above 38 ° C, erythrocyte sedimentation rate over 30 mm/h and monoarthralgia were defined as minor diagnostic criteria in middle and high risk populations. The aim of this study is to determine the effect of new diagnostic criterias on our clinical practice.

Methods Among a total of 1132 patients hospitalized in our Pediatric Cardiology Clinic between 2013 and 2019, data of 58 patients were diagnosed with ARF were investigated and the 3-year period before and after the updated Jones criteria were compared.

Results Fifteen of the 493 patients hospitalized between 2013–2015, 43 of the 639 patients hospitalized between 2016–2019 were diagnosed as ARF. There was a significant increase in the number of patients with ARF during the updated diagnostic criteria ($p = 0.007$). Joint complaints were in the foreground in both periods and there was no difference between the distribution of patients who were evaluated as clinical and subclinical carditis ($p > 0.05$). In the previous period, there was no patient in whom monoarthritis and polyartralgia were accepted as major diagnostic criteria but, in the last three years, of the 17 patients, 8 had monoarthritis and 9 had polyarthralgia as the major diagnostic criteria. Of the ARF patients with polyartalgia defined as the major diagnostic criteria, 6 patient underwent diagnostic changes; 2 had juvenile idiopathic arthritis, 3 had familial Mediterranean fever and 1 had systemic lupus erythematosus. In all of these patients with high ASO levels, except for two patients with subclinical carditis, the diagnosis of ARF was made by polyartralgia, fever and high acute phase reactants. In a patient with carditis findings besides polyartalgia, the association of ARF and Takayasu arteritis was defined at the 8th month of follow-up.

Conclusion With the updated Jones criteria, the number of patients diagnosed with ARF are increased. However, in the follow-up, some patients diagnoses were changed as collagen tissue diseases and vasculitis. New criterias may lead to diagnosis of some cases of ARF that may be overlooked, but may also lead to over-diagnosis. Therefore, collagen tissue diseases and vasculitis should be considered in the differential diagnosis of patients with polyartalgia, especially in patients with atypical joint involvement without carditis.

GP30 ASSOCIATION OF ACE GENE INSERTION/DELETION POLYMORPHISMS WITH HYPERTENSION IN CHILDREN WITH GLOMERULONEPHRITIS

Lyubov Rychkova*, Elena Belyaeva, Tatyana Bairova, Lyubov Kolesnikova. *Scientific Centre for Family Health and Human Reproduction Problems, Irkutsk, Russian Federation*

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Background Kidney diseases are a common cause of secondary hypertension in children, which determines an unfavourable prognosis of the underlying disease. The secondary hypertension is a multifactorial complex disorder caused both by genetic and environmental factors. According to estimations approximately in 30% of cases, the interindividual variability in blood pressure is genetically determined. Numerous studies focused on the role of genetic variation in genes implicated in the renin-angiotensin system (RAS), particularly the angiotensin-converting enzyme (ACE) gene. The purpose of our study was designed to explore association of angiotensin converting enzyme (ACE) gene insertion/deletion (I/D) polymorphism with secondary hypertension in a group of children with glomerulonephritis.

Methods The study included two groups of children with glomerulonephritis, one of them - hypertensives (cases) and another group - normotensives (controls). Group with hypertension comprised 35 children, age 10.46±2.93 years (20 males and 15 females), group without hypertension included 26 children, age 9.61±3.18 years (18 males and 8 females). The variant genotype of ACE gene (I/D polymorphism; rs4646994) was determined using polymerase chain reaction (PCR). DNA was extracted from blood samples. DNA fragments length analysis after amplification and restriction was carried out in 7% acrylamide gel. Statistical analysis was performed using STATISTICA, version 10.0 StatSoft Inc, USA.

Results We determine the distribution of genotype and allele frequencies in the hypertensives and normotensives groups of children with glomerulonephritis. The analysis showed that the frequencies of genotype II, ID, DD of ACE gene in the group of hypertensives were 14.3%, 25.7%, 60%, in the group of normotensives were 76.9%, 15.4%, 7.7%. We found statistically significant higher frequency of DD genotype ($\chi^2 = 8.44$; d.f.=1; $p=0.004$) and D allele ($z=6.1$; $p=0.001$) in the hypertensives group in comparison with the normotensives group of children. We calculated the probability of DD genotype detection in the group of patients with glomerulonephritis by the odds ratio (OR) was calculated with 95% confidence interval (95% CI). Patients with the DD genotype had a higher risk of hypertensives than patients with the II, ID genotypes (OR: 18; 95% Cl: 3.66 - 88.55).

Conclusions The findings revealed significant influence of I/D polymorphism of ACE gene on hypertension in the groups of children with glomerulonephritis. There are significant associations between DD genotypes and secondary hypertension in children with glomerulonephritis.

GP31 ENDOTHELIAL DYSFUNCTION AS THE MAIN COMPONENT OF ARTERIAL HYPERTENSION IN THE CHILDREN

Olesya Horlenko*, Vasyl Rusyn, Natalia Sochka, Olena Debretseny, Fedir Horlenko, Galina Radovetska. *Uzhhorod National University, Uzhhorod, Ukraine*

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Backgruond and aim First of all, endothelial dysfunction (ED) have an importante role in modulation of cardiovascular risk in healthy individuals and patients with arterial hypertension. It is already known that the formation of hypertension is associated not only with the state of vascular tone, but also associated with a violation of the relaxation properties of the endothelium.

Methods We selected 80 adolescents aged 15–17 years (16,0 ± 0,11). Clinical, laboratory and instrumental methods for identification of endothelial dysfunction were used.

Results In the children with AH in association with endothelial dysfunction (BMI = 16), metabolic changes and increased electrical activity of LV in ECG examination were revealed. The level of IVRT is elevated and indicates a decrease in the active ventricular relaxation (109 ± 9.41 ms versus 72.18 ±

3.41 ms in the control group) according to our data. The ambulatory arterial stiffness index (AASI) was 0.88 ± 0.02, which was exceeded the control group data (0.55 ± 0.01). The increase in diameter of the artery(reactive hyperemia test using) in the study group was less than 10% (8.59 ± 0.36%). Changes of ECO-indicators: reduction of peak speed of wave VE; increase of peak speed of wave VA; reduction of the VE/VA ratio; prolongation of the isovolumic relaxation phase; an increase in delayed early diastolic filling may be considered as initial manifestations of diastolic dysfunction in the examined children.

Conclusion The ambulatory arterial stiffness index was 0.88 ± 0.02 in the study groupe. Increase the diameter of the artery after reactive hyperemia test in the study group was less than 10%, KIM - 0,56 ± 0,03 mm). The use of integral morfo-functional parameters provides an objective criterium to optimize diagnosis, for the purpose of risk evaluation in the children with AH and allows controlling the development of cardiovascular pathology

GP32 ECHOCARDIOGRAPHIC FOLLOW-UP OF CHILDREN WITH SUBAORTIC STENOSIS

[1]Betül Çınar*, Sezen Ugan Atik [2], [2]Ayşe Güler Eroğlu, [2]Levent Saltık. *[1]Istanbul University Cerrahpaşa Medical Faculty, Department of Pediatrics, istanbul, Turkey; [2]Istanbul University Cerrahpaşa Medical Faculty, Department of Pediatric Cardiology, istanbul, Turkey*

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Introduction Subaortic stenosis(SAS) accounts for 8–30% of patients with congenital left ventricular outflow tract obstructions. Allthough progressive SAS occurs in many patients, the exact etiology and factors contributing to progression remains unknown. In this study, we evaluated the natural course of SAS, associated aortic regurgitation (AR), the factors affecting the progression of SAS and AR and the outcomes of surgery during the long-term follow-up with echocardiography.

Materials and Methods The study included 105 patients who were evaluated and followed with echocardiography at our institution between 1990 and 2017 with SAS, consisting of either a thin ridge or a thicker but discrete obstruction with a muscular base. Patients with incomplete medical records, abnormal ventricular function, and lesions other than AR were excluded. The last examination prior to any surgical intervention provided our final measurements. The level of narrowing of the LVOT, the distance between right coronary cusp and ridge and the anulus of the aortic valve were determined with two-dimensional echocardiography. Continuous-wave Doppler was used through an apical five-chamber view to record the maximum peak and mean systolic instantaneous gradient across the supravalvular narrowing. A multivariate analysis with Cox proportional hazards modeling was performed to adjust for the different distributions of variables between groups. The enter method was used in logistic regression analysis.

Results Among 105 patients (median, 5 years at initial echocardiography), 64% were male and 36% were female. The patients were followed median 6.6 years. Aortic valve morphology was tricuspit in 95.2% and bicuspid in 4.8%. The median distance of discret membran from the right coronary cusp was 6.4 mm. The degree of SAS staid the same in 60%, progressed in 29% and 11% underwent surgery after initial echocardiography. AR did not develope in 21(20%), not deteriorate in 41(39%) and progressed in 43(41%) patients. Surgery was performed in 38(63.8%) patients. 6(5.7%) patients underwent reoperation.

Conclusion We recommend careful and frequent evaluation for patients with moderate stenosis because surgery may be needed depending on the severity of stenosis and AR. Postoperatively, follow-up is required.

GP33 HIGH PREVALENCE NOONAN SYNDROME IN RUSSIAN CHILDREN WITH HYPERTROPHIC CARDIOMYOPATHY, DIAGNOSED BY NEXT GENERATION SEQUENCING

[1]N Zhurkova V*, [1]K Savostyanov V, [1]AA Pushkov, [2]ND Vashakmadze, [1]N Fedorova V, [1]OP Zharova, [1]LA Gandaeva, [1]NA Sdvigova, [3]OB Kondakova, [1]EN Basargina. *[1]National Medical Research Center of Children's Health of the Ministry of Health of the Russian Federation, Moscow, Russian Federation; [2]Pirogov Russian National Research Medical University, Moscow, Russian Federation; [3]Scientific and Practical Center of Pediatric psychoneurology Department of Health of Moscow, Moscow, Russian Federation*

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Backgraund Noonan syndrome - is rare autosomal dominant disorder from RASopathies group, characterized by facial dysmorphism, short stature, hypertrophic cardiomyopathy, congenital heart defects.

Patients and Methods We have examined 47 patients with hypertrophic cardiomyopathy aged 1 to 17 years. Target areas of the exome were investigated by NGS. Bioinformatic analysis was carried out using the Alamut software. Validation of the identified variants was carried out by the Sanger method.

Results The diagnosis was confirmed in 13 patients with hypertrophic cardiomyopathy and cardiologic abnormalities. Short stature and facial features have all our patients. Congenital heart defect, including pulmonic stenosis diagnosed in 6 cases.

RAF1 mutations were identified in 7 patients, *PTPN11* - in 3 patients, *SOS1*- in 1 patient, *SOS 2* - in 1 patient, and *RIT1* - in 1 patient. Most frequent *RAF1* mutation was *c.770C>T, p. S257L* (5 from 7 cases). Girl with mutation in *RIT-1* was with phenotype Noonan syndrome, but she also have left ventricular noncompaction and skin cafe-au-lait spots.

Conclusion Noonan syndrome was diagnosed in 28% hypertrophic cardiomyopathy patients.

Mutation *c.770C>T, p.S257L* in *RAF1* gen is most common in hypertrophic cardiomyopathy patients with Noonan syndrome

GP34 QUALITY OF LIFE IN CARDIOPATHIC CHILDREN AND ADOLESCENTS

Giulia Amodeo*, Teresa Grimaldi Capitello. *Ospedale Pediatrco Bambino gesù, Roma, Italy*

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Introduction The concept of quality of life is broad and multidimensional. The temporal dimension of its evaluation is very important because the quality of life is not a stable and defined condition, but changes over time.

Heart disease is classified among the most severe chronic disabilities in children and as more and more children and adolescents with severe cardiac defects survive into adulthood, the quality of their life becomes increasingly important.

The quality of life in cardiopaths may vary depending on various factors, ranging from the intake of a drug therapy, to