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X Европейский конгресс педиатров

От редакции: С 7 по 9 октября 2021 года в Хорватии, г. Загребе, состоялся юбилейный X Европейский конгресс педиатров — Europaediatrics 2021, организаторами которого были Европейская педиатрическая ассоциация/Союз национальных европейских педиатрических обществ и ассоциаций (EPA/UNEPSA) и Хорватская педиатрическая ассоциация. В важном для детских врачей мероприятии, которое проходило в гибридном формате, приняли участие около 1400 участников из 28 стран со всех континентов мира.

В научных сессиях конгресса эксперты с мировым именем представили актуальную информацию по вопросам охраны здоровья детей и подростков, профилактики заболеваний и инвалидности, основанную на принципах доказательной медицины. Особое внимание было уделено содействию реализации прав детей на здоровье, равенство и социальную справедливость. Учитывая эпидемиологическую обстановку, ключевой темой конгресса была и пандемия COVID-19, поэтому ряд заседаний были посвящены особенностям течения и последствиям новой коронавирусной инфекции у детей.

Российскую делегацию во главе с Президентом Союза педиатров России Лейлой Намазовой-Барановой представили более 35 педиатров и детских медицинских сестер из Москвы, Иркутска, Саранска, Грозного, Кемерово и других регионов нашей страны. В ходе устных и постерных докладов российские детские врачи и медицинские сестры успешно представили результаты своих научных исследований, поделились практическим опытом, приняли активное участие в научных дискуссиях и получили высокую оценку со стороны своих иностранных коллег.

Признанием на международном уровнем было вручение Президенту Союзу педиатров России Лейле Намазовой-Барановой награды X Европейского конгресса педиатров и Хорватского педиатрического общества за выдающийся вклад в проведение X конгресса Европейской педиатрической ассоциации и международное сотрудничество.

Союз педиатров России поздравляет Европейскую педиатрическую ассоциацию (EPA/UNEPSA) и Хорватскую педиатрическую ассоциацию с блестящим проведением юбилейного X Европейского конгресса педиатров и желает дальнейших успехов.

Представляем Вашему вниманию тезисы педиатров и детских медицинских сестер, представлявших российскую делегацию на Конгрессе. Europaediatrics 2021 проходил на английском языке, поэтому тезисы участников представлены на иностранном языке.

Prevalence of smoking and alcohol consumption among Russian adolescents

Julia Levina[1, 2], Leyla Namazova-Baranova[1, 2, 3], Kamilla Efendieva[1, 2], Anna Alekseeva[1], Elena Vishneva[1, 2], Valery Zvonarev[1]

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Objective. Smoking is one of the main preventable causes of death, but nonetheless leads to more than 7 million deaths in the world each year. Worldwide, 3 million deaths every year result from harmful use of alcohol. Russia is facing a serious burden of smoking and alcohol consumption, which affects not only adults, but also adolescents. In order to conduct appropriate tobacco and alcohol control policies, it is important to understand smoking and alcohol consumption prevalence.

Methods. An analysis of the prevalence of smoking and alcohol consumption was carried out in the research center in Moscow, Russian Federation among 303 adolescents (84 boys and 219 girls) 15–18 years old who participated in a study of the prevalence of allergic diseases according to the protocol of the research program GA2LEN. All participants completed the standardized GA2LEN questionnaire.

Results. 25% of respondents (11% of boys and 14% of girls) said they had smoked for as long as a year, among them, most started smoking at the age of 16.68% of teens who said they were still smoking as of one month ago, smoked 10 cigarettes a day. Among participants who had previously smoked, 55.1% already stopped smoking or started to smoke less, most of them at the age of 18. Among those who stopped smoking, they had smoked 12 cigarettes per day in average. 27% of participants were regularly exposed to tobacco smoke in the last 12 months. 34.2% of respondent's fathers smoked regularly during their childhood, 7.9% of respondents had mothers who smoked regularly during their childhood/before they were born. 3.9% of respondents said that they had been hospitalized before the age of two years for lung disease We found a statistically significant association between a mother smoking during childhood or before the birth of a child and that child being hospitalized before the age of two years for lung disease (p = 0,001).

53% of respondents (14% of boys 39% of girls) said they never drank any alcohol. 31% of participants (7% of boys and 24% of girls) said they drank alcohol less than once a week, 9% of respondents (5% of boys and 4% of girls) said they drank alcohol at least once a week but less than once a day.

Conclusion. Further efforts should be devoted to encouraging smoking and alcohol cessation and to develop strategies for preventing tobacco and alcohol use in young patients.

Cognitive functions and lifestyle of Russian schoolchildren in large cities

George Karkashadze[1], Leyla Namazova-Baranova[1, 2, 3], Alexey Molodchenkov[1], Elena Vishneva[1, 2], Tinatin Gogberashvili[1], Tatiana Konstantinidi[1]

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Objective. To determine the relationship between the parameters of cognitive functions, school performance and extracurricular lifestyle.

Method. 1036 children aged 10–12 years who studied in 5-th classes of 40 Russian schools in 8 different major cities were surveyed. All of them underwent cognitive testing, which included a set of six tests. They defined: arbitrary attention, visual-spatial perception, verbal memory, visual-imaginative thinking, constructive praxis, and verbal-logical thinking. Parents of 598 participants filled out questionnaires about their lifestyle. Using machine learning methods, children were divided into clusters based on the success of cognitive tests. Next, we analyzed the links between cognitively successful children, school performance, and lifestyle.

Results. It was found that children are divided into two clusters: those who performed cognitive tasks more successfully and less successfully. A strong direct link was established between the success of cognitive tests and school performance in three main subjects (mathematics (r = 0.875), literature (r = 0.853), and Russian (r = 0.797). Those who spent more time using the Internet and were more interested in computer games did not differ in cognitive parameters from those who used less and played less. But those who didn't play computer games at all during school days were worse at cognitive functions. Also worse in cognitive functions were those who were interested in unorganized sports (outside of sports clubs), hockey, mountain skiing, lawn tennis, a combination of a passion for music and education with Tutors. High cognitive functions are associated with music, non-sports Hobbies, basketball, football, dancing, and summer recreation in camps.Many lifestyle manifestations were not related to the level of cognitive function.

Conclusion. Cognitive functions are strongly associated not only with school performance, but also with certain Hobbies and lifestyles of children. Such data may be interesting for social policy in the field of childhood.

Are primary care pediatricians ready to care for children with autism spectrum disorders (ASD): challenges and tasks

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Objective. Given the prevalence of autism spectrum disorder (1–2%), all primary care pediatricians should be prepared to care for children with ASD and their families. However, children with ASD often have poor access to appropriate medical support all over the world, including the developed countries. In this paper we describe challenges and tasks in providing of primary health care for children with ASD in Russia.

Methods. Serie of foresight-sessions "Pediatric health care service for children with ASD" (4 sessions). During the sessions, working group was created, which included parents of children with ASD, pediatricians, psychiatrists, neurologists, gastroenterologists, immunologists, public health professionals. Working group compared Russian pediatric practice with guidelines for pediatrician's care for children with ASD from different countries (UK, USA, Australia, Israel) and analyzed evidence-based interventions.

Results. The main problems of the primary pediatric care for children with ASD were described.

1. Pediatricians are not competent enough in early identification of ASD.

2. Pediatricians are not involved widely in assessment for comorbidities as possible etiology (metabolic, endocrine diseases, epilepsy, genetic disorders and others). Consequently, can be delayed diagnosis of rare diseases (mitochondrial disorders, etc.).

3. Pediatricians are not familiar with specialty of medical comorbidities in patients with ASD such as gastrointestinal issues, nutrition status, food selectivity, allergy, pain or discomfort in children with ASD. The high comorbidity of gastrointestinal disorders in children with ASD (up to 84%), which can affect the behavior, is confirmed by many studies.

Meanwhile, the Russian clinical guideline for ASD care does not pay attention to this. Eating disorders ("food selectivity", "perverted appetite") are often regarded as psychopathological symptoms. At the same time, proper treatment of these abnormalities reduces the intensity of mental disorder.

4. Pediatricians are not aware how to conduct proper medical exam for children with behavioral problems. There is no special service to support child with ASD and family during medical procedures (blood sampling, ultrasound etc.).

5. There are no references between pediatricians and mental services.

6. Continuing stigma of mental disorders between pediatricians.

Conclusion. It is important to increase significantly pediatrician's activity in identification, evaluation and management for children with ASD. We conduct the initiative in the development of evidence-based guidelines for pediatricians to care children with ASD and families.

Global child rights dialogue (GCRD): what children across the globe think about their right to health

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Objective. Global Child is an international project that is operationalizing the United Nations Convention on the Rights of the Child (CRC) using a comprehensive indicator framework to develop a global child rights monitoring platform.

Methods. The CRC is grounded on four guiding principles: non-discrimination; best interests of the child; the right to life, survival and development; and respect for the views of the child (child participation). Acting upon the principle of child participation as a guiding principle of the CRC, we launched the Global Child Rights Dialogue (GCRD) to seek children's opinions about their rights, with the objective to incorporate this feedback into the Global Child indicators.

Results. We recruited facilitators across the globe to host workshops with children in their local areas. Using specially developed child-friendly versions of all substantive rights of the CRC, children were asked for their opinions about certain rights and how to know whether they were being implemented or not. Children at one of the sites in Russia specifically discussed articles 23 and 24: the rights to health and health services, and the rights of children with disabilities, respectively. Workshops followed Lundy's Model of Participation, which includes the four elements of space, voice, audience and feedback. First, children were informed about the right to be discussed to assist them to form and voice their opinions within the safe space of the workshop. Facilitators (the audience) listened to and recorded children's opinions. Upon analysis of all collected data, emerging themes were incorporated into the Global Child indicators. Finally, a child-friendly report was prepared to provide feedback to the children on how their input was used.

Data collected from 1,836 children in 52 sites across the globe, confirmed that when children have a safe space, time, and a non-judgmental audience, they can share opinions and insights that are often underestimated by adults. These opportunities are conducive to improving development of the children, to teaching them active listening and to instilling values of tolerance and lifelong learning.

Conclusion. This presentation will share the collective input on articles 23 and 24 from children across the globe, as well as some of the specific outcomes from the workshops in Russia.

Clinical and epidemiological features of acute respiratory viral infections in children

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Objective. To assess clinical and epidemiological features of acute respiratory viral infections depending on the viral pathogen in children.

Methods. We retrospectively analyzed 41 patients with symptoms of acute respiratory infection from birth to 18 years. Among the observed patients, there were prevailed children aged 1–5 years (*n* = 20 children, 48.8%), 13 infants (31.7%) of the first year of life, and 8 children (19.5%) over 5 years of age. Most patients were observed with a primary diagnosis of acute nasopharyngitis — 31.7% (*n* = 13), acute bronchitis — 29.3% (*n* = 12), tracheobronchitis — 19.5% (*n* = 8), obstructive bronchitis — 19.5% (*n* = 8). Complicated forms of respiratory infection in the form of catarrhal otitis media, sinusitis were noticed in 3 hildren — 7.3%, pneumonia — in 1 child (2.4%). All patients underwent swab smear by polymerase chain reaction (PCR) to isolate viral pathogens of respiratory infections.

Results. In half of the cases (*n* = 20, 48.8%), the viral etiology was not established.

Rhinoviruses (*n* = 9; 21.9%) and respiratory syncytial viruses (RSV) (*n* = 8; 19.5%) were the most common pathogens detected. Metapneumoviruses (*n* = 4; 9.8%), adenoviruses (*n* = 2; 4.9%) and parainfluenza virus (*n* = 1; 2.4%) were significantly less common. Bocavirus and coronavirus were not detected.

Mixed infection was detected in 3 patients of the first 5 years of life (7.3%), the association was always rhinovirus in combination with MS virus or adenovirus, or parainfluenza virus. The disease was characterized by a more severe course, manifested by bronchitis, in 1 child of the first year of life — with bronchial obstruction, in 1 of the patients-complicated by acute catarrhal otitis. Metapneumovirus infection was detected in all cases in the winter period, causing damage to the lower parts of the lungs with the development of acute bronchitis, obstructive bronchitis or pneumonia.

The main pathogens of obstructive bronchitis (*n* = 5; 12.2%) and tracheobronchitis (*n* = 3; 7.3%) were of unknown etiology. Acute bronchitis was most often caused by a RSV infection (*n* = 3; 7.3%), metapneumovirus (*n* = 2; 4.9%) and rhinovirus (*n* = 2; 4.9%). In two cases acute bronchitis proceeded as a mixed infection: the association of RSV infection or rhinovirus and rhinovirus with adenovirus infection.

Winter seasonality of respiratory infections was prevailed (*n* = 19; 46.3%), children were less ill in spring (*n* = 11; 26.8%), and rarely in summer and autumn (*n* = 6; 14.6%, *n* = 5; 12.2%, respectively). In winter, viral pathogens were detected more often in patients — in 68.4% of cases (*n* = 13), including RSV virus (*n* = 6; 46.1%), rhinovirus (*n* = 4; 30.8%), metapneumovirus (n = 3; 23.1%) and mixed infection (*n* = 1; 7.7%). The spring-summer morbidity rate distinguished the group of children of the first year of life (*n* = 9; 69.2%).

There was no correlation between the type of pathogen and the age of the patients. Children over 5 years old were mostly ill with acute nasopharyngitis and tracheobronchitis (*n* = 6; 75%), whereas in patients of the first 5 years of life, acute respiratory infection often occurred in the form of lower respiratory tract lesions (*n* = 20; 60.1%). We had three infants aged 1–2 months in our study. The main disease was acute nasopharyngitis of undetected etiology (*n* = 2; 4.9%) and in one case caused by RSV infection (*n* = 1; 7.7%). One child had a symptom of acute bronchitis, the other — of obstructive bronchitis.

Conclusion. Among the established viral pathogens, rhinovirus was most often detected in children with symptoms of acute respiratory infection. The most severe course was associated with mixed infection, as well as metapneumovirus infection, mainly in children of the first 5 years of life and was accompanied by damage to the lower respiratory tract, including the addition of a bacterial infection.

Assessment of the physical development of Russian adolescents

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Objective. To study the prevalence of underweight, overweight and obesity among students in the Federal Districts at the age of 11 and 15 years.

Methods. The study includes data from 2023 schoolchildren of 11 and 15 years recruited in nine regions of the Russian Federation. The estimation of body mass index (BMI) was made according to the standard deviation score tables.

Results. In assessing the BMI in boys of 11 years in the regions of Russia, obesity was recorded in 18.6%, overweight in 15.4%, and underweight in 5.4%. Among boys of 15 years, obesity was detected in 10%, overweight in 11.5%, underweight in 8.5%. The total number of girls with overweight was 17% for the younger age group and 11% for the older. The number of girls with underweight was 20% for 11-year-olds and 26% for 15-year-olds.

Among girls of 11 and 15 years, regions from the Siberian (12.3%), Southern (10.9%) and Far Eastern (10.7%) Federal Districts were the leaders in the prevalence of obesity. Among boys aged 15 overweights ranged from 5% to 25%, the largest percentage was in boys of 11 years in the North-West Federal District, and the smallest (4.3%) in girls of 15 years in the Volga Federal District.

Conclusion. The study found that the prevalence rates of obesity are characterized by regional differences. The data obtained can be used in the development of regional programs aimed at preventing obesity in children.

Patterns of internet use by Russian schoolchildren

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

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

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

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

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

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

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

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

Bloch-Sulzberger syndrome — a rare X-linked dermatosis

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Objective. Bloch-Sulzberger syndrome is a rare genetically determined dermatosis, manifested by skin changes in combination with pathology of the eyes, skin appendages, teeth, central nervous and musculoskeletal systems. The disease manifests in the neonatal period and has four stages: vesicular, verruciform, hyperpigmented and hypopigmented. It occurs mainly in girls, since inheritance has an X-linked dominant character.

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

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

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

Sonographic picture of the chronic pancreatitis in an 8 years old child: clinical case

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Objective. Children's chronic pancreatitis is a casuistic rarity; it is an inflammatory and degenerative disease of the pancreas, which can be associated as a reaction to any pathological process or be of a hereditary nature. Chronic pancreatitis, as a rule, is asymptomatic or with unexpressed clinical signs, and can lead to acute pancreatitis.

Case report. An 8-year-old boy turned to the department with complains on intermittent cramp-like pain in the abdomen during a year, mostly after meal, on the rare liquid stool or it was absence during several days. The pain passed off on its own or was relieved with antacids. Last year at the other clinic the ultrasound examination of the abdominal cavity organs was conducted. Any pathological changes of the internal organs were not detected. Biochemical blood test was not conducted; the symptomatic therapy was prescribed by attending medical doctor.

Results. The ultrasound examination of the hepatobiliary system organs was conducted. Significant changes of the pancreas gland were detected: increases sizes, uneven counters, parenchyma was inhomogeneous due to different tissue areas of incorrect shape, of decreased and increased echoicity; multiple hyperechoic inclusions with sizes up to 1,5–8,0 mm with acoustic shadow were visualized in the tail's region. The Wirsung's duct was expanded among along its entire length up to 5,0 mm with a wavy course, calculi and clots of secretion were detected in the lumen. No mass lesions were detected. Multiple lymph nodes wit sizes up to 1 cm of normal shape with preserved architectonics were detected in the part of the pancreas head. No changes of peripancreatic mass were detected.

Moderate quantity of free liquid was visualized in the abdominal cavity.

Sonographic data correspond to the chronic pancreatitis. Conducted laboratory examinations showed the increase of alpha-amylase up to 624 u/l and lipases up to 202 u/l, that confirmed the inflammatory process in the pancreas gland, the results of computed tomography of the abdominal cavity organs with a contrast enhancement of volumetric formations and foci of abnormal uptake of contrast medium were not detected. The patient was referred to the surgeon's consultation to solve the issue of the further treatment tactics.

Conclusion. Preserved abdominal pain can suggest the chronicity of inflammatory process in the pancreas gland, the patient is recommended to conduct the ultrasound examination not rare than once in 6 months due to the clinical data.

Familial hypercholesterolemia: a rare case of early diagnosis

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Introduction. Familial hypercholesterolemia (FH) is a common life-threatening genetic condition that causes high cholesterol and leads to a much higher-than-normal risk of coronary heart disease (CHD). The heterozygous type is found in about 1 out of 300–500 people, the homozygous type is quite rare in 1 out of 1 million people.

Objective. To analyze the clinical case of family hypercholesterolemia in sibs.

Methods. In 3 siblings (from triplets) at the age of 7 years randomly detected new-onset hypercholesterolemia 5.97 mmol/L, 5.65 mmol/L, 6.43 mmol/L. In a second study after 14 days, hypercholesterolemia persists (6.11 mmol/L, 5.67 mmol/L and 6.49 mmol/L, respectively). Two siblings (identical) had high cholesterol levels (4.060 mmol/L and 4.413 mmol/L) due to low density lipoprotein (LDL). The third child with the lowest level of hypercholesterolemia had normal LDL and a high level of high-density lipoprotein (HDL). No evidence of secondary hypercholesterolemia (diabetes mellitus, chronic renal insufficiency, hypothyroidism, cholestatic hepatitis, iatrogenic illness) was found.

A mother (42 years old) sticks to a strict diet with a reduced fat content, but hypercholesterolemia persists, statins therapy is not conducted, recommendations for examining children have not been received.

Results. Plasma LDL cholesterol level of 4.0 mmol/L or higher in follow-up blood test, provided parents with hypercholesterolemia, confirms the FH in two children from triplets.

At the same time, there are no external physical signs of the disease in children (xanthomas, corneal arch, xanthelasma). This, along with relatively low hypercholesterolemia, suggests a prognostically favorable Heterozygous Familial Hypercholesterolemia. Genetic screening for the presence of FH is not required to confirm the diagnosis but may be useful if the diagnosis is ambiguous.

A strict diet with a reduced fat content was recommended to patients, as well as supervision of a cardiologist and lipid screening. It was decided that at the age of 8–10 years, while maintaining LDL cholesterol > 4.0 mmol/l in follow-up blood test would be observed even on the recommended diet, the treatment with low doses of statins would be discussed.

Conclusion. Despite the prevalence of FH and the availability of effective treatment, FH is rarely diagnosed in children. This emphasizes the importance of lipid screening in childhood and cascading screening of all members of the patient 's family for the prevention of CHD.

A clinical case of a no evidence-based medical treatment of cytomegalovirus infection in an infant

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Objective. Cytomegalovirus, which belongs to the herpes viruses' group, is the most frequent cause of congenital infection. The fetus may be silently infected in utero, as a result of initial infection or reactivation of a chronic infection in the mother. This may lead to different failures of the child's organs and systems. The presence of antibodies to cytomegalovirus (CMV) in infants, even without clinical signs of infection, often leads to unreasonable medical treatment.

Methods. The parents of the 2-month-old child appealed to the department to verify the correctness of prescribed treatment. A high level of CMV IgG was detected in maternal blood after pregnancy. That's why congenital CMV infection was suspected in 1-month-old infant. His laboratory tests revealed high level of CMV IgG, but CMV DNA wasn't found by PCR in blood, urine, saliva. Despite this, the baby was diagnosed with congenital CMV infection and was treated with anti-human anticytomegalovirus immunoglobulin (2 doses). However, antibodies titer was at the same level on repeat testing. On physical examination in our department: the condition of the child was satisfactory, cognitive development was normal.

Results. Diagnosis of congenital CMV infection isn't correct, according to negative CMV DNA PCR in blood, saliva, urine and lack of clinical manifestation (microcephaly, jaundice, petechial rash, hepatosplenomegaly, hepatitis, pneumonitis, sensorineural hearing loss, etc.). Therefore, further examination and specific immunoglobulin therapy aren't needed, dynamic observation is recommended. The child's condition remains satisfactory at the age of 4 months, there aren't any complaints from his parents.

Conclusion. The main diagnostic test of congenital CMV infection is PCR of body fluids, which means that serological research should not be used in routine diagnostics. The detection of CMV IgG in clinically healthy infants isn't a criterion for this diagnose and does not require specific treatment.

Don’t forget about risk of pertussis in children with asthma

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Objective. Pertussis, also known as “whooping cough”, is a highly contagious respiratory disease, caused by Bordetella pertussis. Nowadays it primarily affects children too young to have completed the full course of vaccinations, children with chronic lung diseases, teenagers and adults whose immunity has faded. Incidence of pertussis among children 7–14 years old in Russia increased twice in 2019, compared with 2018, possibly due to fading immunity among vaccinated children. However, there is still no pertussis booster vaccination for children over 6 years of age in the Russian National Immunization Schedule.

Case Report. A 10-year-old boy, appealed to the department with complaints of paroxysmal nonproductive cough, becoming nocturnal for the last 14 days, without fever. The boy was diagnosed with asthma at age of 7 years, for the last 1,5 years he had asthma remission and hadn’t received controller therapy. The boy was vaccinated according to the National Immunization Schedule of the Russian Federation. At the previous pediatrician examination there wasn’t any additional sounds, normal breath during the lungs’ auscultation. However, the doctor decided that it was asthma exacerbation caused by a viral disease. Boy was treated with inhaled salmeterol/fluticasone propionate combination without any significant effect: the cough became worse, hacking, nocturnal with “whooping” sounds.

On physical examination difficulty breathing through the nose was detected. On auscultation, normal breath without any additional sounds were audible over both lungs. The remainder of the physical examination was unremarkable.

Epidemiological anamnesis: The mother of the child had frequent coughing paroxysms for the last 2 weeks.

Results. Pertussis was suspected because of the long-term coughing paroxysms, lack of response to bronchial asthma controller therapy, positive epidemiological anamnesis. Bordetella pertussis DNA was detected by PCR in nasopharynx mucus. Also, positive IgM (12.6 IU/mL) and IgG (60.8 IU/mL) antibodies to B. pertussis were found in the blood serum. The boy was treated with clarithromycin. Since the beginning of treatment there was significant improvement in health: the cough became rare without whooping.

Conclusion. Children with bronchial asthma and lack of response to controller therapy ought to be suspected with whooping cough. Particular attention should be given to schoolchildren and adolescents, especially with chronic lung diseases: vaccination against pertussis at the age of 6 and 12–13 years is recommended, possibly due to fading immunity among vaccinated children over 6 years of age.

Heart and cardiovascular involvement in Russian patients with mucopolysaccharidosis: effects of enzyme replacement therapy

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Objective. Mucopolysaccharidosis is a rare lysosomal storage disorder. Clinical phenotype is very variable. Patients usually have short stature, multiplex dysostosis, facial dysmorphism, cardiovascular abnormalities and other changes.

Methods. We study cardiovascular and heart findings in patients with mucopolysaccharidosis II (MPS II) and effect after enzyme replacement therapy (ERT). 55 boys were included in our study. Cardiac problem has 49 patients (median age 85,00 ± 7,26 (3–207) month). ERT was given to 46 patients since 2008 (median age 81,54 ± 50,24 month).

Results. Before ERT, mitral regurgitation had 27 patients, thickening of the mitral valve — 17, stenosis mitral valve 1, aortic regurgitation — 20 patients, thickening of the aortic valve — 6, stenosis aortic valve — 1, tricuspid regurgitation 13 patients, pulmonary valve regurgitation — 4.

After ERT — mitral regurgitation had 35 patients, thickening of the mitral valve — 20, stenosis mitral valve — 0, aortic regurgitation — 23 patients, thickening of the aortic valve — 35, stenosis aortic valve — 0, tricuspid regurgitation — 14 patients, pulmonary valve regurgitation — 3.

There were no statistically significant deterioration or improvement of the valve heart apparatus in the examined group of patients ($P > 0,05$).

Cardiomyopathy (left ventricular hypertrophy) have 6 boys with MPS II before and after ERT, lung hypertension — 2 cases before ERT, 4 — after. The course of heart failure in MPS is progressive and we evaluated the heart condition by functional class. We have identified significantly high-performance functional class at first patient visit (I class — 44,4% patients, II class — 44,4%). After ERT (median age 81,54 ± 50,24 month) 89% patients have no negative dynamics. It should be noted that most patients began receiving ERT at age 6–8 years old, they had severe somatic and neurological symptoms.

Conclusion. ERT is not able to reverse the cardiac damage but provides stabilization of heart failure. Early initiation of ERT is a factor preventing severe heart disease in MPSII patients.

Parvovirus infection under the mask of an allergy: a case report

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Background. The prevalence of parvovirus infection is quite high, the frequency of detection of serological markers increases with age — from 2–10% in the group of children under 5 years old, to 40–60% in young and middle-aged people. Widespread maculopapular rashes on the body are the most significant and often the only clinical sign of the disease among children and are frequently misdiagnosed as allergic reaction.

Objective. To analyze a clinical case of parvovirus infection in a boy with allergy.

Case report. An 8-year-old boy appealed to the department in December on the 13th day of the disease with complaints of widespread nonpruritic lacy rash.

The boy suffered from pollinosis in spring and cross-food allergy to stone fruits every year from 2 years old. He has heredity burdened by allergy diseases.

The illness began with nonspecific prodromal symptoms, such as rhinitis, sore throat and subfebrile fever, lasted for 3 days. On day 4 erythematous rash on cheeks appeared, 2 days later non-itchy, maculopapular rash developed on the trunk and limbs. Ambulance service regarded these symptoms as a toxic-allergic reaction. The child got dexamethasone IM, chloropyramine IM without any significant effect. Then the boy was treated with oral cetirizine, but the rash spread to the upper and lower extremities. The child also followed a strict hypoallergenic diet. On physical examination the patient's condition was satisfactory. There was red rash on the face ('slapped cheek 'rash) and pink nonpruritic maculopapular lacy rash on the entire body, except the feet and palms. According to the results of the blood serum anti-B19 IgM was detected. Symptoms were relief without treatment for 5 days.

Conclusion. Maculopapular rashes, the lack of effect of antiallergic therapy necessitate advanced diagnostic. It is necessary to conduct differential diagnosis with infectious diseases despite an aggravated allergy history. Setting the correct diagnosis avoids the prescribing of unnecessary drugs and diets.

The long-term monitoring and analysis of outcomes of different approaches to the management of chronic spontaneous in adolescents

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Background. Second-generation of H1-antihistamines (H1-AH) is the main therapy for the chronic spontaneous urticaria (CSU). Omalisumab (Omab) is the only biological, approved for the severe H1-AH resistant CSU in adolescents over 12 years.

Objective. To evaluate different approaches to severe CSU therapy in adolescents and assess achieving of disease control in 3 y period.

Methods. The long-term prospective observation study of 34 children with severe CSU (55% boys, average age 13 yo (min 3; max 17,0), the duration of disease — 33 mo (min 3; max 144); UAS7 — 18 points (min 16; max 24,0) was conducted. All patients received H1-AH for minimum 3 mo.

Patients were randomized in 2 groups. 17 patients of the 1st group were added with Omab to therapy: 55,6% girls, average age — 15 y (min 12,0; max 17,0); disease duration was 45,2 mo (min 3,0; max 144,0), the average total IgE level — 348,2 IU/mL (min 0,8; max 2041,0); the average UAS7 at debut — 17,2 points (min16; max 24). The course of Omab therapy was 6 mo, 300 mg/mo subcutaneously.

17 patients of the 2 nd group maintained alone H1-AH therapy: 64,7% boys, average age — 10,8 y (min 3,0; max 15,0); disease duration was 20,5 (min 3; max 72) mo, the average total IgE level — 182 IU/mL (min 20; max 1050); UAS7 at debut — 18 (min 16; max 28) points.

The efficacy of therapy assessed by urticaria activity score for the 7 days (UAS7).

Results. In the 1st group of patients in 6 mo of Omab therapy UAS7 was 1,6 (min 0; max 20) points, $p < 0,05$. After 3 y of the course Omab therapy UAS7 was 4,5 points, $p < 0,05$.

In the 2 nd group of patients, who received alone H1-AH, in 6 mo UAS7 remained at the same level — 18 points ($p < 0,05$). The average UAS7 in 3 y was 12 (min 0; max 26) points ($p < 0,05$).

Thus, in patients receiving Omab UAS7 significantly decreased after 6 mo.

The UAS7 level in the Omab group indicates a greater proportion of children who have achieved disease control. The proportion of children, who have achieved remission during 3 y (UAS7 = 0): in Omab group 52,9%, in H1-AH — 29,4% ($p = 0,163$).

Conclusion. Our results indicate the efficacy of Omab in adolescents with CSU: rapid relief of urticaria symptoms and a greater proportion of adolescents who have achieved disease control, compared with therapy alone H1-AH.

Seroprevalence of Cytomegalovirus among school-aged children in Russian Federation

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Background. Cytomegalovirus (CMV) is an important human pathogen in case of immature or compromised immune system, such as the unborn child have. We thus aimed to examine risk factors for CMV infection in young people in Russia and, in order to improve our understanding of CMV epidemiology and guide future disorder prophylaxis strategies.

Objectives. To explore cytomegalovirus (CMV) seroprevalence among school-aged children in different age groups.

Methods. We conducted retrospective evaluation of the seroprevalence of CMV IgG antibodies among immunocompetent school-aged children (n = 1315), age group from 10 to 15 years, from different regions in Russia (n = 7). Children were divided into 2 groups; in the first group was children under 13 years old, in the second group — over 13 years old. Comparison of two independent groups was determined using the Mann-Whitney test and the Kruskal-Wallis test. We analyzed the prevalence of CMV serotype and risk factors for infection.

Results. We estimated a total CMV seroprevalence of 74.6% (n = 981). The median of age in the 1st group was 10.9 (10.6; 11.3), the median of age in the 2nd group was 14.9 (14.6; 15.1). CMV seroprevalence was strongly associated with age, increasing from 71,8% in the first group, throughout adolescence (77.2% in the second group) p = 0,048. There were no statistically significant gender differences between regions.

Conclusion. The results are consistent with global data and require further study. These estimates of the CMV distribution will help develop national and regional models and algorithms for disorder prophylaxis in target populations.

ENT conditions and disorders in children with Hunter syndrome

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Objective. To describe the features of the ENT conditions and disorders in children with Hunter syndrome. Hunter syndrome, also known as mucopolysaccharidosis type II (MPS II), is a rare X-linked recessive disease. The MPS II is a hereditary metabolic disorder caused by accumulation of glycosaminoglycans (GAG) in organs and tissues due to mutations in the genes, which encode intralysosomal hydrolysis of macromolecules. Hunter syndrome is a progressive, multisystem disease. At the same time a child may have mental retardation and speech development delay, skeletal bone deformities, loss of vision, hearing loss.

Methods. The study included 17 boys aged from 2 to 12 years old with genetically confirmed Hunter syndrome. All patients underwent examination by otolaryngologist, also tympanometry, diagnostic nasopharyngolaryngoscopy, registration of otoacoustic emission, audiometry, cardiorespiratory monitoring were performed.

Results. The accumulation of GAG leads to a gradual narrowing of the nasopharynx and larynx lumen, a thickening of the tongue and vocal folds, laryngeal cartilages deformation, an enlarged of pharyngeal, palatine and lingual tonsils.

11 children (64.7%) had chronic inflammation of the nasal mucosa and pharyngeal tonsil with frequent exacerbations. Pharyngeal tonsil hypertrophy was detected in 10 patients (58.8%).

14 children (82%) had a wide, thickened tongue. Hypertrophy of the tonsils was diagnosed in 8 children (47%).

The voice had changed (hoarseness) in 16 children (94%). Deformation of the epiglottis was detected in 4 people (23.5%), tracheomalacia — in 1 child (5.8%).

Among the complications acute sinusitis and exudative otitis media were detectable in 52.9% of patients. 10 children (58.8%) had obstructive sleep apnea (OSA) syndrome, two of them (11.7%) had a severe degree of the disease, which was an indication for adenotonsillectomy. Hearing loss was found in 10 boys (58.8%).

In most patients (82%) a combined pathology of the ear, throat and nose was detected, 2 children (11.7%) had only adenoid hypertrophy, and only 1 child (5.8%) did not have any pathology of ENT organs.

Conclusion. Functional disorders and diseases of the ear, throat and nose are found in most children with Hunter syndrome, which could be one of its early manifestations. An ENT specialist may suspect MPS type II according to the presence of an ENT pathology that is difficult to treat in a standard manner, combined with the pathology of other organs and systems.

Early diagnosis and the possibility of enzyme replacement therapy can control the disease progression and avoid early disability.

Analysis of the prevalence of allergic diseases in adolescents in the Russian federation

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Objective. To assess prevalence of allergic diseases among schoolchildren in the Russian Federation (RF) in the age groups of 11 and 15 years.

Methods. The data obtained during the screening of schoolchildren (which included surveys of parents and school doctors with specially designed questionnaires, an allergist's examination, spirometry) — the project of the RF and the WHO European Office for the development of the school medicine network. 2114 schoolchildren were examined in the following federal districts: Central Federal District — CFD, Southern Federal District — SFD, North-Western Federal District — NWFD, Volga Federal District — VFD, Ural Federal District — UFD, Siberian Federal District — SibFD, Far Eastern Federal District — FEFD.

Results. Analysis of the results suggests that the smallest number of children with manifestations of allergic diseases was noted in the SibFD *n* = 81 (24%), and the largest in the UFD *n* = 107 (36%); by districts: in the CFD *n* = 83 (31%), NWFD *n* = 87 (32%), FEFD *n* = 91 (30%), SFD *n* = 100 (30%), VFD *n* = 102 (32%).

At the same time, the average of bronchial asthma (BA) was 2%, seasonal allergic rhinitis (SAR) — 8%, allergic rhinitis (AR) — 8%, atopic dermatitis — 2%, food allergy (FA) — 13%, drug allergy (DA) — 3%. Regional peculiarities were revealed: the prevalence of BA varied from 1–1.5% in the NWFD, VFD, FEFD, SFD to 6% in the UFD. The prevalence of seasonal AR ranged from 2% in the NWFD and 3% in the FEFD to 12% in the UFD and 16% in the North Caucasus Federal District. Perennial AR accounted for from 3% in the CFD to 11% in the UFO and 16% of the FEFD. The highest incidence of food allergy was registered in the Volga Federal District and CFD, 17% and 16%, respectively, and only 10% in the SFD and in the SibFD. AD was diagnosed in equal shares in the CFD and NWFD (3%), in the UFD — 2%, in the SFD, VFD, SFD and FEFD — 1%.

Conclusion. The true prevalence of allergic diseases among schoolchildren of the Russian Federation is significantly higher than data of official statistics but differs by regional peculiarities. The true prevalence has appeared to be in 2–20 times above the data of official statistics: data of official statistics for BA in 2015 amounted to 2,7%, for AR — 0,9%.

Vitamin D supply for children and adolescents with cancer in the South of Russia

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Objective. To assess the supply of vitamin D in children and adolescents with cancer in the Stavropol Territory (45° north latitude).

Methods. We examined 36 children with cancer (18 boys and 18 girls). There were 11 children (30.6%) aged from 0 to 3 years, 16 (44.4%) — from 4 to 10 years, 9 (25.0%) children from 11 to 18 years. The average age is 7.9 ± 0.9 years.

Results. Leukemia was revealed in 19 (52.8%) children, solid tumors — in 9 (25.0%), lymphomas — in 8 (22.2%) children.

The median calcidiol level was 17.5 [12.6–23.4] ng/ml. A deficiency of vitamin D (less than 20 ng/ml) was detected in 21 (58.3%) children, a deficiency at the level of 20–30 ng/ml — in 10 (27.7%), a level of more than 30 ng/ml — in 5 (13, 9%) of examined children.

Median 25 (OH) D in children with leukemia was 21.2 [14.6–22.4] ng/ml, with solid tumors — 16.2 [9.3–16.4] ng/ml, with lymphomas — 15.8 [11.7–22.8] ng/ml. Nine (47.4%) children with leukemia, 7 (77.8%) with solid tumors and 5 (62.5%) with lymphomas had a serum calcidiol level of less than 20 ng/ml, deficiency of 20–30 ng/ml was detected in 6 (31.6%), 1 (11.1%) and 3 (37.5%) children, respectively, the level of more than 30 ng/ml was detected in 4 (21.0%) children with leukemia and 1 (11.1%) of a patient with a solid tumor.

The median vitamin D supply in children from 0 to 3 years old was 16.6 [9.7–21.7] ng/ml, from 4 to 10 years old — 19.8 [14.7–24.8] ng/ml, from 11 to 18 years old — 16.8 [10.5–17.2] ng/ml. Vitamin D deficiency (less than 20 ng/ml) was detected in 6 (54.6%) children under 4 years old, in 8 (50.0%) children from 4–10 years old and 7 (77.8%) adolescents of 11 years old and older ($p < 0.05$); insufficiency with the level from 20 to 30 ng/ml was detected in 3 (27.3%), 6 (37.5%) and 1 (11.1%) children, and the level of more than 30 ng/ml was found in 2 (18.1%), 2 (12.5%) and 1 (11.1%) children, respectively.

Conclusion. In the south of Russia, the majority (86.0%) of children and adolescents with cancer have a 25 (OH) D level of less than 30 ng/ml. Level of 25 (OH) D did not significantly depend on the type of cancer. Adolescents with cancer are at risk for vitamin D deficiency (less than 20 ng/ml).

Clinical and preventive efficacy of phytopreparations in pre-school children with adenotonsillar pathology

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Objective. Assessing efficacy of Tonsilgon N in pre-school children as an etiotropic drug in complex therapy of adenotonsillar pathology at the rehabilitation stage.

Methods. Within the period from 2011 to 2015, 1,076 children (aged from 2 to 5 years) with a history of pathology of nasopharyngeal and palatine tonsils in 100% of cases were reviewed. Pre-school children were given Tonsilgon N phytopreparation in the form of drops in age-specific doses within 30 days.

The control group included 200 children. The phytopreparation efficacy was analyzed before and after the preventive therapy.

Results. A year prior to the preventive therapy, degree II nasopharyngeal tonsil hypertrophy complicated by adenoiditis was noted in 71.3% of children in the treatment group and in 74.5% of children in the control group. Degree III nasopharyngeal tonsil hypertrophy made 28.7% and 25.5%, respectively. A year after the rehabilitation therapy, only 32.9% of patients in the treatment group did not show improvement in the clinical presentation. Symptoms of adenoiditis were practically relieved in half of the patients, nasal breathing was restored in 95.8% of patients, the size of a nasopharyngeal tonsil was reduced from degree II to degree I in 64.9% of patients.

Normalization of the rhinoscopy presentation was noted in 82% of patients, 77.9% of patients showed reduction of the ARI rate from 5–8 to 2–3 times a year. Endoscopic control confirmed reduction of size of both nasopharyngeal and palatine tonsils in 64.9% of patients. The hypertrophy degree of nasopharyngeal and palatine tonsils in pre-school children not receiving Tonsilgon N phytopreparation did not show any change. In addition, hypertrophy demonstrated increase from 74.5% to 83% over time.

Conclusion. Prospective study of children receiving Tonsilgon N as a rehabilitation method demonstrated its efficacy on immune development of the respiratory tract. The obtained data allow us to recommend Tonsilgon N to be included in programs of immune rehabilitation and prevention of exacerbation of adenotonsillar pathology.

The usage of lean (simulation) technologies' tools to evaluate nutrition status and physical development in pediatrics

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Objective. To work out the skill of assessing physical development and nutritional status of children and adolescents in various ways, to carry out comparative analysis of methods.

Methods. Questionnaire data of patients examined in the office of a doctor — a nutritionist with various disorders of nutritive status — 101 people.

Anthropometry was performed, physical development was evaluated by the WHO Anthro program, using centile tables, and the impedance method (body composition). Statistical analysis, which included parametric statistics methods as well as r-Spearman rank correlation, was performed using Statistics 23.1 software.

Results. Physical development of children is a marker of adequacy of nutrition, and affects adaptation, severity of pathological processes, rate of recovery, effectiveness of therapy, terms of hospitalization. Not enough teaching time is devoted to the study and training of nutrition status assessment skills in various ways.

The skill of evaluation of physical development (FR) and nuthritical status (NS) was worked out on the questionnaire data of specific patients. The results of the programme WHO Anthro assessment, the centile tables and the impedance data were compared in 101 patients with eating disorders (54 boys), from 2 m.o to 17 y.o (average 8.5 y.o) Malnutrition 30%, severe malnutrition — 6% cases. Obesity revealed 39% kids. The results of the WHO Anthro assessment, the percentile tables and the impedance data were compared. Direct correlation links between the child 's percentile weight corridor, the WHO Anthro (z-score), and the percentage of deficiency or excess of fat and active cell mass are obtained ($p = 0.05$). The child 's percentile growth corridor and body mass index also correlate with the growth z-score, fat, and active cell mass percent according to body impedance ($p = 0.05$).

Conclusion. The skill of assessing physical development and nuthritical status by students can be formed using simulation technologies. Physical development, as well as the nuthritical status of the child, can be investigated by the WHO Anthro program using a computer application, or by using technologically complex equipment — impedance, or manually — with the help of centile tables. Deviations of nutritive status from physiological norm are determined by all proposed methods.

Socio-demographic and family determinants of school bullying among young adolescents

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Objective. To determine socio-demographic and family determinants of school bullying among young adolescents.

Methods. Data for this study were collected during the 2017/2018 Health Behavior in School-aged Children (HBSC) survey on one of the Russian sites. The sample included 307 urban schoolchildren aged 11 years. We selected the questionnaires with the responses about school bullying. Demographic characteristics of families, outcomes and relationships in the families were analyzed as variables of interest.

Results. 293 questionnaires were selected (148 boys). All adolescents were divided into 3 groups depending on their replies about school bullying: group 1 (n = 167) included adolescents, who had never faced bullying at school; group 2 (n = 90) included adolescents, who had faced bullying at school 1–2 times during last 2 month; group 3 (n = 36) had faced bullying several times per month and oftener.

It was shown that 10 (6.1%) adolescents of the 1st group, 14 (15.9%) adolescents of the 2d group and 7 (19.4%) adolescents of the 3d group did not hope for parents' help in solving school problems ($p1–2$ = 0.023; $p1–3$ = 0.025). Twenty-one (13.2%) people in the 1 group, 21 (23.9%) in the 2d group and 10 (27.7%) in the 3d group were not sure their parents were willing to go to school to talk to teachers ($p1–2$ = 0.033; $p1–3$ = 0.057); 10 (6.2%) schoolers of the 1st group, 16 (18.4%) of the 2d group and 7 (19.4%) of the 3d group believed that their parents were scarcely involved or not at all involved in their school life ($p1–2$ = 0.006; $p1–3$ = 0.025). Adolescents, who had suffered from bullying, relied on parents' help in decision-making to a lesser extent ($p1–2$ = 0.031; $p1–3$ = 0.052).

Eighty (90.9%) adolescents from the 2d group and 28 (80%) from the 3d group thought they looked "normal" and "good" in comparison with 162 (97.6%) adolescents, who had never experienced school bullying ($p1–2$ = 0.038; $p1–3$ < 0.001). Moreover adolescents, who had suffered from bullying, had lower life satisfaction ($p1–2$ < 0.001; $p1–3$ = 0.002).

The family composition, material well-being, occupation (employed/unemployed) did not show significant connections with school bullying of adolescents.

Conclusion. Young adolescents, suffering from school bullying, do not feel they get adequate support from their parents. As a result, they have low self-esteem and low life satisfaction.

Therefore, work of a teacher and a psychologist with adolescents, suffering from school bullying, should be aimed at not only normalizing the situation at school, but also at determination and correction of imbalanced relationships in the family.

Modern view of a patient with bronchopulmonary dysplasia

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Objective. To analyze the new model of a patient with bronchopulmonary dysplasia (BPD) based on the analysis of 70 case histories of infants hospitalized at the National Medical Research Center for Children's Health Federal state autonomous institution of the Russian Federation Ministry of Health from 2020 to 2021.

Methods. Collection and analysis of anamnestic data, as well as clinical and laboratory examination of 70 infants with BPD. The analysis of the results was carried out using STATISTICA 10.0 program.

Results. In total, all studied infants (*n* = 70) were diagnosed with a new form of bronchopulmonary dysplasia. Among them there were 35 boys (50%) and 35 girls (50%). The mean gestational age was 26,4 weeks [SD, 1.9 weeks]. All premature infants were born with extremely low and very low birth weight (85,7% and 14,3%, respectively). The median birth weight was 745 g (Interquartile range (IQR): 650–920). The median Apgar score at the 1st minute was 5 (IQR: 4–5), at the 5th minute — 6 (IQR: 5–6). 67 infants were intubated after birth, the median of duration of mechanical ventilation was 21 days (IQR: 6–36); CPAP treatment was initiated in 33 children, the median was 8 days (IQR: 5–13), BIPAP — 39 patients, nasal cannulas — 47 infants, nasal mask — 14, incubator — 19. Median duration of oxygen dependence was 54 days (IQR: 45–70]. The period of oxygen dependence had an inverse relationship with anthropometric data and gestational age. All patients received surfactant therapy, postnatal corticosteroids — 32 infants (46%). In addition, pneumonia was observed in 35 children (50%), intraventricular hemorrhage in 60 patients (86%), necrotizing enterocolitis in 34 children (49%). Pulmonary hypertension, as one of the complications of BPD, occurred only in 7 patients (10%).

Conclusion. The current model of a patient with a new form of BPD is a premature infant with extremely low or very low body weight, born at a gestational age of no more than 28 weeks. The morphological and functional immaturity of these children, combined with the impact of new resuscitation technologies on their respiratory system, led to the pathomorphosis of BPD and the predominance of a new form in the population. The study of the clinical features and long-term outcomes of BPD is an urgent problem of pediatric pulmonology and requires close attention in the future.

Genetic predictors of a new form of bronchopulmonary dysplasia

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Objective. Bronchopulmonary dysplasia (BPD) is a multifactorial disease with a significant genetic component. Novel genes and associated pathways may play an important role in susceptibility for the development of bronchopulmonary dysplasia in preterm infants. Our aim was to identify rare genetic variants contributing to the new form of BPD phenotype by full exome sequencing.

Methods. Full exome sequencing was performed on 39 DNA samples from patients with moderate and severe new BPD and 30 DNA samples from control group without clinical signs of BPD. After mapping and annotation, each sample showed an average of 40,000 genetic variants with a reading depth of at least 70x.

Results. All autosomal variants were filtered for allelic frequency < 1% according to the gnomAD database (version 2.1). Among them, 821 variants were found the most common (≥10%) in both the control and experimental group. Wherein 280 variants were presented with an alternative allele frequency of more than 10% in the experimental group but were found with a lower frequency in the control. Interestingly, 10 of these 280 variants were in the *ZNF717* gene. This gene encodes a Kruppel-associated box (KRAB) zinc-finger protein, which belongs to a large group of transcriptional regulators in mammals and play important roles in various cellular functions, including cell proliferation, differentiation and apoptosis. Extended bioinformatics analysis showed that 34 unique variants were found in the experimental group and were absent in the control group, which may indicate both the characteristics of Russian children with BPD and the insufficient representativeness of the control group and, in turn, requires a more in-depth analysis.

Conclusion. For the first time in Russia, large-scale studies have been carried out to identify the molecular genetic characteristics of Russian children with BPD using full exome sequencing. Our study indicates *ZNF717* gene may be relevant in BPD pathogenesis, but further research is required. These preliminary results may contribute to improving knowledge of the pathogenesis of bronchopulmonary dysplasia and targeting therapeutic interventions.

Dyslipidemia as an atherogenic factor in patients with different forms of juvenile arthritis

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Objective. Close connection between autoimmune inflammation and proatherogenic lipid changes in rheumatoid arthritis has been well established, while for juvenile idiopathic arthritis (JIA) is under discussion. The aim of our work was to study the incidence and intensity of lipid disturbances in patients with different forms of JIA.

Methods. 90 children with JIA 6–18 years were examined using clinical, biochemical methods, ultrasonic duplex scanning of vessels, thin-layer chromatography, bioimpedance measurement. 49 children without chronic diseases made up the control group.

Results. Dyslipidemia was revealed in 48.9% of patients, mainly with systemic and polyarthritis Most often an increase in atherogenic coefficient (AC) was noted. Average AC value in JIA patients was higher than in control group (2.9 + 0.2 versus 2.0 + 0.1, $p < 0.05$) and correlated with the disease activity index according to JADAS71 ($r = 0.78$), a doctor's global assessment of the disease severity according to VAS ($r = 0.61$), the degree of joints dysfunction ($r = 0.55$), C-reactive protein ($r = 0.53$) and ESR ($r = 0.68$) level. An increase in the concentration of total cholesterol was observed in 28 (31.1%) children with JIA, commonly with a long-lasting disease, and in case of CS intake. In some JIA patients atherogenic changes were detected due to apolipoprotein A1 (ApoA1) deficiency (26.7%). Patients with JIA (maximum with polyarthritis) showed a decrease in the content of the main fractions of polar phospholipids and an increase in the phospholipid lysoforms and free fatty acids by 8.9% 21.4% and 9% compared with the control group. This leads to an increase erythrocyte membranes microviscosity. Dyslipidemia was combined with body composition changes. In this patients IMCT was higher than in control group (0.73 + 0.03 versus 0.45 + 0.02, $p < 0.05$).

Conclusion. In high disease activity both systemic onset and polyarticular JIA proatherogenic lipid disturbances and vascular disorders took place.

The prevalence of functional diseases of the gastrointestinal tract in the first year of life children living in the Belgorod region (Russian Federation)

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Goal. To study the prevalence of functional disorders of the gastrointestinal tract in children of the first year of life living in the Belgorod region, according to the questionnaire data.

Methods. We conducted a survey by questioning 348 women with children aged 1 year to 4 years. All children were born full-term. The ratio of boys to girls was 1: 1 (47.7%/166 boys and 52.3%/ 182 girls).

Results. According to the questionnaire, gastroenterological complaints were found in 82.8%/ 288 children. Most often among functional disorders were detected regurgitation syndrome (18.3%/64), infantile colic (74,4%/259) and constipation (33%/115). In 25.9% cases children had two or more functional disorders, more commonly infantile colic plus constipation.

Among the factors predisposing to the development of functional disorders of the gastrointestinal tract, the following were identified: aggravated pregnancy (31.3%) and childbirth (operative delivery — 25.3%), features of the feeding (early termination of breastfeeding up to 3 months of life in 29.8%), antibiotic therapy in the first months of life — 23.6%.

For medical attention/treatment applied 56.25% parents of children with regurgitation, 67,2% parents of children with infantile colic and 57,4% with constipation.

The use of diet in treatment of functional disorders was most effective in patients with regurgitation syndrome (57%) and constipation (41.7%), while in the case of intestinal colic — only in 31.7% of cases.

Conclusion. Based on a questionnaire survey of 384 parents of children from 1 year to 4 years, living in the Belgorod region, we found a high frequency of detecting of gastroenterological complaints — in 82.8% of cases. Among the functional diseases of the gastrointestinal tract, the most common were regurgitation syndrome (18.3%), infantile colic (74.4%), and functional constipation (33%). Most often sought medical help parents of children with infantile colic. The diet was effective in most cases of functional gastrointestinal disorders except in cases of infantile colic.

Lipid metabolism parameters and levels of antioxidants in Mongoloid girls with obesity

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Objective. To analyze the state of lipid metabolism and antioxidant status in Mongoloid girls with obesity.

Methods. Studies were conducted in 22 girls (mean age — 15.06 ± 1.53 years) with the first degree exogenously constitutional obesity of and in 48 girls of control group (mean age — 14.25 ± 2.42 years). All girls by ethnicity were Mongoloids. Lipid components (total cholesterol, triglycerides, high-density lipoproteins, low-density lipoproteins) and components of antioxidant defense (total antioxidant activity, α-tocopherol, retinol, reduced and oxidized glutathione, superoxide dismutase activity, glutathione peroxidase activity, glutathione reductase activity and glutathione S-transferase activity) in the blood were determinated. Spectrophotometric and fluorometric methods were used.

Results. Mongoloid girls with obesity had higher values of total cholesterol (1.22 times higher, $p = 0.017$), triglycerides (2.16 times higher, $p < 0.0001$) and lower values of HDL (1.26 times, $p = 0.0018$), compared to the control. In the antioxidant defense system, a decrease in α-tocopherol (1.41 times, $p = 0.0262$), retinol (1.12 times, $p = 0.0306$), superoxide dismutase activity (1.28 times, $p = 0.0004$) and glutathione S-transferase activity (1.71 times, $p < 0.0001$) were noted in comparison with the control group with the absence of statistically significant changes in other components.

Conclusion. The study revealed changes in lipid metabolism and antioxidant defense parameters in Mongoloid girls with exogenously constitutional obesity, consisting of the presence of dyslipidemia, a decrease in fat-soluble vitamins and antioxidant-enzymes activity. Based on results obtained corrective measures recommended for Mongoloid girls with exogenously constitutional obesity to stabilize the lipid metabolism and antioxidant status by increasing the content of products containing polyunsaturated fatty acids in the diet and administration of antioxidants complex.

Determinants of energy under-reporting in rural adolescents

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Objective. To assess the extent of under-reporting (UR) in rural adolescents and investigate associated covariates.

Methods. A total of 150 adolescents aged 11–17 years were included. Food intake was reported in a 3–d diet record. Socio-economic status, sedentary behavior and physical activity were collected by questionnaires. Weight height and waist circumference were measured. A body mass index (BMI) was calculated.

Plausibility cut-offs for reported energy intake as a percentage of predicted energy requirements were used to identify under-reporters.

Multivariate logistic regressions investigated the associations between UR and covariates.

Results. The percentages of under- and over-reporters of energy intake were 35.3% and 3.3% respectively. Energy intake was under-reported more in older adolescents and boys. In multivariate analysis UR was associated with BMI and waist circumference Z-scores (OR 1,7 [95% CI 1,1–2,4] and OR 1,5 [95% CI 1,1–2,2] respectively) and participation in sports groups (OR 2,5 [95% CI 1,1–5,4]).

Conclusion. In conclusion, in rural sample under-reporters differ from plausible reporters in several characteristics related to age, sex, weight status and organized physical activity. Therefore, it is important to consider this differential UR bias when investigating diet-disease associations in adolescents.

Cluster breastfeeding syndrome in infants

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Objective. Cluster breastfeeding syndrome ("cluster feedings", "bunch feedings") is when baby has several feedings close together during a certain period, in some cases during the day. The purpose of this study is to examine the frequency and to identify the reasons for the cluster breastfeeding syndrome in lactating women.

Methods. Questioning was conducted with 223 lactating women aged 19–44 years.

Questioning including questions regarding the use of the delivery methods and the lactation period. Psychological testing of lactating women was also conducted. The anxiety level was evaluated by Hamilton Anxiety Rating Scale (HAM-A) (score of 14 points is borderline). Lactating women were divided into two groups: without Cluster Breastfeeding syndrome (group I) and with Cluster Breastfeeding syndrome (control group). Statistical analysis was performed using Microsoft Excel 2007, SPSS Statistics v 24.0.0.0. Spearman's correlation coefficient (r) and Pearson's correlation coefficient (*rxy*) were calculated. Data was compared using chi-square test and $p \leq 0.05$ was regarded as statistically significant.

Results. Among the women surveyed, women with one child prevailed (63%). The average duration of the lactation period was 11.1 ± 5 months. The average time the baby was at the breast was 19 ± 4.1 minutes. Syndrome of prolonged, continuous feeding was noted in 5% of cases when the baby was at the chest continuously for a day, with short breaks at night sleep. In this group, in women (90%), labor was performed by Caesarean section. In the group I (cluster breastfeeding syndrome group), the average age of women was 35.8 ± 5.5 years, in the control group 25.0 ± 4.6 ($p < 0.001$). In group I, the average score on the Hamilton scale was 28.4 ± 6.5 (level of symptomatic anxiety), in the control group — 12.9 ± 9.7 ($p = 0.0003$). All baby in the group I have been gaining enough weight and producing sufficient dirty and wet diapers. Correlation analysis revealed a direct strong correlation between the presence of cluster breastfeeding syndrome in a child born by Caesarean section ($rxy = 0.97$) from mothers who gave birth over the age of 35 and have a level of symptomatic anxiety ($r = 1$).

Conclusion. Cluster breastfeeding syndrome (cluster feedings, bunch feedings) occurs in 5% of cases and is associated with the late birth of the first child, high anxiety of the mother and the birth of a child by Caesarean section.

Screening for antibodies, associated with autoimmune liver diseases in children with celiac disease

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Objective. To determine the prevalence of autoimmune liver diseases in children with CD.

Methods. We observed 45 children aged 3 to 16 years. CD in all patients was diagnosed according to the ESPHGAN criteria. Serological examination with the determination of antibodies to tissue transglutaminase (anti-tTG IgG, IgA); histological examination of the duodenal mucosa; genetic typing for HLA-DQ2/DQ8 were carried out. Duodenal histology having Marsh grade III features were eligible for the study. Antibodies to hepatic antigen cell nuclei, skeletal muscle, cell nuclei, mitochondria, smooth muscles of IgG class were determined by indirect immunofluorescence (nRIF) using reagent kits. The antigen were biochips of primate's liver, primate's musculus iliopsoas, human Hep2 epithelial cells, liver, kidney, and stomach of rats.

Normal titer < 1:80. Anti-Parietal Cell Antibody (PCA), IgG was determined by nRIF using biochips primate's stomach as antigen. This kit of reagents detects antibodies for the diagnosis of such diseases as autoimmune hepatitis (AIH) 1, 2 and 3 types, primary biliary cirrhosis, primary sclerosing cholangitis (PSC), overlap syndrome (combination of AIH and PSC), autoimmune gastritis.

Results. We didn't obtain elevated levels of antibodies associated with autoimmune liver disease in all children with CD. In 1 person we observed elevated levels of anti-parietal cell antibodies. It was a 15-year-old girl with a typical form of celiac disease, additionally suffering from primary oligomenorrhea, autoimmune diabetes mellitus (type 1). Further examination revealed non- Helicobacter pylori atrophic gastritis. Thus, autoimmune gastritis was diagnosed.

Conclusion. Antibodies, associated with autoimmune liver diseases were uncommon in children with CD. Probably, due to insufficient number of participants of the study. On the other hand, anti-parietal cell antibodies have been found.

Imbalance of IL-10 and IL-13 umbilical cord blood in children born to mothers with asthma

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Introduction. Genetic predisposition to atopy persons responding to allergens by the rapid proliferation of Th2 lymphocytes, which secrete cytokines that accelerate the synthesis of IgE antibodies.

Objective. Purpose of the research is to study cytokines rate in umbilical cord blood of children born to mothers suffering severe bronchial asthma (BA).

Methods. Umbilical cord blood samples were taken from 22 full-term babies born to mothers with BA. The comparison group consisted of

66 children born to mothers without allergies. The ratio of boys and girls in the studied groups was the same. The average age of mothers in the groups was the same (31.26 ± 2.32 years and 32.87 ± 2.03 years; $p > 0.05$).

Cytokine's rate (TNF-α, TGF-β1, IL-18, IL-13, IL-10 and IFN-γ) was quantified by IFA. Statistical processing of data was performed on a personal computer using licensed computer software "Microsoft Excel 2016" and "STATISTICA 12". The student t-test value was determined while analyzing the distribution of quantitative data. The criterion of statistical significance level was $p < 0.05$.

Results. A significant rate decrease of IL-13 was found (6.67 ± 1.6 pg/ml versus 2.0 ± 0.23 pg/ml in the comparison group, respectively, $p < 0.05$) in umbilical cord blood of children born to mothers suffering from BA, which is associated with respiratory allergy. IL-13 rate decrease can be judged as a factor of the long-term intrauterine persistence of the inflammatory process in a child.

In addition, rate increase of IL-10 was found (66.69 ± 14.5 pg/ml versus 25.52 ± 3.1 pg/ml in the comparison group, respectively, $p < 0.05$), which is synthesized by T-and B-lymphocytes, monocytes and macrophages, reduces the activity of the Th-1 immune response more than Th-2.

Conclusion. There is an imbalance of the main anti-inflammatory cytokines: a decrease in IL-13 and an increase in IL-10 in umbilical cord blood of children born to mothers with BA. That can reflect the systemic reaction of body to local damage to organs and can be served as one of the indicators of the intensity and duration of the inflammatory process, as well as disease progression.

Inadequate sleep habits ARE associated with obesity in high school children

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Objective. To examine sleep habits and associations between inadequate sleep habits and measures of obesity in high school adolescents.

Methods. A total of 127 adolescents (61% male) aged 15–17 years were recruited from patients of the Children's hospital referred to due to obesity and other conditions in 2017–2019 years. The sample was evaluated to determine if a relationship existed between inadequate sleep habits and measures of obesity (percentile weight-for-length — body mass index — BMI, kg/m2; neck circumference — NC, cm; waist circumference — WC, cm).

Participants provided demographic information and completed the Adolescent Sleep Habits Survey (ASHS). Adolescent weight/length, NC and WC were measured; obesity defined as BMI ≥ 95th percentile, NC ≥ 90th percentile and WC ≥ 90th percentile for age and sex. Measures of inadequate sleep habits: possible activities in bed and possible activities if difficulty falling asleep in the last two weeks. All associations were analyzed using a method of odds ratios (ORs) with 95% confidence intervals [CIs]. All differences were considered significant at $p < 0.05$.

Results. A greater proportion of males than females (52.6% versus 43.7%, $p < 0.05$) and of obese adolescents compared with lean peers (64.2% versus 39.1%, $p < 0.05$) reported inadequate sleep habits. Sleep habits named «ASHS activity in bed» such as "Eat every night" was associated with BMI ≥ 95th percentile (OR 1.30, 95% CI 1.25–1.35), NC ≥ 90th percentile (OR 1.65, 95% CI 1.59–1.71) and WC ≥ 90th percentile (OR 1.72, 95% CI 1.65–1.88). Possible activities if difficulty falling asleep such as "Wake up and to do something (e.g., eat; drink warm milk/coffee) every night" were also associated with measures of obesity: BMI ≥ 95th percentile (OR 1.35, 95% CI 1.29–1.41 and OR 1.21, 95% CI 1.17–1.25, respectively), NC ≥ 90th percentile (OR 2.10, 95% CI 1.8–2.65 and OR 1.75, 95% CI 1.59–1.91, respectively) and WC ≥ 90th percentile (OR 2.5, 95% CI 1.89–3.1 and OR 2.2, 95% CI 1.28–2.99, respectively). Inadequate sleep habit such as "Watch TV in bed every night" was found to be associated with measures of obesity after adjusting for several covariates. No statistical significance was found if the above measures of inadequate sleep habits were noted as "twice or once in the last two weeks" in relation to BMI, NC or WC.

Conclusion. In addition to existing literature, this study found that inadequate sleep habits every night as well as short sleep duration in late adolescence may relate to the development of obesity as measured by BMI, NC, and WC.

Obesity in children in the regions of Russia

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Introduction. The upward steady trend in the prevalence of obesity in the pediatric population during the last decades is a significant medical and social problem.

Objective. Purpose of our research was to study the prevalence of obesity among schoolchildren living in various regions of Russia.

Methods. Totally 5701 schoolchildren (2668 boys and 3033 girls) had been enrolled in the cross-sectional study with the use of random sampling techniques at the All-Russian Children's Center "Orlyonok". All investigated children were subdivided into two subgroups: I — elementary school children aged 7 to 11 years; II — middle school children aged 12 to 16 years. We measured length and weight of the body and calculated body mass index (BMI). We diagnosed obesity if BMI value was above 95 percentiles in accordance with the standards centile scale 'WHO Growth Reference 2007'. Data was analyzed using the statistical package "STATISTICAv.7.0 © STATSOFT, USA". The results are presented as P [CI]%, where P is the percentage, CI is the 95% confidence interval for share. Analysis of statistical significance of differences performed using Pearson χ^2 test (with Yates's correction).

Results. Obesity was diagnosed in 5.6 [5.3–5.9] %. of schoolchildren. We have identified the following relationships between age, gender, place of residence and obesity. There were more than 16.9 [16.2–17.6] % of boys with obesity than 5.2 [4.8–5.6] % of girls ($p < 0.001$). There were more girls with obesity in group I (7.8 [6.6–9.0] %) than in group II (4.7 [4.4–5.1] %; $p = 0.0056$); the boys have no differences between the groups. More obese children were from Ural (15.4%), Southern (12.5%) and North Caucasian (12.1%) federal districts; lower in Far East (6.2%), Volga (7.1%) federal districts.

Conclusion. It is important to continue analysis of the factors leading to deviations in the nutritional status of children.

Evaluation of the of lipid peroxidation reactions and regional blood flow of periodontal tissues in adolescents with arterial hypertension and periodontal diseases

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Objective. To study lipid peroxidation and periodontal tissues regional blood flow parameters in adolescents with arterial hypertension and periodontal diseases.

Methods. 94 adolescents' group with arterial hypertension (AH) and periodontal disease (n = 57, mean age of 14.7 ± 1.89 years) and a group without AH and with periodontal disease (n = 37, mean age of 14.57 ± 2.01 years) (comparison group) were examined. The intensity of lipid peroxidation (LPO) processes was assessed by spectrophotometric and fluorometric methods. The method of Doppler ultrasound was used for the assessment of periodontium hemodynamic and microvascularization.

Results. The group of adolescents with AH and periodontal diseases characterized by the statistically significant differences with the comparison group in primary products of LPO — diene conjugates (1.53 times lower, p = 0.0002), thiobarbituric acid reactive products (TBA-active products) increase (by 1.4 times, p = 0.0018), decrease of of α-tocopherol level (by 1.3 times, p = 0.0013) and retinol level increase (by 1.25 times, p = 0.014). The group with AH and with periodontal disease had an increased values of the blood flow velocity in the systole (1.12 time, p = 0.010) and index of peripheral resistance (1.19 times, p < 0.0001) compared with the data of the comparison group. In the group of adolescents with AH with periodontal disease the appearance of pathological dependencies between toxic metabolites of the lipid peroxidation process and indices of vascular blood flow was noted.

Conclusion. Imbalance in the LPO-AOD system as well as the appearance of pathological relationships between the parameters of lipid peroxidation and the parameters characterizing reduction of vascular blood flow in adolescents with hypertension and periodontal disease was proved and it can be the new comorbid association.

Clinical efficacy of BAC-SET® forte multistrain probiotics complex in the prevention of adenotonsillar pathology in preschool children

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Objective. To study BAC-SET® Forte multistrain probiotic complex efficacy and safety in humans in preschool children with adenotonsillar pathology.

Methods. For the period 2016–2019, 346 children (3–6 years old) with a history of chronic pathology of pharyngeal and palatine tonsil and suffering from frequent recurrent respiratory infections were observed.

The average age was 4.53 ± 2.71 years. Preschool children of the treatment group (n = 230) received multistrain probiotic complex BAC-SET®Forte as a prevention of exacerbations of chronic nasopharyngeal pathology daily, in addition to irrigation and elimination therapy (nasal shower with 0.9% NaCl solution); 1 capsule a day was prescribed for 30 days. The control group (n = 116) was on irrigation and elimination therapy only. The analysis of the efficiency and safety of the multistrain probiotic complex was carried out before and after the preventive course.

Results. A year before the start of the preventive course with BAC-SET®Forte multistrain probiotic complex, the pharyngeal tonsil hypertrophy of degree 2 and with complication by adenoiditis was observed in 76.3% of children in the treatment group and in 75.8% of children in the control group (p = 0.2376). By the end of the study, only 30.4% of patients in the treatment group showed no improvement in the clinical picture (p = 0.000). A year after the recovery stage, 62.7% of patients which were on the multistrain probiotic complex had recovered nasal breathing (p = 0.001); the symptoms of adenoiditis were almost stopped in 51.8% of patients (p = 0.000); 82.7% of patients had a decrease in the volume of the pharyngeal tonsil from degree 2 to 1 (p = 0.000); 78.9% of patients had a normalization of the rhinoscopic picture (p = 0.000); endoscopic control confirmed a decrease in the size of the palatine tonsils in 56.8% of patients (p = 0.000). The degree of hypertrophy of the pharyngeal and palatine tonsils in preschool children who did not receive the multistrain probiotic complex did not change and even increased in dynamics in 81.4% of patients.

Conclusions. The study results have confirmed the efficiency and high tolerability of BAC-SET®Forte multistrain probiotic complex. Prospective observation of children who was on BAC-SET® as a prevention of exacerbations of adenotonsillar pathology have confirmed its efficacy in the formation of respiratory tract immunity. The obtained data allow us to recommend the addition of BAC-SET®Forte multistrain probiotic complex in programs for the prevention of exacerbations of adenotonsillar pathology in children.

Anthropometric indicators and vitamin D level in newborns from women with gestational diabetesmellitus

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Introduction. Maternal hyperglycemia during pregnancy is one of the factors of epigenetic modifications.

Objective. A comparative analysis of anthropometric data and 25(OH)D level in newborns depending on the glycemic target level of the mother with GDM.

Methods. 66 newborns were examined: first group — 16 (24.2%) babies from mothers with GDM with glycemia in the III trimester of less than 5.1 mmol/L; second group — 20 (30.3%) from mothers with GDM with glycemia more than 5.1 mmol/L. Control group — 30 (45.5%) babies.

Results. First and control groups: maternal glucose — 4.2 [4.0–4.3] mmol/L and 4.1 [3.6–4.6] mmol/L ($p > 0.05$) respectively; body weight of newborns was 3,650 [2,350–4,280] grams and 3,345 [3,050–3,600] grams ($p > 0.05$), length 53.0 [47.0–54.0] cm and 51.5 [50.0–53.0] cm ($p > 0.05$), head circumference — 36.0 [33.0–37.0] cm and 35.5 [35.0–36.0] cm ($p > 0.05$) and 25(OH)D level is 14.6 [4.6–17.3] ng/ml and 14.5 [7.9–21.7] ng/ml ($p > 0.05$) respectively.

Second group versus the control group: maternal glucose is 6.9 [5.7–7.8] mmol/L ($p < 0.001$); weight of newborns — 3,830 [3,150–4,220] grams ($p < 0.05$); length — 53.5 [50.5–55.0] cm ($p > 0.05$), head circumference — 36.0 [35.0–38.0] cm ($p > 0.05$); 25(OH)D — 6.9 [5.7–7.8] ng/ml ($p < 0.05$).

Severe deficiency of 25(OH)D in newborns from the first and second groups was detected in 5 (33.3%) and 10 (50.0%), deficiency — in 6 (40.0%) and 7 (35.0%), insufficiency — in 4 (26.7%) and 3 (15.0%) babies respectively.

Conclusion. Newborns from mothers with GDM with glucose more than 5.1 mmol/L had significantly higher body weight and a low level of 25(OH)D than in the control group. Babies from mothers with GDM have poorer vitamin D level than healthy newborn babies.

Level of erythropoietin, sVCAM-1 and VEGF in blood of obese adolescents

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Introduction. Erythropoietin is considered as a protective tissue cytokine that increases angiogenesis. Obesity is associated with the development endothelial dysfunction, playing a key role in the pathogenesis of metabolic syndrome complications.

Objective. To determine the level of erythropoietin and presence of markers of endothelial cell dysfunction sVCAM-1 and VEGF-A in the blood of adolescents with obesity.

Methods. We examined 22 teenagers with obesity (body mass index — BMI — from 30.1 to 42.87) and 22 teenagers with normal BMI. The age of patients ranged from 13 to 18 years (average of 14.25 ± 1.2). We analyzed serum concentrations of vascular cell adhesion molecule 1 (sVCAM-1) and vascular endothelial growth factor A VEGF-A — markers that indicate the presence of endothelial dysfunction. Data was analyzed with the use of statistical package Statistica 10.0 for Windows-10. The significance of the differences was determined at P value < 0.05.

Results. Concentration of sVCAM-1 (1395.23 ± 264.73 ng/ml vs 847.44 ± 190.23 ng/ml; $p < 0.0001$) and VEGF-A (75.89 ± 54.79 pg/ml vs 6.22 ± 5.74 pg/ml; $p < 0.0001$) was higher in patients with obesity compared to the adolescents with the normal BMI. The correlation between the level of sVCAM-1 and BMI ($r = 0,45$; $p < 0,05$).

Erythropoietin level in obese children was lower than in children with normal BMI (17,24 ± 10.9 and 36,31 ± 31,41; $p < 0,001$), a negative correlation between BMI and erythropoietin level ($r = -0.26$; $p < 0,05$).

Obese children revealed a negative correlation between the level of sVCAM-1 and the level of erythropoietin in the blood serum ($r = 0,48$; $p < 0,05$).

Conclusion. Obesity in adolescents characterized by decreased erythropoietin and increased level of endothelial dysfunction markers sVCAM-1 more than 2 times, VEGF-A — more than 12 times compared to adolescents with a normal BMI. Evaluation of the protective role of erythropoietin in the prevention of endothelial dysfunction and its complications is necessary.

Transportation of children born with severe asphyxia from medical institutions of the first and second level of the Perm region to a specialized center

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Objective. To evaluate the results of early transportation to a specialized center of children born with severe asphyxia in level I and II medical institutions.

Methods. The medical histories and transport maps of 20 children born with severe asphyxia in medical institutions of the first and second level of the Perm Region and transported to the neonatal intensive care unit of the GDKB PR number 13 in 2020 were analyzed. Transportation was carried out by the resuscitation team of the department of emergency advisory medical care of the Perm regional children clinic hospital.

Results. 19 (95% of children) were full — term, 1 — premature (at 35 weeks) — 1 (5%); the average body weight was 3363.9 ± 15.6 g. All patients were on a mechanical ventilation of lungs from the birth. Taking into account the severity of the condition and the need for specialized care, these patients required emergency transportation to a specialized center in Perm. In the first day of life, 16 (80%) children were taken out; 3 (15%) were consulted on the spot due to their non-transportable condition, and later 2 children were transported when their condition was stabilized on the 2nd and 3rd days of life. The fatal outcome occurred in 2 (10%) children, 1 patient died an hour after birth, 1-on the 3rd day. Thus, 18 patients (90%) were transported within the first 3 days. Transportation was carried out after the assessment of the child's condition by the transporting resuscitation team after the necessary preparation and correction of treatment, mechanical ventilation of lungs, inotropic support, infusion therapy with elements of parenteral nutrition. There was no deterioration in the condition of the children during transportation. When studying the catamnesis, it was revealed that among the transported children, the mortality rate was 5.5%.

Conclusion. Transportation to a specialized center of children born with severe asphyxia in level I and II medical institutions in the first day of life improves the prognosis and helps to reduce neonatal mortality.

The antiarrhythmic efficacy of H1 — histamine receptor blocker quifenadine in children with frequent extrasystoles

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Objective. To determine safety and efficacy of quifenadine versus amiodarone in children with premature beats (PB).

Methods. 84 patients (mean age 10.4 ± 3.8 yrs) with frequent (> 10000 during 24 h) ventricular (n = 45) and supraventricular (n = 39) PB were randomized 1:1 to quifenadine (1–3 mg/kg/day, n = 54) or amiodarone (8–10 mg/kg/day, n = 30) arms. The therapeutic efficacy was evaluated by 24-hour Holter monitoring at 2–4 and 9–12 weeks of treatment.

Results. Complete antiarrhythmic effect (PB < 50% from baseline) has been achieved in 23/54 (43%) of quifenadine-treated patients, which was less than in amiodarone groupe (24/30, 80%, p = 0.02). Quifenadine was mostly beneficial in children with supraventricular PB and/or bradycardia. Quifenadine therapy led to moderate QTc interval prolongation without exceeding of clinically meaningful values. The side effect incidence in quifenadine group (drowsiness and headache) was significantly lower (2%) than in amiodarone group (40%, p < 0.05). In case of lack of quifenadine and amiodarone alone effect, combination therapy was used (quifenadine 1–2 mg/kg/day and amiodarone 4–6 mg/kg/day). The combination therapy showed complete antiarrhythmic effect in 10/12 (83%) of patient without significant QT prolongation or sinus node depression (probably due to quifenadine anticholinergic properties). The only side effect was thyroid dysfunction (8,3%) in this group.

Conclusion. The obtained data have shown quifenadine antiarrhythmic activity in children with premature beats. Quifenadine with amiodarone combination led to decrease antiarrhythmic side effects incidence while maintaining it therapeutic efficacy.

Very rare case of Noonan syndrome, type 2

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Background. Noonan syndrome, type 2 (NS2) is rare autosomal recessive disorder of RASopathies group, caused by mutations in the *LZTR1* gene. NS2 characterized by a typical face, short stature, broad, short neck, congenital heart disease, developmental delay. The most common heart disease in children with NS2 is hypertrophic cardiomyopathy.

Methods. Patient is a boy, 15 years old with short stature, developmental delay at 1 first year of life and heart disease. He had distinctive facial features of NS: downslanting palpebral fissures, epicanthic folds, hypertelorism, low-set ears short neck, wing-like folds on the neck, pectus deformity. Hypertrophic cardiomyopathy was identified at 1 month old. Now patient have obstructive, hypertrophic cardiomyopathy, cardiac arrhythmia: ventricular extrasystoles, 4A Lown, intraventricular block combined with bundle branch block, transient WPW. Syncopal episodes.

Surgical correction of obstructive hypertrophic cardiomyopathy: septal myomectomy was performed on the child due to the high risk of developing sudden death syndrome. Target areas of the exome were investigated by next generation sequencing (NGS). Bioinformatic analysis was carried out using ACMG recommendation. Validation of the identified variants was carried out by the Sanger method.

Results. We revealed nucleotide missens VUS: c.1259A>G, p.Q420R and c.2051T>C, p.I684T in the heterozygous state in *LZTR1* gene. All variants were absent in HGMD professional and genome aggregational database.

Conclusion. Child with severe hypertrophic cardiomyopathy and typical phenotype of Noonan syndrome was detected NS 2, caused by compound heterozygous missense variants c.2051T>C, p.I684T and c.1259A>G, p.Q420R in *LZTR1* gene.

ROHHAD syndrome: clinical case and literature review

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Background. Sleep-related breathing disorders are a potentially lethal conditions characterized by multiple episodes of sleep apnea. Study of these conditions seems to be relevant for pediatrics due to the difficulties of diagnosis and the lack of actual effective treatment strategies. This article describes the clinical case of the rare ROHHAD syndrome which represents a condition related to central sleep apnea.

Objective. To review the literature data on sleep-related breathing disorders and present our own clinical case of the rare ROHHAD syndrome.

Methods. More than 100 literature sources were reviewed. Presented the case of a 6-year-old child with sleep apnea hospitalized to the Mordovian Republic Clinical Hospital.

Results. ROHHAD syndrome is a very rare disease (about 100 cases are described worldwide). In most cases it presented as rapid obesity development for 3–5 years of age as well as hypoventilation, endocrine dysfunction, electrolyte disturbances. Autonomic nervous system dysfunction reveled in ROHHAD pediatric patients rarely. Own observation demonstrated a typical clinical presentation of ROHHAD syndrome. Boy D., 6 years 11 months was admitted to the pediatric department of the Children's republican clinical hospital with severe weakness, fatigue during physical exertion, obesity, shortness of breath at rest, orthopnea, inability to sleep on the back (sleeps with an elevated head end or on the right side); lower extremities weakness during walking and climbing upstairs during the last year, changes in walking and in behavioral reactions. Cyanotic attacks and tachycardia have appeared during sleep in previous six months before admission. Firstly, the condition manifested with sudden and rapidly progressive obesity. Ventilation disorders became clinically evident after carried over pneumonia. Episodes of sleep apnea lasted up to 20 seconds, were accompanied by cyanotic attacks and tachycardia. Multiple investigations were performed to exclude systemic connective tissue diseases, neurological disturbances and brain pathology, muscular tissue pathology and metabolic diseases. Finally, the diagnosis was confirmed by the polysomnography and genetic tests. The ventilation support during sleep was prescribed.

Conclusion. The rarity of the disease leads to a decreased awareness of pediatricians. The diagnosis is made by clinical picture and polysomnography, and timely diagnostics can increase the duration and improve the quality of life of patients.

Clinical and Genetic Spectrum of Dystroglycanopathy Due to *POMGNT1* Mutations in Russian Patients

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Objective. Dystroglycanopathies are the heterogeneous group of hereditary disorders, caused by the abnormal glycosylation of α-dystroglycan. The most common dystroglycanopathy is muscle-eye-brain disease (MEB) associated with mutations in the *POMGNT1* gene. MEB is autosomal recessive disease characterized by congenital muscular dystrophy, ocular abnormalities and brain malformation. The goal of the study is an analysis of clinical findings, laboratory features and results of instrumental research.

Methods. Molecular genetic diagnostics was performed using full exome sequencing. All patients and all parents were confirmed by Sanger sequinning.

Results. We observed 3 boys with MEB disease aged from 25 to 118 months, averaging about 7 years (83 months). The one patient had exceeded average values of height and weight at birth, two the other children had normal ranges. All children had severe motor development delay, only one patient could walk without support. Two older patients had mental retardation and lack of speech development. Language skills represented as vocalizations. Physical growth of our patients fluctuated from 3 to 75 percentiles for height and weight. The other clinical features include hypotonia and strabismus (all patients), autistic behavior (2 patients), ataxia, seizures, hepatomegaly (one in each patient). Dysmorphic features were non-specific.

Ophthalmological examination revealed congenital high myopia (2 patients), partial atrophy of an optic nerve (2 patient), nystagmus (1 patient), astigmatism (1 patient), retinal atrophy (1 patient).

Biochemical analysis showed elevated creatine kinase from 1874 to 6266 (averaging 3754 U/L), ALT from 42 to 93(68 U/L), AST from 53 to 106 (80 U/L), LDH from 370 to 503 (437 U/L).

Electromyographic examination has showed that all children had signs of primary muscle lesion. Muscle MRI has displayed a severe atrophy muscles and fatty infiltration in one patient. MRI findings have reported pachygyria and ventriculomegaly (all patients), hypoplasia cerebellum, corpus callosum, pons (2 patients), hypoplasia of temporal lobes (1 patients), cerebellum cysts (2 patients). EEG did not reveal epileptic form activity.

We revealed compound heterozygous mutations in all three patients. These were five different mutations: missense c.385C > T (p.R129W) and c.1325G > A (p.R442H), nonsense c.643C > T (p.R215X), splicing c.1539+1G > A and frameshift duplication c.453_456dup (p.S153Vfs*5). The last one was not previously described in the HGMD database.

Conclusion. Clinical and genetic features were described in Russian patients with dystroglycanopathy due to *POMGNT1* mutations.

Umbilical cord blood cytokines TNFα and IFN-γ levels increased in children born to mothers who are obese

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Background. Maternal obesity is considered one of the several key factors that affect development of the immune system of newborns. Experimental and clinical data indicate an increased risk of developing autoimmune, allergic diseases and obesity in the offspring of obese mothers. The main mechanisms of the relationship between mother's body weight and the immune system of a newborn person remain poorly understood.

Objective. Aim of this study was to analyze the cytokine status of umbilical cord blood of children born to mothers with obesity.

Methods. Umbilical cord blood samples were taken from 65 children born to thin (n = 24), with overweight (n = 9) and obese mothers (n = 32). The levels of TNF-α, TGF-β1, IL-18, IL-13, IL-10 and IFN-γ were quantified by IFA. Statistical processing of data was performed on a personal computer using licensed computer software "Microsoft Excel 2016" and "STATISTICA 12". The student t-test value was determined while analyzing the distribution of quantitative data. The criterion of statistical significance level was $p < 0.05$.

Results. Compared to children born to thin mothers, children born to obese mothers had higher levels of umbilical cord blood plasma TNF-α (12,75 ± 10,80 pg/ml and 4,94 ± 3,55 pg/ml; $P1,3$ = 0,005408) and IFN-γ (798,90 ± 565,96 pg/ml and 311,05 ± 249,08 pg/ml; $P1, 3$ = 0,014947).

Conclusion. These results confirm the hypothesis that maternal obesity affects programming of the immune system of newborns providing a potential connection with an increase in the incidence of chronic inflammatory diseases and obesity in offspring.

Fabry's disease with minimal manifestations in girls

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Objective. Fabry disease (Anderson-Fabry disease) is a rare inherited lysosomal multisystem disease with X-linked inheritance due to the deficiency of lysosomal enzyme α-galactosidase A and leads to accumulation of sphingolipids (globoside, globotriaosylceramide) in the walls of blood vessels in all organs.

Methods. Clinical cases of low-symptomatic Fabry's disease in 2 girls (9 and 10 y.o.) with positive family history of the underlying disease are described.

Results. Girl 9 y.o. is granddaughter and girl 10 y.o. is daughter of a male patient with Fabry's disease. Girls had complaints of reduced sweating and febrile acroparastesia only.

Physical and sexual development according to age, the skin was clean.

Echocardiography, abdominal USE, pulmonary function test, cornea and fundus, ear, nose and throat examination were normal. Urine examination showed 1+ proteinuria both girls have. Glomerular filtration rates were normal. The lyso-Gb3 and galactosidase A levels analyzed in dried blood spots (DBS) by tandem mass spectrometry and molecular genetic analysis was performed. An increase lyso-Gb3 concentration and a change in the nucleotide sequence c.983G > C, leading to the replacement of p.G328A, described in the international database on *HGMD* mutations (CM 930337) in the hemizygous state has been identified.

Conclusion. These cases confirm that it is very important that the pediatricians become aware of the importance of genealogical anamnesis findings and clinical features of Fabry's disease, so they can participate in the identification of unrecognized patients.

Markers of inflammatory response of the intestine in newborns whose mothers received a probiotic during the 6 weeks before delivery

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Objective. To determine the concentration of the fecal eosinophil-derived neurotoxin (EDN) and fecal calprotectin as intestinal markers of inflammatory response in newborns whose mothers received a probiotic during the 6 weeks before delivery.

Methods. Pregnant women (n = 115), depending on the number of detected chronic diseases, were divided into two groups. The main group included women with a history of two or more chronic diseases, including the genitourinary sphere — 63/54.8%. The comparison group included pregnant women without a history of chronic diseases or having one chronic disease, except for the pathology of the urogenital sphere — 52/46.2%. Pregnant women of the main group with a preventive purpose for 6 weeks before delivery used a probiotic containing Bifidobacterium longum и Streptococcus thermophiles. The concentration of fecal EDN and fecal calprotectin was assayed using an ELISA method (Immundiagnostik, Bensheim, Germany and Calprest, manufactured by Nycomed).

Results. We found the following changes from the intestinal markers of inflammation in newborns depending on the use of probiotic by women before delivery: in children of the main group (mothers used probiotics) both markers were significantly lower than in comparison group (mothers not used probiotics).

Concentration of fecal EDN was 163.4 ± 58.2 vs 224.1 ± 83.4 ng/g (P < 0.001) and fecal calprotectin was 240.3 ± 78.6 vs 315.6 ± 101.2 mcg/g (P < 0.001).

Subsequent follow-up (during the first year of life) showed that in children of the main group were less frequently detected functional disorders of the gastrointestinal tract (15.8% vs 36.5%, P = 0.034) and allergic diseases (1.2% vs 5.8%, P = 0.056).

Conclusion. In newborns whose mothers received a probiotic during the 6 weeks before delivery, after birth levels of both markers of intestinal inflammation (fecal eosinophil-derived protein and fecal calprotectin) were significantly lower than in comparison group (children whose mothers not used probiotics).

In addition, in children of the main group in the first year of life were less than half detected such conditions as functional disorders of the gastrointestinal tract and manifestations of food allergies.

Pathomorphological features of rhabdomyosarcomas in pediatric patients

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Objective. We want to explore the specific morphological manifestations, determine the frequency of different histological variants, as well as identify local mutations of genes determining the development of these tumors.

Methods. Medical records and histological preparations of 7 children treated with the diagnosis were analyzed at the pathologoanatomic department of St. Petersburg State Medical University: "Rhabdomyosarcoma" from 2001 to 2018. Histological preparations were photographed using a Pannoramic Midi 2 scanning microscope.

Results.

- Numerous star-shaped and elongated, spindle-shaped cells irregularly distributed among the stroma were found in embryonic RMS (ERMS) preparations. Elongated cells with eosinophilic cytoplasm with transverse striations in the cytoplasm were also detected.
- In pleomorphic RMS, tumor cells were determined to have a variety of shapes, including rocket-shaped, with oval nuclei and small nuclei. Large foci of myxomatosis of tumor stroma were observed.
- Alveolar rhabdomyosarcoma (ARMS) was characterized by the presence or formation of alveolar structures lined by rounded or oval-shaped tumor cells. The nuclei of these cells were kidney-shaped and/or lobulated, with a well-defined cytoplasm. Hyalinized fibrous septa were detected.
- Alveolar and pleomorphic RMS are formed from elements of muscle tissue, and in rare cases alveolar from endothelial progenitor cells after reprogramming and myogenic transdifferentiation, while ERMS has a dysontogenic origin, that is arises from detached rudiments of muscle tissue and are transverse muscular tissue hamartoblastomas.
- Alveolar RMS is a more malignant and less differentiated tumor, for which reason the mortality rate in patients with ARMS is significantly higher than in other types of RMS.
- ARMS are associated with the *FOXO1* gene fusion return system found in 90% of cases.
- 56 mutations were found in 28% of ERMS cases and included 7 mutations in rate in the RAS family, 4 mutations in *FGFR4*, 3 mutations in *PIK3CA*, 2 mutations in *CTNNB1* and single mutations in *BRAF* and *PTPN11*.

Experience of the use of herbal medicinal product Tonsilgon N in preschool children with Waldeyer's tonsillar ring medical condition

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Objective. To evaluate efficacy and safety of the use of the herbal medicinal product Tonsilgon N (HMP) in clinical practice in preschool children with Waldeyer's tonsillar ring medical condition.

Methods. 628 children (aged 2–5 years, median age — 3.34 ± 2.78, girls — 316 (50.4%), boys — 312 (49.6%)) with chronic conditions of the pharyngeal and palatine tonsils were enrolled in the study. All children were divided into two comparable groups. Children in group I (n = 317) received HMP in drops during 60 days in overall.

Preschoolers in the group II (n = 311) did not receive HMP. Clinical efficacy and safety evaluation of the drug was performed before and after the treatment course.

Results. HMP therapy (group I) eliminated the signs of adenoiditis and led to a decrease in the number of children with second degree tonsil hypertrophy from 75.0–6.84% to 36.1–4.62% (p < 0.001). Correspondingly, the number of children with I degree hypertrophy of pharyngeal tonsils increased (p < 0.01). Adenoiditis signs and symptoms were almost resolved in 75.8%–5.34% of patients; symptomatic treatment of adenoiditis was not effective (p > 0.05) The degree of hypertrophy of pharyngeal and palatine tonsils in preschoolers of the 2nd group did not change and even showed a tendency to increase to 83.6%.

By the end of HMP therapy, children in group I demonstrated a good overall well-being, the swelling of palatine tonsil subsided, adequate nasal breathing was restored, there was no pathological discharge from the pharyngeal tonsils. One year after the end of treatment, nasal breathing was restored in 62.7% of patients who received HMP (p < 0.01); snoring stopped in 81.8% of patients (p < 0.01). Treatment with HMP was accompanied by an increase in the level of lysozyme in nasal secretions from 56.9–0.88% to 69.8–0.45% (p < 0.001) and SIgA from 0.18–0.005 g/L to 0.20–0.003 g/L (p < 0.01). In group II, no significant change was observed in the indicators characterizing the state of local immunity. Among those who received HMP, 90% showed excellent and good results. Symptomatic treatment in group II did not affect the size of the pharyngeal tonsil and local immunity parameters.

Conclusion. Study results confirmed efficacy and good tolerance of the HMP. Evaluation of the frequency of exacerbations of chronic ENT pathology in children who received this HMP during rehabilitation confirmed its effectiveness in boosting of respiratory tract immunity.

Typical problems of parents of children with cleft lip and palate

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Objective. Presented review of literature data (medline, eLIBRARY.RU, PubMed) identifies typical problems of parents whose children were born with cleft lip and palate (RGN), as well as research directions in improving the organization of medical care for parents and patients with UAH in solving these problems.

Methods. Analysis of domestic and foreign authors on MEDLINE databases, eLIBRARY.RU, PubMed.

Results. Research on the experiences of parents with RSN (what typical problems they encountered in the maternity hospital and, especially, at home) still lacks. It is necessary now to carry out research that has been traditionally rely not only on psychological approaches, but also on broader prospects for research in the field sociology, social policy, nursing and health care using both qualitative and quantitative methods.

Conclusion. In addition, there is a lack of research in the field of cleft lip and palate to study the experience and needs of parents at different stages of their children's lives.

The first clinical case of rare form of focal epilepsy caused by the novel mutation in the *NPRL3* gene in Russian federation and Kazakhstan

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Objective. Mutations in the *NPRL3* gene (OMIM 600928) are described predominantly in patients with autosomal dominant focal epilepsy according to the HGMD database.

Methods. We want to introduce the clinical case of the female patient, 2 years old, with focal epilepsy from healthy parents. She was born full-term from the 1st pregnancy by caesarian delivery. The cerebellum hypoplasia was suspected during ultrasound and MRI diagnostic at 17th week of gestation.

Weight at birth — 3.780 kg, 7/8 on APGAR scale. Neurosonography at birth has shown left sided ventriculomegaly. First recurrent afebrile tonic extensor epileptic spasms with head and eyes turns to the left side were noted at the age of 2 months. Eyelid and tongue myoclonus were revealed as well. Later the frequency of seizures has increased to 150 per day. The video electroencephalogram (VEEG) showed sharp, spike waves with partial origin at left temporal region, while clonic seizures in the left arm were recorded. The brain MRI has shown focal cortical dysplasia in the left insular region. Signs of intellectual impairment and behavioral disturbances were revealed during the neurological examination at the age of 24 months. The patient could not walk without support. We have performed full exome sequencing for our patient to identify the molecular genetic causes of the disease after medical genetic counseling.

Results. The nucleotide variant c.481C > T which leads to stop codon p.Q161* in heterozygous state was revealed in exon 5 of the *NPRL3* gene. This variant was not described in gnomAD and HGMD databases and was considered as pathogenic according to ACMG criteria. An interesting fact is that the most frequent pathogenic variants in the NPRL3 gene (among the 21 described variants in the HGMD) are nonsense mutations and frameshift deletions.

Conclusion. This paper describes the first clinical case of rare form of focal epilepsy caused by the novel mutation in the *NPRL3* gene in Russian Federation and Kazakhstan.

Exogenic and endogenic factors affecting the supply of vitamin D in healthy children and adolescents of the south of Russia in winter-spring periods

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Objective. To analyze the influence of exogenous and endogenous factors on the supply of vitamin D in children and adolescents living in southern Russia in the winter-spring period.

Methods. The study included 27 healthy children and adolescents, aged 3 months to 16 years, living in Stavropol (45°02′ N 41°58′E). Children under 3 years old were 9 (33.3%), 4 to 7 years old — 5 (18.5%), 8 to 11 years old — 6 (22.3%), 12 to 16 years old — 7 (25.9%) people. Blood sampling was performed in February-March 2018. Vitamin D provision was assessed by the serum calcidiol level. Satisfactory supply was diagnosed at a level of 25 (OH) D 30–100 ng/ml, insufficiency — 20 to 30 ng/ml, deficiency — less than 20 ng/ml.

Results. The study of polymorphism of biotransformation genes was carried out by PCR and subsequent RFLP analysis. Polymorphisms selected for study included *CYP2C9*2* (430C > T; R144C), *CYP2C9*3* (1075A > C; I359L), *CYP2D6*4* (1846G > A), *CYP3A4*1B* (–392C > T).

Median vitamin D availability was 34.6 [23.5–44.6] ng/ml. Vitamin D deficiency was detected in 5 (18.5%) children, deficiency was also found in 5 (18.5%) children, satisfactory provision was revealed in 17 (63.0%) children.

There is a negative correlation between the age of children and the level of 25(OH)D: $r = -0.69$, $p = 0.0001$, as well as between body weight and calcidiol level $r = -0.64$, $p = 0.0004$.

Direct correlation was found between the dose of cholecalciferol products and serum 25(OH)D level, $r = -0.60$, $p = 0.001$.

With CC polymorphism (*CYP2C9*2*) 25(OH) D < 30 ng/ml was detected in 8 (80.0%) children, and more than 30 ng/ml — in 2 (20.0%), with CT — in 2 (20.0%) and in 2 (11.8%) respectively. With polymorphism AA (*CYP2C9*3*) 25(OH) D < 30 ng/ml was in 9 (90.0%) children, more than 30 ng/ml — in 15 (88.2%), with CA — in 1 (10.0%) child and in 2 (11.8%) children respectively. With TT polymorphism (*CYP3A4*1B*) 25(OH) D < 30 ng/ml was detected in 9 (90.0%) children, and more than 30 ng/ml — in 16 (94.1%) children, with TC — in 1 (10.0%) and in 1 (5.9%) respectively. With polymorphism GG (*CYP2D6*4*) 25(OH)D < 30 ng/ml was detected in 7 (70.0%) children, and more than 30 ng/ml — in 13 (76.5%) children, with GA 25(OH)D < 30 ng/ml was in 2 (20.0%), more than 30 ng/ml — in 4 (23.5%), AA was only in 1 (10.0%) child with vitamin D less than 30 ng/ml.

Conclusion. Insufficient vitamin D levels were found in 37.0% of healthy children and adolescents living in the Southern Russia. The serum calcidiol level depends on age; the older is the child, the higher is the likelihood of hypovitaminosis D. Intake of cholecalciferol product is an effective way to prevent vitamin D deficiency and insufficiency, while hypovitaminosis D was not convincingly associated with any of the studied polymorphisms of biotransformation genes: *CYP2C9*2*, *CYP2C9*3*, *CYP2D6*4*, *CYP3A4*1B*.