CHD over the perioperative period by comparing them with genetically normal controls, matched for cardiac lesion.

Infants with DS and with CHD of atrioventricular septal defect (AVSD) and ventricular septal defect (VSD) were prospectively recruited to undergo three echocardiograms (pre-operatively, post-operatively within 24 hours and pre-discharge from hospital). Left (LV) and right (RV) ventricular function was measured using deformation analysis to derive LV global longitudinal strain (LV GLS) and RV free wall longitudinal strain (RV FwLS). LV end systolic wall stress (ESWS) and RV systolic pressure (RVSp) were also measured. A non-DS control group with VSD and AVSD were recruited for comparison. Data on cardiorespiratory parameters, complications, and length of stay were compared.

34 infants with DS were compared with 17 controls. LVGLS was significantly higher in the DS group at echo 1 and 3. LVGLS decreased in both groups post-operatively with neither group recovering to preoperative values pre-discharge. RV FwLS significantly decreased in both groups post-operatively with controls demonstrating better recovery by hospital discharge. ESWS was lower and RVSp was higher in the DS group throughout the study period. Post-operatively, infants with DS had higher vasoactive inotropic scores, longer duration of inotropic support and total duration of PICU stay. Chylothorax was seen exclusively in the DS population.

Infants with DS and CHD demonstrate increased LVGLS and lower ESWS peri-operatively. There was a lower recovery in RV function observed pre-discharge in DS infants with a higher RVSp. They also exhibit an increase disease burden post-operatively. This may reflect the autonomic dysfunction described in infants with DS who have lower systemic but higher pulmonary vascular resistance, although this requires further evaluation.

179

EVALUATION OF THE OF LIPID PEROXIDATION REACTIONS AND REGIONAL BLOOD FLOW OF PERIODONTAL TISSUES IN ADOLESCENTS WITH ARTERIAL HYPERTENSION AND PERIODONTAL DISEASES

Marina Darenskaya, Lyubov Rychkova*, Larisa Kolesnikova, Anna Pogodina, Lyudmila Grebenkina, Sergei Kolesnikov, Lyubov Kolesnikova. *Scientific Centre for Family Health and Human Reproduction Problems, Irkutsk, the Russian Federation*

10.1136/archdischild-2021-europaediatrics.179

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

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 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.

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.

180

THE ASSOCIATION OF PRENATALLY DIAGNOSED CARDIAC RHABDOMYOMA AND TUBEROUS SCLEROSIS COMPLEX

Ana Ćorić*, Tomislav Čaleta, Iva Vukšić, Petra Džepina, Dorotea Ninković, Andrea Dasović Buljević, Vesna Benjak, Boris Filipović-Grčić, Dalibor Šarić, Daniel Dilber, Nada Sindičić Dessardo, Ruža Grizelj. *Department of Pediatrics, University Hospital Centre Zagreb, University of Zagreb, School of Medicine*

10.1136/archdischild-2021-europaediatrics.180

Although rare, cardiac rhabdomyomas (CRs) are the most common fetal cardiac tumors. They may be the earliest manifestation of tuberous sclerosis complex (TSC). TSC is diagnosed in 75-80% cases of multiple fetal CRs and in 30% of single cases. We retrospectively reviewed the clinical outcome of fetal CR cases.

All cases of prenatally diagnosed rhabdomyoma in a single tertiary centre from 2009 to 2019 were ascertained from medical records.

We identified 14 fetuses with prenatally diagnosed CRs. Mean period of follow-up was 4 years (range 4m-9y). Two fetuses were twins (BC/BA), both affected by CR with family history of TSC, and one fetus of the dichorionic pair was not affected by rhabdomyoma. The mean GA at diagnosis was 29 weeks (range 21-35). There were eleven fetuses with multiple, and three with a solitary tumor. The right and left ventricle as well as the intraventricular septum were equally affected with a slight predominance of the left ventricle. The cardiac atrium was affected in two cases. The size ranged from small tumors of several mm in diameter up to dimensions of 20x20 mm.

Sixty percent were <20 mm in diameter. Two of fetuses were affected by hemodynamically relevant cardiac obstruction (LVOT), and in four arrhythmias (SVES, VES, WPW syndrome, and AV block) were observed. No hydrops fetalis or fetal perinatal demise were observed. In 11 (79%) TSC was confirmed clinically or by mutational analysis. After birth, most rhabdomyomas demonstrated a stable (14%, n=2) or spontaneous regressive growth pattern (57%, n=8). In four children (29%) tumors entirely disappeared. No progression of tumor size or number was observed. After birth in five (36%) children the conduction abnormalities were confirmed by ECG and responded well to the anti-arrhythmic medication and two children were affected by LVOT obstruction. Cerebral manifestations (tubers and/or nodules) were detected by brain MRI in 9 patients (3 prenatally). Subependymal giant cell

astrocytoma was diagnosed in 3 cases, 2 received treatment with mTOR inhibitor, and one child underwent VP shunt due to enlarging SEGA causing obstructive hydrocephalus. Six (55%) of the children with TSC suffered from epilepsy and psychomotoric development delay.

Although CRs are benign from the cardiovascular standpoint, and have a natural history of spontaneous regression, their close association with TSC prompt for early prenatal diagnosis and family counselling regarding the dismal long-term prognosis. Recent literature suggests that early therapy with mTOR inhibitors may prevent the development of TS manifestations.

181

THE ANTIARRHYTHMIC EFFICACY OF H1 – HISTAMINE RECEPTOR BLOCKER QUIFENADINE IN CHILDREN WITH FREQUENT EXTRASYSTOLES

Larisa Balykova*, Makarov Leonid, Soldatov Oleg, Soldatov Yuri, Shekina Nataliya. *National Research Ogarev Mordovia state University*

10.1136/archdischild-2021-europaediatrics.181

Goal 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 group (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.

182

TWENTY YEARS OF HYPOPLASTIC LEFT HEART SYNDROME IN CROATIA

Viktorija Ana Buljević*, Andrea Dasović Buljević, Dorotea Bartoniček, Dalibor Šarić, Daniel Dilber. *University Hospital Centre Zagreb*

10.1136/archdischild-2021-europaediatrics.182

Hypoplastic left heart syndrome (HLHS) is rare congenital heart defect in which the left side of the heart is severely underdeveloped. It has been a lethal congenital heart anomaly until the last four decades until three palliative surgeries were established (operation by Norwood, Glenn and Fontan). The aim of this study was to evaluate the outcomes of treating patients with HLHS.

The main methods were statistical, and the clinical characteristics were retrospectively reviewed.

We included 132 patients in 20-year period who have been treated at University Hospital Centre Zagreb, operated in our and in foreign centers.

We followed them before, in the meantime and after final operation. Of all patients, 69 survived and 63 died. The highest mortality was in period between operation by Norwood and Glenn in early infancy and accounts for almost 48% all lethal outcomes.

The most common anatomic variant is the mitral atresia and aortic atresia

(MA-AA) subtype and the rarest is mitral stenosis and aortic atresia (MS-AA) subtype.

Apart of a three – staged operation procedures, 53 patients required one or more interventions involving implantation of the stent into the pulmonary branches, isthmus the aorta, the Sano or mBT junction, and the dilatation of the same, then coiling of the arteriovenous malformations, electrostimulator implantation and the closure of fenestra. The most common interventions are stent implantation into the pulmonary branches and dilatation of the aortic recoarctation and stenosis of the pulmonary branch stent. In twelve patients, fenestra was closed with an Amplatzer occluder.

The mean follow-up age operation by Fontan (TCPC) is 7.64 years (1.1 –

16.5 years). Two patients were transferred to the GUCH population.

This retrospective study included 132 patients with hypoplastic left heart syndrome in twenty-year period who have been treated at University Hospital Centre Zagreb. The overall survival of all patients is 52.2%. The highest mortality was in period between operation by Norwood and Glenn in early infancy. The most common interventions are stent implantation into the pulmonary branches and dilatation of recoarctation of the aorta. Mean follow-up age operation by Fontan (TCPC) is 7.64 years.

183

CHILDREN WITH BRAIN DEATH – FINDINGS ON ECHOCARDIOGRAPHY

Vesna Herceg-Čavrak*, Sanja Pek, Ivančica Škarić, Višnja Tokić Pivac. *Children's Hospital Zagreb*

10.1136/archdischild-2021-europaediatrics.183

Brain death is frequently associated with systolic dysfunction. The actual mechanisms are not yet completely understood.

The goal of this study is to assess echocardiography findings among organ donors in our institution.

Methods we conducted a retrospective study for the period of 17 years (October 2001- December 2018). A total of 20 patients under 18 years with declared brain death were identified. The mean age was 8.8 years and 70% (14 patients) were male. One patient had a history of cardiac disease – ventricular septal defect- and his heart was not accepted for donation.