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% ($8.59 \pm 0.36\%$). 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 group. 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 morpho-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

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Introduction Subaortic stenosis(SAS) accounts for 8–30% of patients with congenital left ventricular outflow tract obstructions. Although 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 annulus 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 tricuspid in 95.2% and bicuspid in 4.8%. The median distance of discrete membranous from the right coronary cusp was 6.4 mm. The degree of SAS stayed the same in 60%, progressed in 29% and 11% underwent surgery after initial echocardiography. AR did not develop 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

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Background 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* gene is most common in hypertrophic cardiomyopathy patients with Noonan syndrome

GP34 QUALITY OF LIFE IN CARDIOPATHIC CHILDREN AND ADOLESCENTS

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Introduction The concept of quality of life is broad and multi-dimensional. 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

the possible prohibition of performing sports, to the results of surgery as scars, a possible source of shame.

Parents of children with heart disease may experience higher stress levels than normal parental function and may feel very stressed about issues related to accountability and social integration.

Methods This research aims to investigate the quality of life in pediatric age both in terms of purely cardiological aspects (CardioPeds) and of general quality of life (PedsQL), also evaluating, starting from 12 years, the possible presence of depressive symptoms (PHQ-9) or anxious symptoms (GAD-7). These tests were administered both to the children and the parents, with the addition for these last ones of the compilation of the PSI, to investigate the parental distress.

The research allowed to divide the sample of 500 patients into 6 predominant pathologies, in order to compare the quality of life of children in different diseases and to understand if a specific pathology is associated or not with a lower quality of life.

Results Through an adequate statistical analysis it was found that the quality of life perceived by the subjects included in the study is significantly better than that perceived by the parents about the same children and adolescents.

Conclusions This study was one of the first to investigate the quality of life in congenital and/or acquired pediatric heart disease. However, the results obtained require further studies, in order to deepen what has emerged.

GP35

STUDY OF THE STIFFNESS OF THE VASCULAR WALL IN CHILDREN FROM FAMILIES WITH A BURDENED HISTORY OF CARDIOVASCULAR DISEASES

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Medical emergency center in Kazan conducted targeted screening to select patients with ischemic strokes, myocardial infarctions, angina pectoris, widespread atherosclerosis of peripheral vessels, angiographically confirmed coronary atherosclerosis, ischemic heart disease and dyslipidemia. 230 patients were selected (men ≤55 years, women ≤60 years). Their children, grandchildren (144 people) (Group 1) aged 5–17 years were examined. Control group of conditionally healthy children (112 people) was formed (Group 2), their family history was collected with the aim of excluding heredity burdened by CVD. A biochemical blood test was performed to determine the lipid profile; thus, a group with dyslipidemia (Group 1a) was selected among children with burdened family history, which included 76 people (53%), lipidogram indicators (Group 1b) that were normal (68 people, 47%). Rigidity indices of main vessels were determined: PTT (pulse wave propagation time), PWVao (pulse wave propagation speed in the aorta), AIx (augmentation index), (dP/dt) max (max blood pressure rise rate). An ambulatory daily monitor, BPLab® software using Vasotens® technology was used.

Results In general, in Group 1, the rigidity indicators of the vascular wall were similar (average daily PTT 145.3 ± 15.9 ms, PWVao 6.7 ± 0.9 m/s, AIx $4.9 \pm 8.1\%$, (dP/dt) max

667.9 ± 119.8 mm Hg/s) with those of Group 2 (average daily PTT 140.5 ± 15.2 ms, PWVao 6.4 ± 1.1 m/s, AIx $5.1 \pm 8.2\%$, (dP/dt) max 668.1 ± 121.3 mm Hg/s) and were statistically significant ($p < 0.05$). However, it was found that if we consider non-maximum daily indicators, then in Group 1 they were significantly higher ($p < 0.05$) (max daily PTT 199.9 ± 10.2 ms, PWVao 16.3 ± 1.4 m/s, AIx $35.3 \pm 3.4\%$, (dP/dt) max 1098.1 ± 120.5 mm Hg/s) than in Group 2 (max daily PTT 181.7 ± 9.6 ms, PWVao 13.1 ± 1.9 m/s, AIx $28.3 \pm 1.4\%$, (dP/dt) max 1005.1 ± 110.3 mm Hg/s). Comparing the parameters between Group 1a and 1b, no statistically significant difference was found between the mean values, but statistically significant difference was found between the maximum values.

Conclusions Direct relationship has been revealed between the presence of a burdened family history of CVDs, changes in the lipid profile of parents, relatives of the 1st and 2nd lines, and changes in the rigidity of the wall of the great vessels in their children. The rigidity of the vascular wall was significantly higher in Group 1 than in Group 2, differed between children from Group 1a and Group 1b.

GP36

FEEDING DIFFICULTIES IN CHILDREN WHO UNDERWENT CARDIAC SURGERY FOR COMPLEX CONGENITAL HEART DISEASE IN KOSOVO

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Introduction A feeding disorder in infancy and during childhood is a complex condition involving different symptoms such as food refusal and faddiness, both leading to a decreased food intake. It often results from abnormal feeding development. Also, adequate nutrition is crucial and challenge in children after surgery for congenital heart disease. There is a worldwide reason for attention to lesion or specific feeding problems, supplementation of trace elements and minerals, and an organized approach to pace, timing, and type of feeding are beneficial.

Aim Of presentation is assessing the prevalence and predictors factors of feedings difficulties in children who underwent cardiac open heart surgery in neonatal period and infancy. We address selected nutritional and caloric requirements for children after cardiac surgery and explore nutritional interdependence with other system functions.

Method This was a retrospective study in a tertiary referral hospital, and prior approval from the institutional ethics committee was obtained. Information for 78 children (42 male and 36 female) was taken from patients charts. The presence of feeding difficulties or disorders was assessed by a questionnaire when the child was 3 years old. As a feeding disorder was defined as an inadequate food intake for age, failure of thrive or for few cases need for tube feeding. Data were analysed with descriptive statistics and logistic regression.

Results From cohort of analysed children feeding problems occurred in 23%. At the time of study, refusal to eat or poor appetite was reported as a significant problem in 19 children and subnormal height and/or weight were recorded in 11 children. Early neonatal intervention and reoperation were identified as a risk factors for latter feeding difficulties