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.

184 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 Talernans, 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 testis, 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.

185 CORONARY ANEURYSM – A REALITY IN KAWASAKI DISEASE ASSOCIATED TO COVID 19

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Introduction Kawasaki disease is an acute febrile vasculitis, primarily affecting children, that is epidemiologically and clinically closely linked to a viral infection (2). This entity can sometimes have severe forms. Numerous clinical studies published in the last two years have shown that infection with SARS-CoV2 virus (COVID 19) in children is frequently associated with this entity, which can sometimes be severe.

We will present the case of a 9-month-old infant, without personal history, who was hospitalized for fever.

The onset of the disease was 14 days earlier, with high fever and rash. He was hospitalized in another clinic, where the diagnosis of Covid 19-associated Kawasaki Disease was established (the criteria were as follows: fever for longer than 5 days, papulo-erythematous rash on the chest and limbs, changes in the oral and lip mucosa, palmo-plantar edema, bilateral conjunctivitis, Ig G anti SARS CoV2=4.52u/ml).

He received treatment with Gamma globulin 2g/kg in single dose, corticosteroid therapy, Aspirin.

The evolution was initially favorable. But after 7 days, high fever reappeared. Then, he is hospitalized in our clinic. At admission he had high fever, hyperemic lips, bilateral conjunctivitis, no manifestations of cardiac dysfunction, leukocytosis: 26,700/mm³, marked inflammatory syndrome (ESR=130 mm/h, C-reactive protein=8.8 mg/dl). Cardiac ultrasound detects an aneurysm of the coronary arteries (RCA=3.5mm, Z score=6, LCA=2.5mm, Z score=1.43).

The diagnosis of Kawasaki disease refractory to immunoglobulin treatment has been established, resuming administration in a dose of 400 mg/kg of body weight/day for 3 days.

Treatment was supplemented with intravenous methylprednisolone pulse therapy, aspirin and enoxaparin.(1) The evolution was favorable with the remission of clinical symptoms and inflammatory syndrome, but with the persistence of coronary dilation.

He was discharged from hospital with good general condition, normal inflammatory markers, changes in lipid profile (total cholesterol=306 mg/dl, total lipids=951 mg/dl,

triglycerides=172 mg/dl) and indications of treatment with Medrol 2 mg/kg with gradual reduction of dosage and Aspirin 4mg/kg.

After about 1 month he returns for revaluation, showing good general condition, upward weight curve, biological samples within normal limits.

Ultrasound cardiological reassessment and CT angiography further reveal significant aneurysmal dilatation of the left coronary artery. Anticoagulant treatment was completed with enoxaparin.

Conclusions KAWASAKI disease associated with COVID 19, may present an unfavorable outcome with lack of response to the initial immunoglobulin treatment and evolution to coronary aneurysm.

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CARDIAC INVOLVEMENT AND SHORT-TERM OUTCOMES OF SARS COV 2 INFECTED PATIENTS WITH PEDIATRIC MULTI-SYSTEMIC INFLAMMATORY SYNDROME AT AN EMERGENCY CLINICAL SETTING IN BUCHAREST

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Pediatric multi-systemic inflammatory syndrome (PIMS) has gained attention throughout the medical world due to the ongoing SARS COV 2 pandemic. Being a systemic inflammatory response, cardiovascular complications are no exception, thus awareness of such conditions must be raised, as well as of prompt and accurate diagnosis and treatment. Attention is particularly brought to PIMS due to the fact that symptoms of it overlap with Kawasaki disease and toxic shock syndrome. The aim of this paper is to present cardiac findings and short-term outcomes in children with PIMS admitted to one of the main pediatric emergency health care units in Romania.

This current paper draws data from a single center, and is an observational, prospective study. The number of patients that were included in this study is 26, with age range between 1 month and 17 years, hospitalized between December 2020 and April 2021 in the 'Grigore Alexandrescu' Emergency Hospital for Children, Bucharest. The main criterion for inclusion was PIMS as defined by CDC/WHO case definition. Out of all symptoms, persistent fever and gastrointestinal conditions were the most frequent ones (88% and 69%, respectively).

Cardiac involvement was found in 11 patients (42%), being represented by: left ventricular dysfunction (26%), coronary artery abnormalities (15%), atrioventricular valve regurgitation (30%), with only one patient showing pericardial effusion and one an ECG anomaly. Improvement of initial clinical symptoms was paralleled by alleviation of cardiac symptomatology, as well as normalization of cardiac and inflammatory laboratory findings.

Our study shows that cardiac involvement is frequent in pediatric population with systemic inflammatory syndrome and we believe that PIMS in SARS COV 2 infected patients should be thoroughly screened and treated by a multi-disciplinary team. 24 out of 26 patients were fully recovered and discharged without sequelae, but two patients with persistent coronary artery aneurysmal findings.

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KAWASAKI DISEASE TREATMENT EXPERIENCE IN UHC SESTRE MILOSRDNICE

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Introduction Kawasaki disease (KD) is one of the most common systemic vasculitis predominantly involving coronary arteries with unknown etiology. It is generally assumed as inflammation of blood vessels predominantly affecting the cardiovascular system and possibility of developing long-term complications (primarily aneurysms and coronary artery thrombosis). It most often occurs in infants and preschool children, especially boys with much higher incidence of complications in younger children, notably if the disease remains unrecognized and untreated. In the period from 2012 to 2019, we hospitalized and treated five children with Kawasaki disease, three girls and two boys aged four to seven years. The disease was diagnosed in all patients based on clinical criteria. Echocardiographical changes in the heart and coronary arteries were registered (primarily pericardial effusion, mitral regurgitation and perivascular hyperechogenicity in the coronary arteries of varying intensity, in one child ectasia of the initial part of the LCA was noted). After IVIG therapy, all patients became afebrile within 24 hours, with no long-term cardiovascular complications.

Case Report S.K., a 4-year-old girl was treated as an outpatient with peroral penicillin for suspected scarlet fever. On the fourth day of the disease abnormal urine testing was noted and cephalosporin antibiotic introduced into the therapy. On the fifth day of the illness, she was admitted to the Gastroenterology Section of our Department due to suspected pancreatitis with hepatic lesion and acholic stools. Abdominal ultrasound revealed gallbladder hydrops. The day after that conjunctival hyperemia was noted. Because of the suspected KD echocardiography was performed with mild mitral regurgitation, enlargement of the ostium LCA (+3 SD for body surface) and mild pericardial effusion registered.

A.S., a 5-years-old boy was admitted to the Respiratory Section due to generalized urticaria with periobital edema and bloated hands. Initially treated with antihistamines, he received a single dose of parenteral corticosteroid. With the appearance of scarlatiniform rash and raspberry tongue, scarlet fever was suspected and peroral penicillin introduced. While being unresponsive to antimicrobial therapy, and with the appearance of conjunctival hyperemia, swelling of the feet and elevated liver enzymes, Kawasaki's disease was suspected. Echocardiographically pericardial effusion and negative T waves in the ECG were found.

D.M., a 5-year-old boy. His disease began with high fever, sore throat, dysphagia, neck pain and neck stiffness. On the third day of the disease visible swelling on the right side of the neck (3x5cm) was noted with erythema of the feet, palms and axillae. He was hospitalized with aim to treat parapharyngeal abscess as a complication after unsuccessful penicillin therapy for Streptococcal pharyngitis and scarlet fever. After the initial work-up (ENT examination, MSCT of the neck), surgical removal of the lymph node was performed and ceftriaxone introduced. On the fourth day of the disease, edema