Child neurology
Sleep disorders

P 1155

Ascorbic acid and gluthathione CSF concentration in newborns with bacterial meningitis

M. T. E. S. Lukavac
Institute for Neonatology, Belgrade, YUGOSLAVIA

Introduction Pathophysiological mechanisms of meningeal inflammations are very complex. Free oxygen radicals play an important role. Non-enzymatic antioxidants are also important for prognosis of illness. Two of them are ascorbic acid and gluthathione.
Objective We tested the hypothesis that ascorbic acid and gluthathione are important non-enzymatic protective factors.
Methods We tested CSF and blood serum of 32 newborns with bacterial meningitis in the first seven days of illness. We used a method with 2.4 dynithrophenil-hydrasine for ascorbic acid and a method with Elmans substance for gluthathione. We formed control groups of newborns with high risk for bacterial meningitis.
Results The mean value of CSF ascorbate concentration in groups of sick newborns is 112.93uM/L and in control, group's 102.79uM/L. There is no statistical difference. Dehydroascorbate concentration in CSF is 75.42uM/L and in control group 75.10/L, $p>0.05$. Blood serum concentration of ascorbic is 161.00-ump/L in the group of sick newborns and 128/L in the control group<0.05.
But ascorbat/dehydroaskorbat ratio (which is constant) shows statistical important changes between two groups. There is no statistical significance in correlation between concentration of ascorbic acid with protheinorachy and numbers of leukocytes. Mean CSF concentration of gluthathione is 13.37uM/L in groups of sick newborns and 14.50uM/l in control groups. There is no statistical correlation between protheinorachy, and number of leukocytes with gluthatione concentration.
Conclusion Ascorbic acid and gluthathione are not important antioxidant protectors in early stages of neonatal bacterial meningitis. Change in ascorbat/dehydroascorbat ratio shows that dynamic has been changed but that only as reparative protectors they maybe more important in another stage of illness.

P 1156

Arnold-Chiari malformation, the character of epileptic seizures, particular features of EEG

O. I. Pavlova[1], S. G. Pantuchov[2], O. A. Kuznetsova[2]
[1]St-Petersburg Paediatric Medical Academy, St-Petersburg, RUSSIAN FEDERATION, [2]Pediatric Medical Academy, St-Petersburg, RUSSIAN FEDERATION

Introduction Arnold-Chiari malformation (ACM) becomes a problem of paediatric neurology because of the increasing frequency in clinical practice.
Methods 28 patients aging from 4 to 13 years have been examined. MRI and MR-angiography proved the diagnosis of ACM.
The first degree of ACM was in 5 cases, the second in 19 and the third in 4 patients. EEG registration was performed. International System 10 – 20 of electrode positioning.
Results On the basis of clinical and physiological data several types of epileptic attacks were detected: simple partial (sensory; with vegeto-visceral manifestation) complex partial, with secondary generalisation.
Particular features of EEG pattern:
1) Basic activity is slow, increasing percentage of slow waves in posterior area;
2) Registration of slow rhythmic waves, tracing more than 10% of registration time in caudal area (there is no reaction to eyes opening);
3) Generalised bilateral paroxysmal activity (polymorphic sharp slow waves, sharp waves).
Conclusions In all groups of patients with frequency of epileptic seizures, the EEG pattern correlated with the degree of ACM.

P 1157

Hypothalamic hamartoma presenting as true precocious puberty and gelastic seizures

M. Monteiro[1], C. Horta[1], J. Reis[2], R. Rangel[2], H. Cardoso[1], B. Serra[1], H. Ramos[1]
[1]Dept. Endocrinology, Hospital Geral Santo Antonio, Porto, PORTUGAL, [2]Dept. Neurosurgery, Hospital Geral Santo Antonio, Porto, PORTUGAL

Hypothalamic hamartomas (HH) are congenital lesions usually located at the floor of the third ventricle, containing LHRH-secreting cells, that can cause true precocious puberty (TPP) and in some cases gelastic seizures.

The authors present the clinical case of a female patient first observed aged 5 years old, presenting with pubertal development and gelastic seizures. The hormonal findings were compatible with central activation of the GnRH pulse generator (LHRH test: basal and post-stimulation LH of 12.4 mUI/ml and 100.89 mUI/ml, respectively). MRI demonstrated a 3 cm isointense hypothalamic lesion with retroclival expansion showing no gadolinium enhancement. EEG and SPECT confirmed temporal hyperactivity. These signs were highly suggestive for the diagnosis of HH, as the cause for TPP and gelastic seizures.
Aged 20 years old, in spite of multiple anti-convulsive therapies, the patient still presented several daily seizures, so that surgical excision of the lesion was performed in an attempt to control the epilepsy. Post-operatively seizures became rare but the patient developed ACTH, TSH and gonadotrophin insufficiency. Three months later, although adrenal and thyroid function recovered she remains with secondary hypogonadism.
Conclusion HH are benign lesions associated with seizures often resistant to anticonvulsive therapy. Neurosurgical treatment presents considerable risk for endocrine and neurological morbidity, although controversy should be considered whenever medical therapy failed and patients present already high morbidity.

P 1158

Cerebrovascular reactivity in migraine in children

A. N. Trubacheva, V. I. Guzeva, M. Y. Sharf
St-Petersburg Paediatric Medical Academy, St-Petersburg, RUSSIAN FEDERATION

Migraine headaches are relatively common, affecting approximately 5% of all children.
Alteration of intracranial vessel tone has been implicated in the pathophysiology of migraine. The cerebrovascular reactivity was measured by means of transcranial Doppler in 45 migraine patients with (15 patients) or without aura during headaches-free intervals and in 50 healthy controls (age 10–16 years). The vasomotor response was evaluated during hypercapnia. Induced by inhalation of a mixture of CO2 5% and O2 95% and during hypocapnia obtained after voluntary hyperventilation. Reactivity index values during CO2 inhalation and during hyperventilation were significantly higher in patients suffering from migraine with aura and without aura.
Our data suggest an increased cerebrovascular reactivity in children suffering from migraine with respect to controls that might be related to baseline disturbance cerebrovascular regulation.

P 1159

Sleep disturbance of affected and at risk individuals from the pallido-ponto nigral degeneration (PPND) family with N279K mutation on chromosome 17 (FTDP-17)

S. Lin, Z. K. Wszolek
Mayo Clinic, Jacksonville, FL, USA

Sleep disturbances are commonly seen in neurodegenerative diseases such as Alzheimer's, Parkinson's disease and multiple system atrophy. PPND is a progressive disorder characterized by papallido-ponto-nigral degeneration. PPND belongs to frontotemporal dementia and parkinsonism linked to chromosome 17 (FTDP-17) and is due to N279k mutation. Cognitive decline and motor symptoms of extrapyramidal dysfunction with paucity of resting tremor occurs in the fifth decade of life. Dementia usually ensues and death can occur within eight years. Clinical manifestation and neuropathological finding of FTDP-17, has been discussed well characterized except for sleep disturbance. We report the polysomnographic findings of five affected subjects and five at risk individual from this family.
Results 1. The two most advanced affected subjects showed severe insomnia. No sleep was achieved by one of them and the other achieved only 43 minute of very disrupted sleep. The other three affected subjects also had difficulty initiating and maintaining sleep.
2. Disordered breathing was not observed in any of the subject, affected or at risk.
3. Periodic leg movement was elevated in the three less affected and one of the at risk subjects.
4. No parasomnia, REM or NREM, was observed.
Discussion Pontine tegmentum, the caudal solitary track nucleus, the preoptic area and basal forebrain, among others, have been implicated in sleep generation. Neuropathological findings of PPND subjects have shown that pontine and mesencephalic tegmentum are areas the most involved. Some of our findings of sleep disturbance in affected subjects correspond with pathological abnormalities seen in this family.

P 1160

Excessive daytime sleepiness and unintended sleep episodes in Parkinson's disease patients treated with dopaminergic agents

L. D. Borchert[1], T. Roth[2], D. L. Bliwise[3], C. Cantor[4], J. M. Gorell[2], J. P. Hubble[5], B. Musch[1], C. Pollak[5], D. B. Rye[3], M. B. Stern[4], R. L. Watts[3]
[1]Pharmacia Corporation, Peapack, NJ, USA, [2]Henry Ford Hospital, Detroit, MI, USA, [3]Wesley Woods Health Center, Atlanta, GA, USA, [4]Pennsylvania Hospital, Philadelphia, PA, USA, [5]Ohio State University, Columbus, OH, USA

Introduction Unintended sleep episodes (SEs) have been reported in Parkinson's disease (PD) patients taking dopamine agonists (DAs)[1]. This observational study was undertaken to determine if patients reporting SEs and taking DAs are sleepy during the day and/or sleepier than PD patients not reporting SEs.
Methods This study includes 24 patients (5 women, 19 men) who received DAs for idiopathic PD and had abnormal Significant Other Epworth Sleepiness Scale (SOESS) scores of $\geq$10. Patients were divided into 2 groups: those with SEs (SE+, n=16) and those without SEs (SE–, n=8). Patients underwent 2 consecutive nights of polysomnography followed by multiple sleep latency testing (MSLT).
Results The overall frequency of pathological sleepiness (MSLT<5) was 42% (10/24). No significant differences between the SE+ and SE– groups in mean level of sleepiness, frequency of pathological sleepiness, frequency of naps with stage 2 sleep, or frequency of REM naps were observed. Interestingly, no correlation was found between level of sleepiness and nocturnal sleep parameters or specific DAs.
Conclusions PD patients with SEs have a history of excessive daytime sleepiness: therefore, SEs are not simply the result of insufficient sleep or the effects of a specific DA. Additional studies are needed to assess contributions of other drug effects, disease progression, and other as yet unidentified factors in the aetiology of SEs among PD patients.
Reference
1. Frucht S, Rogers JD, Greene PF, Gordon MF, Fahn S (1999). Falling asleep at the wheel: motor vehicle mishaps in per-sons taking pramipexole and ropinirole. Neurology; 52:1908–1910.

P 1161

Sleep abnormalities in neurodegenerative diseases

J. Vankova
1st Medical Faculty Charles University, Prague, CZECH REPUBLIC

Five patients with Niemann-Pick disease, type C (NPC) were included in the study. The clinical diagnosis was confirmed biochemically and by the presence of foam cells on bone marrow. Deterioration of intellectual function, the presence of pyramidal, dystonic and cerebellar features and splenomegaly were observed in all cases. Only one patient reported cataplexy. Nocturnal polysomnography revealed interrupted and disorganized night sleep in all patients. The total sleep time was significantly shortened; sleep efficiency was lowered compared to age-matched controls. A decreased amount of REM and delta sleep was seen in all the patients. Altered sleep patterns included EEG sigma activity connected with rapid eye movements and muscle atonia, atypical K-complexes and spindle activity, and the presence of alpha-delta sleep. All NPC cases exhibited fragmental myoclonus. Shortened mean sleep latency was observed during MSLT, SOREMs were found only in the one case with cataplexy. Human leukocyte antigen typing and CSF hypocretin-1 levels were tested in 4 patients. The patient with cataplexy was HLA DQB1 * 0602 positive, while other subjects were negative. The hypocretin levels were reduced in 2 patients (one with cataplexy), in the remaining 2 patients the levels were at the lower limit of the norm. The mean value of hypocretin level (204.8±39.3pg/ml) in the NPC group was significantly lower than the controls (265.8±48.8 pg/ml). The findings suggest that lysozomal storage affects brain structures including lateral parts of hypothalamus; these changes may be partially responsible for sleep abnormalities and a manifestation of cataplexy in NPC patients.

P 1162

Cancelled

P 1163

The effects of electrical stimulation on the H-reflex, F-response and muscle hypertonus in children with hemiplegic cerebral palsy (CP).

E. Kavlak[1], T. Sahiner[2], U. Cavlak[3]
[1]Pamukkale University, School of Physical Therapy and Rehabilitation, Denizli, TURKEY, [2]Pamukkale University School of Medicine Department of Neurology, Denizli, TURKEY, [3]Pamukkale University School of Physical Therapy, Denizli, TURKEY

Introduction This study was planned to search the effects of electrical stimulation on the H-Reflex, F-Response and muscle hypertonus in children with hemiplegic CP.
Methods 16 children (8 boys and 8 girls) who suffered from hemiplegic CP were included in this study. The average of age of the subjects was 6.25±2.89 years. All the subjects were treated with Bobath therapy plus tetanic Faradic stimulation (TFS). TFS was applied on the tibialis anterior muscle, on the affected side. The subjects received TFS during five weeks (four days a week, 20 minutes). The effects of TFS on H- reflex, F and M responses of the m. gastro- soleus on affected side were evaluated using electromyography. Modified Ashworth scale was used so as to evaluate muscle hypertonus.
All the data obtained from this study was calculated using SPSS for WINDOWS' 98 version.
Results At the end of the study, after TFS we observed a decreasing in spasticity according to the modified Ashworth scale ($p<0.001$). In the other hand, there was no significant difference about H- Reflex, F and M responses of the gastro-soleus muscle between affected and unaffected sides ($p>0.05$).
Conclusion The use of TFS in non-spastic muscle (m. tibialis anterior) did not provide any difference H- reflex, F and M responses of the spastic muscle in children with hemiplegic CP.

P 1164

Fixated set in children with various types of tics

S. Natriashvili, G. Natriashvili, N. Geladze
Tbilisi State Medical University, Tbilisi, GEORGIA

The aim of our study was to estimate the role of fixated set in children with various types of tics for revealing the possible correlates with clinical patterns. 90 children from 5 to 15 years were investigated (40 girls, 50 boys). The diagnosis was based on the findings of subtle anamnestic, laboratory, electroencephalographic and cardio logical investigations. In 19 patients, tics were manifested on the background of sub acute rheumatic encephalopathy (rheumatic tics). In 28 patients-on the background of minimal residual dysfunction (organic tics). In 22 cases tics were observed with some kinds of neurotic disturbances – nocturnal enuresis, logo neurosis, phobias, or only tic disorders without any egzogenity (functional tics), 17 children were diagnosed to have obsessive condition with anancastic patterns and tics (neurotic tics), Tourette syndrome was recognised in 9 cases, with coprolalia in 4 cases. (13–15 years old boys).
All patients underwent the psychological test of set fixation by method of D. Uznadze in haptic and visual modalities. We estimated the level of set excitation, extinction and irradiation and the types of set – dynamic, static, variable sets and their subtypes. Then we defined correlations between this or that type of set and various types of tics.
Set fixation presents not only the supplementary high effective method for the diagnosis of various types of tics but it also gives the possibility of prognosing tics tolerance upon the course of disease and treatment.

P 1165

Effectiveness of tetanic faradic stimulation on range of motion and gait parameters in children with hemiplegic cerebral palsy (CP)

E. Kavlak[1], U. Cavlak[1], T. Sahiner[2]
[1]Pamukkale University, School of Physical Therapy and Rehabilitation, Denizli, TURKEY, [2]Pamukkale University School of Medicine Department of Neurology, Denizli, TURKEY

Introduction The aim of this study was to investigate the effects of electrical stimulation in non-spastic muscle (M. Tibialis Anterior) on spastic Gastro-Soleus muscles on affected side in children with hemiplegic CP.
Methods 16 children (8 boys, 8 girls with mean of age 6.25±2.89 years) diagnosed with hemiplegic CP were included in this study. All the subjects were treated using Bobath therapy plus tetanic faradic stimulation (TFS) on affected side (m. tibialis anterior). TFS was applied during 5 weeks (4 times a week, 20 minutes).
16 subjects were evaluated three times during the study (before treatment, the end of treatment and one month later) using universal goniometer and gait analysis. Range of motion of ankle-foot dorsi flexion on affected side was measured passively. Using goniometer, gait analysis was done to evaluate walking

characteristics such as support surface, step length, step width, heel strike and cadence.
The data obtained from this study was calculated using SPSS for WINDOWS'98 version
Results After treatment, the result of the goniometric measurements of ankle-foot dorsi flexion on the affected side showed high significant improvement statistically ($p<0.001$). However, this improvement didn't continue till first month after treatment ($p>0.05$). When the gait analysis revised, according to the considerations before, the end of treatment and one month later, a significant difference was found about support surface, only ($p>0.05$).
Conclusions As a result of this study, TFS showed just positive effects on range of motion of ankle-foot dorsi flexion in children with hemiplegic CP.

P 1166

Clinical electroencephalographic correlations of stammer in children

A. Mindadze, G. Natriashvili, S. Natriashvili
Tbilisi State Medical University, Tbilisi, GEORGIA

43 children with different forms of stammering have been examined. Age of the patients 3–10 years, 29 – male, 14 – female. Duration of stammering evolution varied from 1 month up to 6 years, and number of relapse amounted to 1–11. In 18 cases, obvious relation of stammer with exogenous factors has been revealed (group I– neurological form of stammering). In the remaining 20 cases, stammer has been insinuating, revealed without exogenous factors (group II– organic form of stammering). 5 patients of the group II have displayed convulsions in anamnesis and 3 other patients suffered epileptic seizures after revealing of stammer.
EEG research was carried out to all patients by 16-channel Medicor and 20 of them where additionally examined by computer EEG of brain mapping by means of Brain Survivor Saico apparatus. EEG patterns of group I patients predominantly varied within the limits of corresponding age norms. Patients of group II revealed paroxysmal activity of different qualities. It is noteworthy that the area of heightened excitability was mostly revealed at frontal-temporal abductions of the left hemisphere.
Drug therapy was carried out either by traditional preparations, tranquillizers, anti-depressant and sodium valproat (Depakin). In patients of group II positive therapeutic results of Depakin was revealed.

P 1167

The diagnostic dilemma in a child with visual loss and clinical symptomatology of demyelinating disease remains unsolved after a 4-year follow-up study

E. Tzamourani-Vontzalidou[1], A. Rossidou[1], A. Kiamili[1], N. Paterakis[2], Z. Parzaconi[3], G. Stavrakas[2]
[1]General Hospital of Elefsina, Neurology Dept., Athens, GREECE, [2]General Hospital of Elefsina, Opthalmology Dept., Athens, GREECE, [3]General Hospital of Elefsina "Thriassio", Athens, GREECE

In 1999 we presented the case, a girl 6.5 years old, who after a viral infection presented unilateral visual failure and spastic paraparesis, by arising the question whether Devic's disease is a unique syndrome of unilateral visual failure or a manifestation of multiple sclerosis.
The girl, 16.5 years old now, had been in therapy with presolone in combination with gamma globulin IV for 2.5 years. At the beginning of the therapeutical course, she presented two attacks, loss of vision 2/10 in the right eye. The last 2 years of the therapy the recovery led to vision 7/10. Two years without therapy – the recovery of vision is stable. Magnetic resonance imaging brain and optic nerves – normal.
If it were Devic's disease, could it be possible to expect recovery of the vision under treatment with presolone-gamma globulin, or the episodes of the loss of vision were a manifestation of multiple sclerosis?

P 1168

Parry-Romberg disease, case report

P. Szymanski, K. Tomczykiewicz, R. Janda, K. Macek, J. Kotowicz
Military Medical Academy, Warsaw, POLAND

The Parry-Romberg disease (progressive facial hemi atrophy-PFH) is a rare neurological disease. The aetiology and pathophysiology is unknown. Symptoms and physical findings associated with Parry-Romberg disease usually become apparent during the first decade of life or early in the second. In rare cases, the disorder is apparent at birth. This entity is characterised by atrophy of the skin, the subcutaneous tissue, sometimes the underlying bony structures, as well as the eye, the muscles, the larynx or pharynx. The authors present an unusual case of 42-year-old man with right side hemi atrophy (face, trunk, hand and leg) that began at the age of 6 and has progressed slowly until now. Family history was negative. Apart from the atrophy of the subcutaneous tissue and muscles of the right side of the body, general and neurological examination revealed no abnormalities. EMG examination showed delayed conduction time in the right personal nerve and the features of regeneration in the right tibialis anterior muscle. MRI, angio-MRI, CT scans, TCD EEG, as well as laboratory tests significant for collagenosis and scleroderma were normal.
Conclusions Sympathetic skin response test and termovision camera imaging, did not confirm damage within the autonomic nervous system, which has so far been reported.

P 1169

Cerebral blood flow velocity acceleration in a sleep apnoea syndrome with intracranial arterial stenosis

S. Behrens[1], K. Spengos[2,1], M. Daffertshofer[1], M. G. Hennerici[1]
[1]Dept of Neurology, University of Heidelberg, Klinikum Mannheim, Mannheim, GERMANY, [2]Dept of Neurology, University of Athens, Medical School, Athens, GREECE

Sleep apnoea syndromes (SA) of different etiologies (central, obstructive or both) are known to induce complications, such as tiredness, daytime sleeping, chronic headache and cardiovascular diseases including stroke. However, the exact mechanisms involved in cerebral ischemia are obscure. We measured the cerebral blood flow velocities (CBFV) by means of transcranial Doppler sonography in an 81-year old patient who presented with an acute ischemic stroke caused by an intracranial middle cerebral artery (MCA) stenosis in the presence of SA-syndrome. During apnoeas simultaneous recordings revealed reduced intraarterial oxygen, but increased carbon-dioxide-saturation, which resulted in an increased CBFV (220–303 cm/s) and suggested intermittent hemodynamic significance of a structurally only moderate MCA stenosis. Intracranial artery stenosis can become hemodynamically significant due to episodic hypercapnia in patients with SA. This may cause ischemic infarction in peripheral vascular territories.

P 1170

Sleep disorders in patients with hypothalamic syndrome of complicated genesis

M. G. Zhestikova
Institute of Postgraduate Training, Novokuznetsk, RUSSIAN FEDERATION

Aim The aim of our research was to study insomnic disorders in patients with hypothalamic syndrome of complicated genesis.
Material 359 of 450 patients studied there were women and 91 were men, ranging in age from 18 to 55 yrs. Hypothalamo-hypophyseal disorders of multifactorial aetiology have been detected in all our patients. Insomnia lasted for 1 wk to 10 yrs, the rate of night sleep disorders was found to be from 1 to 7 times per week.
Methods The following study methods were employed: clinical neurological, neurophysiology, magnetic resonance imaging (MRI), standard questionnaire tests approved by WHO, statistical analysis.
Results The reported complaint of sleep deprivation was rather common, with the prevalence rate of 66.85% in females and 57.14% in males. Frequent night arousals have been detected in 27.85% women and in 38.46% men. It should be noted that patients of the latter group were over 50 yrs of age and total sleep time was 4 hrs and 50 min. Sleep was characterized as "light sleep/direct speech". Many patients, 87% females and 86% males, complained of having dreadful dreams. Morning awakens was reported as "difficult/non-refreshing arousal" by 97% females and 89% males. Prevalence rate of presomnial disorders was 64.88%, of intrasomnial disorders it was 30%, and of postsomnial disorders it was 5.12%. In the elderly group, patients suffered from hypothalamic syndrome in the structure of "sleep-awakeness" circle.
Conclusion Sleep disorder in patients with limbic system dysfunction is found to be of great concern and needs proper diagnosis and treatment.

P 1171

Are the sleep patterns of fatal familial insomnia (FFI) related to different prion protein gene (PRNP) polymorphisms?

A. Garay[1], A. Reder[2], E. Van Cauter[2], S. Blanco[3], O. Rosso[3], J. Spire[2]
[1]CEMIC, Buenos Aires, ARGENTINA, [2]University of Chicago School of Medicine, Chicago, IL, USA, [3]Instituto de Cálculo, FCEyN-UBA, Buenos Aires, ARGENTINA

Objective To re-evaluate sleep patterns reported in a patient with FFI (D178N-129M plus 24bp deletion; (Neurology 1995; 45:1068) based on new molecular genetic evidence.
Methods We analysed extensively polysomnograms (PSGs) totalling over 100 hours performed between 4 to 1 months prior to death.
Results Total Sleep Time (TST) was reduced to 8.3± 3.8% of Total Recording Time (TRT); atypical REM sleep (aREMs) onset (6/6 PSGs) and aREMs preceding atypical NREM sleep (aSWS) devoid of spindling activity (3/6 PSGs) were observed during these studies. Sleep had preserved circadian (Nocturnal PSGs: 71.1±19.2 min.; Diurnal PSGs: 24.0±12.0 min., $p<0.05$) and ultradian periodicity (aREMcycle1: 98.1±30.4 min; aREMcycle2: 96.1±31.6 min.), and aREM presented a cyclic alternating pattern (CAP). Nocturnal dose of gamma-hydroxybutirate (GHB) produced a significant increase of aSWS.
Conclusions Recent data concerning polymorphisms of the PRNP give new insights about the phenotypical expression of pathogenic PRNP: 1) the influence of met/met polymorphism at codon 129 on the decrease of spindling activity (Neurology 2001; 56(3):A9), with the consequent inability to initiate sleep could allow the reversal of NREM-REM sleep presentation; 2) the octapeptide repeat deletions could play a causative role in the modulation of prion replication and disease presentation (Neuron 2000; 27(2):399; Neurology 2001; 57:354). It will be important to see whether other patients carrying these mutations exhibit similar altered sleep patterns.

P 1172

Mirtazapine induces confusion with hallucinations and REM sleep behaviour disorder in parkinsonism

A. Thomas, A. Luciano, D. Iacono, G. D'Andreamatteo, M. Onofrj
Neurophysiopathology, Pescara, ITALY

Mirtazapine is a new noradrenergic and specific serotonergic antidepressant drug. Shortly after the initiation of mirtazapine treatment of depression we observed the appearance of hallucinations, psychomotor agitation and cognitive changes accompanied by REM sleep behaviour disturbance (RBD) with apparent dream enactment in two male patients (1,2) affected by L-Dopa responsive parkinsonism and the appearance of RBD in a third male patient, with rigidity and cogwheeling in the left arm. These disturbances promptly remitted after the drug discontinuation and did not reappear in the following year (patients 2 and 3) and in following 2 years (patient 1). Polysomnography evidenced short onset REM sleep (SOREM) and lack of muscle inhibition with REM sleep during mirtazapine treatment. The observation might suggest that RBD can be triggered also by a drug lacking anticholinergic activity.

P 1173

Delayed diagnosis of the Kleine-Levin syndrome and treatment with Modafinil. Report of two cases

S. Boufidis[1], A. Karlovassitou[1], A. Balla[1], T. Karapanayiotidis[1], E. Vlahoyanni[2], P. Hamlatzis[1], S. Baloyannis[1]
[1]Aristotle University of Thessaloniki, Thessaloniki, GREECE, [2]"Agios Pavlos" General Hospital, Thessaloniki, GREECE

Introduction The Kleine-Levin Syndrome (KLS) is a rare disorder characterized by periodic hypersomnia, compulsive hyperphagia, behavioural-emotional disturbances and sexual disinhibiton lasting from a few days to a few weeks with almost complete remission during the intercritical periods. Very often the syndrome is under-diagnosed.
Method – Results Two cases are reported: a case of a 15 year old boy, in whom the diagnosis was delayed for 2 years, and one of a 34 year old man, who was diagnosed as KLS with a delay of 12 years. Upon their admission, we conducted a clinical assessment, a sleep study, a psychiatric assessment, as well as a brief neuropsychological evaluation which supported the diagnosis. The choice of treatment with modafinil was based on supposition of a hypothalamic dysfunction. Both of our patients responded to the treatment with modafinil and remission of symptoms has continued (for a period for more than 2 years).
Conclusion KLS often is diagnosed with delay and modafinil seems to be a promising help for the remission of symptoms.

P 1174

Polysomnographic findings in 40 patients with Parkinson's disease

N. J. Diederich[1], C. L. Comella[2], A. Bumb[1], J. Tiete[1], P. Lyen[1], M. Friedgen[1]
[1]Centre Hospitalier de Luxembourg, Luxembourg, LUXEMBOURG, [2]Rush Presbyterian St Luke's Medical Center, Chicago, IL, USA

Objective To describe the sleep abnormalities using polysomnography (PSG) in randomly selected patients with idiopathic Parkinson's disease (PD).
Background A variety of sleep abnormalities have been described in PD, ranging from primary involvement of sleep regulating centres to nocturnal recurrence of PD symptoms and side effects of pharmacotherapy. PSG studies have been rarely reported.
Patient sample and methods 40 PD patients (30 men, 10 women; mean age 65 [44–80] years; mean Hoehn-Yahr stage 2.5 [1–4]) underwent a PSG study with a 16-channel montage. All patients were taking their usual medications.
Results The mean sleep latency was 42+/–56 min. The mean percentage of deep sleep was 18+/–13 (17 patients with more than 20%). The average number of macroarousals was 29. The mean REM sleep percentage was 9.6 with a REM sleep latency of 171 min. No patient demonstrated a REM episode at sleep onset. There were polysomnographic or videographic signs of REM sleep behaviour disorder (RBD) in fifteen patients (38%). Nine patients (23%) had a periodic leg movement (PLM) index/ hour greater than 10. Twelve patients (30%), with a mean body mass index of 26, had a sleep apnoea syndrome (SAS) with a respiratory depression index/hour greater than or equal to 10; their mean lowest nocturnal oxygen saturation was 84 [58–90]%.
Conclusion In this series of PD patients, we found high percentages of RBD, SAS and PLM. Further studies assessing the risk factors and the prevalence of these abnormalities are indicated.

P 1175

Polysomnographic evaluation of daytime sleepiness and sleep attacks in Parkinson's disease

R. Manni, C. Pacchetti, M. Terzaghi, I. Sartori, R. Zangaglia, F. Mancini, V. Rustioni, D. Marchese
IRCCS Institute of Neurology "C.Mondino", Pavia, ITALY

Objective to evaluate daytime sleepiness, namely the features of sleep attacks in Parkinson's disease (PD) patients.
Study design A structured clinical interview, the Epworth Sleepiness Scale (ESS) and continuous 24-hour ambulatory polysomnography (A-PSG) were used to assess daytime sleepiness in a cohort of thirty-one PD patients.
Results Absence of abnormal daytime sleepiness was reported in six patients (group A). Rare microsleep and early afternoon voluntary naps were documented on A-PSG monitoring.
Mild to moderate sleepiness not resulting or rarely resulting in sleep only during moments of extreme relaxation was reported in twelve patients (group B).
Moderate to severe subcontinuous daytime sleepiness and an episodic urge to sleep sometimes actually resulting in sleep was reported in thirteen patients (group C).
The features of so-called sleep attacks, i.e., a sudden occurrence against a background of wakefulness, without heralding symptoms, were reported in eight of these patients with instances of falling asleep during the day. Polygraphic patterns of sudden NREM sleep onset were recorded in two patients of group C.
Group C showed a mean global ESS score significantly higher than group B and an higher occurrence of PSG-A documented microsleep and intentional naps.
Conclusions Sleep attacks can be clearly documented by A-PSG monitoring, represent a real, even if rare, occurrence in PD, and seem to represent the extreme of the continuum of daytime sleepiness observed in PD patients.
Patients with long-lasting disease and higher L-Dopa and/or dopamine agonist usage should be carefully evaluated in relation to the risk of developing sleep attacks.