

vitamin B6. The control follow – up at 14 months of age showed normal psychomotor development.

Severe infantile vitamin B12 deficiency is rare and usually due to maternal nutritional deficiency (vegetarian and vegan diet) or malabsorption (gastric resection, pernicious anaemia, Crohn's disease, celiac disease and other).

In this case the origin of mother's normochromic and normocytic anaemia during pregnancy was not diagnosed and as usual, she only was supplemented with folic acid and iron. In conclusion possible mechanisms of the influence of vitamin B12 metabolism on the nervous system based on the literature data are presented. The aim of this case is to draw attention to the importance of adequate supplementation to prevent potentially irreversible neurologic damage.

396 FRONTAL LOBE EPILEPSY WITH GELASTIC SEIZURES

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Gelastic epilepsy are an uncommon seizure type most often symptomatic associated with hypothalamic hamartomas, with a prevalence rate of about 0,5 per 100 000. However, idiopathic and cryptogenic cases with no evidence of cortical structural lesions have also been described. The term gelastic comes from Greek word gelos meaning laughter. Laughter is pathological in nature and can be spontaneous without obvious cause.

We present a 13-year-old girl without structural lesions, manifesting gelastic seizures. The seizures were not associated with feelings of mirth.

Repeated 1,5T MRI revealed no structural abnormality. Interictal EEG showed paroxysms in the right frontal region. Ictal EEG demonstrated diffuse attenuation, followed by fast activities and spike-wave complexes predominantly over the right hemisphere. Unlike the seizure from temporal lobe, semiological investigations revealed that the laughter in our case was not accompanied by a subjective feeling of mirth, and an interictal EEG showed frontal spikes. Results of neurological examination were normal. The Wechsler intelligence scale for children IV (WISC-IV) revealed a mild mental retardation with the intelligence quotient score <70. An interictal FDG-PET CT showed hypometabolism over the right superior frontal lobe and right medial temporal lobe. The seizures were resistant to oxcarbazepine, levetiracetam, valproate and lamotrigine and were suppressed by topiramate monotherapy. Further clinical examinations (high-resolution 3T MRI, ictal and interictal SPECT) will be done.

In conclusion, gelastic seizures are commonly associated with hypothalamic hamartomas but ictal laughing, has been reported with temporal lobe and frontal lobe seizures. Although gelastic seizures have been described as intractable, a few medications including valproic acid, lamotrigine, levetiracetam and steroids were reported to be effective in patients without hypothalamic hamartoma. In our patient topiramate monotherapy proved to be effective in the treatment of gelastic seizures without hypothalamic hamartoma.

397 LONG-TERM MONITORING AND OUTCOME OF HIGH NEURORISK PRETERM INFANTS

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Perinatal brain damage is non-progressive and the processes of maturation and plasticity along with medical procedures can lead to functional recovery. About 3% of newborns are high neurorisk and especially susceptible to brain damage.

The aim of this study is to show the neurodevelopmental outcomes of 77 preterm infants born ≤ 32 nd week of gestation, at the age of nine.

We studied and habilitated 170 high neurorisk children born in Zagreb's largest maternity hospital between 2007 and 2008. At the age of nine, 132 children have remained continuously studied, 77 of whom were preterm infants born ≤ 32 nd week of gestation. All the children were assessed using the Touwen examination. Children with CP were classified according to motor function classifications. Furthermore, all children have undergone EEG, early neonatal brain ultrasound, whereas children with CP had MRI as well.

Infants were classified as 'high risk' according to factors of risk.

At the age of nine years, out of 77, 17 children (22,1%) developed CP. As for associated disabilities, 6 children (35,3%) had an IQ below 70, severe speech impairment had 8/17 (47%), visual impairment 14/17 (82,4%) and 4/17 (23,5%) severe sight loss. 12/17 (70,6%) had refractive error and strabismus. Four children (23,5%) developed epilepsy. Brain ultrasound showed the most severe forms of periventricular leukomalacia for 8 children, another eight children had the most severe forms of peri – intraventricular haemorrhage, while only one child had no abnormality detected. MRI was performed with 13 children, 12 of which suffered from predominant white matter damage, and one of them had additional thalamic haemorrhage. A normal motor outcome was observed in 60/77 children (78%). Of the children with normal motor outcome, one third had refractive error or strabismus, 25/77(32%) suffered from specific language development disorders, while almost a quarter had pathological EEG.

High neurorisk children are particularly susceptible to brain damage in the pre- and perinatal period. Because of that, they need to be detected early, their development needs to be monitored even at school age and they must be included in habilitation programmes. The process of selecting, registering and habilitating children is simple and easily applicable. For that reason, it should be carried out with all preterm infants.

398 LONG-TERM OUTCOME OF EPILEPTIC SEIZURES AND ADAPTIVE BEHAVIOR IN PATIENTS WITH WEST SYNDROME -EXPERIENCES FROM OUR CENTER

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West syndrome is a specific, age dependent electroclinical syndrome and one of the most common and well described form of epileptic encephalopathy. In this study, we describe outcome regarding epilepsy and adaptive behavior in patients with WS treated in our clinic.

Patients with WS treated in our clinic from 2000 -2018 were followed up regularly for at least 2 years. For this study, their adaptive behavior was measured using Vineland-II Adaptive Behavior Scales. Adaptive behavior (AB) refers to the skills needed by individuals to function and be self-sufficient within their everyday environments. Vineland-II assesses adaptive behavior in four domains: communication, daily living skills, socialization and motor skills, each divided in more subdomains. It provides standardized scores in every domain and is applicable from birth throughout life.

A total of 64 patients were enrolled in this study, 38 males (59%) and 26 females (41%). Symptomatic WS had 46 patients (72%) and 18 patients (28%) had cryptogenic WS. One patient with symptomatic WS died during follow up.

Regardless of response to initial treatment and resolution of infantile spasm, 20 patients (31%) developed different kind of seizures during follow up and are still taking antiepileptic therapy, 17 of symptomatic (37%) and 3 of cryptogenic WS (16%).

Among all patients, 4 (6%) have no deficit in adaptive behavior in all measured domains and subdomains while 40% of patients have severe or profound deficit. Significant differences were found between symptomatic and cryptogenic form of WS. In cryptogenic WS, most of the patients have moderately low AB, 66% of them, 17% have AB and 17% have low AB. In symptomatic WS moderately low AB were measured in 37% of patients, low AB in 62% and only 4% had adequate AB. Low AB was also in negative correlation with seizure freedom.

Analyzing each domain and subdomain of adaptive behavior, the lowest scores in almost every patient (patients with adequate AB were excluded) were measured in communication domain, more specific, in expressive speech and in subdomain of community daily living skills.

In this study we confirmed that the most important prognostic factor for outcome of epilepsy and psychomotor development in WS is underlying etiology of the syndrome. Despite the etiology, the great majority (94%) of the patients with WS need some kind of help in everyday functioning throughout lifetime.

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ELECTRICAL STATUS EPILEPTICUS IN SLEEP (ESES): CLINICAL AND EEG CHARACTERISTICS AND RESPONSE TO TREATMENTS

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Electrical status epilepticus in sleep (ESES) is defined as an age related, self-limited epileptic encephalopathy. It is characterized by heterogeneous clinical manifestations and a specific electroencephalographic (EEG) pattern of continuous spikes and waves during slow sleep (CSWS)

Objective The aim of this study was to describe the electroclinical spectrum in children with electrical status epilepticus in sleep (ESES), and assessment of treatment pattern.

Methods Clinical data of 16 patients with ESES/CSWS syndrome who were treated and followed at least two years were analyzed. Inclusion criteria were as follows: (1) Determination of the ESES pattern on the EEG and a (2) follow-up period of two to four years. All patients underwent a clinical evaluation including history, physical and neurological examinations, sleep and awake EEGs, and brain MRI. Patients with an underlying etiology were classified as symptomatic while others were classified as idiopathic.

Records of EEGs of patients were reevaluated to determine two aspects of ESES: (1) The spike-wave index (SWI) on the NREM sleep EEG and (2) the area of maximum amplitude of continuous epileptic activity. The SWI on the NREM sleep EEG during the ESES period was visually calculated. The ranges of the SWI considered were as follows: N85– 100% (typical ESES) and 50–85% (atypical ESES). We also defined the ESES pattern as anterior if the maximum amplitude of spike-waves was in the frontal, frontocentral, or frontotemporal areas and as posterior if the maximum amplitude of spike-waves was in the posterior temporal, temporo-occipital, or occipital areas on the EEG.

Results Complete data were available in 16 children. Age at ESES diagnosis ranged from 36 to 84 months. Antiepileptic drugs were used as first treatment for ESES in 16/16 (100%). Electrical status epilepticus in sleep initially resolved in 87%, but 56% had subsequent relapse. At last follow-up, ESES resolved in 56%. And those children was seizures free.

Conclusion We found high failure rate of first line AEDs in preventing ESES, and high relapse rate. There are no standardizations of management of ESES. We were managed by our experiences, relevant medical records and clinical trials.

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POSTNEONATAL CEREBRAL PALSY – TIMING AND ORIGIN

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Cerebral palsy (CP) is most common severe neurodevelopmental disorder, affecting 2-3/1000 live borns. Postneonatal cerebral palsy is a rare and distinct form of CP in which damage to immature brain occurs after the newborn period until the age of 24 months. Surveillance of Cerebral Palsy in Europe (SCPE) is European project encompassing 26 currently active centers from 21 European countries. Common database of children with CP is one of the main assets, providing better insight even in the rare CP types.

Croatian SCPE register is enrolled since 2012 as „C28 RCP-HR Register of cerebral palsy of Croatia”, and is affiliated to Children's Hospital Zagreb. Object of this study is to address children with postneonatal origin of CP in our National Register and the main causes of this rare CP.

This population-based study included 502 children from C28 RCP-HR born 2003-2009, living at the area of Register at point of registration, with postneonatal cause of cerebral palsy. Patients were stratified according to the timing and origin of the lesion.

Out of total of 502 children with CP (total CP prevalence 2.11/1000), 11 had postneonatal origin, presenting 2.19% of total CP population or 0.46/10000 live borns. Most common