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.

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


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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 antiarrythmic 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 antiarrythmic effect in 10/12 (83%) of patient without significant QT prolongation or sinus node depression (probably due to quifenadine antiholinergic properties). The only side effect was thyroid dysfunction (8,3%) in this group.

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

**TWENTY YEARS OF HYPOPLASTIC LEFT HEART SYNDROME IN CROATIA**

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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.

**CHILDREN WITH BRAIN DEATH – FINDINGS ON ECHOCARDIOGRAPHY**

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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.
The adequate transthoracic echocardiogram was possible in 18 (90%) of potential organ donors. All examinations were performed before evaluation protocol confirmed brain death.

Results echocardiogram was normal in 13 (65%) patients. One had a moderate mitral and tricuspid insufficiency. Minimal pericardial effusion was present in 4 (20%) patients. Two had mild septal dyskinesia with normal left ventricular ejection fraction. Diffuse hypokinesis with ejection fraction of less than 55% was found in 5 (37%) patients, in one of which it was less than 45%. A total of 14 hearts (70%) were harvested for transplantation, including the one with the poorest systolic function.

Conclusion mild left ventricular systolic dysfunction occurs frequently in children with brain death, but these hearts can still be considered for transplantation.

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**LONG QT SYNDROME -A CASE REPORT**

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Timothy syndrome is a rare genetic disorder characterized by an abnormally prolonged cardiac ‘repolarization’ time (long QT interval). This predisposes individuals to arrhythmias, cardiac arrest and sudden death.

Other features associated with this syndrome are dysmorphic facial features, webbing of fingers and/or toes (syndactyly); congenital heart defects, developmental delays and autism.

We want to report a case of Timothy syndrome, incidentally detected post induction for orchidopexy.

**Background**

TS is an extremely rare genetic disorder of the L-type cardiac channel.

Cav1.2 encoded by CACNA1C. The syndrome is characterized by multisystem abnormalities consisting of QT prolongation, congenital heart defects, syndactyly, facial dysmorphism, and neurological symptoms.

**Case Report**

8yr old boy was admitted to hospital for elective left orchidopexy, during induction developed 2degree AV block with T Talternans, maintained reasonable cardiac output throughout, QTc 504 msec. Past medical history an episode of syncope needing hospitalization. Currently being evaluated for autism.

Physical exam was normal. Holter, showed QTc prolongation with T alternans.

Genetic testing showed he is positive gene mutation CACNA1C. Parents have been counselled for the need for implantable defibrillator. He has been given external automated defibrillator in the meantime.

Currently on nadolol 40 mg OD, parents are awaiting gene testing.

**Discussion**

Classic timothy syndrome (TS) is a rare genetic disorder with dysfunction in multiple organ systems, clinically characterized by long QT syndrome and syndactyly. Timothy syndrome was first described in 1992. Classic TS is caused by a single missense mutation G406R of exon 8A of the Cav1.2 L-type calcium channel gene (CACNA1C) and is inherited in an autosomal dominant fashion, although it usually is the result of a de novo mutation. Patients with TS are prone to life threatening ventricular arrhythmias as a consequence of prolonged QT interval.

Since the affected gene is widely expressed in multiple adult and foetal tissues including gastrointestinal system, brain, lungs, immune system and tests, extracardiac manifestations are common.

The risk for life-threatening ventricular tachyarrhythmia is the limiting factor of TS. Since ventricular tachyarrhythmia is the leading cause of death in patients with TS, effective anti-arrhythmic medication and an implantable cardioverter defibrillator are the mainstay of therapy Conclusions Timothy syndrome is a rare congenital arrhythmia disorder with dysfunction in multiple organ systems. Patients are at high risk for sudden death due to life threatening ventricular tachyarrhythmia. Implantation of an ICD at a very young age may be the best means to prevent sudden death.