Linear Scleroderma in Childhood

St. Stefanov, K. Lisichki, M. Bojidarova, A. Telcharova

Ten children with linear scleroderma were observed for a period of 13 years - 6 girls, 4 boys (ratio - 1.5:1), mean age of onset - 7.1 years (range - 2.6 - 13 years). The follow-up study is for a period of 1- 11 years.

Linear type of scleroderma „en coup de sabre" is observed in 2 patients. Eight children have „en bande" type lesions, 5 of them are combined with morphea. A positive ANA titer is detected in 2 patiens (28.8%). Visceral changes and overlap with other connective tissue disease were not observed. Hypotrophy/atrophy affecting also the skull is detected in all cases. Growth retardation and shortening of the affected limb is observed in 40% (4 patients), joint contractures in 6 children (60%).

This consequences in serious orthopaedic, neurological and psychological problems.

Key words: *Linear type, scleroderma, morphea*

Rubella Associated Arthritis in Childhood

St. Stefanov, A. Zvetanska, K. Lisichki, A. Telcharova

Seventeen children with rubella are observed - 10 boys, 7 girls (ratio 1,4 : 1) mean age of onset - 11,5 years (range - 6 - 14 years).

Arthritis/artralgia appeared within 1 - 45 cays (mean 9.6 days) following the rash. The diagnostic assessments is based on: 1. Clinical diagnosis of rubella in non-vaccinated children; 2. Arthritis/artralgia appearing whitin 6 weeks after the onset of rubella; 3. Serological confirmation of rubella; 4. Exlusion of other cases, related to the joint symptomathic.

Oligoarthritis predominated - 47%, followed by polyarthritis - 23.5%, monoarhritis - 17.6%. In 2 children (11.75%) only artralgia is observed. The knee joint most commonly affected - 76.5%, followed by the wrist and ankle - 58,8%, PIPS - 47% and rarely - other joints. Flexor tenosynovitis of the hand is observed in 2 children. The duration of joint symptomps is short and varies whitin 1 - 21 days (mean 6.4 days). Arhritis is presenting temporary and is benign one.

Key words: *rubella virus, arthritis, rubella antibody*

Bronchoobstructive Syndrome /BOS/ and Gastroesophageal Reflux /GER/

I. Galabov, D. Bliznakova, D. Petrov, M. Galabova

For a 4-year period overall 278 children with BOS were admitted at the I[st] Children's Clinic of the Varna Medical University; 2 groups were identified:

- ⇨ Children with BOS - 180 /64.74 %/, aged up to 4 years;
- ⇨ Children with asthma bronchiale /AB/ - 98 /35.25 %/, aged over 3 years.

The highly prevalent night episodes of bronchial obstruction and cough during sleep justified the investigations for GER.

In the Ist group we verified GER in 36.11 % /65 chil and we assumed that the latter is one of the etiological fac leading to bronchial obstruction.

In the children with AB we found GER in 14 chil /14.28 %/, that the GER is of secondary origin.

The retrospective analysis among different age gr found lack of correlation between the severity of GER the severity of bronchial asthma, as well as lack of correl between the GER symptoms and the frequency of bron obstruction attacks. The adequate treatment of the al diagnosed GER leads to positive influence upo bronchoobstructive syndrome in the I[st] group of chi while in the asthmatic children the exact treatment of as leads to decrease in the signs of GER.

Key words: *bronchial obstruction, bronchial asthma,*

Adjustment and Needs in Families v Mentally Retarded Children

V. Stancheva - Popkostadinova

In the process of care for their mentally retarded parents are faced by multivarious problems, which enced their psychosocial functioning. The aim of the is to evaluate the adjustment and problems in paren mentally retarded children. The semi-structured tionnaire is created for the purpose of the study. The show that parents have expressed needs for ad information an specialized help for the child with retardation, as well as for his education; informatio existing services and programs, adequate financial s and consultation, concerning personal problems, the process of the care. The provided questionr recommended as useful in the first meeting with the for development of good communication between and professionals and collection of useful informati

Key words: *mental retardation, family, pro professionals, and social support*

A Combination of Pseudohypopara thyreoidism and Systemic Lupus Erythematosus- Diagnostic and Therapeutic Challenges

V. Iotova, B. Varbanova, Y. Yotov

A 19 year-old patient is presented whose compl dated back to his early childhood with growth an retardation, seizures and characteristic morphologi such as round face, hyperthelorism, saddle-ba divergent strabismus, short and broad fingers an enamel damage. The physical examination and l investigations provided clinical and biochemica hypocalciemia, high serum phosphorus, obesity, lenses, and fibrous osteitis. On that basis the dia pseudohypoparathyreoidism (Albrigth's hereditar dystrophy, AHO) was established.

Since the patients was admitted becaus dominating complaints of fever, arthralgia, rash, we and fatigue, a careful monitoring and anoth investigations was undertaken in that direction. The

SUMMARIES

Overgrowth Syndromes. I Syndromes with Generalized Overgrowth

Z. Stanchev

Overgrowth Syndromes are a large group of diseases, mainly congenital and most frequently hereditary, characterized by a tall stature, malformative signs and tendency of different tumour development. Inspite of the great variety of overgrowth syndromes they may be separated in three basic groups syndromes with generalized overgrowth, syndromes with overgrowth of different body segments and syndromes with overgrowth of different parameters. In the present article are examined the clinic, evolution and prognosis of group with generalized overgrowth, as well as the factors causing it.

Key words: overgrowth - generalized, segmented, genetical factors, hormonal and metabolic factors

Sleep Related Breathing Disorders

S. Filchev

Sleep problems are common in childhood. Sleep-related breathing disorders (SRBD), in the absence of any primary lung abnormalities, are related to a deficiency of respiratory control. They are state dependent and typically cause significant clinical symptoms only during sleep. In this paper we discuss some respiratory control disorders that are related to congenital or acquired abnormalities that will result in substantial morbidity if not appropriately diagnosed and treated. These are mainly central hypoventilation syndrome (CHS), a defect in central respiratory drive, and obstructive sleep apnea (OSAS), an abnormality of control of upper airway patency. Also included are discussion of chronic lung disease and its potential for causing a SRBD. Infants and children at risk for SRBD or with symptoms of SRBD should be investigated for respiratory control disorders. Optimal treatment of SRBD in the developing children is of central importance for preserving normal behavior and neurocognitive development. Oxygen supplementation and/ or ventilatory support during sleep prevent uncontrolled episodes of hypoxemia, sleep fragmentation, and REM sleep reduction.

Key words: sleep, control of breathing, central hypoventilation syndrome, obstructive sleep apnea syndrome, chronic lung disease

udden Infant Death Syndrome

Georgieva

The trend of nowadays birthrate diminishing demands re cares for the life of every newborn. High risk newborns sf be known and qualified specialists should help them e quickly. Sudden infant death syndrome (SIDS) takes the ding place as a cause for child's mortality in many ntries. That is why it is necessary more researches to be ne - pathophysiological, biochemical, genetical, robiological, etc.,to find and explain the reason for the dden death. What is the possibility an apparently healthy infant to die if SIDS, it is still discussed. Ultrastructural anomalies are searched for to discover SIDS-victims earlier and treat them as high risk newborns. In some cases, clear causes for the fatal end are found and that excludes a diagnose SIDS.

Key words: sudden infant death syndrome, high risk newborns

Localized Scleroderma in Childhood - Clinical Observation

St. Stefanov, M. Bojidarova, A. Telcharova, K. Lisichki

Clinical observation of 12 children with localized form of scleroderma was performed. The study encompasses 7 girls, 5 boys - 1,5:1 ratio, mean age of onset 6,5 yrs (range 2-13 yrs). The follow-up study includes observations lasting from 6 month up to 11 yrs. Linear form of scleroderma was observed in 9 patients (75%), morphea - in 3 patients (25%) and combination of morphea and linear changes - in 5 children. Linear form „en coup de sabre" diagnosed in 1 child. Eight children are presenting the „en band" type linear changes. Generalized morphea, an extremely rare form of scleroderma is present in 1 child. Positive ANA titers are detected in 3 of 8 studied patients. Neither visceral changes nor overlap with other systemic disease was observed. Prognosis at vitam is good. One patients get handicapped due to: hypertrophy/atrophy of the affected limb or the skull in „en coup de sabre" forms. This symptoms affected all patients. Growth retardation and limb shortening are observed in 4 patients; joint contracture - in 6 children. All changes result in serious cosmetic, orthopaedic, neurological and psychological problems. Assessment was made in time only in 4 children. Delay of diagnosis (average 2 yrs) is detected in 8 children. Early complex treatment delays skin and muscle fibrosis, hypertrophy and growth disorders.

Key words: localized scleroderma, linear form, morphea

Twins Method for Investigaiton the Genetics of Children with Retinopathy of Prematurity

A. Popova

Purpose: To make clinical-genetic evaluation of ocular phenotype in twin pairs (TP) with Retinopathy of Prematurity (ROP).

Patients: In totally 42 twins with ROP originate from 29 TP in population of 246 children with ROP from 233 pedigree were investigated. All they are from passed the Pediatric Eye Department - Sofia and the School for visually disabled children - Sofia for the last 15 years.

Methods: Two general group of methods are applied:

I. Ophthalmological;

II. Genetical - genealogical analysis and twins method of C. Smith (1972) with modified model (pat. № 63067/23.04.2001). The zygosity of TP is evaluated according to 24 criteria.

Results: 7 TP are monozygotic (MZ), 15 TP are dizygotic

SUMARY

Clinical Laboratory Characteristics of Protein Losing Enteropathy in Children

D. Mondal, Hr. Zhelev, A. Radivenska, R. Koumanova, E. Lazarova, E. Pantaleeva, P. Yanev, K. Kunchev

Protein losing enteropathy (PLE) is being diagnosed by estimating alpha-1-antitrypsin in the stools. This is a non invasive method. In the accessible litrature only single communicationson clinical laboratory symptomatics in PLE related to this new diagnostic method are present. It gives a possibility ot disclose also subclinical forms of PLE by a harmless reliable method. Only a few communications proving the real prevalence of anaemia, hypoalbuminaemia and hypocalcaemia in patients with PLE if subclinical forms are being added, the latter being nowadays diagnosed. Present investigation reveals a lower serum level of totalprotein, albumin, cholesterol, triglycerides as well that of haemoglobin, calcium and iron this being caused by icnreased loss in pateints wuth PLE disclose this increased enteral loss leading to subnormal serum levels. Anaemia, hypocalcaemia, hyposideraemia are being found with a prevalence of 28,5%, 20.90%, 44% respectively. Hypoalbuminaemia, hypoproteinaemia reach 24,4%, hypoalbuminaemia - in 23,40% respectively.

Key words: *protein losing enteropathy, faecal alpha-1-antitrypsin, children*

Clinical Observations in Six Children with Aesthesioneuroblastoma

I. Hristosova, L. Marinova, Ts. Rouseva, Dr. Bobev, I. Shturbanov

Aesthsioneuroblastoma is a rare disease in childhood. For a period of 16 years (1981-1997) six children with olfactory neuroblastoma (from a total of 180 with neuroblastoma) have been diagnosed, treated and followed-up at the Paediatric Oncohaematological Clinic. Clinical Symptomatics is characteristic but scarce knowledge in oncology is causing a delay in diagnosis and treatment. All patients were in an advanced stage (B and C) which enhances prognosis turming to the worse. Complex treatment (surgery, radiology, chemotherapy) has been carried out. Four patients died within a period of 6 to 12 months after therapy. Two children are alive for a period of observation of 16 and 5 2/12 years respectively. The present paper discusses therapeutic approaches which could improve the survival rate.

Key words: *aesthesioneuroblastoma, childhood, radiotherapy*

Antioxidants and Products of Peroxide Oxidation of Lipids in Children with Rheumatologic Disease

A. Maneva, D. Mihailova, V. Gancheva, B. Taleva, K. Lisichki, N. Ivanova, St. Stefanov

Indices for the assessment ofthe system antioxidants / oxidants (general enzyme antioxidant activity, AOA), total serum content of antioxidants with - SH groups, nonprotein and protein thiol antioxidants and products of peroxide oxidation of lipids (POL) have been investigated in the sera of children with juvenile chronic arthritis (JCA), systemic lupus erythematosus (SLE), hypersensitive vasculitis, dermatomyositis and reactive arthritis. Results reveal that indices under investigation present different patterns different in different diseases and their estimation is of different diagnostic value. Disturbances in the system of antioxidants /POL in JCA disclose a deficit of -SH groups in serum proteins as well hyperproduction of POL. SLE discloses an inadequate weak increase in the production or secretion of enzyme antioxidants (AOA), deficit in the serum content of nonprotein thiol antioxidants and disturbance in the formation of POL. Changes in the system antioxidants/ oxidants in reactive arthritis reveal an increase in AOA which is two times weaker compared to that in JCA and much more presented compared to SLE. Deficit in this system of thiol antioxidants in reactive arthritis is commensurable by a normal content of POL. Submitted data reveal that in the different rheumatologic diseases it is appropriate to use parallel to antirheumatic additional natural or synthetic antioxidants. Their import would corect the antioxidant deficit and normalize serum POL content, especially in chidlren with collagen disease.

Key words: *antioxidants, products of peroxide oxidation of lipids, juvenile chronic arthritis, systemic lupus erythematodes, hyperreactive vasculitis, dermatomyositis, reactive arthritis*

Clinical Manifestations in Cerebrovascular Hypoplasiae in Childhood

V. Bozhinova, P. Dimova, L. Belopitova

Present study is being aimed at the analysis of clinical manifestations of cerebrovascular hypoplasiae in childhood. The clinical contingent includes 205 children aged 3 to 14 years of age. Cerebrovascular hypoplasiae are being diagnosed with the aid of angiography (n=63), with transcranial Doppler sonography (n=188) and with MRI angiography (n=4). In the teritory of the internal carotid artery (ICA) are localized 198 of them (97%) and in the vertebrobasilar system only 6 (3%). With transient ishaemic attacks (TIA) are 43 children (21%), with cerebral infarctions 35 children (17%), with migraine-like headache - 10 children (4,9%), with unilateral cerebral atrophic process - one child (0,5) and with local and/ or secondary generalized epileptic seizures 116 children (56,6%). Hypoplasiae in the ICA, often combined with loops and stenoses (n=86 or 41,9%) determine TIA (n=23) cerebral infarctions (n=12, 8 of whom during intranatal asphyxia with following hemiplegic form of cerebral palsy), migraine-like headache (n=6), focal or secondary generalized epileptic seizures (n=44). Hypoplasiae in the middle cerebral artery (n=111 or 54,1%) determine TIA (n=17), cerebral infarctions (n=18),, 12 of which in the neonatal period, migraine like haedache (n=4) and focal or secondary generalized epileptic seizures (n=12). Hypoplasiae in the anterior cerebral artery (n=2) and in the basilar artery (n=3) are the cause for cerebral infarctions in the vertebral arteries for TIA (n=3). The role of additional factors in childhood is being discussed (i.e. increased metabolic neds, hypovolaemia, intranatal asphyxia) for the haemodynamic significant failure of the cerebral circulation in the territory of the hypoplastic artery, as well as the possibilities of the noninvasive neuroimagig methods (transcranial Doppler sonography, MRI angiography) for the diagnostics of cerebrovascular hypoplasiae.

Key words: *cerebrovascular hypoplasiae*

Subacute Sclerosing Panencephalitis with Atypical Onset. Clinical-Computor-Tomographic and Nuclear Magnetic Reasonance Correlations

V. Bozhinova, L. Belopitova, P. Dimova, V. Gergelcheva

Clinical CT and MRI correlations on three children with subacute sclerosing panencephalitis (SSPE) with atypical onset have been corried out, The clinical contingent includes two girls aged 6 and 11 years and one boy aged 4 11/12 years. All of

SUMMARIES

Macrophage Activation Syndrome in Pediatric Patients - Basic Characteristics, Diagnostic and Therapeutic Approach

K. Lisichki, V. Kenderova

Macrophage activation syndrome (MAS) is a life-threatening complication of rheumatic disease that, for unknown reasons, occurs more frequently in individuals with systemic juvenile idiopathic arthritis. MAS is a hyper-inflammatory disorder. It is characterized by pancytopenia, liver insufficiency, coagulopathy and neurologic symptoms and is thought to be caused by the activation and uncontrolled proliferation of T lymphocytes and well-differentiated macrophages, leading to widespread hemophagocytosis and cytokine overproduction.

We describe its clinical manifestations, most common triggers, laboratory abnormalities, diagnostic criteria and treatment.

Key words: Macrophage activation syndrome, children;

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Autoimmune liver Diseases in Children and Adolescents

M. Georgieva

The review focuses on autoimmune liver disease in childhood. In 1950, the first description of autoimmune hepatitis was published, and a wealth of knowledge about the etiology, pathogenesis, clinical picture and treatment of autoimmune liver disease in children and adolescents has been accumulated to this day. The differences in the course and treatment of this pathology in children and adults are highlighted.

Key words: autoimmune hepatitis, autoimmune sclerosing cholangitis , children, adolescents

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Diagnostic Methods for Demonstrating Helicobacter Pylori Infection

P. Hadjiiski, L. Boyanova

There are many diagnostic methods to detect Helicobacter pylori infection. The tests are subdivided into two large groups depending on whether or not they require fibrogastroduodenoscopy: invasive and non-invasive tests. Histological methods, rapid urease test, culture for isolating the causative agent and subsequently antibiotic susceptibility testing and molecular methods to detect H. pylori or H. pylori virulence or resistance genes are included in the invasive methods. Non-invasive techniques include serological tests, urea breath test and faecal antigen test. Any diagnostic method for H. pylori infection has its own specific advantages and disadvantages that have to be tailored to the intended goals, for instance the choice of the rapid urease test mainly for untreated patients, histological examination especially in the presence of alarm symptoms for detection of ulceration or malignancy, serology mostly for epidemiological studies, urea breath test, or fecal antigen test with monoclonal antibodies to control the success of eradication, culture method with strain susceptibility testing against a wide range of antibiotics, or molecular assays for resistance genes for the optimal choice of the treatment regimen in case of unsuccessful treatments. It is important to keep in mind that the use of antibiotics up to one month before the study or proton pump inhibitors up to two weeks before the study can worsen the performance of most of the diagnostic tests.

Key words: Helicobacter pylori, diagnostics, histology, culture, PCR, stool antigen test, urea breath test

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Twenty Years of Experience in a Multidisciplinary Approach in the Treatment of Children with Hypophosphatemic Rickets

M. Dikova, E. Stefanova, D. Yordanova, Z. Todorova, B. Hristov

Hypophosphatemic rickets are a rare, genetic disease that causes significant skeletal deformities in young children, as well as lifelong disability and pain. Conventional therapy aims to manage symptoms by taking oral phosphorus and active vitamin D. Treatment for progressive bone disease also includes orthopedic surgery.

We announce our 20 years of experience and make a retrospective analysis evaluating the evolution of the treatment approach of 14 patients at the Children's Orthopedic Clinic of the University Hospital of Orthopedics, Medical University, Sofia. The results of preoperative studies of calcium phosphorus exchange reflect the biological constellation of the disease. For the past three years, we have also been looking for a level of Fibroblast Growth Factor 23 that has been found elevated in 4 out of the studied 7 patients. Drug treatment is the main treatment but it is not well controlled in the first thirteen years of the period under review. It is performed according to world medical standards and at doses used by other authors. Orthopedic surgery is performed after radiography of the entire lower limbs in the upright position, measuring the deviation of the mechanical axis and the femurotibial angle, and with modern surgical techniques after 2012.

The evolution in our behavior regarding this treatment is a result of the general development of medical science and its application under the available to us conditions.

psychological study of the patients by the methods of the Thematic Aperceptive Test (TAT); personalized questionnaire of Izenk, H. (E.P.O.) and projective methods. From the results, obtained by the study, it is evident the necessity of a joint strategy and interdisciplinary partnership between doctor, clinical psychologist and family therapist.

Key words: ALL, relapse, family group, multidisciplinary team, psychological approaches.

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Childhood Hemolytic Uremic Syndrome

S. Todorov, A. Anadoliyska, D. Rusinov, P. Miteva, A. Anadoliyski

HUS is the most important clinical effect of STEC infection in humans, and young children are more vulnerable than any other age group.

We conducted retrospective analysis of childhood hemolytic uremic syndrome (HUS) from 1992 to 2002 to describe overall survival, outcome, incidence and clinical characteristics of the patients.

Key words: *childhood, hemolytic uremic syndrome, outcome, incidence, clinical characteristics*

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A Study on Some Aspects of the Epidemiology of Nosocomial Infections in a Pediatric Ward - General Profile

Mitova Y., A. Gladilova, N. Ribarova, N. Modeva, R. Semkova, D. Nikolov

During the last few years, between 65 000 and 95 000 patients have been treated in the pediatric wards - general profile in Bulgaria. The official statistics show that between 2 and 6% of the patients develop nosocomial infections. The purpose of the present study is to examine some aspects of the epidemiology of NI in a pediatric ward - general profile. The study is conducted in the period 01.08.2005 - 06.12.2005, during which in the pediatric ward - general profile there has been evidence that 22 patients have developed nosocomia clinical form of infection while in the hospital, during the present or past hospitalizations.

The most frequent clinical form of activity of the NI during the studied period is pneumonia. In twenty-two of the patients or 96% there has been evidence of nosocomial pneumonia. In 9 of the cases the etiological agent that has been isolated is K.pneumoniae (42,86%). For the studied period in the observed pediatric ward - general admittance, the second frequent clinical form is the nosocomial sepsis. In our study 38,46% the K.pneumoniae isolates are ESBL- producing.

Key words: *nosocomial infections, pediatric ward, clinical form, etiological agent;*

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Transient Anterior Opercular Syndrome in a Child with Benign Epilepsy with Centrotemporal Spikes Complicated with Continuous Spike-waves During Sleep

P. Dimova, V. Bojinova

Transient anterior opercular syndrome has been rarely described in the course of benign epilepsy with centrotemporal spikes (BECTS) with continuous electrographic focal or bilateral seizure activity with rolandic predominance. The condition of continuous spike-waves during slow wave sleep (CSWS) is also known as a relatively rare complication of BECTS. Here, we report on a 6-year old child with the diagnosis of BECTS who developed transient oro-motor deficit. After a 6-month period of poorly controlled right-sided rolandic seizures with typical centrotemporal spikes on the left in the electroencephalogram (EEG), magnetic resonance imaging revealed a temporo-polar angioma on the right. The child was operated on, and shortly thereafter experienced a progressively worsening speech difficulties and drooling, while some increase in seizure frequency. Awake EEG demonstrated bilateral independent rolandic discharges. The condition deteriorated further with unceasing facial, mainly perioral myoclonia more prominent on the right, and sleep EEG revealed CSWS. After a short-term IV treatment with corticosteroids, the condition improved dramatically with complete resolution of CSWS and gradual neurological normalization shortly thereafter. Antiepileptic drug therapy was continued adding levetiracetam to the baseline valproate and resulting in seizure freedom. This case confirms that transient anterior opercular syndrome is a possible, although rare complication in BECTS and could be related to an increase in the epileptiform EEG activity up to CSWS.

Key words: *anterior opercular, BECTS, CSWS, EEG, epilepsy, rolandic, levetiracetam*

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Amyopathic Dermatomyositis Diagnostic and Therapeutical Problems

K. Lisichki , Kr. Grozdeva

The amyopathic dermatomyositis is an extremely rare form of juvenile dermatomyositis (68 cases in the literature up to now)

We present a 7 years old boy with two years history of juvenile dermatomyositis, fulfilling the criteria of classical juvenile amyopathic dermatomyositis - lack of muscle tenderness and weakness, normal serum levels of muscle- derived enzymes and normal EMG findings. The patient presents also other characteristic findings - periorbital violaceous erythema, livid-erythic skin changes, reflecting the systemic vasculopathy, Gottron papules palpable over joins, deep skin and muscle calcification, arthritis with severe join contractures.

The treatment with steroids, methotrexate and diltiazem has showed a good control of the disease.

Key words: *amyopathic dermatomyositis, calcinosis, treatment*

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Neonatal Diabetes Mellitus - a Rare Reason for Neonatal Hyperglycemia

R. Stancheva

Some rare forms of diabetes result from mutations in a single gene and are called monogenic. Neonatal diabetes mellitus (NDM) and maturity onset diabetes of young (MODY), are the two main forms of monogenic diabetes. NDM occurs in the first 6 months of life. It is a rare condition with frequency in only one in 400 000 to 500 000 live births. NDM can be mistaken for the much more common type 1, but type 1 diabetes usually occurs later than the first 6 months of life and is associated with markers of autoimmunity. In about half of those with NDM the condition is lifelong and is called permanent neonatal diabetes mellitus (PNDM). In the rest of those with NDM, the condition is transient neonatal diabetes mellitus (TNDM). NDM can occur as an isolated phenotype or as a part of a syndrome: Walcott-Rallison - syndrome (NDM and spondyloepyphiseal dysplasia); NDM and hyperuricemia (von Mühlendah). TNDM cannot be distinguished from PNDM based on clinical features. Molecular analysis of chromosome 6 anomalies, and the KCNJ11 and ABCC8 genes, provides a tool to identify TNDM from PNDM in the neonatal period. This analysis also has potentially important therapeutic consequences leading to transfer some patients from insulin therapy to sulfonylureas.

We report one case of probable neonatal diabetes mellitus.

Key words: *neonatal hyperglycemia, neonatal diabetes mellitus*

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Use of Cardiac Natriuretic Peptides in Children with Congestive Heart Failure

R. Marinov, V. Pilosoff

Neseretide,human B-type natriuretic peptide,has hemodinamic effects that may be beneficial in pediatric patients after cardiac surgery and native congestive heart failure.Expirience with neseretide and pediatric is limited.The purpose of this study was to evaluate effects and safety of neseretide in pediatric patients.BNP is cardiac neurohormone produced from the cardiac ventricles in pathophysiologic states ,such as systolic or diastolic function,increased left ventricular mass , and low ejection fraction.

Key words: *neseretid, heart failure, hemodinamic effect*

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Multicenter Study of The Quality of the β-Thalassemia Treatment for Bulgarian Patients

G. Petkov, P. Chakarova, D. Stoianova, M. Rangelova, V. Kaleva, A. Stoianova, J. Marinova, K. Peeva, B. Parashkevova, Zh. Zdravkova, G. Jordanov, D. Lazarov, T. Radevska

The aim of this multicenter study was to assess the treatment quality of the β-thalassemia patients and their survival. The study includes 167 β-thalassemia cases from hospitals in Sofia, Varna, Plovdiv and Stara Zagora. Patients' medical records from hospitals pointed above were used as a source of information as well as expert assessment of the team led this treatment. Appropriate methods of statistical analyses were applied and especially survival analysis. The haemotransfusion therapy among most of the cases studied was received regularly but receiving substitutive and chelation therapy were insufficient. Survival rate among studied cases was 34.5 years and there was a direct relation with the shown expert assessment. The level of medical services had been not optimal yet. The weaknesses especialy from subjective nature were considered as a main reason for the presented situation with substitutive and chelation therapy. The study results were compared with such from similar studies led by distinguished research workers world-wide. The need to initiate the National Program for improvement of treatment and prevention of the β-thalassemia in Bulgaria was obvious.

Key words: *β-thalassemia, multicenter study, survival, haemotransfusion, chelation, substitutive therapy*

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The aim of the present study was to determine the presence of enterovirus RNA in serum of children with newly diagnosed type 1 diabetes in Bulgaria. Serum samples were collected from 18 children with newly diagnosed type 1 diabetes and they were tested by RT-PCR. We also studied 18 matched control subjects. Enteroviral RNA was detected more frecuently in sera from diabetic children than in healthy control children ($P < 0.05$). Our results suggest that enteroviral infections are associated with the induction of autoimmune diabetes.

In this paper we also review the epidemiologic data concerning type I diabetes and discuss potential mechanisms of pathogenesis.

Key words: enteroviruses, type I diabetes, RT-PCR

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Cross-sectional Survey of Vaccination Coverage with Routine Immunizations in Children Born in 2006 in Sofia Region

K. Parmakova, M. Kojouharova, M. Borisova, A. Kurchatova

Timely administration of routine childhood immunizations as well as maintaining of high vaccination coverage are the main factors in preventing the emergence and spread of vaccine preventable diseases. The aim of this study is to assess and compare the vaccination coverage and timeliness of vaccine administration in children from Bulgarian and Roma origin in Sofia region from 6 to 18 months of age and to analyze the reasons for delay or missed immune prophylaxis. The number of immunized infants is decreasing by each following vaccine dose and in older children. The highest immunization coverage is observed for HBV1 and HBV2, and for OPV1 and DTP1. Significant difference is found out for the 3rd dose administration for all vaccines, where the coverage is less than 95%. Roma infants are immunized later compared with Bulgarian ones. The highest immunization coverage is found in maternity hospitals where infants receive vaccination, except in cases of temporary contraindications or written parents refusal.

Key words: imunization coverage, timeliness of vaccine administration, subject to immunization

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Case Report of an Infant Presenting with Salt Wasting with Late Determination the Nature of the Disease

D. Iliev, A. Anadoliyska

The article presents a case report of an infant with salt wasting manifested during the neonatal period. Two plausible diagnoses are considered at the maternity hospital – salt-wasting form of congenital adrenal hyperplasia and pseudohypoaldosteronism. Methylprednisolone therapy introduced via an intravenous route is started. During the course of this treatment the plasma level of aldosterone is measured. It turns out to be elevated. Pseudohypoaldosteronism is accepted to be the correct diagnosis and sodium chloride substitution is initiated. At the age of nine months, the child is admitted at the University Children's Hospital, with clinical and laboratory signs of chronic malnutrition, refractory loss of appetite, dehydration, metabolic acidosis, dyselectrolytaemia, and salt wasting in spite of the therapy with sodium chloride. It is important to note the presence of enlarged external genitals a fact which makes the diagnosis pseudohypoaldosteronism less probable. Therefore, the level of 17-OH Progesterone is measured which turns out to be very elevated. Salt wasting form of congenital adrenal hyperplasia is diagnosed. Substitutional and pathogenetic treatment with glucocorticoids and mineralocorticoids is initiated. As a result, the child's condition rapidly improves, it gains weight, and both electrolyte and acid-base homeostasis are restored.

Key words: salt-wasting form of congenital adrenal hyperplasia, pseudohypoaldosteronism, diagnostic criteria

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A Clinical Case of Primary Septic Myositis

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An eight years old girl presenting with fever, edema, hyperemia and pain in the left shank. Routine laboratory tests were with mild proinflamatory activity. A septic myositis is diagnosticated after magnetic resonance imaging.

The treatment with antibiotic and symptomatic therapy within five weeks showed good control of the disease.

Pyomyositis is a primary infection of the skeletal muscle usually caused by Staphylococcus aureus. This infectious disease is endemic in tropical areas and sporadic in temperate climates. Primary pyomiositis is probably the result of a transient bacteriemia. Trauma to the affected muscle has been proposed as a possible etiology. Primary pyomiositis can involve any muscle group in the body, but the most commonly affected anatomic sites are large muscles of the lower extremities. There are three consecutive clinical stages: diffuse muscle infection, abscess formation and sepsis. Magnetic resonance imaging is the preferred diagnostic imaging modality. The choice of treatment for pyomyositis depends on its stage and includes complete drainage of any abscess cavity combined with appropriate antibiotic therapy. Most patient have complete recovery with no long-term sequelae.

Key words: Primary septic myositis, children.
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Treatment Choice in Basedow's Disease in Childhood and Adolescence

N. Kaleva, T. Shabanova, R. Dimov, M. Spasova

The authors present a case of Basedow's disease in a 17-year-old girl, resistant to conducted antithyroid drug therapy. Besides the lack of effect of high-dose treatment the girl experienced also life-threatening complications - leucopenia and neutropenia, requiring application of colony-stimulating factor - G-CSF and exclusion of other accompanying disorders. The surgery carried increased intraoperative risk during the hyperthyroid phase, but resulted in rapid improvement of the girl's general condition and hematological and hormonal deviations, thus being a good therapeutic choice. The three available treatment methods for Basedow's disease in childhood - antithyroid drugs, surgery and radioiodine are reviewed according to literature data and in relation with the reported case regarding their advantages and disadvantages in this age group.

Key words: *Basedow's disease, childhood, therapy side effects*

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Introducing Insulin Pump Therapy in Bulgaria

M. Konstantinova

Insulin pump treatment has been started in the world diabetic centres more than 15 years ago and has already proven its priority for the possibility of maximal precision of the insulin dose 24 h. daily. The lack of registration of the device in our country till October 2005, was the reason the Bulgarian patients willing to treat themselves with insulin pump to go to diabetic centres abroad.

The aim: To discuss the initial data of the diabetic patients put on insulin pump therapy in our centre.

Material, Methods and Results: Since March 2007 we have started therapy with insulin pump MiniMed-Medtronic in 14 Bulgarian patients aged 3 to 22 yrs, x = 9.09 ± 6.06 yrs. The level of HbA1c before starting the pump was x = 9.01 ± 2 05%.

The control value of HbA1c /in 6 of the patients/ is x =7.7 ± 1.69%.

The most common problems in this group of patients has been the stop of the insulin infusion for different technical mistakes. We had 4 episodes of initial diabetic ketoacidosis.

Conclusions:

1. The therapy with insulin pump is a better option for intensive insulin treatment compared to multiple injections.

2. In order to obtain the best possible control of the disease with a pump, precise education of the patients both for the technical management of the pump and the unceasing adaptation of the dose to the changing modes of life style in the patients.

Key words: *Insulin pump, children, diabet*

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Hypoallergenic Formulas for Dietary Allergy Prevention in Infancy

The prevalence of atopic diseases including atopic dermatitis, allergic asthma, hay fever and food allergy has increased considerably during the past decades. In Europe, about one quarter of the paediatric population are affected by an atopic disease. Specific preventive measures have been aimed at decreasing the incidence of atopic diseases and/or postponing their first occurrence. In this context, it is important to avoid sensitization by reducing the exposure to dietary allergens from an early age, as dietary allergens represent a major part of the allergen load during infancy.

The most frequent allergens present in foods are native proteins. The allergenicity of food proteins including the cows' milk protein component of infant formulas can be reduced by various processing techniques, of which enzymatic hydrolysis is the most commonly used.

In line with expert recommendations, infant formulas with hydrolysed protein are now widely used in many European countries in terms of atopy prevention. They are well suitable for the nutrition of infants with a known or unknown risk of atopic disease and provide an important tool for the reduction of an infant's allergy risk.

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ЮВЕНИЛЕН ХРОНИЧЕН АРТРИТ И АНТИГЕНИТЕ НА HLA СИСТЕМАТА

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Резюме. *Изследвани са 148 български деца с ювенилен хроничен артрит (ЮХА) на възраст от 8 месеца до 17 години. HLA типизирането на антигените от локуси А и В е извършено посредством стандартен микролимфоцитотоксичен тест на Terasaki, а на антигените от локус DR – чрез пролонгиран микролимфоцитотоксичен тест. Използвани са анти-HLA типизиращи серуми, покриващи 12 специфичности от локус А, 15 специфичности от локус В и 11 специфичности от локус DR. Контролната група здрави лица за HLA-A и В антигените обхваща 1085 здрави лица, а за DR антигените – 170 лица. Резултатите от изследването показват значението на носителството на HLA антигените DR4, B27 (и CREG – B7, B22, B40) и B18 като предиспониращи фактори за развитието на ЮХА. Изследванията на HLA-DR4 показват сигнификантна асоциация както в общата група, така и в отделните подгрупи (с изключение на групата „разширен олигоартрит", при която HLA от II клас не са изследвани): 34.17% срещу 18.82% при здрави контролни лица (χ^2 =7.21, $p < 0.01$, RR = 2.24). Установена е сигнификантна асоциация с HLA-B27 при олигоартритните случаи, особено при тези с олигоартрит II тип. Носителството на HLA-B27 е сигнификантно високо и при децата с разширен олигоартрит – позитивен при 5 от 8 деца (62.5%) (χ^2 = 21.68, $p < 0.001$, RR = 13.79). За първи път при нашите изследвания се установява асоциация на полиартритния тип ЮХА с HLA-B18: 25.58% срещу 10.69% при контролните здрави (χ^2 = 9.34, $p < 0.01$). Асоциациите на ЮХА с А2, DR1 и DR3, показани в литературата, и установеното от нас носителство с по-голяма, но несигнификантна честота подлежат на по-нататъшни изследвания при повече пациенти.*

Ключови думи: *HLA антигени I и II клас, ювенилен хроничен артрит*

St. Stefanov, F. Martinova, I. Boikinov, K. Lisitchki, N. Ivanova, D. Mihailova and V. Trunkova,
JUVENILE CHRONIC ARTHRITIS AND THE ANTIGENS OF HLA SYSTEM

Summary. *148 Bulgarian children with juvenile chronic arthritis (JCA), aged from 8 months to 17 years, have been investigated. HLA typifying of antigens from locci A and B has been performed by means of a standard microlymphocytotoxic test of Terasaki and that of the antigens from locus DR – by means of a prolonged microlymphocytotoxic test. Anti-HLA typifying sera have been used to identify 12 specificities from locus A, 15 specificities from locus B and 11 specificities from locus DR. The control group includes 1085 healthy persons for HLA-A and B antigens and 170 persons for DR antigens. The results of the study reveal the significance of the carriage of HLA antigens DR4, B27 (and CREG – B7, B22, B40) and B18 as predisposing factors for development of JCA. The examination of HLA-DR4 shows the presence of a significant association both in the common group and in the separate subgroups (excluding the group with „extended oligoarthritis" where HLA antigens from class II have not been examined): 34.17% against 18.82% in the control group (χ^2 = 7.21, $p < 0.01$, RR = 2.24). A significant association is found with HLA-B27 in cases with oligoarthritis, especially in those with type II oligoarthritis. The carriage of HLA-B27 is significantly higher in children with „extended oligoarthritis" – positive in 5 out of 8 children (62.5%) (χ^2 = 21.68, $p < 0.001$, RR = 13.79). An association of polyarticular type of JCA with HLA-B18 was found out for the first time in our study: 25.58% against 10.69% in the control group (χ^2 = 9.34, $p < 0.01$). The associations of JCA with A2, DR1 and DR3 reported in literature and the established by us carriage with greater but non-significant incidence, have to be submitted to further examination in a larger number of patients.*

Key words: *HLA antigens I and II class; juvenile chronic arthritis*

След като Brewerton и сътр. и Schlosstein и сътр. (1973) доказаха сигнификантната асоциация на анкилозиращия спондилит с HLA-B27, последваха много изследвания при различни заболявания. Целта е да се търси носителство на определени HLA антигени от I и II клас като израз на генетична предиспозиция за възникването и развитието на дадено заболяване. Там, където се открива висока сигнификантна асоциация, съответните HLA антигени влизат като допълнителни критерии за диагнозата – напр. DR4 при ревматоидния артрит [7].

С такава задача са проведени и продължават да се провеждат изследвания на антигените от системата на HLA при деца с ювенилен хроничен артрит (ЮХА). Ние си поставихме за цел

SUMMARIES

Prophylactic X-Ray Treatment of the Lungs in Ewing's Sarcoma

L. Marinova, Ts. Rouseva, I. Hristozova, Dr. Bovev, Iv. Shturbanov, P. Perenovska

Prophylactic X-ray treatment of the lungs in the complex therapy of Ewing's sarcoma is being discussed. Good pulmonary tolerance has been observed after reaching GRD - 15 Gy in combination with polychemotherapy (VAC or VACA). Ewing's sarcoma presents a systemic character and metastatic progress in the lungs. Alternative therapeutic methods are being looked after at aiming an increase of survival witout any disease.

Key words: Ewing's sarcoma, prophylactic X-ray treatment

Clinical Heterogeneity of Diabetes Mellitus and its Manifestation in Childhood

R. Savova, V. Gancheva, K. Koprivarova, G. Popova, M. Konstantinova, B. Angelova

In 1248 children with recently revealed diabetes mellitus during the diagnostic period clinical and laboratory data are being analysed. Preceding symptoms in this period have been assessed in 1100 Blood glucose and urinalysis have been examined in 1022 children. HbA1c has been followed up in 243 children, acid-base status - in 558 children with disclosed ketonuria. Further 82 patients presented acidotic breathing. Education of the mothers has been checked in 1226 cases. In 13,5% of 148 children with known urinalysis no ketonuria has been found (148 children, 12 of whom present fasting blooc glucose ≤ 6,4 mmol/L). Of all children 18,2% have been admitted with severe ketoacidosis (pH - 7,2 or acompanying acidotic breathing, mentioned in the epicrisis).

Mean duration of polydipsia-polyuria syndrome is 28±33 days. Children without ketonuria with a level of blood glucose ≤ 6,4 mmol/L show significantly shorter duration of symptoms compared to those with a blood glucose level ≥ 6,4 mmol/L (17±25 against 25±31 days; p=0,0991). Cases with severe ketoacidosis pH ≤ 7,2 compared to those with less pronounced ketoacidosis (pH 7,21 - 7,34) reveal shorter duration of preceding symptoms (24±25 against 34±42 days; p=0,0658). Moderate positive age dependence and duration of symptoms has been established at the moment of diagnosis (x^2 43,28, DF=8; p 0,0000). Percentage of severe ketoacidoses is higher in the age group 0-4 years. Present feverish disease preceding diabetic symptoms is more often observed in groups with shorter duration of polydipsia-polyuria syndrome. It doesn't however change the percentage of severe ketoacidoses. There are no significant differences between the education level of the mother and the duration of the period preceding the disclosure of diabetes.

Level of metabolic failure at the moment of diagnosis of diabetes in childhood is not being influenced by the duration of preceding simptoms. Clinical heterogeneity is probably based on different genetic background related to class II HLA genes which condition the immune response and B-cell destruction as well the gravity of insulinopenia respectively.

Key words: insulin dependent (type I) diabetes mellitus in childhood, age, morbidity, polydipsia, polyuria, ketoacidosis, clinical characteristics.

Carrier State for HLA B27 and HLA B7-CREG Antigens in Children with Juvenile Ankylosing Spondylarthritis

K. Lisichki, F. Martinova, St. Stefanov, V. Trunkova, A. Telcharova N. Ivanova

This investigation covers 36 children with ankylosing spondylarthritis - 32 boys and 4 girls (interrelation 8:1). Their mean age is 10 10/12 years, (i.e. 4 6/12 to 16 years) at the onset of the disease. All patients disclosed peripheric oligoarthritis on the lower extremities, entesopathia, one-sided or bilateral sacroilitis (proved by X-ray or/and scintigraphy).

No one of the patients disclosed present antinuclear antibodies (ANA) and rheumatiod factor (IgM-RF).

All patients have been examined for HLA B7 antigen, in 6 ot them - the full formula of HLA B locus.

Among six children with the examination of the full HLA B locus two of them are carriers of B_w and two of B13 antigen. Prevalence of HLA B 27 carrier state among our patients is 83,3% or if all CREG-positive cases are taken into account (33 from a total of 36 children) it reaches 91,7%. It is significantly higher ($p < 0,001$) than the prevalence in the national population - 10%.

Carrier state of CREG-antigens (B7, B12, B13, B27, Bw22, B40, Bw42) can be taken into account as an additional diagnostic criterium in JAS.

Key words: ankylosing juvenile spondylarthritis, HLA B27, HLA B27-CREG

24-Hours Noninvasive Dispensary Monitoring of Arterial Blood Pressure in Children

T. Vasileva, V. Petrova, L. Spasov, E. Dimova, R. Rahneva

Present investigation submits data analysis during a 24-hours dispensary monitoring of arterial pressure in three groups separated in accordance to casualy measured arterial blood pressure, clinical and paraclinical characteristics: 1) with normal arterial blood pressure (n=19); 2) with primary arterial hypertension (n=16) and 3) with primary arterial hypotension (n=21). Monitoring has been carried out with the aid of the device MEDITECH-ABPM, measuring arterial blood pressure by oscillometric method at 15 minutes intervals during day hours (6-22 h) and every 30 minutes at night, (22 to 6 h). A mean 75,4 measurements have been carried out per child during 24 hours.

- restlessness, anxiety, laryngospasm occurring after deep breathing, suggested hypocalcaemia as the cause for apnoea. Hypocalcaemia was supposed to be due to vitamin D deficiency. Our hypothesis was based on the incorrect feeding of the infant, the lack of vitamin D prophylaxis, as well as the infrequent sunlight exposure. The child was urgently treated with calcium administered via an intravenous route. The diagnosis infantile hypocalcaemia was supported by the laboratory findings: total calcium 1.13 mmol/L. (2.12-2.62), phosphates 1.30 mmol/L. (1.4-2.3), and alkaline phosphatase 1000 IU/L. (82-383). The diagnosis and treatment of hypocalcaemia is crucial. Its most serious complication - infantile hypocalcaemic tetany, can have a fatal outcome if not treated correctly.

Key words: *Apnoea, hypocalcaemia, rickets*

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A Clinical Case of Acute Hemorrhagic Edema

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Acute hemorrhagic edema of infancy (AHEI) is a rarely form of leukocytoclastic vasculitis. Onset is often dramatic with the cutaneous findings and the edema.The cause of this disorder is unknown. The differential diagnosis includes Henoch-Schönlein purpura , meningococcemia, erythema multiforme, urticarial vasculitis, Kawasaki disease and necrotizing fasciitis. Treatment is symptomatic. AHEI is a self-limited disease.

We present a 2 years old boy with normal physical development. The disease started acutely with respiratory tract infections. After 4-day appeared edema the forehead and the head that goes for 24 hours; appeared the pain and edema in ankle joints, the left hand , the foots and haemorrhagies in surrouding skin;the trouble in the walk until the impossibility of the motion.

Physical examination revealed: the edema and the pain on the backstoke to his foots and hands,the left knee. Impossibility of the motion in two ankle joints. For two days appeared edema the upper eyelid and the right buttock issue, the same haemorrhagic edema in the base to the penis, in the left-half the scrotum and appeared the left-side foniculocele. Routine laboratory tests were with mild proinflamatory activity.

The treatment with antibiotic and symptomatic therapy showed good control of the disease. Physical status of patient was normal after week.

The clinical follow up for 6 months showed normal physical status and routine laboratory tests was normal, too.

Key words: *Acute hemorrhagic edema, self-limited disease*

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Epilepsy due to Hypothalamic Hamartomas - Diagnosis and Treatment.

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Hypothalamic hamartomas are rare but proven cause of medically resistant epilepsy. These lesions are common for the pediatric age group and occupy 3 - 4% of pediatric epilepsy surgeries in countries with developed epilepsy surgery. The authors found only one case of hypothalamic hamartoma with gelastic seizures in the Bulgarian literature. The present paper review the actual data regarding etiology, pathology, epileptogenesis, clinical presentation, diagnostic and treatment options for hypothalamic hamartomas with medically resistant epilepsy. The aim of this review is to stimulate the search for etiology of epilepsies and particularly for etiology of gelastic seizures. Diagnosis of hypothalamic hamartomas could change treatment strategy in surgical direction because surgery has considerable advantages in comparison with medical treatment in patients with medically resistant epilepsy due to hypothalamic hamartomas.

Key words: *epilepsy surgery, hypothalamic hamartomas, pharmacoresistant epilepsy*

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Treatment of Gastroesophageal Reflux Disease in Infants

M., Georgieva, R. Pancheva, B. Varbanova

Gastroesophageal reflux (GER)and gastroesophageal reflux disease (GERD) (symptoms & complications of GER) are common problems in infants & children. In infants the prevalence rate is 67% in the age of 4 month. The review describes the pathogenesis of GER. There are several options for infant GERD treatment, including life-style, feeding (thickened and hypoallergic milk formula) and sleeping modifications, medications (H2-blockers, PPI, prokinetics) or surgery. The treatment recommended will depend on the infant's age and severity of symptoms. In most cases, medications or surgery are not needed, as many infants outgrow reflux by 1 or 2 years of age.

Key words: *gastroesophagela reflux, gastroesophageal reflux disease, infants, thickened milk formula, procinetics, proton inhibitors, H2 blockers*

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able factors, first and foremost insufficient direction of pregnancies at risk for investigation as well inadequate provision with staff and financial means. A considerable subjective disadvantage of this activity is the relative low part of chorion biopsy. Practical significance of cordocentesis is being highly approved. It has been introduced in the practice of the Centre in 1990. Authors conclude that results till now are encouraging and prenatal diagnosis may have an important significance for concrete families at risk, as well an evident effect in relation to the incidence of a series of genetic defects. For this porpose well considered managerial decisions relating to education and organization are necessary.

Key words: prenatal invasive diagnosis of genetic defect, analysis of results

Congenital Anomalies in Perinatal Mortality

I. Todorov, E. Simeonov, R. Kadirova

Authors submit retrospective and prospective data on dynamics and components of perinatal infant mortality related to the material of the Obstetric Clinic at the Medical Faculty Stara Zagora during the period 1984-1989. Despite a constant trend towards a decrease these parameters remain as being higher than the average-ones for this country. Prenatal diagnosis of fetal anomalies, introduced 1987 has led to a decrease of perinatal infant mortality by 1,78 %. During 1987 a decrease in birth has been established accompanied by an increase in the relative part of congenital anomalies. It is assumed that these facts may by in causal relation to the Chernobil disaster.

Key words: perinatal infant mortality, congenital anomalies

Prenatal Echocardiographic Diagnostics

Sn. Tomova

Analysis of results with prenatal echocardiography in this country for a period of two years (1987-1989) is being submitted. Investigations cover observation on the fetal heart with the aid of ultrasound in 238 pregnant women. An own methodology has been applied by means of monodimensional echocardiography and Doppler-echocardiography for the estimation of numerous parameters of the normal fetal heart anatomy and function. In nine pregnancies (3,7 %) congenital heart disease has been disclosed. As a whole this investigation ensure an easier performance of fetal echocardiography in this country.

Key words: prenatal echocardiographic diagnostics, fetal echocardiography, congenital cardiac malformations

Aortic Valve Stenosis in the Neonate and Early Infancy

M. Pavlova, A. Kunova, A. Todorov, M. Tsonzarova, V. Pilosof, N. Arnaoudova

The investigation covers 20 infants (8 girls and 12 boys) with a critical stenosis of the aorta in an utmost degree. It was manifested by cardiac failure during the neonatal period and early infancy. Patients were hospitalized at the paediatric Clinic of the National Centre for Cardiovascular Diseases for the period 1986 – May 1991. Average age at hospitalization was 1 month and 27 days (1 day – 6 months and 7 days). Nine children (45 %) disclosed accompanying congenital heart disease, i.e. coarctation of the aorta – 3, persisting arterial channel – 8, ventricular septal defect – 2, mitral anomaly – 2, discontinued aortic arch – 1. Diagnosis has been established by means of clinical noninvasive methods of investigation.

Surgery has been carried out in 13 children (65 %), mean age 2 months and 25 days (18 days – 6 months and 22 days). In all cases valvulotomia has been carried out under ECC and in 4 patients accompanying coarctation of the aorta has been removed (2 cases) and persisting arterial channel (4 cases). Death rate due to surgery reach 53,8 %. Results after surgery proved very good – residual gradient left chamber/aorta disclossd an average of 25 mm, minimal insufficiency of the aorta, regression of left chamber hypertrophy as well loading according to EchoCG and ECG in all cases surviving surgery checked during a period of 1 year 5 months (3 months – 3 years). The discussion covers peculiarities of clinical manifestations, precision of diagnosis, indications for surgery and its results.

Key words: aortic valve stenosis, accompanying congenital heart disease, results of surgery

Idiopathic Granulomatous Hepatitis in a Nine Years Old Boy

Dr. Bobev, K. Lisichki, A. Telcharova

Feverishness, myalgyae, arthralgiae, pericarditis, hepatosplenomegalia as well laboratory data for biological activity were present in this case with idiopathic granulomatous hepatitis. Numerous investigations in order to achieve and aetiological elucidation of diagnosis no causal injurious agents were revealed.

Treatment with antibacterial and antimycotic means remained without effect. Subsequent evolution of the disease proved favourable ending with spontaneous remission. Follow up in the course of one year proved normal relating to the clinical and paraclinical status.

Key words: idiopathic granulomatous hepatitis

Pan(Poly)-Arteriitis Nodosa

M. Ivanova, I. Boikinov

The report deals with two children 4 and 15 years of age with classic pan(poly)-arteriitis nodosa, diagnosed during life time and verified at post mortem (histology). Onset proved non characteristic in both children. It resembled respiratory and gastrointestinal disease. Development of characteristic variegated symptomatics and increasing blood pressure – high values of a reanal-vascular type made diagnosis possible. The fifteen years old patient developed infarction of the intestinal wall which led to perforation and peritonitis – indication for a subtotal resection of the gut. Discussion covers the significance of a series of laboratory tests as well the therapeutic possibilities.

Key words: panarteriitis nodosa, laboratory tests, therapeutic possibilities.

Two Cases with Mucha-Habermann Disease

K. Lisichki, V. Kenderova

Mucha-Habermann disease or pityriasis lichencides et varioliformis acuta (PLEVA) is a rare cutaneous disorder of unknown etiology. Pityriasis lichenoides encompasses a spectrum of clinical presentations ranging from acute erythematous papular lesions that rapidly evolve into pseudovesicles and central necrosis. It is accompanied in a few cases by: low grade fever, arthralgias, arthritis, abdominal pain.

We present a six years old boy and a three years old boy with typical cutaneous lesions. The first boy has abdominal pains and arthralgias in the knees and the ankles, too.

The treatment with antibiotic (lincomycin), low dose steroids and UV radiation in the first case and UV radiation only in the second case has a good effect.

Key words: *Mucha-Habermann, pityriasis lichenordes et varioliformis acuta (PLEVA)*

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Hypokalemic Periodic Paralysis: a Case Report

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Hypokalemic periodic paralysis (HPP) is an inherited disease, affecting mainly males. In two third of the cases it is inherited in autosome dominant manner, while the remaining one third of the cases of this disease are sporadic. It is supposed that de novo mutation could appear.

The main symptoms of this disease are muscle weakness, paresis or paralysis, following by periods of normal muscle function. Disease crises usually appear in early hours of the day, during or immediately after night dream or rest.

The exposure to cold, the inappropriate diet, and the excess carbohydrate, salt or alcohol consumption are considered to be some of the most important triggering factors of this disease.

The pathogenesis of HPP is still a question of disputation and it is not well clarified. Usually the level of serum potassium is under 2,5-3 mmol/l, which is found to be as a result of impaired function of Ca2+-activated K+ channels. This disturbance in channel function is determined by a mutation in the gene located on the short arm of chromosome 1 (1p 31-32).

The therapy aims to normalize the serum potassium level. The disease crises are avoided by taking Acetazolomice. Triamterene or Spironolacton could be prescribed if the effect of Acetazolomide is not sufficient.

A ground for this short review on HPP is a 14 years old boy, with symptoms with severe form of HPP, which was overcome by substitutional therapy with potassium.

Key words: *Hypokalemic periodic, paralysis.*

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A Case of Late Diagnosticated Neurological Form of M. Wilson

I. Litvinenko, M. Uzunova

Wilson's disease is presented as progressive autonomic recessive disorder characterized by disorder of transport and excessive accumulation of copper in liver, CNS, in the eyes and in other organs. The defect is in the ATP7B gene produce a protein with decreased ability to bind copper. People who have liver-related symptoms first, do so at a younger age than do those who first present with neuropsychiatric symptoms[15 years and 25 years on average, respectively. Patient I. P. 20 years old was diagnosed for the first time for Myasthenia, after that for Neurosis. When she comes in our clinic (1,5 years after beginning of the disease) we diagnosed Wilson's disease with neurological manifestations. The symptoms include dystonia, dysarthria, dysphagia. The blood level of ceruloplasmin was markedly decreased. A newer copper chelating agent currently being investigated is Tetrathiomolybdate. The hope is that it will prove to have fewer side effects than penicillamine, yet be more effective than Trientine. We treated our patient with tetrathiomolybdate with good results.

Key words: *Wilson's disease, ceruloplasmin, Tetrathiomolybdate*

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Tamiflu - the Therapeutic Choice by Influenza Virus Infection

Dr. Bobev, M. Yordanova

Influenza is a common acute respiratory disease due to a virus that causes annual seasonal epidemics. Vaccination with closely matched epidemic strains is the main preventive measure. Three antiviral drugs are currently approved worldwide for prevention and treatment of influenza: amantadine, zanamivir and oseltamivir. In situations warranting antiviral therapy oseltamivir (Tamiflu) is the drug with the best risk-benefit balance. Its use by children shortened the median duration of illness by about 1,5 days, reduced the time to return to normal activity and the complications of influenza (particularly otitis media). Tamiflu is effective also in preventing naturally occurring cases of clinically defined influenza when the treatment is started within 48 hours after exposure to the infection. The drug is safe, but can cause vomiting. In practice, the use of antiviral drugs in otherwise healthy adults and children is not generally recommended, but their use for treatment of persons at risk for complications and as short-term prophylaxis for unvaccinated individuals at risk in epidemic situations are effective.

Key words: *influenza, antiviral, drugs, Oseltamivir*

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Hydatid Disease of Lungs - Unusual Cause for Pulmonary Artery Thrombosis in a Child

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Epidemiological studies and statistical data in recent years show, what echinococcouis morbidity in our country, including children, has increased significantly.

The authors report a case of plural echinococcosis in a gipsy girl 12 years of age with left pulmonary artery thrombosis. The thromb was composed of a great number of small echinococcous cysts - estabished out only patholo-goanatomically.

Concomitant disease mediastinal lymphadenomegaly, established many times with different image techniques turn the diagnosis first to tuberculosis and later to malignant hemopathy lymphom.

Inspite of echographic data for cystic forms in the right lung still the first hospitalization and several hemopthoes, echinococcosis was not suspected and the necessary investigations were not carried out. The diseased girl died from massive lung hemorrhage, result of big blood vessel rupture.

Key words: *echinococcosis, pulmonary artery thrombosis*
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Clinical Variant of Coxsackievirus Infection

K. Lisichki

The coxsackie infections with clinical features which imitate autoimmune connective tissue diseases are discribed rarely. They are caused by coxsackie virus B.

We describe a six years old boy who diseased in July, practically three months before the hospitalization. The boy has a fever 40°C, he has spiking fever, evarescent macular rash, pericarditis, vomiting, diarrhoea, hepatosplenomegaly, arthritis.

The laboratory findings establish an expressed syndrome of inflammation. The virological investigation demonstrates coxsackie virus B 3 infection.

The treatment with antibiotics of the diagnose sepsis is ineffective. The features persisted and required steroids and NSAIDs to control the symptoms. The description in the next 4 years, when the boy is without some therapy, doesn't mark any patological disorder in the status.

Key words: *coxsackievirus, arthritis, Still's disease*
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A Case of Congenital Myotonic Dystrophy

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Congenital myotonic dystrophy is a rare autosomal disease, caused by an increased number of cytosine-thymine-guanine (CTG) trinucleotide on chromosome 19q. The disease is usually inherited from the mother and shows a more severe course than the adult form of dystrophia myotonica. Congenital myotonic dystrophy is associated with increased perinatal mortality. In the neonatal period the most peculiar clinical features are orthopedic abnormalitis, hypotonia, facial diplegia, respiratory and feeding difficulties. Clinical and electrical myotonic discharges are difficult to elicit in the newborn. A decisive criterion for the diagnosis of this form is the occurrence of myotonic dystrophy in the patient's mother. We report a case of congenital myotonic dystrophy in a male newly born presenting with hypotonia, facial diplegia, and talipes, feeding and respiratory difficulties.

Keywords: *congenital myotonic dystrophy, muscle hypotonia*
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Dialysis for Menagement of Pediatric Acute Renal Failure

Todorov, S., A. Anadoliyska

Literature review

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Levetiracetam in Children - Results, Long-term Experience

P. Dimova

Efficacy and tolerability of levetiracetam in children were proved in recent studies. First follow-up data showed a promising long time effect without serious adverse events or undesirable loss of efficacy.

Key words: *anticonvulsant, children, epilepsy, Levetiracetam, Keppra®*
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Topiramate - Efficacy in Epileptic Encephalopathies in Childhood

V. Bojinova, P. Dimova, V. Gergueltcheva, T. Yankov, V. Tomov

The aim of the study was to establish the therapeutic efficacy of на topiramate (TPM, Topamax) as add-on therapy in children with epileptic encephalopathies. . TPM was used in dose 6-9 mg/kg daily on 55 children aged 2 to 16 years. 30 children were with Lennox-Gastaut syndrome (LGS), 73,3%of which with symptomatic etiology; 10 children- with Dravet syndrome (SMEI), 5 children- with electrical status epilepticus during slow sleep (ESES); 8 children- with myoclonic- astatic

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КЛИНИЧНИ И ПРОФИЛАКТИЧНИ НАБЛЮДЕНИЯ

КЛИНИЧНИ ОСОБЕНОСТИ НА ЮВЕНИЛНИЯ АНКИЛОЗИРАЩ СПОНДИЛОАРТРИТ

К. Лисички

МА, Научен институт по педиатрия, директор чл.-кор. Ш. Нинью

К. Лисички — *Клинические особенности ювенильного анкилозирующего спондилоартрита*

Резюме. В клинике ревматических и коллагеновых заболеваний НИП диагностировали лечили 17 детей, больных ювенильным анкилозирующим спондилоартритом (ЮАС) — 16 мальчи и 1 девочку в возрасте начала заболевания от 6 до 15 лет (или в среднем 10,5 лет) — все в акти фазе болезни. Контрольными были взяты 50 детей больных ювенильным ревматоидным артри Изучались следующие клинические и параклинические симптомы: артрит сакроилиачных, тазо ренных и периферических суставов, боли в области спины, наличие энтезопатий, иридоциклита сительство HIA B 27 AG. Для оценки общей активности процесса воспаления использованы ост фазовые показатели — РОЭ, гаптоглобин, сиаловая кислота, церулоплазмин, общий белок с ковыми фракциями, а для иммунологической активности — уровень иммуноглобулинов и цир лирующие иммунные комплексы. При статистическом сравнении данных установлены значи различия в следующих показателях: сакроилит, боли в спине, энтезопатия, возраст в начале левания старше 9 лет. Эти симптомы можно считать основными для диагноза ЮАС. Дополните ными критериями могут служить — мужской пол, поражение менее 5 периферических сустав (чаще всего нижних конечностей) и HIA B 27 позитивности. Использованные клинико-лаборат ные показатели не выявляют значимые различия в обеих группах ввиду чего они не могут б диагностическими критериями, а служат только для оценки активности воспалительного про

Ключевые слова: анкилозирующий спондилоартрит, энтезопатия, сакроилит артрит

K. Lisichki — *Clinical Paculiarities of Juvenile Ankylosing Spondylarthritis*

Summary. The present study covers 17 children, diagnosed and treated for juvenile ankylo spondylarthritis (JAS), between 6 and 15 years of age (mean age 10,5 years), including 16 boys a girl. All of them disclosed an active phase of the disease. Fifty children with juvenile rheumatoi thritis were included as controls. Following clinical and laboratory parameters have been checked thritis of the hip-joint and peripheral articulations, back pain, present entesopathies, iridocycli HLA B 27 antigen carrier state; further — inflammatory activity has been evaluated by mean acute phase reactants, e. g. BSR, haptoglobin, sialic acid, ceruloplasmin, total serum protein protein fractions, as well immunological activity e. g. levels of immunoglobulins and circulat immune complexes. Statistical assessment of respective data discloses significant differences be the following parameters: sacroiliitis, back pain, entesopathy, age at the onset of the disease be the age of 9 years. These symptoms are admitted as basic for the diagnosis of JAS. Additional cri may be — male sex, affection of less than five peripheral articulations (mostly of the lower extremi

ОПРЕДЕЛЯНЕ НА ИНТЕРЛЕВКИН-6 В СЕРУМА ПРИ ДЕЦА С ЮВЕНИЛЕН ХРОНИЧЕН АРТРИТ

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Резюме. *Интерлевкин-6, TNF, IFN-γ и техните инхибитори заемат основно място в патогенезата на ревматоидния артрит. Серумната концентрация на IL-6 е определена при 34 деца на възраст от 1.5 до 18 год. с ювенилен хроничен артрит (ЮХА) в активна фаза на заболяването. От тях с полиартритна форма са 10 деца, с олигоартритна – 14 и със системна форма – 10 деца. Най-висока концентрация на IL-6 в серума се установява при системната форма на ЮХА в сравнение с полиартритната и олигоартритната форма. При децата в активна фаза на заболяването се намират значимо по-високи стойности на IL-6 спрямо тези при неактивна фаза. Заедно с IL-6 се променят еднопосочно и други показатели на острата фаза на възпаление – СУЕ и CRP. Установява се корелация на IL-6 с клиничната активност и при трите форми на ЮХА. Мониторирането на IL-6 при отделния пациент е важно с оглед прогнозата на заболяването, отговора към терапията и възможността за по-ефективно лечение с цитокинови инхибитори.*

D. Michailova, D. Nikolova, N. Ivanova, E. Marinova, K. Lisitchki, S. Stefanov, V. Genova and V. Tsantcheva. *DETECTION OF INTERLEUKIN-6 IN THE SERUM OF CHILDREN WITH JUVENILE CHRONIC ARTHRITIS*

Summary. *Interleukin-6, TNF, IFN-γ and their inhibitors play a fundamental role in the pathogenesis of rheumatoid arthritis. The serum concentration of IL-6 is determined in 34 children, aged from 1.5 to 18 years, with an active phase of juvenile rheumatoid arthritis (JRA). 10 of the children are with polyarthritic form of the disease, 14 – with oligoarthritic one and 10 – with systemic form. The highest serum concentration of IL-6 is established in the systemic form in comparison with the polyarthritic and oligoarthritic ones. Considerably higher values of IL-6 are found in children with acute phase of the disease versus those with inactive one. Another parameters of the acute phase of inflammation as ESR and CRP react unidirectionally together with IL-6. There is a correlation between IL-6 and the clinical activity of the three forms of JRA. The monitoring of IL-6, TNF, IL-1 and other cytokines in the patient is important for the prognosis of the disease, the therapeutic response and the possibility for more effective treatment with cytokine inhibitors.*

Key words: *arthritis, juvenile rheumatoid/immunology; interleukin-6/blood*

Цитокините са биологичноактивни полипептиди и участват във всички фази на автоимунната възпалителна реакция. Съотношението между цитокините и техните естествени природни инхибитори определя активността, развитието и изхода на заболяването [3, 5]. Интерлевкин-1, интерлевкин-6, тумор-некротизиращият фактор и интерферон-γ, и техните инхибитори заемат основно място в патогенезата на ревматоидния артрит [4]. Серумното ниво на посочените цитокини се свързва със системните прояви на заболяването, а нивото им в ставната течност – с локалната активност на артрита [5]. Интерлевкин-6 (IL-6) стимулира чернодробната синтеза на острофазови белтъци, като С-реактивен протеин (CRP), α-1-гликопротеин, фибриноген, хаптоглобин и синтезата на имуноглобулини от В-лимфоцитите [3]. Измерването на IL-6 в серума и синовиалната течност при болни с ревматоиден артрит може да се използва за оценка на активността, еволюцията и прогнозата на процеса и за отчитане ефекта от лечението [1, 5].

Целта на проучването е да се определи серумната концентрация на IL-6 при деца с ювенилен хроничен артрит (ЮХА) в зависимост от формата и активността на заболяването и да се установи корелацията между IL-6 и някои показатели на възпалителната и имунологичната активност.

Материал и методи

Концентрацията на IL-6 в серума е определена при 34 деца с ЮХА, 15 момчета и 19 момичета, на възраст от 1.5 до 18 год. В активна фаза на заболяването са 25 деца, в неактивна – 9. Пациентите са разделени на три групи в зависимост от формата на заболяването. С полиартритна форма са 10 от децата, с олигоартритна – 14, а системна форма е установена при 10 деца. Приложеното лечение включва: нестероидни антиревматици – при всички пациенти, глюкокортикостероиди – при 11 от децата, резохин и сулфасалазин са получавали общо 22 деца, метотрексат е използван като базисна терапия при 11 деца.

T1D boys and healthy controls yet in the first 5 years and this trend continues during the diabetes evolution: I-21,1+/-5,7% / 7,1+/-3,1 kg/, II-25,5+/-5,5% /11,9+/-2,9 kg/, III- 28,1+/-5,8% / 13,1+/-3,2 kg/, IV- 30,9+/-3,3% /15,7+/-2,4 kg/ to 23,3+/-3,1% / 10,7+/-3,2 kg/, 20,8+/-5,4%/9,2+/-2,6 kg/, 21,5+/-5,4%/9,9+/-2,3 kg/, 25,8+/-1,5%/13,7+/-1,6 kg/ in controls /p<0,001/. There were no significant changes in FFM during diabetes development /p>0,100/. This proves that insulin treatment during adolescence is associated with changes in body composition only in girls referring FM. FM is normal at the onset of T1D and yet the first 5 years it increases to obesity as an additional health problem.

Key words: *fat tissue /fat mass/, fat free mass, type 1 diabetes, insulin, children*

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Glucose Tolerance and Polycystic Ovary Syndrome In Childhood

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It is well known that Polycystic ovary syndrome /PCOS/ initiates in puberty and is associated with insulin resistance / IR/, obesity, type 2 diabetes and cardiac complications. This study aims to define whether impaired glucose tolerance / IGT/ exists in the early pubertal phase of PCOS, analyzing serum levels of glucose and insulin as factors for insulin resistance and beta-cell dysfunction. For this aim 31 girls with PCOS, 11-18 years of age /14,8+/-1,8 yrs/ divided in 2 groups: I - 12 nonobese /BMI=20,5+/-2,7 kg/m²/ and II-19 obese / BMI=32,4+/-4,6 kg/m²/ were tested and compared to a control group of 10 healthy girls at the same age /BMI=19,3+/-2,0 kg/ m²/. Glucose tolerance /GT/ is analyzed along OGTT at 0 and 120 minute according to WHO criteria, 1985, by serum levels of blood glucose /BG/, immunoreactive insulin /IRI/ and insulin to glucose ratio /IRI/BG/. Results show increased BG in PCOS girls after glucose load compared to control group /p<0,001/ . Considering the levels of IRI in PCOS we found basal hyperinsulinemia /HI/ in 1/3 of first and 2/3 of second group and after stimulation in only 1/3 of the obese girls /p<0,001/. IRI/BG ratio shows in PCOS the presence of IR more often at fasting stage and obesity /p<0,001/. IGT is diagnosed in 8,3% /n=1/ of first and 15,8% /n=2/ of second group /p>0,100/. Fasting hyperglycemia and type 2 diabetes are not found. The study proves that IGT exists in the initial pubertal phase of PCOS, it is more often in obese girls and is associated mainly with basal HI and IR that requires their early diagnosis whenever PCOS is suspected.

Key words: *Polycystic ovary syndrome, blood glucose, serum insulin levels, insulin resistance, beta-cell dysfunction, glucose tolerance, girls, puberty*

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Familial Predisposition to Hypertension in Children and Adolescent Diabetics with Nephropathy

V. Madjova, V. Tzaneva, V. Iotova

Arterial Hypertension (AH) is very frequent in Diabetes Mellitus (DM) type 2 patients' and their relatives and it's the main risk factor for deteriorating renal function. The question for the role in arising and development of renal damage in young diabetics hasn't been solved yet. The aim of this study is to establish the correlation between the familial predisposition to AH and the initial diabetic nephropathy (DNP) in children and adolescents. The patients in a 10-years prospective study are 146 young diabetics, mean age 14, 25±3,75 years, duration of DM 8,48 ±3,6 years, 84 boys and 62 girls. They have been examined by: questionnaire, traced for microalbuminuria (MAU), metabolic control, lipid profile and sphygmomanometry. We establish that adolescent diabetics with initial DNP have in 56,75% AH in their families versus 21,1% AH in children's families without renal involvement. Patients with MAU with familial predisposition to AH have in 52,38% both mother and father hypertensive simultaneously. Their parents have earlier appearance of AH and greater % of them are treated with antihypertensive drugs (36,48% versus 24,32% in children without DNP).

Key words: *diabetes Mellitus (DM), diabetic nephropathy (DNP), microalbuminuria (MAU), familial predisposition, arterial hypertension (AH); children and adolescent diabetics, risk factors.*

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Reiter's Syndrome In Childhood. A Retrospective Study of 39 Patients

St. Stefanov, I. Boykinov, K. Lisichki

39 children with Reiter's syndrome are observed - 26 boys, 13 girls (ratio 2:1), mean age of onset - 10,8 years (range 3-17 years).

Arthritis was present in all patients - predominantly oligoarthritis in 23 cases (59%), polyarthritis - in 11 cases (28,2%) and monoarthritis in 5 cases (12,8%). Dactilitis was observed in 3 (7,7%) cases and sacroiliitis - in 9 (23%) cases.

Diarrhea prior to onset was observed in 29 cases (74,4%), urethritis - in 25 patients (64,1%) and conjunctivitis in 29 patients (74,4%). Gram negative bowel infections was detected in 21 out of the 29 cases with diarrhea. Balanitis was present in 17 out of the 26 male (23%) cases and keratoderma blennorrhagicum - only in one (2,6%) patient.

21 out of 32 patients tested(65,6%) proved to be HLA B27 positive.

Arthritis persisted to 3 months in 24 (61,5%) cases and from 3 months to 1,10 years in 15 (38,5%) cases. Short recurrences of arthritis was observed in 2 childrens.

Key words: *Reiter's syndrome, HLA B27, reactive arthr*

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SUMMARIES

Faecal Alpha-1-Antitrypsin in Healthy Children

D. Mondal, Hr. Zheleb, A. Radivenska

Alpha-1-Antitrypsin is a marker for proteins in the gut. n 90 healthy children its level has been estimated (576, ug/g non lyophilized faeces). Beyond that value the loss is to be assessed as pathological. Neither significant differences in alpha-1-antitrypsin faecal concentration has been been revealed for the different age groups nor in relation to dietary dependence.

Key words; protein loosing enteropathy, faecal alpha-1-antitrypsin

Protein Loosing Enteropathy - an Unusual and Scarecely Known Manifestation of Lupus Erythematosus

K. Lisichki, D. Mondal

The report deals with a 16 years old girl with systemic lupus erythematosus, diagnosed according the criteria of the American Rheumatologic Association. The disease is with a course of four years. Following signs have been observed at clinical admission, i.e. increased fatigue, dyspnoea, painful cough, edema at the lower extremities, anuria, hypoprozeinaemia and hypalbuminaemia. By revealing increased levels of alpha-1-antitrypsin in the faeces protein loosing enteropathy has been demonstrated. Investigation ins including biopsy of the small intestines cardiac or hepatic causes as well as lymphangiectasis have been rejectec.

Treatment with endoxan, corticosteroids, symptomatic medicines and haemodialysis has been carried out. Fatal outcome is ensuing despite the treatment during the course of a severe virus infection (influenza tupe B demonstrated by imunofluorescence).

Key words: protein loosing enteropathy, systemic lupus erythematosus

Diagnostics and Treatment of Helicobacter Pylori Infection Linked to Gastroduodenal Disease in Children and Adolescents

M. Georgieva-Shakola

The investigation covers 180 children and acolescents with a mean age of 12,8 years with upper dyspeptic complaints and/or haemorrhages in the upper part of the gastrointestinal tract, investigated for Helicobacter pylori (H.p.). The study is being aimed to elucidate the capacities of some invasive methods in order to demonstrate present H.p. as well as the effect of a therapeutic scheme applied in adult patients, i.e. ampicillin and metronidazone in children. The cytological method is quick and representative with a high sensitivity and precision. It is especially suitable for the investivity and precision. It is especially suitable for the investigation of children and aoolescents. Eradication of H.p. with the reported therapeutic scheme during 10 days reaches 83,33%. In conclusion - the authors recommend the cytomorphologic test for the disclosure of H.p. and the combination with H2 blocker and ampicillin+metronidazole for the treatment of the infection.

Key words: Helicobacter pylory, histiomorphologic and cytomorphologic investigation, urease test, ampicilin metronidazole

Entesopathy in Children - Special Clinical Features

K. Lisichki, St. Stefanov, M. Bozhidarova

Twenty-four children with entesopathy and arthritis are being presented. Clear cut prevalence of males is present (22 boys, 2 girls; quotient 11:1). Mean age of the patients 13,8 years. Oligoarthritis - in 5 (20,8%). Entesopathy is most frequent on the calcaneus in 19 children, at the patellar ligament for the tuberositas tibiae - 5 children; the plantas fascia - 44 children= Acute iridocyclitis has been observed in 3, IgA-nephritis - in 1 child. Carriers of HLA B27and B40 are 7 of 22 children under investigation (77,3%). Family loading is being established in 21,7%. Clinical follow up characterizes 45,8% of the cases as idiopathic SEA syndrome, Reiter's syndrome - 16%, psoriatic arthritis - 4.2%.

Key words: entesopathy, SEA syndrome.

Structural Chromosomal Disturbances in Solitary Cells

A. Andreev

Results from 2657 patients from the routine contingent of the cytogenetic laboratory for the period 1987-1991 are being reported. In 27 of a total 16090 assessed cells single chromosomes disturbances 7 and 14, in another six - chromosomes 7 and 14 are also taking part. Results from this investigation are being consolidated with results from reports in the available literature. Eventual causes and mechanisms are being discussed for the non casual participation of chromosomes 7 and 14 in the chromosomal distubances in single cells (CDSC). Characteristic peculiarities of the phenomenon CDSC in the present communication and its appearance in routine practice after the Chernobil disaster without any visible causes are being discussed. Dynamics discloses an increase up to 1990 and a decrease in the following years.

Key words: radiation, single cells, translocation, chromosomes 7 and 14.

Arterial Blood Pressure Values in Children 2 between 7 and 14 Years of Age for the Period 1992-12993 in Sofia

T. Vasileva, R. Rahneva, E. Molle

The investigation cover 4861 children from Sofia, i.e. 2473 boys and 2388 girls between 7 and 14 years of age. Measurements have been carried out by the means of mercury sphygmomanometer. Present day standard method has been aplied. Statistical assessment is based on mean values for systolic and diastolic values of three measurements. Age distribution of arterial blood pressure values is presented by the method of persentiles and by variation analysis. Low (hypotensive) values of arterial blood pressure are being admitted as P_s - P_5, normal-ones - in the range P_5 - P_{95}, high values (hypotensive) - $\geq P_{95}$. Authors' results disclose that arterial blood pressure val-

National Pediatric Centre for Hemodialysis. The children with ARF were between 7 and 16 years of age.General mean prevalence is new 9 cases yearly. In 10 Patients (11%) ARF has been non oliguric, a result of medicinal toxicity. In 54 children (60%) dialysis has been applied, as haemodialysis in 50 cases, in 4 as peritoneal dialysis. Total recovery of renal function has been observed in 67 children, 3 remained in a stadium preceding haemodialysis of chronic renal failure, 9 passed into terminal renal failure remaining for cotinued dialysis treatment. Eleven children died. Lower lethality is being observed in those children at dialysis. Authors established acute glomerulonephritis and haemolytic uraemic syndrome, followed by acute tubular necrosis as the most frequent cause for ARF in childhood. Chronic glomerulonephritis and vasculitis are responsible for the severe ARF and chronified renal failure in about 50%.

Key words: ucte renal failure, dialysis, intensive treatment

Acqured Renal Cysts in Children at Dialysis Treatment

A. Boueva, A. Anadoliyska

Acquired renal cysts are being observed in patients with terminal renal failure. They have been for the first time described in 1978. This status is being diagnosed basing on 5 or more cysts in each kidney.

Ultrasound investigations have been carried out in 21 children in order to disclose acquired cysts in the kidneys. The latter were present in 4 children (19%) before starting dialysis therapy. After a mean duration of 21 months acquired cysts have been observed in 18 children (61,9%).

Distribution of the patients in relation to the basic illness revealed that after dialysis treatment acquired cysts are prevalent in patients with pyelonephritis leading to terminal renal failure. After three years of dialysis treatment pronounced increase of prevalence is being observed.

Key words: acquired renal cysts, renal failure. echography

Early Manifestation of Adult Type Renal Polycystosis

A. Anadoliyska, Sv. Marinova, A. Boueva

Adult type renal polycystosis is in general clinically manifested during the fourth or fifth decade of life. In recent years this disease in being prenatally diagnosed as well during the neonatal period and infancy. This imposes its differentation from the infantile type of renal polycystosis.

The present case report deals with the first clinical features during the neonatal period thus presenting some differential diagnostic problems. Besides increased dimension of the kidneys containing cysts, transitional renal failure, severe changes in the heart with extreme dilatation, third degree of mitral insufficiency resulting from developing renin type arterial hypertensinon have been observed.

Key words: adult type renal polycystosis, early clinical manifestation. diagnosis

Nephrotic Syndrome with Minimal Lesions (NSML) Retrospective Follow up Study, Clinical Picture, Laboratory Indexes, Diagnosis

D. Farhudy

The present study covers children with NSML (88 with a male to female ratio 1,58:1) examined at the Paediatric Nephrology Clinic for the period 1990-1994. In the age group 0 - 3 years NSML affects the immune system, 17.5% of the examined patients present associated allergic disease. In 81,25% NSML is preceded by different infectious diseases mainly of the respiratory system. Edemas of different intensity and incidence are the main symptom at the beginning. Laboratory indexes reveal albuminuria. hypoproteinaemia. hypalbuminaemia (18,7 ± 3,4 g/L), hypercholesterolaemia, hypertriglyceridaemia decreased IgG ans IgA as well increased IgM levels.

Diagnosis of NSML is determined by the trid: massive albuminuria, hypoproteinaemia and edemas.

Key words: hephrotic sindrome with minimal lesions

Echography through the Fontanelle in a Neonate with Herpes Simplex Meningoencephalitis

R. Georgieva

In a neonate with herpes simplex meningoencephlits evolution of morphological changes in the brain estabilised with the aid of echography have been discussed. They have been compared to the dynamics of the clinical features and progressing EEG abnormalities. Initial slightly abnormal findings are being followed by the periventricular and parietal development of massive hyperechogenic zones on both sides. Their morphological substrate is haemorrhagic necrosis and cerebral edema. Later-on multiple pseidocysts are taking shape (multicystic encephalomalacia is developing), dilatation and deformation of the lateral ventricles. Rough abnormal echogaphic picture correlates with progressing changes to the worse of neurological symptomatics and total inhibition of the basic bioelectrical activity in the EEG. Echography through the fontanelle if continuously checked is highly informative in relation to the gravity, evolution of the process. effect of the treatment and prognosis in neonates with herpes simplex meningoencephalitis.

Key words: meningoencephalitis. herpes simplex. neonate

Faecal Alpha-1-Antitrypsin as Endogenic Marker of Protein Loosing Enteropathy in Children with Extraintestinal Disease

D. Mondal. R. Koumanova, A. Koufardzhieva, R. Slavova, K. Lisichki

Present investigation is checking pathological enteral loss of serum proteins by measuring enteral faecal concentrations of alpha-1-antitrypsin (FA-1-AT) in children with mucoviscidosis, diabetes mellitus, purpura Schonlein-Henoch, low height. lupus erythematosus. Authors establish that children with mucoviscidosis and constitutional low height do not reveal any increased intestinal loss of serum proteins. Pathological value of FA-1-AT has been established in one of the seven children with diabetes mellitus in whom later coeliac disease has been revealed, further on in one child with coeliac disease presenting low height without diarrhoea as well in the four examined children with purpura Schonlein-Henoch, as also in the four girls with lupus erythematosus.

Key words: faecal alpha-1-antitrypsin, mucoviscidosis, diabetes mellitus, lupus erythematosus, purpura Schonlein-Henoch

Arterial Pressure Norms According to Height and Body Mass in Children from Sofia

T. Vasileva, R. Rahneva, E. Dimova, V. Tsancheva

In accordance with modern requirements criteria for the assessment of arterial pressure should be based rather on norms of height and body mass. Present investigation is aimed at elaborating norms for arterial pressure according to height and body massin children from the City of Sofia. The investigation has been accried out during the years 1992-1993 including 2705 children from Sofia between 6 to 15 years

of immunological memory and protection for 5 - 15 years after initial vaccination scheme with recombinant hepatitis B vaccine.

Key words: *hepatitis B vaccine, protection, immunological memory, children*

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Recognising Kawasaki Disease and the Following Coronary Risk - Experience of Paediatric Rheumocardiology Clinic, Paediatric University Hospital, Sofia

A. Telcharova, St. Stevanov, K. Lisichki, Sn. Tomova

We report 52 cases of KAWASAKI DISEASE /KD/ hospitalized in pediatric rheumatology department from 1997 to 2009 yr. The age group includes children from

2 months to 13 years, 39 of them (75%) presented the typical form of the disease and 13 - (25%) had incomplete or atypical presentation. The disease was unrecognised until the hospital admission and was misdiagnosed with acute infection diseases including scarlet fever, septic conditions, UTI, or other diseases presenting with fever and rash like Still disease, allergic reactions to antibiotics after treatment of tonsillitis or acute polyarthritis.

According to our studies 41 patients (72.7%) were referred in the subacute stage of the disease, and only 11 (21.1%) in the acute stage. We report a very high percent of coronary complications - 22 kids - (43%) had coronary dilatations or aneurysms verified by 2D-ECHO.

Patients with atypical onset or incomplete cases of KD are at higher risk to develop coronary alterations (53.8% versus 38.4%) since they do not receive the appropriate treatment or they do not receive it timely. Patients referred in the subacute stage of the disease are also at higher risk to develop coronary alterations - 48.8% versus 18.8% admitted in the acute stage, where treatment was performed.

Conclusions: There are some diagnostic difficulties which delay the diagnosis of KD and the early appropriate therapy.

Missing atypical or incomplete forms of KD results in higher percentage coronary involvement because lack of proper treatment.

Atypical forms of KD are more common in age group under 1 yr and over 4 yrs, but they still are present in the typical for the disease age group 1-4 yrs.

Key words: *Kawasaki disease (KD), incomplete KD, atypical KD, coronary alterations in KD*

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Emotional Characteristics in Early Childhood

T. Tatiozov

Initial early established bonding between patents and child are based on the frequent contacts between them, starting immediately after childbirth and lasting for several months. The first affectionate words, parents' tender physical caresses, their positive energy, body temperature, mother's emollient hands, child's quick orientation towards mother's breast, cooing exchange and visual interactions are considered to be preconditions for bonding.

Bonding contributes to eliminate mother's strain and anxiety and thus the child becomes more tranquil. Parents are more active when taking care of their child. They take a greater responsibility for the rearing and development of the child.

Bonding provides also a basis for the initial model of child-parents and child-other next-of-kin relationships.

Key words: *early childhood, bonding, reciprocity, children, parents.*

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Congenital Heart Disease (Data Based on the Registry of Congenital Anomalies in the Pleven Region)

K. Kovacheva, M. Simeonova, V. Nedkova, M. Yonov, A. Valkova, V. Atanasova, A. Velkova, G. Tzankova, R. Rosmanova

Congenital heart disease (CHD) is the largest group of congenital structural defects with a great impact on the health and mortality of affected children. ***The aim*** of the study was to present some data concerning epidemiology, genetics and prevention of CHD, data based on the registry of congenital anomalies (CAs) in the Pleven region.

Materials and methods: The source of the data was the regional population-based registry of CAs using criteria according to EUROCAT recommendations. During the study period 1988-2006, 47 622 births were surveyed in the University hospital, City of Pleven.

Results: During the study period, a total of 1425 cases of CAs and 363 cases of CHD were ascertained. Most of the CHD cases (67%) were isolated defects and 1/3 of the cases were cardiac defect combined with other CAs (as a part of multiple malformation syndromes). The isolated CHD were the most common congenital defects, with a proportion of 17% of all CAs registered in the newborn babies and a prevalence of 4.3 per 1000 births. The follow-up of the children up to the end of the first year of the life gives an opportunity to discover additional cases of CAs with a manifestation in postneonatal period. The use of the additional sources of information influenced the final prevalence rate to 5.8 per 1000 births. The genetic counseling revealed familial data (other affected child or parent) in 2% of families with CHD. Prenatal diagnosis by using fetal echocar-

МЕТОД ЗА ЛЕЧЕНИЕ НА ОВЕНИЛНИЯ ХРОНИЧЕН АРТРИТ ПОСРЕДСТВОМ МАГНИТНО-ЛАЗЕРНО ВЪЗДЕЙСТВИЕ

(Предварително съобщение)

Б.Тонев, М.Николова, Д.Михайлова, К.Лисички, А.Телчарова

METHOD FOR TREATMENT OF JUVENILE CHRONIC ARTHRITIS BY MAGNETIC-LASER INFLUENCE

(a preliminary report)

B. Tonev, M. Nikolova, D. Mihailova, K. Lisichki, A. Telcharova

Р е з ю м е. Проследен е лечебният ефект на комбинираното въздействие на постоянно магнитно поле с инфрачервен лазер на фона на медикаментозното лечение.

Изследвани и лекувани са 11 деца с ЮХА, в активна фаза, на възраст от 1 година и 2 месеца до 15 години. За лечение е използван апарат за комбинирано магнитно-лазерно въздействие "Изель" (Русия), с дължина на вълната на ИЧЛ 850 nm, мощност на излъчване 2 mW и магнитна индукция на изхода на излъчвателя - 40 mT. Приложени са две методики: локално-ставна и комбинирана: надбъбречна област и локално.

Резултатите са отчитани въз основа на локалната активност (болка, оток, сутрешна схованост) и общата биологична активност.

Отчетени са подобрение на ставния синдром при 8 деца и липса на динамика при 3.

S u m m a r y. The therapeutical effect combined influence of constant magnetic field with infrared laser at the background of drug therapy have been traced.

Eleven children with active phase juvenile chronic arthritis from a year and two months to 15 years old have been studied and treated.

The device for combined magnetic-laser influence "Izel" (Russia) have been used for treatment. It has wave length of 850 nm, radiation power of 2mW and magnetic induction in outlet - 40 mT. Two methods have been applied: local joint and combined - suprarenal area and local.

The result have been read by local activity (pain, edema, morning stiffness) and total activity.

Improvement of joint syndrome at 8 children and lack of changes at 3 children have been established.

Ювенилният хроничен артрит (ЮХА) е едно от честите заболявания на опорно-двигателната система с хронично рецидивиращ ход в детската възраст.

Проблемите на това заболяване се определят от неизвестната етиология, многообразието на клиничните прояви и социалната му значимост - рано настъпваща инвалидизация.

В комплексното лечение на ЮХА съществено място заемат физикалните фактори.

В сектора по физикална терапия на детските заболявания на база - Факултетска детска болница, с успех се прилагат нискочестотно импулсно магнитно поле, електрофореза с цитостатици, ултразвук, хелий-неонова лазерна акупунктура и др.

По литературни данни (1,2,3,4,5,9,10,11) магнитното поле (МП) е с изразено противовъзпалително, противоотечно и обезболяващо действие. Постоянното магнитно поле (ПМП), комбинирано с инфрачервен лазер (ИЧЛ) с дължина на вълната от 800 до 1200 nm, освен че подобрява микро-, хемо- и лимфоциркулацията, скъсява фазите на възпалителния процес и активизира процеса на регенерация. В тъканите възникват допълнителни физични явления. ПМП чрез преориентиране на диполите успоредно на лазерния лъч увеличава дълбочината на проникването му, повишава светопоглъщащата способност на биологичните структури и засилва метаболитните процеси (6,7).

В достъпната литература не сме срещнали данни за комбинирано въздействие на ПМП и ИЧЛ при заболявания в детската възраст.

Цел на настоящото съобщение е да проследим лечебния ефект при комбинирано приложение на тези два физикални фактора при деца с хроничен артрит (ХА).

Материал и методика

Изследвани и лекувани са 11 деца с ЮХА на възраст от 1 година и 2 месеца до 15

examination had confirmed the diagnosis: Primary angiitis of CNS.

Key words: child, primary angiitis, CNS, criteria of diagnosis

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Double Cortex Syndrome: Genetically Determined Childhood Epilepsy with Unusual Clinical Characteristics and New DCX Mutation

P. Dimova, S. Bonakdar, D. Moris-Rosendahl

We present a case with *double cortex syndrome*, in which refractory late-onset epileptic spasms and moderate cognitive deficit were observed. The interictal electroencephalographic (EEG) finding revealed diffuse slow wave changes, while the ictal EEG demonstrated paroxysms of high multiphasic slow waves accompanying the epileptic spasms. Magnetic resonance imaging showed a global band heterotopia. A novel *DCX* mutation has been found. Levetiracetam add-on to the basic valproate therapy led to full seizure control (> 3 months).

Key words: *double cortex, West syndrome, EEG, epileptic spasms*

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Polyneuritic Syndrome at the Beginning of Polyarteritis Nodosa

I. Litvinenko, K. Lisichki, D. Hristova

Polyarteritis nodosa (PAN), the prototype of systemic vasculitis, is a rare condition characterized by necrotizing inflammation of medium-sized or small arteries. Signs and symptoms of this disease are primarily attributable to diffuse vascular inflammation and ischemia of affected organs. Virtually any organ with the exception of the lungs may be affected, with peripheral neuropathy and symptoms from osteoarticular, renal artery, and gastrointestinal tract involvement being the most frequent clinical manifestations. Diagnosis in childhood requires a high degree of clinical suspicion as serological tests are at best non-specific, and absolute biopsy proof often requires examination of multiple sites. Early diagnosis is important, as treatment with corticosteriods usually induces symptomatic relief and provides a better chance of long term survival. If an acute remission is obtained the patient not only should have a good quality of life but in a proportion of cases, where the remission is prolonged, steroid therapy may be ceased. A case of polyarteritis nodosa presenting as peripheral neuropathy is reported. The nature of the patient's signs and symptoms led to the diagnosis of polyarteritis nodosa, which was supported by biopsy. Therapy with corticosteroids and endoxan resulted in rapid improvement.

Keywords: polyarteritis nodosa (PAN), peripheral neuropathy

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Case of Ponto - Cerebellar Angioreticuloma

I. Litvinenko

The author report 11 year-old girl with neoplasm within the ponto-cerebellar angle. She has dysphonia and dysphagia as alone clinical manifestation of the disease. Computed tomography (CT) of the brain showed a normal imagination. Magnetic resonance imaging (MRI) showed a heterogeneous mass in the ponto-cerebellar angle, with isointense signal on T1 and hyper intense signal on T2-weighted images and gadolinium enhancement. The tumor was histologically diagnosed as angioreticuloma. According to literature, this type of hamartoma does not occur or are weary rare in children of prepubertal age. CONCLUSIONS: Pineal region tumors have no pathognomonic imaging pattern. CT and MRI are complementary in diagnosis and are important to determine localization and extension.

Keywords: *angioreticuloma, neoplasm, ponto-cerebellar angle, CT, MRI*

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Topiramate - Efficacy in Epileptic Encephalopathies in Childhood

V. Bojinova, P. Dimova, V. Gergueltcheva, T. Yankov, V. Tomov

The aim of the study was to establish the therapeutic efficacy of на topiramate (TPM, Topamax) as add-on therapy in children with epileptic encephalopathies. . TPM was used in dose 6-9 mg/kg daily on 55 children aged 2 to 16 years. 30 children were with Lennox-Gastaut syndrome (LGS), 73,3%of which with symptomatic etiology; 10 children- with Dravet syndrome (SMEI), 5 children- with electrical status epilepticus during slow sleep (ESES); 8 children- with myoclonic- astatic epilepsy (MAE); 2 children- with Sturge-Weber syndrome.

Results: In SMEI TPM was effective in 8 children (80%): seizure-free were 3, with seizure reduction over 50%- 5 (50%). The clinical effect on the generalized tonic-clonic seizures (GTCS) (seizure-free were 5, with seizure reduction over 50%-5), secondary generalized tonic-clonic seizures (SGTCS) (seizure-free were 4, with seizure reduction over 50%-4) and on the myoclonic seizures (seizure-free were 3, with seizure reduction over 50%- 5) was the most significant; the clinical effect for the absences (reduction over 50% in 4 children) was the most insignificant. In LGS TPM was effective in 23/30 children (77%): 4 (13%) were seizure-free during the follow-up

Mean age at evaluation was 20.6 years (range 9-32) and mean follow-up duration was 13.2 years (range 5-25). The control group included 35 age- and sex-matched healthy volunteers.

Auxologic (height, weight, BMI and waist circumference (WC)) and physiological parameters (arterial pressure) were measured. Biochemical tests included serum glucose and lipid profile. The MS was defined according to the International Diabetes Federation's consensus.

MS was more frequent among long-term ALL survivors as compared with healthy controls - 14.9% vs 0% ($p<0.007$). Male survivors above and under 18 years of age were more likely to present with MS than male controls of the same age groups (24% vs 0%, $p<0.007$ and 17.8% vs 0%, $p<0.02$), respectively. Most prevalent components of MS in both survivors and controls were WC (31,9% vs 5,7% $p<0,001$) and (53,2% vs 14,3% $p<0,001$). The frequency rates of WC and HDL-cholesterol were also significantly higher among younger (under 18) and older (above 18) survivors as compared to controls. Survivors presented with one, two, or three MS signs frequently than controls (34% vs 14,3%, 17% vs 8,6% and 12,8 vs 0%, respectively). There was a weak positive nonsignificant relationship between cranial radiotherapy and MS ($p=0,097$) in males.

MS is more frequent among childhood ALL survivors, especially among men and those above 18 years of age. MC signs appear shortly after treatment, usually through abdominal obesity and lipid profile abnormalities. Routine follow-up of childhood ALL patients in long-term remission should include an assessment of metabolic risk.

Key words: *metabolic syndrome, childhood acute lymphoblastic leukemia survivors*

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Collagen type VI-related Myopathy. The First Genetically Verified Family with Bethlem Myopathy in Bulgaria.

T. Chamova, A. Taneva, S. Atemin, T. Todorov, M. Gospodinova, A.Todorova, I. Tournev

Collagen VI-related myopathies encompass a group of conditions with a spectrum of clinical severity ranging from mild Bethlem myopathy (BM) to severe Ullrichmuscular dystrophy. The disease is a result of mutations in the COL6A1,COL6A2, and COL6A3 genes that encode 3 of the collagen6 (COL6) chains. BM can be inherited in autosomal dominant trait,although recessive inheritance has also been described.BM is a benign myopathy with onset in the first two decades of life. It is characterized by contractures (commonly finger flexors and ankles) and slowly progressive proximal muscle weakness. The majority of patients remain ambulatory into their fifth decade of life,although muscle weakness may occur after this time.

We present the first genetically verified family with BM, due to heterozygous mutation c.1056+G>Ain COL6A1 gene. The clinical onset was in the first decade of life with limb-girdle muscle weakness, rigid spine, ankle contractures, slightly increased creatine kinase, spared ventillatory and cardiac functions.

Key words: *COL6, Bethlem myopathy, contractures*

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Benign Acute Childhood Myositis with Rhabdomyolysis

K. Lisichki, Kr. Petrova, V. Kenderova

Acute muscle pain and walking difficulty are symptoms compatible with both benign and severe degenerative diseases. Benign acute childhood myositis (BACM) is a rare, acute, transitory, self-containing muscle disease which affects predominantly school-aged children. We present a clinical case of a 12-year-old boy with BACM with muscle pains and walking difficulty, extremely high levels of muscle enzymes and myoglobinuria, in whom we diagnosed an enteroviral infection, followed by a complete recovery. A daily urine analysis and intravenous rehydration are of vital importance regarding the prevention of the most severe complication of rhabdomyolysis - the acute renal failure. Knowledge of the typical clinical course of the disease allows the opportunity to avoid unnecessary highly-specialized and expensive diagnostic tests in a portion of the patients.

Key words: *benign myositis, rhabdomyolysis, children*
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Portal Vein Thrombosis in Childhood: a Clinical Case

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The portal vein thrombosis in childhood is a rare disease, ranging from 1,1 / 100 000 to 0,72 / 1 000 000 live births, and the age and clinical manifestation is quite variable. It is the most common cause of preheatal non-cirrhotic, non-malignant portal hypertension in childhood and is defined as extrahepatic portal vein obstruction (EHPVO).

The etiology is not completely clear. The disease often develops slowly and manifests clinically with bleeding from the esophagus and / or splenomegaly. The low frequency of the disease, the difficulties in diagnosis and the need to prevent bleeding in the light of the developed guidelines, argue the authors to present and comment on their clinical experience.

A boy aged 1 year and 5 months presented with hematemesis and melaena. The fibrogastroscopy found in the lower third of the esophagus moderate varices without active bleeding at the moment. The article presents the diagnostic searches and the therapeutic activities in the described clinical case.

Key words: *portal vein thrombosis, risk factors for thrombosis, portal hipertesion, children.*

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МУЛТИСИСТЕМЕН ВЪЗПАЛИТЕЛЕН СИНДРОМ – КЪСНА ПРОЯВА НА COVID-19

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РЕЗЮМЕ

Мултисистемният възпалителен синдром при деца (MIS-C) е усложнение, което се наблюдава средно 2–6 седмици след развитие на SARS-CoV-2 инфекция в детската възраст. Характеризира се с фебрилитет, клинико-лабораторен синдром на възпаление и мултиорганна дисфункция, като показва известни сходства с други хиперинфламаторни състояния при децата като Болест на Кавазаки/Кавазаки шок синдром, Токсичен шок синдром и Синдром на макрофагеална активация (MAC)/Хемофагоцитна лимфохистиоцитоза. Разгръща се хиперимунен отговор с развитие на антиген-антитяло медирана цитокинова буря и изява най-често на кожно-лигавичен синдром, гастроинтестинална и кардиогенна увреда, като в част от случаите се развиват коронарни аневризми, миокардна фиброза и фиксирани ритъмно-проводни нарушения. Лечебният подход включва флуидотерапия, респираторно подпомагане, приложение на имуномодулатори, антиагреганти и антибиотици, като прогнозата в повечето случаи е добра. Бъдещите наблюдения ще покажат какъв ще бъде дългосрочният ефект на MIS-C върху детското здраве.

ВЪВЕДЕНИЕ

В литературата до момента съществуват голям брой публикации, описващи протичането на COVID-19 в детската възраст и асоциирането му с развитието на синдром на свръхвъзпалителен отговор в някои от случаите. Това състояние се обозначава с различни названия:
– Multisystem inflammatory syndrome in children (MIS-C) – Centers for Disease Control and Preventions (CDC, USA)
– Pediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2 (PIMS-TS) – Royal College of Pediatrics and Child Health (RCPCH, UK)
– Multisystem inflammatory disorder in children and adolescents – World Health Organization (WHO).

Между различните определения има малки възрастови, клинични, клинико-лабораторни и епидемиологични разлики (Табл.1) [1].

АВТОВЪЗПАЛИТЕЛНИ ЗАБОЛЯВАНИЯ ПРЕД ГАСТРОЕНТЕРОЛОГА

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РЕЗЮМЕ: Системните автовъзпалителни забслявания са хетерогенна група редки и тежки възпалителни заболявания, характеризиращи се с епизоди на непровокирано и неконтролирано системно възпаление поради дисрегулация на вродената имунна система. През последните години се повишава както тяхната честота, така и разпознаването им, което повишава клиничния интерес към тях. Поради тази причина ние предлагаме кратко представяне на най-честите автовъзпалителни заболявания в детска възраст, като акцентът е върху най-характерните им симптоми от страна на гастроинтестиналния тракт.

КЛЮЧОВИ ДУМИ: *автовъзпалителни заболявания, коремна болка, афтозни лезии, повръщане, диария*

ТЕРМИНЪТ АВТОВЪЗПАЛИТЕЛНИ ЗАБОЛЯВАНИЯ (AID) СЕ ИЗПОЛЗВА ЗА ПЪРВИ ПЪТ ПРЕЗ 1999 Г. ГРУПАТА НА АВТОВЪЗПАЛИТЕЛНИТЕ ЗАБОЛЯВАНИЯ ВКЛЮЧВА КАКТО МОНОГЕННИ ЗАБОЛЯВАНИЯ – ФАМИЛНА СРЕДИЗЕМНОМОРСКА ТРЕСКА (FMF), КРИОПИРИН-АСОЦИИРАНИ ПЕРИОДИЧНИ СИНДРОМИ (CAPS), МЕВАЛОНАТ КИНАЗЕН ДЕФИЦИТ (MKD), ТУМОР НЕКРОЗИС ФАКТОР РЕЦЕПТОР АСОЦИИРАН ПЕРИОДИЧЕН СИНДРОМ (TRAPS), ТАКА И МУЛТИФАКТОРНИ ЗАБОЛЯВАНИЯ КАТО НАПРИМЕР СИНДРОМА НА ВЕНÇЕТ.

AID са хетерогенна група редки и тежки заболявания, характеризиращи се с епизоди на непровокирано и неконтролирано системно възпаление, поради дисрегулация на вродената имунна система, което води до отделяне на цитокини. При тези заболявания няма наличие на антиген-специфични Т-лимфоцити и автоантитела, което ги различава от класическите автоимунни заболявания.
AID са група от генетично различни, но клинично сходни заболявания, засягащи няколко органи и системи. Общите им характеристики са представени в табл. 1.

АВТОВЪЗПАЛИТЕЛНИ ЗАБОЛЯВАНИЯ, ПРОТИЧАЩИ С КОРЕМНА БОЛКА

Диференциалната диагноза на острата коремна болка е трудна и включва много групи заболявания, ангажиращи ГИТ, пикочно-половата и дихателната системи. При анамнестични данни за повтарящи се епизоди на силна коремна болка, съпътстващ фебрилитет и повишение на маркерите на възпаление, при липса на отклонение в образните и микробиологичните изследвания, трябва да се обсъди и възможността коремната болка да е в контекста на някои от следните автовъзпалителни заболявания:

Фамилната средиземноморска треска (Familial Mediterranean Fever, FMF)
За първи път заболяването е описано през 1945 г. като самостоятелна болест. FMF е най-честото автовъзпалително заболяване. Предава се автозомно-рецесивно, свързано е с мутация на MEFV гена. Този ген кодира пирина, който участва в регулацията на т.нар. инфлазоми, играещи основна роля в продукцията и секрецията на проинфламаторни цитокини, като интерлевкин-1β (IL-1β). Инфлазомите са включени в патогенезата на различни автовъзпалителни заболявания.
FMF се среща по-често при народите, населяващи сушата около Средиземно море – евреи, араби, гърци, арменци, населението на северна Африка.
Най-често заболяването започва в периода на детството: при 25% до 60% от пациентите преди 10-годиш-

СИНДРОМ НА МАКРОФАГЕАЛНА АКТИВАЦИЯ, ПРОВОКИРАН ОТ MYCOPLASMA PNEUMONIAE В ДЕТСКА ВЪЗРАСТ – КЛИНИЧЕН СЛУЧАЙ

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РЕЗЮМЕ: Синдромът на макрофагеална активация е рядко, в повечето случаи животозастрашаващо състояние, което се характеризира с ексцесивна възпалителна реакция, дължаща се на неконтролиран и дисфункционален имунен отговор, включващ продължителна активация на Т-лимфоцити и макрофаги, което води до масивна хиперсекреция на проинфламаторни цитокини. Развива се най-често като усложнение на автоимунни ревматични заболявания, макар че и редица инфекциозни агенти могат да доведат до развитие на MAS.

Представяме момиче на 12 год., с вродена хипоплазия на очния нерв, при което по повод на фебрилитет кашлица, рентгенови промени за десностранна пневмония и положителни IgM антитела срещу Mycoplasma pneumoniae се прие, че се касае за микоплазмена пневмония. В допълнение се установиха обрив, хепатомегалия, лимфаденомегалия, както и промени в лабораторните показатели: тромбоцитопения (Platelet count 89 × 109/L), трансаминаземия (Aspartate aminotransferase 228 U/L), хиперферитинемия (1844 ng/ml), триглицериди – 1,1 mmol/l, фибриноген – 3,2 g/l, положителни D-димери. Въз основа на описаните промени се прие, че се касае за синдром на макрофагеална активация, провокиран от Mycoplasma pneumoniae. Започнатата терапия с пулсови дози кортикостероид и циклоспорин, в съчетание с етиологична терапия с хинулонов антибиотик доведоха до стабилизиране в клиничното състояние и нормализиране на променените лабораторните показатели за MAS, с изключение на феритин и тромбоцити. Включването на анти-IL-1-рецепторен антагонист доведе до трайно нормализиране на серумните нива на феритин и позволи спиране на лечението с циклоспорин.

КЛЮЧОВИ ДУМИ: *синдром на макрофагеална активация, Mycoplasma pneumoniae, детска възраст*

Синдромът на макрофагеална активация (СМА) е тежко, животозастрашаващо състояние, дължащо се на активирането и неконтролираната пролиферация на Т-лимфоцити и макрофаги, което води до ексцесивна хемофагоцитоза и свръхпродукция на цитокини. Наблюдава се като усложнение на автоимунни ревматични заболявания. Чести тригери на СМА са инфекциозни причинители – вируси, бактерии, гъбички, паразити.

КЛИНИЧЕН СЛУЧАЙ

Представяме момиче на 12 год. и 6 мес., с вродена двустранна хипоплазия на очния нерв.

Анамнеза на настоящото заболяване

Започва с остро начало – фебрилитет до 38,4-39°C, постепенно засилваща се кашлица, дисфоничен глас. Поради неповлияване на описаните симптоми от амбулаторно започнатото антибиотично лечение с цефиксим, е проведена рентгенова графия на бял дроб, установена е десностранна пневмония. Детето е хоспитализирано в детско отделение по местоживеене и е проведено венозно антибиотично лечение с фортум и амикацин, последвано от меронем. Персистирането на фебрилитета, кашлицата и изявата на генерализиран обрив налагат прекъсване на описаната терапия и насочване за хоспитализация към нашата детска клиника.

[illegible]

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[illegible]: Синдромът на макрофагеална активация (СМА) е животозастрашаващо усложнение на ревматичните заболявания, което по неясни причини се проявява много по-често при лица със системен ювенилен идиопатичен артрит. СМА е хиперинфламаторно нарушение. Характеризира се с панцитопения, чернодробна недостатъчност, коагулопатия и неврологични симптоми и се смята, че е причинен от активация и неконтролируема пролиферация на Т-лимфоцити и добре диференцирани макрофаги, което води хемофагоцитоза и свръхпродукция на цитокини.

Ние описваме неговите клинични прояви, най-честите тригери, лабораторните отклонения, диагностичните критерии и лечение.

[illegible]: синдром на макрофагеална активация, деца

СИНДРОМЪТ НА МАКРОФАГЕАЛНА АКТИВАЦИЯ (СМА) Е СЕРИОЗНО, ЖИВОТОЗАСТРАШАВАЩО УСЛОЖНЕНИЕ НА НЯКОИ СИСТЕМНИ ВЪЗПАЛИТЕЛНИ ЗАБОЛЯВАНИЯ. С НАЙ-ВИСОКА ЧЕСТОТА СЕ НАБЛЮДАВА ПРИ СИСТЕМНАТА ФОРМА НА ЮВЕНИЛЕН ИДИОПАТИЧЕН АРТРИТ (СЮИА), НО МОЖЕ ДА СЕ РАЗВИЕ И ПРИ ПАЦИЕНТИ С ДРУГИ АВТОИМУННИ И/ИЛИ АВТОВЪЗПАЛИТЕЛНИ ЗАБОЛЯВАНИЯ, ТАБЛ. 1 [5, 6, 15]. СМА СЕ ХАРАКТЕРИЗИРА С ЕКСЦЕСИВНА ВЪЗПАЛИТЕЛНА РЕАКЦИЯ, ДЪЛЖАЩА СЕ НА НЕКОНТРОЛИРУЕМ И ДИСФУНКЦИОНАЛЕН ИМУНЕН ОТГОВОР, ВКЛЮЧВАЩ ПРОДЪЛЖИТЕЛНА АКТИВАЦИЯ НА Т-ЛИМФОЦИТИ И МАКРОФАГИ, КОЕТО ВОДИ ДО МАСИВНА ХИПЕРСЕКРЕЦИЯ НА ПРОИНФЛАМАТОРНИ ЦИТОКИНИ. КЛАСИФИЦИРА СЕ КАТО ВТОРИЧНА ХЕМОФАГОЦИТНА ЛИМФОХИСТИОЦИТОЗА (ХЛХ) [6, 19, 24].

ЕПИДЕМИОЛОГИЯ

[illegible]

Ретроспективни мултицентрови проучвания за 20-годишен период установяват, че заболяването се наблюдава при 25% от пациентите със сЮИА, а субклиничните форми са до 40% [18, 20]. При системния лупус честотата е 5%, а при болестта на Kawasaki – 1.1%-1.9% [3, 9, 22].

СМА възниква най-често в началото на заболяването, но са описани случаи до 14 години от дебюта на болестта.

ЕТИОЛОГИЯ

[illegible]

Най-честият тригер е изостряне на основното заболяване, следван от смяна на терапевтичния план, инфекции, табл. 1.

ПАТОГЕНЕЗА

В патогенезата участват както вродената, така и адаптивната имунна система. Заболяването често се развива на фона на определен генетичен терен (носителство на хетерозиготни мутации, които в хомозиготитен тип се срещат при фамилната ХЛХ) и терен на хронично възпаление. Дължи се на ексцесивна активация на Т-лимфоцити и макрофаги, които изявяват хемофагоцитна активност.

[illegible]

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Ваксините са едно от големите постижения на съвременната медицина. Те служат за предотвратяване на заболявания, част от които протичат тежко, а за други няма ефективна терапия. Основен проблем е недостатъчният имунизационен обхват. Той трябва да е над 90-95%, за да се предотвратят инфекциозни взривове. Освобождаването на специфични категории деца с определени заболявания, както и антиваксиналните нагласи не позволяват да се постигне тази цел.

ПОРАДИ АВТОМУННАТА ПАТОГЕНЕЗА И ПРОВЕЖДАНАТА ТЕРАПИЯ С ИМУНОСУПРЕСИВНИ МЕДИКАМЕНТИ ДЕЦАТА С РЕВМАТИЧНИ ЗАБОЛЯВАНИЯ (РЗ) СА С УВЕЛИЧЕН РИСК ОТ ИНФЕКЦИИ, ПО-ВИСОКА ЗАБОЛЯЕМОСТ И СМЪРТНОСТ. ЕФЕКТИВНАТА, НО И БЕЗОПАСНА ВАКСИНАЦИЯ Е ОТ КЛЮЧОВО ЗНАЧЕНИЕ ЗА НАМАЛЯВАНЕ НА ВАКСИНОПРЕДОТВРАТИМИТЕ ИНФЕКЦИИ ПРИ ТЯХ. ЕТО ЗАЩО ПРЕЗ 2011 Г. EULAR (EUROPEAN LEAGUE AGAINST RHEUMATISM) ПУБЛИКУВА ПРЕПОРЪКИ ЗА ВАКСИНИРАНЕ НА ДЕЦА С РЗ. В СЛЕДВАЩИТЕ ГОДИНИ ИМА МНОЖЕСТВО ПУБЛИКАЦИИ, КОИТО ДОПЪЛВАТ ТЕЗИ ПРЕПОРЪКИ И СА ОТРАЗЕНИ В НАСТОЯЩИЯ ОБЗОР.

Преди да вземем решение за имунизиране на дете с РЗ, трябва да отговорим на две групи въпроси, свързани с ефективността и безопасността на ваксините.

ЕФЕКТИВНОСТ

Най-точен показател за ефикасността на една ваксина е честотата на заболяванията при имунизираните пациенти. Такива проучвания се правят рядко, защото изискват големи групи пациенти и са скъпи. Затова се използва заместващ показател като имуногенност. Имуногенността се отнася до имунната реакция, предизвикана от ваксинацията. Това обикновено се измерва чрез специфични за ваксината геометрични титри на антителата или геометрични концентрации, процент на сероконверсия и/или нива на серопротекция. Освен тези краткосрочни ефекти е важно персистирането на имунологичната памет, т.е. колко време след имунизацията децата с РЗ са защитени срещу съответното заболяване. Друга група въпроси са свързани с това каква е ефективността на имунизациите при различните РЗ и как се влияе от прилаганото лечение с кортикостероиди, цитостатици, антималарици, сулфасалазин-, биологични медикаменти.

БЕЗОПАСНОСТ

Безопасността на ваксините при деца с РЗ се определя от честотата на нежеланите поствакцинални реакции в сравнение със здрави контроли, повишаване на активността или рецидив на основното заболяване, поява на локални или генерализирани инфекции, предизвикани от живи атенюирани патогени във ваксините. Има ли разлики в профила на безопасност при различните РЗ и влияе ли се от провежданата терапия? Могат ли ваксините или техните съставки, вкл. адюванти, да причинят автоимунно заболяване.

ИМУНОГЕННОСТ И ГЛЮКОКОРТИКОСТЕРОИДИ

Децата, лекувани с глюкокортикостероиди (ГКС), обикновено са с по-ниски концентрации на антиваксинални антитела, но достигат серопротективни титри. Имуногенността е по-добра при дози, съизмерими с метилпреднизолон до 0.5 мг/кг/т, но не повече от 20 мг/24 ч. Дози под 10 мг/24 ч. осигуряват по-добра серопротекция. Едновременното приложение на ГКС и други имуносупресори се свързва с по-нисък, но все още достатъчен защитен отговор. Малко са проучванията върху персистирането на ваксиноспецифичните антитела, затова се препоръчва рутинното им изследване и при спадане – приложение на бустерни дози. В заключение можем да отбележим, че няма общ вреден ефект на ниски дози ГКС върху имуногенността или установените нива на антитела.

ИМУНОГЕННОСТ И ДРУГИ ИМУНОСУПРЕСИВНИ МЕДИКАМЕНТИ

Не е установен негативен ефект върху краткосрочната имуногенност и персистирането на поствакциналните антитела при лечение с имуносупресори, ако се спазват следните дози: метотрексат под 15 мг/кв.м/седмично или 0.4 мг/кг/т/седмично, азатиоприн 1-3 мг/кг/т/24 ч., циклос-

Постстрептококов реактивен артрит

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Концепцията за реактивните артрити ги определя като разнородна група възпалителни несептични артрити, включващи ревматичната болест (РБ), постстрептококовия реактивен артрит (ПСРА) и артритите, свързани с инфекции на генитоуринарния и ентералния тракт, предизвикани от специфични микроорганизми. В доантибиотичната ера основно внимание се обръща на РБ поради голямата й честота - 100-200/100 000. Едва след широкото приложение на пеницилина и намаляването на РБ до 0.5-3/100 000 се обръща внимание на артритите, възникнали след стрептококов фарингит, но непокриващи напълно ревизираните диагностични критерии на Jones.

В исторически план още през XVIII-ти век са описани различни клинични синдроми, свързани с преболедуване от скарлатина. В тази традиция през 1959 год. *Crea и Mortimer* описват "скарлатинозен артрит" при деца, който не отговаря на критериите за РБ. В същата година *Friedburg* наблюдава РБ при възрастни, която протича далеч по-често с артрит, без прояви на кардит, хорея, подкожни възли, еритема маргинатум. Той предлага тази болест да се отдели от РБ при децата и я дефинира като "недеформиращ фебрилен полиартрит без кардит след инфекция с група А стрептококи" (ГАС)[5,7]. Това са първите съобщения на пациенти с ПСРА. *Goldsmith и Long* 23 години по-късно публикуват случаи на деца с постстрептококов симетричен артрит, който се влияе лошо от лечение с аспирин, както и възрастни със сакроилиит. Така постепенно характерната клинична картина на ПСРА се очертава като сигнификантно различна от тази на РБ. *Deighton* (1993 год.) предлага отличителни белези на ПСРА, а през 1997 год. *Ayoub и Ahmed* ги допълват и създават първите диагностични критерии *(Табл. 4)*. В контраст с артрита при РБ, ПСРА се определя като рано започващ след стрептококов фарингит, немигриращ, с протрахирано протичане, лошо отговарящ на лечението с аспирин или други нестероидни противовъзпалителни средства (НСПВС). Въпреки тези различия някои автори смятат, че ПСРА се включва в разширения спектър на РБ. През последните години все повече проучвания показват, че този синдром е самостоятелна категория с различна патогенеза и клинична картина от тази на ревматичния артрит.

Епидемиология

Трудно е да се определи честотата на ПСРА поради факта, че се публикуват единични случаи или малки групи пациенти. Анализът на рискови групи, както и ретроспективните проучвания, я определят на 1-2/100 000, т.е. два пъти по-висока от тази на РБ. В смесени групи от деца и възрастни няма ясно изразена полова предиспозиция. При селектирани групи в детска възраст има леко преобладаване на момчетата, а над 18-годишна възраст - на женския пол.

Възрастовото разпределение е различно от това при РБ, която е с пик около осмата година и не се наблюдава след 25-годишна възраст. При ПСРА възрастовият спектър е много широк - от 3 до 70 год. и е с бимодална крива на заболяемостта с първи връх между 8 и 14-годишна възраст, средно 9.7 години, и втори - между 21 и 37 год., като при втория пик 60% от заболелите са жени.

Честотата е различна по географски райони. Мнозинството от случаите са в САЩ и Европа, значително по-малко в Азия и Австралия и нито един в Африка. В САЩ преобладават децата, а в Европа - възрастните. Няма точно обяснение на тези факти най-вероятно значение имат честотата на ГАС фарингитите, както и информираността и подготовката на лекарите.

Етиология и патогенеза

При всички пациенти се доказва предшестваща стрептококова инфекция чрез изследване на култури от различни материали, бързи антигенни тестове, антистрептолизинов титър (АСО), антидезоксирибонуклеаза В (АДНаза В). Положителни култури се откриват при 37% от изследванията, като в 95% са от гърло, в 4% от хемокултури и в 1% - вагинален и цервикален секрет. Докато при РБ единствен етиологичен агент са ГАС, то при ПСРА се установяват и стрептококи от други групи *(Табл. 1)*.

Латентният период между началото на инфекцията и ПСРА е значително по-къс (10 дни) в сравнение с този при РБ. При деца има съобщения за начало на артрита на четвъртия ден от началото на стрептококовия фарингит[8,9].

Патогенезата не е изяснена. Предполага се участието на имунологични механизми по аналогия с РБ: общи анти

Recommendations for Electronic Health Records in Practice of Pediatricians

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EHR solutions designed specifically for pediatricians often include features which are not found in other more generic medical records packages. These features must include many of the following: time of birth as well as date of birth, allowing important decisions to be made during the first days of life; unique patient identifier methodology, which allows entire families to be indexed together; the abilily to deal with various family structures, including adoption, step-children, foste·care and child abuse, etc.; immunization tracking and preventive service reminders; age based normal findings, such as for vital signs and other laboratory tests; medication prescribing information which offers dosing assistance which is age and weight based, allergy and medication lists, and clinical decision support; maintenance of genetic information, including assistance with genetic counseling; special privacy issues; electronic connection to other sources of clinical data (i.e., hospital, pharmacy or laboratory), registry linkages and exchange of information at regional, national and European levels; pediatric specific templates and plans, integration with medical equipment and automation of document creation

Keywords: *Electronic Health Record (EHR), pediatrics, clinical practice*

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Computer Estimated GRF and Its Implementation in Adolescent Children

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We implemented Cockcroft,s equation for estimation of creatinine clearance and glomerular filtration rate in the Clinical Laboratory of the University Hospital - Pleven. Thus we investigated the possibilities for applying the equation in adolescent children by comparing the achieved values with those estimated by using both Schwartz's classic equation for children and Conant-Barraat's equation. In this article we present GRF average values calculated according to the three equations and correlation coefficients, which show statistically significant correlation - r =0.818 in the boys' group, and r = 0.948 in the girls' group.

Key words: *glomerular filtration rate, creatinine, Cockcroft - Gault*

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Diastrophic Dysplasia in Newborn Baby

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Skeletal dysplasias are congenital disorders of morphogenesis, development and growth of the skeleton. Their clinical manifestations include short stature, abnormal body proportions and/or limb shape, deformity, and various complications in different organs.

Skeletal dysplasias are genetically determined. Although most dysplasias are individually rare, there is great heterogeneity, and over 200 different phenotypes of dysplasias and dysostoses have been distinguished by a combination of clinical features and skeletal appearance on radiography.

Diastrophic dysplasia was one of the first skeletal dysplasias to be clearly delineated on a phenotypic and radiographicground. It was described by Lamy and Maroteaux in 1960.

We present a 25day old child from third pathological pregnancy with dysmorphic feature, brachicephaly, marked short stature (both the trunk and the limbs areshortened, but the limbs more severely so), bilateral clubfoot, cleft palate, characteristic hand deformities, changes of the ear pinnae, progressive kyphoscoliosis of the spine, and joint stiffness.

Key words: *skeletal dysplasias, diastrophic dysplasia, abnormal body proportions, cleft palate*

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Drug-induced Lupus by Thionamide Therapy with Description of Two Cases

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Side-effects during the treatment for thyrotoxicosis in childhood occur in 3-5% of the patients. Drug-induced lupus /DIL/ is one of the rarest adverse reactions- in less than 0.1 % of the patients. Two such cases are presented, induced by Propylthiouracil and Methimazole. The first patient was a 16 years old girl, with thyrotoxicosis of 3 years duration, treated with Propylthiouracil with good effect. The first symptom of DIL /arthritis/ was associated with an increase of the dose after a relapse. She was referred to our hospital 7 months later with a full-blown clinical picture: photosensitivity, oral ulcerations, serositis, arthritis, confirmed by the immunologic investigations. After a course with corticosteroids and resolution of the symptoms, the antithyroid treatment was resumed with Methimazole. 3 months later the symptoms of DIL returned. Thyroid surgery was performed. In the other case, a 12 years old girl with first presentation of thyrotoxicosis, DIL began with skin rash and arthralgia 2 weeks after introducing a treatment with Methimazole. Propylthiouracil was continued a month after, without any adverse effects for the next 15 one and a half year. Children treated with thionamides should be followed carefully and in cases suspicious for DIL immunological investigations should be carried out.

Key words: *graves disease, children, LED, drug-induced lupus*

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recessive. It is estimated that 9% to 10% of the total population of the Western world is homozygous, and up to 42% of the population is heterozygous. For a period of 5 years (2016-2020), in the gastroenterological clinics of the University Hospital "St. Marina", Gilbert's syndrome was proved in 35 patients by molecular genetic analysis. This metabolic disorder is an extremely common cause of unconjugated hyperbilirubinemia that is easily diagnosed. With clinical thinking about it, unnecessary examinations of the patient and the family can be avoided, carriers can be found in the family and the manifestation of the pharmacogenetic defect in the treatment of solid tumors with irinotecan (medication used to treat colon cancer, and small cell lung cancer) can be prevented.

Key words: Gilbert's syndrome, liver, genetic analysis, children, adults

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COVID-19 Induced Myosistis - a Clinical Case

K. Lisichki, V. Kenderova, M. Ganeva

Coronavirus disease - 2019 (COVID-19) was first identified in Wuhan, China. When compared with adults, children are less commonly symptomatic when having acute Covid-19 infection In contrast to adults, most infected children appear to have a milder course and have better outcomes overall. Myositis is a rarely seen in COVID-19 patients. Its pathogenesis is probably multifactorial with direct myocyte invasion, toxin-mediated muscle damage, dysregulated immune response, and immune-mediated mechanisms playing a role.

We report the clinical case of a 7-year-old boy with COVID-19 induced myositis with favorable outcome. At the fourth day of treatment the child is without clinical signs of myositis. At the sixth day creatine kinase levels normalized. The evolution of the myositis is favorable and corresponds to the reports in the literature. This was a second episode of myositis for the boy in the time frame of nine months. The first episode was defined as self-limiting benign acute childhood myositis with unknown trigger.

COVID-19 myositis might develop at any time during the course of the infection, usually during the second week but it can be also the presenting symptom. COVID-19 induced myositis, although rare, might be accompanied by rhabdomyolysis and might be a life-threatening complication. Early recognition and effective management with intravenous hydration are crucial for the effective management and prevention of development of acute kidney failure.

Key words: COVID-19, myositis, children

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Multisystem Inflammatory Syndrome Associated with COVID-19 Infection in Children - Experience with First Patients in Varna

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Multisystem Inflammatory Syndrome associated with COVID-19 infection in children (MIS-C) is a severe, life-threatening, rapidly evolving condition that occurs 3 to 4 weeks after infection with the SARS-CoV-2 virus. It results in persistent fever for more than 4 days and severe inflammation of many organs - heart, lungs, eyes, skin, kidneys, brain, blood vessels, gastrointestinal tract, etc. mainly in older children and adolescents without previous underlying chronic diseases. This syndrome requires rapid and accurate diagnostics in order to initiate treatment with properly selected immunomodulatory, anticoagulant, antiplatelet, infusion, antibiotic and cardiotonic therapy. The proper assessment of the clinical and laboratory values together with adequate treatment leads to complete recovery without complications.

The aim of this study is to share our experience in diagnosis, treatment and follow up of the first four children with MIS-C, who were treated at the Paediatric clinics of the Department of Paediatrics at the University Hospital „Sveta Marina", Varna. Detailed knowledge of this novel disease is responsibility of every Paediatric clinic in the current Corona virus pandemic situation.

Key words: multisystem inflammatory syndrome (MIS-C), COVID-19 infection, children

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Post-Infectious Myopathy in a Child with a Manifestation of COVID-19 Infection

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Myopathy and myalgias are relatively rare in adult patients with COVID-19 and there are insufficient cases of children in the available literature. We present a case of a 10-year-old girl with initial symptoms such as fever, headache, subsequent myalgia and muscle weakness of the four limbs and paraspinal muscles, who was diagnosed with inflammatory myopathy developed during SaRS-CoV-2 infection with diagnosed with Covid-19. The diagnosis of myositis or inflammatory myopathy was based on anamnesis data, a constellation of laboratory data for elevated creatinine kinase (CPK) level: by 195 U/l and the presence of electromyographic changes in the affected muscles characteristic of myogenic damage. After symptomatic treatment, there was an improvement in gait and a significant improvement in the muscular strength of the four limbs. Clinical suspicion of viral myositis-induced rhabdomy-

Key words: bronchodilation, corticosteroid, immunotherapy, biology therapy, history of asthma

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Therapy of Pediatric Asthma - Millennial Challenge - Part Two

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Bronchial asthma as a common and well-known old disease is a wonderful example for evolution of medical knowledge over the years and its impact on the therapeutic approach. Although the initially applied anti-allergic measures are still relevant today, the greatest advances in therapy have been made in the last century due to the central role of bronchodilators and the powerful anti-inflammatory effect of inhaled corticosteroids.

This is the second part of our extensive review focusing on the change in therapeutic approach, in line with the expanded scientific knowledge on the pathogenesis of bronchial asthma.

Key words: bronchodilation, corticosteroid, immunotherapy, biology therapy, history of asthma

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Consensus Guidelines for Growth Hormone Treatment in Children with Prader-Willi Syndrome

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This updated guideline provides evidence-based recommendations regarding the diagnosis, and growth hormone treatment for children with Prader-Willi syndrome (PWS).

PWS is a genetic condition caused by loss of the paternal copy of a region of imprinted genes on chromosome 15 (15q11-q13). PWS is characterized by severe muscular hypotonia and failure to thrive in the neonatal period, followed by hyperphagia, obesity, short stature, hypogonadism, mild to moderate intellectual impairment and behavioral problems. The phenotype is mainly due to hypothalamic dysfunction which may lead to multiple hormonal disturbances. Recombinant human growth hormone (rhGH) is the first treatment for the PWS patients that radically changed the care of these children. Main benefits of rhGH treatment are improved body composition and linear growth, increased levels of physical activity, which can help with the aim of preventing severe obesity. GH therapy is contraindicated in the presence of severe obesity, untreated severe obstructive sleep apnea, active cancer and uncontroiled diabetes mellitus. The rhGH treatment is only effective in conjunction with dietary and life-style measures.

Key words: Growth hormone treatment, Prader-Willi syndrome, childhood

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Kawasaki Disease in two 3 Month Old Infants

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Kawasaki disease (KD), also known as mucocutaneous lymph node syndrome and Kawasaki syndrome, is an acute febrile illness of early childhood characterized by vasculitis of the medium-sized arteries with predilection for the coronary arteries, leading to the development of coronary artery aneurysms (CAAs). KD is the most commonly acquired heart disease in pediatric age. The acute phase is characterized by inflammation of the medium-sized arteries of different organs, which leads to various clinical symptoms.

KD is most often met in the age group from 6 month to 5 years. Only 10% of the patients are below the age of 3 months, and only 1.67% of the patients are below the age of 1 month.

The etiology is unknown. There is no specific diagnostic marker, therefore the diagnosis is based on diagnostic criteria.

This article aims to outline the difficulties in the diagnostic process of two 3-month old infants with incomplete form of KD, the current treatment options and the recommendations for follow-up observation according to the American Heart Association.

Key words: Kawasaki disease, infants, diagnosis

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Симптоматично лечение на острите инфекции на горните дихателни пътища в детска възраст

Остри инфекции на горните дихателни пътища (ОИГДП) е термин, който обхваща полиетиологичен симптомокомплекс с разнообразно клинично протичане, обединен от анатомичната локализация на процеса. Горните дихателни пътища включват назалните ходове, параназалните кухини, фаринкса и ларинкса, които свързват външната среда с трахеята, бронхите и белодробните алвеоларни пространства. Специфични нозологични категории са ринит, синуит, фарингит, епиглотит, ларингити трахеит. Възможно е едновременното засягане на две или повече анатомични области.

Най-честите ОИГДП са:

- Ринит - възпаление на назалната лигавица;
- Риносинуит или синуит - възпаление на носа и параназалните синуси (фронтални, максиларни, етмоидални и сфеноидален);
- Ринофарингит - възпаление на носа, фаринкса, хипофаринкса, увулата, и тонзилите;
- Фарингит - възпаление на фаринкса, хипофаринкса, увулата, и тонзилите;
- Стафилит или увулит - изолирано възпаление на увулата, което понякога се засяга и меките части на твърдото небце;
- Епиглотит - възпаление на епиглотиса, горната част на ларинкса и супраглотисната област;
- Ларингит - възпаление на ларинкса;
- Ларинготрахеит - възпаление на ларинкса, трахеята и субглотисната област;
- Трахеит - възпаление на трахеята и субглотисната област.

ЕПИДЕМИОЛОГИЯ

Острите инфекции на горните дихателни пътища са най-честите инфекции. Заболяемостта от тях и от грип е по-висока, в сравнение с всички други инфекциозни заболявания. Имат подчертано сезонно разпределение - започват през есента и постепенно се увеличават през есенно-зимния период, но се срещат и през лятото (ентеровирусни инфекции).

Полова предиспозиция. Хормоналните промени по време на менструалния цикъл при момичетата водят до хиперемия и оток на лигавицата на носа и синусите и хиперсекреция, което рефлектира в по-тежко протичане на ринита и/или по-лесното му възникване. При ларингит, ларинготрахеит и епиглотит превалира мъжкият пол в съотношение момчета:момичета - 3:2.

Възрастова предиспозиция. Честотата на ОИГДП е най-голяма при децата под 5-год. възраст. Те боледуват прибилизително 3-8 пъти годишно от вирусни респираторни заболявания, юношите и възрастните - 2-4 пъти, а индивидите над 60 години - веднъж годишно. Вирусните и бактериални фарингити са най-чести във възрастта от 4-7 години, епиглотитите - между 2-7 години, с пик на третата година. Ларингитът се наблюдава във всяка възраст, но е най-чест при деца на възраст от 6 месеца до 6 години, с пик през втората година от живота.

ЕТИОЛОГИЯ

Вирусите са най-честите причинители на ОИГДП - до 98%. Най-честите причинители са Rhinovirus, Coronavirus, Adenovirus, Coxsackievirus. Риновирусите се установяват при 50% от заболелите деца. По-рядко се доказват Adenovirus, Orthomyxovirus (вкл. grippe A и B virus), Paramyxovirus (paragrippe virus), Respiratory syncytial

ЕСТ НА КАВАСАКИ С ПРИНОС НА ЕДИН СЛУЧАЙ

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К. Лисички, Н. Иванова — *Боледзнь Кавасаки с описанием одного*

Резюме. Описан новый случай болезни Кавасаки у мальчика 7 месяцев
овных симптомов заболевания — фебрильности, длящейся более 5 дней, к
ной в орофарингсе (покрасневшие сухие, растрескавшиеся, с рагадами губы
ная слизистая оболочка полости рта), кожной сыпи, отека ладоней и стоп, у
еских узлов, наблюдали также и некоторые нетипичные реже встречаемы
гащают клиническую картину заболевания. Таковы: ишемии периферическ
атомегалия са слегка повышенной активностью трансаминазы, энтерит. Пор
системы выражено преходящей альбуминурией и стерильной лейкоцитури
ю. Параклиническим исследованиями установлены анемия, лейкоцитоз с
нуклеоз, тромбоцитоз, повышение СОЭ, увеличение альфа$_2$ и гамма
альбуминов. После проведенного лечения НСПВС клинические явления
вого месяца число лейкоцитов, тромбоцитов и уренень гемоглобино пр
ерживается слегка повышенным до конца 4-го месяца, что вызвало необход
держивающего амбулаторного лечения. Проведенный спустя 6 месяцев к
азал, что у ребенка соматический статус в норме и налицо отсутствие пат
ий при лабораторных исследованиях.
Таких детей необходимо диспансеризовывать и наблюдать за ними в детских
ких кабинетах, чтобы избежать опасности наступающих позднее осложнени

Ключевые слова: болезнь Кавасаки, ишемии периферических сосудов.

K. Lisichki, N. Ivanova — *Kawasaki Disease: Case Report*

Summary. A boy, seven months of age, developed the main symptoms of
shness during more than five days, conjunctivitis, changes in the rhinopharyn
rhagades, dry and hyperaemic oral mucosa), skin rach, edema of the hands an
s, as well some atypical and rarely observed manifestations which enrich the cli
— peripheral vascular ischaemiae, arthritis, hepatomegaly accompanied by sl
inase activity enteritis. Involvement of the urinary system was marked by
and sterile leukosyturia. No involvement of the heart has been recorded.
ngs include — anaemia, leukocytosis with a shift to the left and polynucleo
ased ESR, increased alpha$_2$ — and gammaglobulins, desreased albumin. Cli
ded after treatment with non-steroid antiinflammatory drugs. At the end of
te and thrombosyte count as well haemoglobin returned to normal. ESR dosclc
nd of the fourth month this leading to continiation of treatment as outpatient. /
been found that the child is in a normal state with no pathological deviatio