anomalies, and a typical number of other phenotypes, including occurrence of cardiac fibroma. The syndrome is caused by microdeletion of the long arm of chromosome 9, in the region q22.3-q31 which includes the PTCH tumor suppressor gene. The diagnosis is made clinically, through large and small criteria that include the already mentioned clinical characteristics.

Case Report We will present the case of a 5 – year – old girl diagnosed with Gorlin – Goltz syndrome with the presence of fibroma in the left ventricular wall. The patient has an uneven psychomotor development and shows atypicalities in the field of socio-emotional functioning. Phenotypically, we find increased neurocranium, rough facial features, divergent strabismus, and a wider nasal root. The girl was initially hospitalized for the clinical presentation of heart failure: she was clinically tachydyspnoic, with audible crepitations in the lungs, enlarged liver, and pretibial edema. Echocardiographically, it was diagnosed with dilated cardiomyopathy, with an ejection fraction of 25%, while the formation of an unclear etiology was seen in the left ventricle. The patient was treated with anticongestive therapy with low molecular weight heparin and further treatment was performed. MRI of the heart showed a formation that according to radiological criteria corresponds to a large fibroma. Due to the opinion that dilated cardiomyopathy and fibroma with phenotypic characteristics could be parts of systemic disease, molecular karyotyping was performed which found microdeletion of the long arm of chromosome 9 in the q22.3 region, which includes the PTCH gene that regulates cell growth and functions as a tumor suppressor gene. Haploinsufficiency of this gene has been described as Gorlin – Goltz syndrome, which is characterized by phenotypic traits such as those found in our patient. Among patients with Gorlin-Goltz syndrome, 10% develop cardiac fibroma with the most common localization in the left ventricular cavity. Symptoms of cardiac fibroma depend on the size of the tumor, the involvement of the conduction system, and the possible existence of intracavitary obstruction. The patient we present has an extensive tumor located intramurally in the anterolateral wall of the left ventricle, measuring 5.8x4.8 cm. Measured values of cardiac pressures obtained by invasive cardiac treatment indicate impaired systolic and diastolic heart function and increased pulmonary pressure, as a result of dilated cardiomyopathy with clinical signs of heart failure.

Discussion The presence of dilated cardiomyopathy with markedly impaired systolic function may be secondary to the tumor or as primary disease, that is why genetic processing of cardiomyopathy is ongoing. Cardiac fibroma can be treated by surgical resection, but in a situation of severely impaired systolic function, transplantation treatment is more likely, with the risk of immunosuppression in a patient with a tumor suppressor gene disorder.

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FIRST CROATIAN GUIDELINES FOR DIAGNOSIS AND TREATMENT OF ARTERIAL HYPERTENSION IN CHILDREN AND ADOLESCENTS

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Hypertension in children and adolescents is a growing public health problem. The risk of developing hypertension in children is higher than ever before due to adverse circumstances such as obesity, excessive salt and processed food intake, reduced physical activity and stress. Hypertension is transmitted from childhood, and especially adolescence, into adulthood and thus increases the risk for cardiovascular disease in adulthood. As information regarding pediatric hypertension has changed over the last few decades, we have developed the first Croatian guidelines for the diagnosis and treatment of hypertension in children and adolescents based on recent literature and guidelines of international societies adapted to our circumstances. Fourteen doctors from different centers – members of the pediatric hypertension working group from the Croatian Society of Hypertension – participated in the development of our guidelines. We tried to cover various aspects of pediatric hypertension – epidemiology, prevention, primary and secondary hypertension, diagnosis, vascular phenotype factors, athlete hypertension, neonatal hypertension, treatment of hypertension and hypertensive urgency and emergency. With these recommendations, we tried to make it easier for doctors to cope with the issue of pediatric hypertension, as well as offer practical instructions on how to deal with such patients. These are all reasons why we believe that the guidelines, written by members of the Croatian Society of Pediatric Cardiology and Rheumatology, the Croatian Society of Pediatric Nephrology, the Croatian Society of School and University Medicine and the Croatian Society of Hypertension, will be useful in prevention, active search for hypertension in children and its treatment. The guidelines can be found on the website of the Croatian Society for Hypertension https://hdh.emed.hr/upload/hdh_smjernica/dokument_1581600283.pdf

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ALAGILLE SYNDROME IN INFANT WITH FALLOT TETRALOGY

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Introduction Alagille syndrome is an autosomal dominant disorder that primarily affects the liver and heart. The genetic basis for the development of the syndrome is a mutation in the JAG1 gene (in most cases) or the NOTCH2 receptor. The syndrome is clinically characterized by chronic cholestasis, congenital heart defect (most often pulmonary stenosis), abnormalities in the skeletal system ('butterfly' vertebrae), changes on the eyes (posterior embryotoxon) and characteristic phenotype. The prevalence of the syndrome is estimated at 1:30 000 live births.

Case Report We will present a male infant aged 10 months with a diagnosis of Alagille syndrome. The boy was born from his mother's first, regularly controlled pregnancy. The boy was vital at birth with mild neonatal jaundice. On the third day of life, a systolic murmur was observed over the precordium, louder along the left edge of the sternum, and a diagnosis of tetralogy of Fallot was made by echocardiography. During the next examination, at the age of one month, the classic form of tetralogy of Fallot was verified by ultrasound