an outline of the basic division of seizure type according to SSC began using this classification at their own center after the launching of their Epileptological Monitoring Unit.

7 Parietal lobe syndrome and EEG in dementia – M. Brunovsky, M. Matousek and L. Edman (Prague)

The purpose of the study was to find out to what degree examination aimed at regional syndromes could improve EEG diagnosis in dementia. Seventy-seven dements aged 47–83 years were included in the study in compliance with DSM-III-R criteria. Clinical investigation made use of so-called step-by-step analysis of the patients' clinical condition designed to estimate the incidence and degree of intensity of the parietal, frontal, subcortical or the less regionally expressed global syndrome. EEG records were assessed visually and by means of spectral analysis. After comparisons with other regional syndromes or with the depth of dementia, the intensity of the parietal lobe syndrome was found to be the most closely correlated with slow EEG activity. These observations may account for some of the discrepancies between EEG findings and the results of clinical examination in cases of dementia. In practical terms, EEG can be expected to help the most in the diagnosis of early-onset dementia where the parietal lobe syndrome is a dominant feature.

8 Sleep disorders and melatonin – S. Nevsimalova, K. Blazejova, H. Illnerova, J. Hajek, K. Sonka and M. Pretl (Prague)

Clinical examination and a sleep study (all-night video-polysomnography, multiple sleep latency test (MSLT5), actigraphic monitoring) with a 24 h circadian hormones profile (melatonin, cortisol) were performed in 105 patients (29 men, 76 women, mean age 32.1 $\pm$ 13.5 years) with primary or secondary sleep disorders. The melatonin and cortisol profile was estimated from saliva using the method of radioimmunoassay. The *T* test and the method of ANOVA were used for statistical evaluation of the diagnostic and control groups. The 32 patients with idiopathic and non-idiopathic hypersomnia exhibited a significantly delayed onset of melatonin secretion, signs of a shift into the early morning hours, and a slightly prolonged signal of sleep hormone secretion. The finding correlated with prolonged nocturnal sleep in all-night PSG. The narcolepsy-cataplexy group (24 patients) tended to have a multi-peak melatonin signal in daytime with markedly shortened sleep onset latency on MSLT5 records, and a significantly lowered cortisol level, possibly relative to the emotional flatness and inhibited affectivity characteristic of the narcoleptic personality. Patients with the delayed sleep onset syndrome (7 patients) exhibited a non-significant shift in melatonin secretion and a significant cortisol shift into early morning hours corresponding to the results of actigraphic monitoring. The shape of the melatonin secretion curve of periodic in-sleep movements depended on the sleep disorder type. Insufficient sleep was marked by signs of decreased melatonin secretion like that in the primary insomnia group (7 patients). The group of secondary sleep disorders in neurodegenerative or neurometabolic diseases affecting children and adults was made up by 29 patients with pathological findings dependent on the gravity of the clinical picture and localization of changes.

9 Hypocretin/orexin role in the development of narcolepsy – S. Nevsimalova, J. Faraco, C. Peyron, S. Nishino, J. Vankova, K. Sonka and E. Mignot (Prague, Palo Alto, CA)

Hypocretin/orexin deficiency (primary or secondary) has a major role to play in the development of narcolepsy-cataplexy. Narcolepsy is the only sleep disorder associated with pathological REM sleep penetrance and with related changes in the structural cyclic arrangement of sleep stages. Clinical, genetic and pathophysiological research into the problem has been attracting attention for decades now. A breakthrough came with experimental works. Animal experiments showed the hypocretin/orexin complex to be the main neurotransmitter of sleep mechanisms regulation. Signal mutation in the Hcrtr locus leading to hypocretin/orexin deficit is the cause of genetically induced narcolepsy-cataplexy in dogs and mice. Evidence of the hypocretin/orexin role can also be read from the mediator's deficiency in the CSF of most narcoleptics and from its absence in autopsied narcoleptics. Seventy-four patients with narcolepsy-cataplexy had genetic tests performed. A Hcrtr mutation similar to that in an animal model was found in only one patient, an 18-year-old man followed up for the disease since infancy. His clinical picture shows all 4 signs of the narcoleptic syndrome (imperative somnolence, cataplexy, sleep paralysis, and hypnagogic hallucinations). Also present are periodic limb movements, disorders of behaviour and pronounced bulimia, mainly in nighttime. Repeated MSLT5, round-the-clock monitoring and all-night polysomnography showing the presence of SOREMs are in full accordance with the clinical diagnosis. The patient is HLA-DQB1*0602-negative. This is the first and so far the only case of genetic hypocretin/orexin mutation in clinical medicine with the narcoleptic phenotype fully expressed. The picture of hypocretin/orexin deficiency is complemented by its absence in the cerebrospinal fluid.

10 Melatonin, a synchronizer of biological rhythms – V. Hancinova and S. Mehesova (Bratislava)

Melatonin, a neurohormone produced by pinealocytes of the pineal body, is regarded as a universal mediator of circadian oscillations in all mammals including man. Pineal body neurons synthetize melatonin from tryptophan depending on exposure to light incident on the retina with maximum levels between midnight and 02:00 h. Newly synthetized melatonin is released into circulation with a short half-life of disintegration lasting from 30 to 50 min. In the target tissues the effect of melatonin is mediated by its bond to the melatonin receptor, where the antioxidation effect at the subcellular level need not be bound to any specific receptor. For its ability to shift the circadian rhythm phase, melatonin is used therapeutically in the treatment of sleep disorders bound to circadian rhythms in the blind, in sleep disorders with delayed or advanced phases, and in jet lag syndrome cases. In addition to that, it is also used as a sedative, hypnotic, antioxidant and adjuvant antiepileptic. Despite its alleged benignity, exogenous melatonin, which is not registered as a medicament but merely as a food additive, must be treated as a neurohormone with receptors throughout the organism. Of crucial importance is its action on GABA-ergic transmission where melatonin is active by way of the benzodiazepine receptor, though its effects on serotoninergic, glutamatergic as well as opioid transmission are of no minor relevance either. The hazards of melatonin overdosage in uncontrolled intake cannot be ruled out. The scope for using melatonin more often in clinical practice, as well as the risks of uncontrolled application, are also discussed.

11 REM sleep density in patients with idiopathic hypersomnia and narcolepsy – J. Vankova, S. Nevsimalova and K. Sonka (Prague)

All-night polysomnography (PSG) was employed in order to examine 28 patients with narcolepsy-cataplexy (mean age 34 ± 11.2 years, disease onset at 22.6 ± 10.1 years). Sleep paralysis was found in 43% of the patients, with hypnagogic hallucinations in 53% of them. Group 2 was made up of 10 patients with idiopathic hypersomnia (mean age 37.5 ± 8.3, disease onset at 16.4 ± 7.2 years). Sleep drunkenness was present in all of them. The control group consisted of 28 age- and sex-matched healthy adults. Clinical diagnosis was confirmed by means of the multiple sleep latency test and all-night PSG. PSG records were studied for the frequency of two phasic REM sleep parameters: number of REMs/min, mental muscles phasic activity/min, Tws). The results showed significantly higher values of the above parameters in both groups of patients with this type of dyssomnia (for REMs $P < 0.1$, Tws $P < 0.5$). Also found were changes in the number of REMs and twitches of muscles of the chin in the subsequent periods of nocturnal paradoxical sleep. The REM sleep density expressed in those values kept rising continuously during the night. The controls exhibited a similar tendency, albeit with significantly lower values ($P < 0.5$).

12 Polysomnographic findings in patients with Parkinson's disease – M. Jakoubkova, K. Sonka, E. Ruzicka, J. Roth, B. Michalcikova, R. Jech, P. Mecir and S. Nevsimalova (Prague)

Some patients with Parkinson's disease suffer from sleep disorders, some of them considerably troublesome and conspicuous. Eleven patients (7 men, 4 women, mean age 59.8 ± 7.2 years) with Parkinson's disease underwent polysomnographic examination at the sleep laboratory of the Department of Neurology, 1st Medical Faculty, Charles University, over the past year. The duration of the disease was 9.6 years (ranging from 1 to 14 years), and the mean period of treatment was 6.6 years (ranging from 1 to 10 years). Ten patients were treated with L-DOPA, one patient was treated with a dopamine agonist. All-night polysomnography (PSG) was performed using a complete PSG array (6 EEG leads, electrooculography, EMG of the muscles of the chin and mm. tibiales anteriores bilat., ECG, air stream outside the mouth, respiratory movements of the chest and abdomen, haemoglobin saturation with oxygen). The results confirmed the frequency and gravity of sleep disorders in patients with Parkinson's disease. Ten of them were found to have a poor-quality sleep architecture with cyclization disorders, delta sleep as well as REM sleep reduction. Signs of the sleep apnoea syndrome were noted in 3 patients. Two exhibited periodic LE movements, and 4 were diagnosed as having disordered muscle atonia during REM sleep, a pathophysiological groundwork for a disorder with abnormal behaviour during REM sleep.

13 Sleep architecture in children with neuromuscular involvement – M. Pretl, I. Stepanova, K. Sonka, S. Nevsimalova, M. Havlova, M. Jakoubkova, P. Hnidkova and J. Vankova (Prague)

Thirteen patients with progressive muscular dystrophy (Duchenne and Becker) and 13 age-matched healthy controls were examined polysomnographically for the purpose of sleep architecture and microstructure objectivization. The mean age of both groups was 12.69 ± 3.75 years. Of the parameters under scrutiny, no prolonged sleep onset latency was found in the patients group, whose sleep effectivity was lower (91.78 ± 4.90, $P < 0.023$). Comparisons of the patients' sleep stage proportional representation revealed a significant increase in vigilance and NREM stage 1 ($P < 0.005$ and 0.039, respectively) and REM sleep reduction ($P < 0.009$). The two groups showed no difference in the proportion of the rest of the sleep stages (NREM 2, 3, 4). The sleep apnoea syndrome was noted in 4 patients (30.77%), and the mean respiratory disturbance index (RDI) was 17.75 ± 1.79. One patient had apnoea solely in REM sleep (RDI in REM, 29; RDI for total sleep time, 5). Periodic LE movements were seen in 3 patients (23.08%). Abnormal intermittent tonic EMG activity was noted in one patient. Four patients (30.77%) were found to have sporadic sleep time spontaneous discharges in one motor unit resembling fragmentary myoclonus.

14 Rhonchopathy and sleep apnoea syndrome in a Brno sleep laboratory – M. Moran (Brno)

The author presents admission screening tests of patients referred to this sleep laboratory for snoring and sleep time respiratory disorders in the October 1998 to October 2000 period. The spectrum of referring medical centres is analyzed (ENT, neurology, pneumology). The predominance of otorhinolaryngology can probably be put down to the highest rate of information among ENT specialists. The proportion of the patients is examined in terms of sex and age; women made up about one-third of the group in what was a greater representation than that reported in the literature. An analysis is presented of rhonchopaths (220) and apnoeics (110) examined for SAS (AI10, AHI15, DI10) for different parameters characteristic of both groups (AI, apnoea index; AHI, apnoea-hypopnoea index; DI, desaturation index; BMI, body mass index; heart-rate variation index; body position-related complaints). The more specific indication of SAS and, subsequently, treatment with CPAP from other than the Brno centres can be explained by the existence of a number of other ENT centres performing plastic operations as distinct from only two centres indicating and monitoring CPAP treatment. The following therapeutical methods were indicated and carried out on the basis of the admission tests: CPAP, 50 patients; ENT plastic surgery in the upper airways, 247 patients; both methods of treatment, 9 patients. As regards indications for CPAP therapy, no major differences were found from other laboratories or from literary reports.

15 Fear-anxiety: neuroanatomical network, neurotransmitters, neurophysiological targets – P. Kukumberg (Bratislava)

Fear, specifically experienced, definable apprehension, is a phylogenetically ancient reaction of prompt defence and protection against threat. Its concomitant biochemical and physiological manifestations can be registered in animals as well as in man. Anxiety, a 'charge-free' type of apprehension of variable intensity, is typical of man, with the amygdala as the center for the perception and, at the same time, for the efferentation of fear-anxiety. The amygdala receives information from the thalamus (direct communication) which reaches it closely ahead of cortical areas to correlate the data with hippocampal engrams and, immediately afterwards, with processed cortical signals. Using predilection pathways (hypothalamus, locus coeruleus, periaqueductal gray, nc. parabrachialis, etc.), it sets off physiological or pathological responses to emotional challenges of fear-anxiety (universal hypothalamo-pituitary-adrenergic axis activation; panic fear). These processes are mediated by neurotransmitter operations and

marked by perceptible visualizable neurobiological and neurophysiological changes.

16 The current concept of the Guillain–Barré syndrome – Z. Ambler (Plzen)

The Guillain–Barré syndrome (GBS) is an acute, self-limited motor-dominant polyneuropathy, often preceded by infection (viral or bacterial), surgery or immunization. Its pathogenesis is believed to be immune-mediated with humoral and cell-mediated immune mechanisms as contributing factors. The disease is clinically and electrophysiologically heterogeneous. Electrophysiological examination provides the most sensitive tests for an early detection of abnormalities and for GBS pattern characterization. The most frequent pattern is that of acute inflammatory demyelinating polyneuropathy (AIDP). Though the clinical findings are typically symmetrical, the signs of demyelination are multifocal. The following subtypes with primary axonal degeneration are currently recognized: acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN). The Miller–Fisher syndrome is another variant of GBS. Antibodies against various gangliosides (GM1, GM1b, GD1b and others) have been found in a number of GBS cases; at present, they are diagnostically useful mainly in the Fisher syndrome (GQ1b). Treatment for GBS should start as soon as possible in any patient who is too weak to walk unassisted or whose condition shows rapid progression. Plasma exchange and intravenous human immunoglobulin (IVIG) are equivalent there, the latter as a therapy of first choice. Corticosteroids should be avoided. The IVIG-methylprednisolone combination is currently under trial.

17 The piriform muscle syndrome – a controversial clinical entity – Z. Kadanka (Brno)

The first literary reports about ischiadic nerve compression by the piriform muscle came from Yeoman in 1928. In 1937, Freiberg defined the diagnostic triad of: ischiadic point sensitivity, positive Lasegue's manoeuvre, and improvement after conservative treatment. In 1947, Robinson added the following: gluteal region trauma, gluteal muscle atrophy, and a piriform muscle mass with tenderness on palpation. The syndrome went into oblivion for a number of years; now, its presence is presumed to account for 'radicular' syndromes without signs of root compression in the spinal canal region. Characteristic symptoms comprise pain in the gluteal region and palpation tenderness of the ischiadic point, pain provoked by prolonged sitting particularly on a hard pad (wallet) and by bending forward, and aggravated in activities requiring femoral adduction and internal rotation (ski running), with relief in walking or standing. There is no pain in the small of the back. Parasthesia appears in the gluteal and ischiadic innervation regions. There are frequent cases of gluteal region trauma in the patient's history. In the absence of paresis, the patient tends to walk with the lower extremity in external rotation. The Trendelenburg sign, Freiberg symptom and PACE and AIF tests are usually positive. The piriform muscle tends to show tenderness on palpation per rectum and during examination of the pelvis. Needle EMG is normal. There is often asymmetry in the H reflex in the neutral position and during the AIF test. Abnormal vessels and connective tissue stripes are often identified by magnetic resonance imaging. There is scope for both conservative and surgical treatment.

18 Chronic motor axonal neuropathy – D. Vesela, E. Klimova and E. Kahancova (Kosice)

The numerous groups of motor neuron disorders affecting the anterior horns of the spinal chord include a number of chronic dysimmune neuropathies. Though their clinical course resembles degenerative diseases, they are potentially treatable. Present-day immunological and electrophysiological methods help to differentiate some of them, including neuropathies associated with increased titres of autoantibodies against GM-1 gangliosides. Typical diagnostic features include: axonal lesions without motor conduction block in EMG tests, progressive, symmetrical flaccidity of the girdle muscles of extremities, and breathlessness. The titres of anti-GM-1 antibodies are unincreased. Final diagnosis is facilitated by physiological values of the CSF. The cases are presented of two patients, men aged 63 and 68 years, respectively, who met the above criteria despite the impossibility of anti-GM-1 antibodies estimation. As the disease took a subacute course, the therapeutical decision was in favour of repeated application of IVIG as one of the immunomodulatory treatment options. The clinical condition of the two patients is now stabilized.

19 Neuropathy of the critically ill – F. Vlcek, J. Hromada, R. Höffer and I. Woznicova (Ostrava)

Since the 1980s, a new clinical entity has been appearing at intensive care units under the name of 'neuropathy of the critically ill'. It is marked by declining reflexes, flaccid paralysis or even quadriplegia, no scope for disconnection from the respirator, a need for prolonged intensive care, and also a high death rate. Systematic clinical and EMG research and muscle biopsy have resulted in this entity's subdivision into a group of neuropathies (neuropathy of the critically ill and axonal motor neuropathy), and a group of myopathies (myopathy of the critically ill, cachectic myopathy, and acute narcotizing myopathy). Recent views presuppose a measure of overlapping of the components of myopathy and neuropathy with a predominance of one of the two, hence the newly coined term polyneuromyopathy. The root cause of this entity is in the development of a state of shock and sepsis (chronic systemic inflammatory response syndrome, SIRS), followed by multiorgan dysfunction syndrome (MODS) leading to failure of different organs. SIRS and MODS are co-responsible for disordered microcirculation affecting peripheral nerves, neuromuscular transmission, and muscles.

20 Neuromuscular disorders in critically ill patients – M. Schreiber, H. Matulova, F. Para, P. Kunc and J. Manak (Hradec Kralove)

Pathological changes of the neuromuscular system in critically ill patients have been receiving increased attention in the past 20 years with growing numbers of patients successfully surviving serious conditions of sepsis, multiorgan dysfunction and failure. The authors sum up their experience of early diagnosis of newly developed neuromuscular disorders. Seventeen patients in a critical state marked by flaccid quadriplegia and difficult weaning from ventilation were examined at the intensive care unit of the Department of Gerontology and Metabolism, Hradec Králové Teaching Hospital, in the years 1998–2000 and found to have myopathy and polyneuropathy of the critically ill. These clinical entities are the most frequent causes of skeletal muscle flaccidity in critically ill patients due mainly to multiorgan failure and sepsis. Detailed electromyography including direct muscle stimulation according to rich and muscle biopsy prove to be the most productive methods.

21 EMG findings in inflammatory myopathy – K. Kalous (Prague)

Inflammatory myopathy is made up of a group of muscular diseases arising from skeletal muscle inflammation. Regarded as autoimmune diseases, the group is clinically subdivided into cases of polymyositis, dermatomyositis and myositis with inclusion bodies. Diagnosis proceeds from clinical examination, laboratory biochemical tests, EMG and muscle biopsy. In terms of EMG, inflammatory myopathies can be divided into 4 groups: normal EMG, myogenic EMG, myogenic findings with fibrillation potentials, and mixed EMG findings. In conjunction with the Institute of Rheumatology in Prague, a total of 224 patients (53 men, 171 women, mean age in men 50 years and in women 50.6 years) were examined in 1999 up to the end of September 2000. EMG and other laboratory findings were negative in 24 men (45%) with clinically suspect inflammatory myopathy. Out of the dermatomyositis group, 8 patients (80%) were EMG-positive, and two (20%) were EMG-negative. The polymyositis group comprised 10 patients (77%) with EMG positivity, and 3 (23%) were EMG-negative. In the female group, 67 patients (39%) with suspect inflammatory myopathy had negative EMG and laboratory test findings. In the dermatomyositis group, 24 women (86%) were EMG-positive and 4 (14%) were EMG-negative. Out of the polymyositis group, 37 women (82%) were EMG-positive, and 8 (18%) were EMG-negative.

Conclusions: (a) negative EMG does not rule out the presence of inflammatory myopathy; (b) positive EMG can provide information on the activity of the complaint, its gravity, distribution and ongoing regeneration.

22 Is there a correlation between myasthenic complaints and treatment with interferons alpha and beta? – H. Matulova, F. Para, M. Valis and J. Simko (Hradec Kalove)

The case is presented of a 48-year-old crime squad officer who underwent, at the age of 33, right-sided lumbar sympathectomy for ischaemic disease of LE, melipranol treatment for arterial hypertension from the age of 41, and therapy for polycythemia vera from the age of 45, first with venipuncture, later with erythocytophoresis and hydroxycarbamide and, as from September 1999, with interferon alpha (Intron A 5 MIU s.c. twice weekly). After 20 injections, the patient developed bilateral blepharoptosis, diplopia, flaccidity of the right UE, dysphagia, dysarthria, and dyspnoea. Myasthenia gravis was diagnosed on the basis of typical clinical findings, decreased amplitude of the muscle action potential evoked by repetitive nerve stimulation of 3 Hz, and a positive neostigmine test. The level of antibodies against acetyl choline receptors was found to be low, and CT of the mediastinum was negative. The treatment was changed to 5× plasmapheresis, azathioprine, corticoids, pyridostimine and IVIG. Because of only minor improvement, the patient is being prepared for thymectomy. The authors warn of the need for great caution before treatment with interferon in non-cancer patients since experience of INF therapy is still relatively limited, and further undesirable effects cannot be ruled out in the future.

23 Correlation of clinical and neurophysiological findings in a patient with type 7 autosomal dominant spinocerebellar ataxia – P. Bauer, V. Matoska, A. Zumrova, J. Kraus, R. Mazanec, A. Boday and T. Marikova (Prague)

A number of hereditary as well as non-hereditary metabolic disorders may become manifest through spinocerebellar ataxia. Differential diagnostic progress has been made within the scope for molecular genetic proof of pathological expansion of trinucleotides at different sites of the genome. Spinocerebellar ataxias of type 7 (SCA7) are relatively rare in the group of autosomal dominant hereditary ataxia – a mere 6–13% of the patients so far diagnosed. The authors present the first confirmed case of type 7 spinocerebellar ataxia in the Czech Republic. Patient I.J., formerly believed to suffer from Friedreich's ataxia, was found to have an expansion of CAG repeats in the region of the short arm of chromosome 3 (3p12-13). The patient's clinical and electrophysiological values are compared with cases published in world literature to date. His family tree is presented to demonstrate the difficulties in determining the mode of heredity in repeat diseases, with the phenomenon of anticipation obscuring the dominant transmission of the pathological allele.

24 Electrophysiological findings in hereditary neuropathy CMT 1A, HNPP and CMT1 X – R. Mazanec, P. Seeman, M. Ctvrteckova and M. Bojar (Prague)

With its incidence of 1:2500–5000, Charcot–Marie–Tooth (CMT) is one of the most frequent cases of hereditary neuropathy. A similar phenotype is conditional upon mutations of 10 known genes in more than 18 chromosomal localizations. The most frequent mutation is that of duplication of 1.5 Mb on chromosome 17 p11.2-12 affecting the gene for peripheral myelin protein PMP 22. Duplication is demonstrable in 84% of CMT type 1 with AD heredity. Deletion in the same region was found in 86% of tomaculous neuropathy (HNPP). Mutation in the X-linked gene for connexin 32 is the cause of 10% of all hereditary myelinopathy cases. Besides the typical clinical finding, the diagnostic protocol includes EMG. Conduction studies complemented with specific-target needle EMG serve mainly to verify neuropathy and to differentiate between the demyelination and axonal types of CMT, but also to detect the oligosymptomatic forms of CMT in other family members, and to follow up the disease progression. Myelin gene mutation is caused by seemingly homogeneous myelinopathy; in spite of that the authors tried to find differences between the most frequent types of hereditary neuropathy by evaluating some of the conduction parameters of the nerve fibres. These help to make a more precise identification of the phenotype of different myelinopathies and, thereby, to indicate DNA analysis for a particular myelin gene. Twenty-three patients with CMT 1A, 10 patients with HNPP and 6 patients with CMT X were examined electrophysiologically for the purpose of defining the parameters of each of the neuropathy types.

25 Electrophysiological findings in myotonic dystrophy – J. Kraus, A. Boday, V. Matoska and T. Marikova (Prague)

Myotonic dystrophy (MD) is a multisystem disease of marked intra- and interfamilial variability, with an incidence of 1:8000, and autosomal dominant heredity with the phenomenon of anticipation. At the molecular level, DM1 is caused by gene mutation on chromosome 19 in the 19q13.2-13.3 area. The gene encodes myotonin protein kinase. Out of 74 patients with clinically suspect MD and 150 members of their families, amplification was found in 43 cases (in 17 families). Conduction study findings in the minor forms of DM1 tend to be normal except for a lower CMAP amplitude and a variable CMAP decrement in response to repetitive stimulation. At lower frequencies of stimulation the decrease may suggest the presence of neuromuscular transmission disorders. Patients with more serious involvement exhibit a lower rate of conduction along the motor fibres of n. medianus and n. peroneus of as much as 60% of their healthy relations' values. Tests with concentric needle electrodes will show the presence of positive waves, myotonic discharges and motor unit action potentials (MUPs) of short duration, lower amplitude and rapid recruitment. MUPs are polyphasic, though to a lesser degree than in other types of dystrophy.

26 Neurogenetic diagnostics – our experience and options – J. Kraus, A. Boday, V. Matoska, A. Zumrova and T. Marikova (Prague)

Molecular genetic analysis as a routine diagnostic method may help to corroborate or, with regard to the frequency of mutation, rule out suspicion of a specific involvement of, in particular, the most frequent hereditary diseases. The authors make routine neurogenetic diagnoses of neuromuscular diseases (deletion forms of DMD/BMD and SMA5q, myotonic dystrophy DM1, HMSN Ia, and CMT-X), and ataxia (Friedreich's ataxia with amplification of GAA repetitions, and of late also the diagnosis of ADCA/SCA1,2,3,6,7,8 as 95% of triplet mutations are due to amplification). Neurogenetic diagnosis of mental retardation is another field of application (FRAXA, Angelman's syndrome, and Prader–Willi syndrome). The group plan to introduce neurogenetic diagnosis of the distal type of myotonic dystrophy DM2, m. Pelizaeus–Merzbacher, the familial type of febrile convulsions and Rett's syndrome. Diagnosis is often established in conjunction with other centres, especially for the girdle forms of muscular atrophy, FSHD, Emery–Dreifuss muscular dystrophy of types 1 and 2, congenital myopathy with merosin deficiency and neurodegenerative diseases (ataxia telangiectasia, neuronal ceroid-lipofuscinosis and leucodystrophy involving diagnosis of homozygotes of excessive residual enzyme activity, and identification of carriers).

27 The significance of neurophysiological methods in neurotoxicology – P. Urban and E. Lukáš (Prague)

While the number of cases of acute poisoning is on the decline in present-day clinical neurotoxicology, the significance of long-term exposure to low-concentration neurotoxic substances has been rising. The health consequences of such exposure take the form of non-characteristic subjective complaints marked by scanty neurological findings. These subtle manifestations of neurotoxic damage can be objectivized by means of neurophysiological methods. Neurophysiological abnormality can be seen as an electrophysiological marker of CNS generator dysfunction. VEP study appears particularly promising, especially as regards exposure to organic solvents and heavy metals. The former cause bilateral, rather symmetrical prolongation of VEP latency and/or reduction of amplitude. In pathophysiological terms, these changes are due to myelin sheath lipid structural alteration under the effect of lipophilic organic solvents. With the exposure discontinued, VEP changes in alcohol abuse appear to be easier to reverse than those after exposure to toluene. As for exposure to mercury vapours,

signs of latency shortening are characteristic of VEP changes in the initial stage, correlating with the clinical picture of erethism. In advanced cases, however, there may even be latency prolongation. VEP changes are typically associated with slowed down sural nerve conduction as a sign of incipient mercury-induced sensory polyneuropathy. While aetiologically non-specific, these neurophysiological changes may in individual cases be due to exposure to a neurotoxic noxa provided that (a) there is evidence of significant exposure, (b) the clinical picture is compatible with intoxication, and (c) differential diagnosis has ruled out other causes. In group estimation of exposure, any possible connection between neurophysiological anomalies and neurotoxic exposure should be considered relative to evidence of a dose–response interdependence.

28 Standard examination VEP procedures are insufficient – J. Chlubnova, J. Kremlacek and M. Kuba (Hradec Kralove)

In an effort to improve neuro-ophthalmological diagnostic procedures, the authors continue developing more specific stimuli for selective stimulation of separate visual pathway subsystems. Hence, reactions to centrifugal radial motion 'expansion' (E-VEPs) were added to their currently used set of visual evoked potentials, pattern-reversal visual evoked potentials (P-VEPs) and motion-onset VEPs (M-VEPs). The stimulus was designed so as to decrease the pattern spatial frequency and to increase the motion velocity towards the visual field periphery (with respect to the physiological properties of the retina). To prove the stimulus efficiency, the authors compared E-VEP parameters with VEP results to monocular stimuli in a selected group of 100 patients (in 6 unipolar leads, Oz, Or, Ol, 5 cm to the right and left of the Oz position, Cz, Pz and Fz). The E-VEPs were found to have the highest interpeak amplitudes of all the VEPs tested ($P < 0.001$). Moreover, in 5% of the cases the E-VEP was the only distinct (and reproducible) reaction. Despite the high correlation of the dominant negative peaks in the M-VEPs and E-VEPs (unidirectional and expanding motion), 12% of the patients were found to have only the E-VEPs selectively impaired (prolonged latency). Thus, additional examination of the reactions to expanding motion (E-VEPs) can increase the sensitivity of VEP examination for about 12%. However, the specificity of the E-VEP changes has yet to be proved.

29 Electrophysiological changes related to magnesium therapy in anxious-depressive disorders – D. Gayer, E. Libigerova, J. Chlubnova, J. Kremlacek and M. Kuba (Hradec Kralove)

Electrophysiological readings were evaluated in anxious-depressive patients as part of a randomized double-blind placebo-controlled study relative to magnesium therapy. Twenty-nine patients (mean age 42 ± 10 years, 16 on placebo, 13 on magnesium) were examined twice, once before and once after a 5 week period of treatment. VEPs of the primary and secondary visual cortex (pattern-reversal VEPs, motion-onset VEPs in response to unidirectional linear motion and to centrifugal motion, 'expanding' radial pattern) were recorded from Oz and lateral temporo-occipital leads. VEPs from cognitive cortical areas were tested in unipolar Pz, Cz and Fz leads. EEG frequency analysis in the eyes-closed mode was also employed. The cognitive task (oddball paradigm) consisted of the recognition of either non-coherent motion or a normal face (a scrambled human face was displayed as a non-target stimulus). Twenty-seven sex- and age-matched healthy subjects served as controls. Significant differences in the group of patients and changes related to magnesium therapy were only found in the cognitive VEP parameters. An increase of the P300 amplitude was found in the human-face target stimuli in patients after treatment with magnesium (from 9.1 ± 3.8 to 11.2 ± 3.5 μV) together with a significant lowering of the non-target P300 amplitude (from 7.2 ± 4.1 to 3.8 ± 3.8 μV) in coherent/non-coherent motion stimuli (not found in the placebo group). Both changes can be regarded as a positive influence of magnesium over the patients' cognitive functions. The intensity of reactions to the target stimuli was found to be increased as distinct from decreased reactions to non-important stimuli which should be filtered out during the cognition process.

30 Utilization of visual evoked cognitive potentials – M. Kuba, J. Kremlacek, J. Szanyi, J. Chlubnova, D. Gayer and F. Vit (Hradec Kralove)

The use of cognitive evoked (event-related) potentials (ERP) can improve the diagnosis of some psychiatric disorders and help to evaluate therapeutical effects. Unlike auditory ERP, a wide enough spectrum of visual stimuli can offer a more complex insight into cognitive processes. However, there are some limitations. The most serious problem is posed by considerable inter- and intra-individual variability (mainly in highly complex cognitive tasks), the variation coefficient being twice as large as in primary VEPs, e.g. pattern reversal. It is essential to maintain standard conditions of examination (time, glycaemia, and constant psychic state). Protracted ERP examination increases the number of blinking-related artifacts, more so in the fronto-central areas than in occipital primary VEPs. In the oddball paradigm, target stimulus signalling with any kind of motor activity (pushing a button) leads to P300 contamination. Hence, emotionally potent stimuli (e.g. human faces) which can do without the subjects' active co-operation are preferable. According to the authors' data including the simultaneously recorded time of reaction to target stimuli, the cognitive process is better represented by the negative peak preceding the P300, the latency of which is much more constant. As specification of a 'norm' in ERP is still rather complicated, longitudinal intra-individual evaluation of changes seems to be more reliable for clinical purposes.

31 Electrophysiological findings in migraine – J. Szanyi, G. Waberzinek, J. Kremlacek, M. Kuba and J. Chlubnova (Hradec Kralove)

According to recent EP studies, abnormal cortical information processing is in progress in the migraine-affected brain during attack-free periods. A fundamental, probably protective, feature of the brain function – response habituation while the stimulus is repeated – is thus impaired. Visual evoked potentials (VEP) and EEG frequency spectrum were examined in 26 patients suffering from migraine (5 with and 21 without aura) during attack-free periods, as well as in 27 normal controls. Transient pattern-reversal VEPs and two variants of motion-onset VEPs failed to display any significant differences between the migraine group and controls. Since the standard recording time was less than 1 min for each of the specified VEP examinations, it may have been too short to exhibit the envisaged VEP habituation in normal subjects. No correlation was found between the EEG frequency spectrum parameters and the migraine descriptors in patients. Using the oddball paradigm, visual evoked event-related potential (ERP) (recognition of coherent/non-coherent motion) and emotional passive ERP (recognition of normal and scrambled face pictures) were also studied. In standard ERP, no significant differences in P300 latencies or peak-to-peak amplitudes were noted between migraineurs and controls. The only intergroup dissimilarity observed was found in the emotional passive ERP. The control group exhibited a significant amplitude reduction in the non-target response as distinct from the target response ($P < 0.004$). However, this lowering was not present in migraine sufferers ($P < 0.2$), probably due to decreased habituation selectively present in the higher cortical processes.

32 Functional MRI estimation of ERP generators – R. Jech, E. Ruzicka, A. Nebuzelsky, J. Krasensky and Z. Seidl (Prague)

To localize ERP (event-related potentials) generators with more accuracy, the authors compared ERP and fMRI (functional MRI) results. Ten healthy volunteers (6 men, 4 women) were assigned an oddball type task and asked to focus attention on rare target stimuli (horizontal black and white stripes, 22%) pseudo-randomly appearing among standard stimuli (vertical black and white stripes, 78%), proceeding in two ways on spotting the target stimulus by (a) subtracting the number 2 from 100 (mental task), or (b) performing rhythmical movements of the right-hand fingers (motor task). Functional MRI was analyzed using cross-correlation between the 'box-car' function and the dynamic course of the signal in

each voxel. The fMRI and ERP data were co-registered by means of 3 reference points: nasion, left and right tragi. The group mean of fMRI and ERP was used for synthesis. The deployment of ERP generators was estimated with the method of equivalent current dipoles manually deployed in cortical areas activated by fMRI. A pair of dipoles in the visual cortex helped to account for up to 95% of the signal while the P1 ERP component was culminating. In the mental task, P3 genesis was explicable by means of 6 dipoles at more than 99% (lobulus parietalis superior bilateral, SMA bilateral, and dorsolateral prefrontal cortex bilateral). During work on the motor task, the activation was also observed of the primary SM cortex on the left, SMA bilateral, and cerebellum on the right. The MP component calculated from the difference in ERP curves registered during work on the motor and mental tasks proved explicable by means of 3 dipoles at a rate of 99%. ECD-aided modelling restricted to fMRI-detected areas may help localize cortical EP generators. Admittedly, fMRI may not be sensitive enough to detect some generators activated too briefly or subliminally.

33 Nervous evoked potentials after transcranial magnetic stimulation – J. Benetin, D. Richter, A. Glubocky and P. Valkovic (Bratislava)

The recording of MEP (motor evoked potential) after transcranial magnetic stimulation from the respective muscle may be rendered difficult or even impossible. Hence, the authors tried to find out whether or not evoked electrical activity in the nerve can be recorded after transcranial magnetic stimulation. Using 5 healthy volunteers they recorded MEP from the m. abductor digiti minimi and, simultaneously, from surface electrodes over the ulnar nerve in the cubital canal area where an amplitude polyphasic potential of some 0.1 mV was regularly registered. The latency of the response in the cubital canal region ranged between 14.7 and 17.4 ms, while the MEP latency ranged between 21.6 and 23.6 ms. The difference between the response recorded in the elbow region and MEP was within the range of 5.7–6.9 ms. The response recorded over the ulnar nerve appears to correspond to the nervous action potential induced by transcranial magnetic stimulation. More research is needed to verify this hypothesis.