The syndrome was first described by American pediatrician-endocrinologist Di George Angelo Mario in 1965. In clinical practice this syndrome is associated with a primary immune deficiency with defective T-cell immunity in combination with hypo- or aplasia of the parathyroid glands, congenital heart disease and frequent occurrence of the face skull malformations. The delay of physical and psycho-motor development, cognitive impairment are observed in most patients.

The disease is caused by disembyogenesis of the 3-4 pharyngeal pouches, from which the parathyroid glands and thymus are formed. As neighboring organs are affected, the facial dysmorphism appears subsequently. The combined CHD - truncus arteriosus, tetralogy of Fallot's, coarctation of the aorta, ventrical septal defect are marked in 2/3 of children with Di Giorgi syndrome (DGS). Anomalies of the great vessels are common. [1, 2].

In 1981, for the first time it was suggested that there is a significant part of 22-nd chromosome deletions in the occurrence of Di George syndrome. With the introduction of the method of fluorescent hybridization (FISH in situ), this chromosomal abnormality is identified in more than 90% of patients with Di George syndrome. Cases with similar symptoms DGS have been described in the localization of the deletion of other chromosomes: 10p13, 17p13, 18q21. [3, 4, 5].

The rearrangement of the chromosome 22 is the most common chromosomal abnormality in children with congenital heart disease (CHD). [2, 3]. Early detection of microdeletions 22q11.2 in children with CHD is important, as these patients have an increased risk for the development of hypocalcemia, immunodeficiency, post-operative complications [6].

DGS is a failure, mainly of cellular immunity. This syndrome is caused by primary impairment proliferation and differentiation of B- and T-lymphocytes. When there is aplasia or hypoplasia of the thymus, defective T-cells appear, but there are no T-suppressors. The leveling of immunological disorders by the age of 5 is possible [1, 4]. However, the "full" Di George syndrome with severe abnormalities of the immune system is extremely rare.

The most persistent manifestations of the disease is hypothyroidism. The frequent debut DGS are tonicoclonic convulsions in the neonatal period as a result of hypocalcaemia. Long-term reduced blood calcium level can lead to the violation of child growth and development, pathology of the skeletal system, multiple dental caries. Symptomatic hypocalcemia greatly complicates the course of CHD and may cause secondary syndrome elongated interval QT, ventricular arrhythmias and sudden cardiac death [7, 8].

Due to the variety of clinical symptoms, the patients with DGS can be observed by doctors of different specialties.
**Own observation:** During one year we observed the child Sh., born in 2013, with the diagnosis: Di George syndrome. The patient was repeatedly admitted to cardio-pulmonary department for routine inspection due to multiple congenital malformations: CHD (pulmonary atresia 2 type with expressed pulmonary artery hypoplasia, subaortic ventricular septal defect (VSD). Large aorto-pulmonary collateral arteries.). Congenital lung anomaly - cystic hypoplasia of the left lung. Tracheal bronchus right. Subsegmentary atelectasis of upper right lobe.

It’s known from anamnesis that early neonatal period was characterized by a syndrom of heightened of nervous-reflectory irritability, frequent regurgitation, hemodynamic disorders. The congenital heart disease and blood vessels (VSD, pulmonary artery atresia) and subsegmentary atelectasis of the upper lobe of the right lung were diagnosed during the investigation. At the same time, hypocalcemia and potassium-sodium imbalance have been identified. The severe lymphopenia is marked during repeated hematological investigations (within 22-24%). The Di Georgie syndrome was suspected. The study of parathyroid hormone was carried out once and found no deviations from the normative values (PTH - 11.79 pg / ml). Attention was drawn to the absence of thymus visualization during the CT scan.

A child under the age of 4 months has been operated in the Medical Center of Pediatric Cardiology and Cardiac Surgery of Ukrainian Health Ministry, left-sided modified Bleloka anastomosis has been imposed. The second stage of the surgical correction of cardiovascular malformations was anastomoses from the descending aorta to ensure adequate blood supply to the right lung at the age of 9 months.

During repeated haematology tests of pre- and post-operative stages, the expressed lymphopenia was constantly detected. It made us think about immune deficiency syndrome in a patient again. Minimum values of absolute and relative number of lymphocytes in peripheral blood were observed in the age of 4.5 months and up to 16% (1,552 • 10^3 /mcL). The immunological study in 6 months age revealed the violations of T-cell and phagocytosis. The decrease of the relative number of lymphocytes, absolute values of B-lymphocytes (CD19), NCT-test, phagocytic activity and phagocytic number was noted. Absolute figures of T-suppressor (CD8) were at the lower limit of normal. We conducted a study of molecular cytogenetic method of FISH-analysis. The karyotype nuc ish (TUPLE1x1) [39/100], (ARSAx2) [100] was detected. The cells with deletion 22q11.2 were observed in 39% of cases, which is typical for the Di Giorgio syndrome.

Thus, clinical criteria and the results of the research allowed us to think about the presence of incomplete syndrome Di Giorgi (syndrome of deletion 22q11.2 chromosome) in the patient.
This case demonstrates the difficulty of diagnosing of incomplete syndrome Di Giorgi with moderate immunological disorders and transient hypocalcemia when CHD goes to the foreground of the clinical picture. It is necessary to draw the attention of doctors and parents of patients to a comprehensive monitoring of a child by pediatrician, immunologist, endocrinologist and a cardiologist with carrying out ultrasound thymus, monitoring of immunological parameters, levels of PTH, calcium and phosphorus.

Patient management tactics should be focused not only on a surgical correction of CHD, but also on the prevention of recurrent infections and hypocalcemia, which prevents the complicated course of a disease and determine a favorable prognosis.