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CLINICAL MANIFESTATIONS OF PATIENTS WITH PANDAS IN PATIENTS FOLLOWED UP AT SREBRNJAK CHILDREN'S HOSPITAL IN 5-YEAR PERIOD

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PANDAS, or pediatric autoimmune neuropsychiatric disorder associated with *Streptococcus pyogenes*, is characterized by an acute onset of neuropsychiatric symptoms which seem unexplainable. These symptoms usually include obsessive-compulsive disorder, and a variety of tics, that appear in children before puberty.

The aim of this study was to compare the clinical manifestations of our patients with PANDAS to the latest studies and report our findings.

This retrospective study included 20 patients with valid clinical features of PANDAS who were treated and followed at Srebrnjak Children's Hospital between 2014 and 2019.

Our examinees were children before puberty which fulfilled the main criterion of early-onset and at least two major criteria – obsessions/compulsions (OCD) and neuro-motor dysfunction (tics, hyperactivity). We retrospectively analyzed all the data with special focus on neuropsychiatric symptoms.

In 5-year period a total of 20 children, 16 (80%) male and 4 (20%) female, aged 3 to 11 years (median age 6,5) were followed up under the diagnosis of PANDAS. Only one patient had a burdened perinatal history (prematurity, short-term mechanical ventilation, neuromotor habilitation). Family history of tics and autoimmune disorders was positive in N=5 (25%). All patients had OCD and tics with acute onset of symptoms.

Elevated levels of antistreptolysin titers were found in all but one patient (95%). Anti-DNAse-B levels were tested in 7 patients and was elevated in 4 (57%).

Attention disorders were present in N=17 (85%) and 4 (20%) patients had sleeping difficulties. Speech difficulties were detected in N=5 (25%).

Decline in school performance was also common in children attending school N=10 (71%).

None of the patients had pathological changes in EEG, and those with neuroimaging studies (N=12 or 60%) did not have any specific findings.

First-line antibiotics were given to 15 (75%) patients. In 5 (25%) patients a full regression of symptoms was registered, 10 (50%) had partial improvement, 5 (25%) had no response. In 7 (35%) patients intravenous immunoglobulins (IVIG) were given after initial antibiotic treatment with incomplete regression. Tics persisted in 11 patients and 4 had tics with behavioral difficulties. In 7 patients treated with IVIG we proceeded with parenteral antibiotics because of partial symptom regression and 2 were given corticosteroids (still followed).

We observed a higher rate of full symptom regression with first-line parenteral antibiotics and some of them improved after IVIG. Further research on larger cohorts of patients are necessary for better management of this clinical entity.

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PATTERNS OF INTERNET USE BY RUSSIAN SCHOOLCHILDREN

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Study the amount of internet use by Russian schoolchildren and its connection to the other factors of social functioning.

Schoolchildren from 9th grade from 5 major cities in Russia participated in the research. The questionnaire concerning out-of-school life and well-being was used. The questionnaire was filled in by the parents.

A total of 598 children were examined, with an average age of 14,83 years, 48,7% of them being girls.

It was found that during the schooldays 26% of the children do not use the internet at all, 16% – spend less than an hour a day; 20% – spend around 1-2 hours a day on the internet; 15% – 2-3 hours a day; 23% – 3-4 hours a day. A direct connection was revealed between internet use and interest in computer games ($r = 0,95$).

Clear gender distinction was found: among the children using the internet more than 3 hours a day – 64,1% were boys. Among the children not using the internet at all or spending less than an hour a day on the internet – 71,8% were girls.

Children who use the internet a lot (more than 3 hours a day) are more likely to sleep less than the others: less than 8 hours (50,8% against 42,6%). However, there is no significant distinction between those who do not use the internet at all or use the internet 1, 2, or 3 hours a day.

Further, children who use the internet for more than 4 hours a day participate less in sports-related activities and clubs (25,8% against 37,9%). However, there is no significant distinction in terms of the involvement in sports between those who do not use the internet at all or use the internet for 1, 2, or 3 hours a day.

There is a direct connection between internet usage and interest in computer games. Boys spend significantly more time on the internet than girls. This could be explained by the faster development of higher mental functions for girls and less interest in computer games. There are no significant distinctions between the children who use the internet moderately and the children who do not use the internet at all. However, the children who use the internet a lot, sleep reduced hours, and are less interested in sports.

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BLOCH-SULZBERGER SYNDROME – A RARE X-LINKED DERMATOSIS

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Bloch-Sulzberger syndrome is a rare genetically determined dermatosis, manifested by skin changes in combination with

pathology of the eyes, skin appendages, teeth, central nervous and musculoskeletal systems. The disease manifests in the neonatal period and has four stages: vesicular, verruciform, hyperpigmented and hypopigmented. It occurs mainly in girls, since inheritance has an X-linked dominant character.

A 7-year-old girl, appealed to the department in satisfactory condition, with complaints of mental deficiency, reduced memory, behavior disorders, multiple hyperpigmented spots on the limbs and body. The child after birth had multiple vesicles on an erythematous background on the body, localized linearly, mainly on the flexor surface of the limbs, as well as on the skin of the chest and back. After birth, he was diagnosed with progressive erythema and vesicular rash. In connection with skin manifestations, she was examined for intrauterine infections – pathology wasn't detected. The patient had no family history of skin diseases. Subsequently, the appearance of hyperkeratosis in the form of plaques, warts and lichenoid growths along the Blaschko lines was noted in the affected areas of the body. The girl was under the supervision of a pediatrician. During the examination in the Department revealed: mental retardation, hyperpigmented spots of gray-brown color with light edges, on the extremities of the rash in the form of 'mud splashes', on the trunk – in the form of a 'spiral', ring 'or' marble cake', with the location of pigmentation along the lines of Blaschko.

The course of Bloch-Sulzberger syndrome is suspected in a child with clinical manifestations of mental retardation, stages of the skin process with a debut in the newborn period. To confirm the diagnosis, a molecular genetic examination was performed: mutations of the IKBKG gene, which is located on the X-chromosome, were detected.

Typical skin manifestations allow you to clinically suspect the presence of Bloch-Sulzberger syndrome in newborns, to conduct timely molecular genetic diagnostics to make the correct diagnosis. In view, the importance of differential diagnosis in the neonatal period and the rare disease, a multidisciplinary team of neonatologists, pediatricians and neurologists to this disease is necessary.

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FIRST EPISODE OF SPORADIC HEMIPLEGIC MIGRAINE IN PEDIATRIC PATIENT- DIAGNOSTIC PHANTOM

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The aim of this case report is to describe rare form of migraine in pediatric patient and to point out difficulties of making the right diagnose and classification according to ICHD-III (International Headache Society 2018.)

Case Report A 14-year old girl driven by emergency team to pediatric emergency from school where she had severe unilateral headache with nausea and paresthesia in the right hand and leg about an hour before arrival. At arrival she was confused, unknowing the name of her school and dysphasic. Neurological exam showed dysphasia and right hemiparesis, in the next half an hour she was somnolent, only opening her eyes when shaken and started vomiting.

After two hours (three from the first symptoms) all clinical signs vanished, she was in her full consciousness, ambulatory with no neurological deficits!

-Brain CT, EEG and MRI/MRA were normal, so did D-dimers and coagulogram, we transported her to tertiary level center

Differential Diagnoses 1. cerebrovascular disease- ischemic or hemorrhagic stroke; transient ischemic attack 2. CNS infections/encephalopathy/brain tumor 3. syndrome of transient headache and neurological deficits with cerebrospinal fluid lymphocytosis (HaNDL) 4. migraine

Diagnosis and Classification -Diagnosis is clinical, TIME has major role -It is the diagnosis of exclusion- when we exclude diagnosis from above and time shows that the attack was episodic and reversible, we can than afford ourselves to think about migraine -which type of migraine? According to ICHD -III it does not fulfill criteria for hemiplegic migraine since it takes at least two attacks with motor deficit, so we can only classify this case as 1.5.2: probable migraine with aura.

Conclusion Diagnosis of a first episode of sporadic hemiplegic migraine (severe attack) in a pediatric patient is very challenging for a clinician, especially in a first few hours What makes already dramatic clinical picture even harder to diagnose:

-children and adolescents find it hard to describe their aura symptoms- motor aura is necessary feature for the diagnosis, clinician must establish as clearly as possible whether the patient actually experienced motor weakness as an aura symptom -lack of literature about this cases; no guidelines for the management of acute attacks -signs and symptoms described in all of the other aura types- heterogeneous symptoms Regardless of the new technologies in diagnostic, accessible medical data bases and several headache classifications, some conditions will always remain challenging

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VESTIBULAR SCHWANNOMA IN A TEENAGER: CASE REPORT

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Relevance Vestibular Schwannoma (VS) – is a benign encapsulated tumor, initially arises in the inner auditory canal from the neurolemma of the vestibular nerve with further growth towards the base of the skull. Basically, they are associated with neurofibromatosis type 2 (NF2). In children sporadic cases of VS are uncommon. We report a case of VS in teenager.

Methods A 15-year-old girl with a viral infection was diagnosed with unilateral acute sensorineural hearing loss (SNHL). There was no NF2 family history. General medical examination revealed no ENT-pathology. MRI showed no tumor and vascular malformations in the brain and temporal region. After the standard course of dexamethasone effect was not obtained. SNHL progressed over the next 9 months. Evaluation of advanced hearing tests showed: tympanometry (probe-ton 226 Hz) demonstrated Type A bilateral, TEOAE were documented as 'Pass' bilateral, pure tone audiometry determined profound hearing loss on the right, Auditory Brainstem Response (ABR) was defined wave V latency on the right, ABR thresholds were detected with intensity stimulus level 85 dB SPL on the right and 30 dB SPL on the left. At the series X-ray CT images of temporal region were determined inner auditory canal deformation and extension. A second MRI scan revealed a bulky tumor coming from the inner auditory canal. The