

of leukemia treatment. Practice shows that one of the organs affected by chemotherapy is the liver, and its damage directly depends on the toxicity and duration of chemotherapy. Our task was to conduct research work in this area, and to study toxic liver lesions in patients with leukemia. Before that, we carried out similar work at the stages of induction and consolidation of treatment of acute leukemia in children. And this period of research work is devoted to the supportive stage of therapy in patients. Objective: To study the frequency of toxic liver damage in children with acute leukemia during support therapy, to choose treatment tactics according to the severity of toxic hepatitis. **Methodology:** The study group included 51 children with primary acute lymphoblastic leukemia who completed the induction stage with complete remission and retained this result for the entire period of consolidation. The age of the patients ranged from 2.5 years to 15 years. Of these, there were 28 boys and 23 girls. The patients were from Baku and the regions of the republic. Treatment of acute lymphoblastic leukemia was carried out according to two branches of the Moscow – Berlin – 2015 program: B and T ÌmRG. The protocols of maintenance therapy of these branches do not differ, and both begin with the 31st week of the general program, end on the 104th. Each protocol consists of 8 stages of a combination of chemotherapy drugs Metotreksat + 6-Mercaptopurine, which last for 6 weeks and alternate with two-week courses of reinduction - Deksametazon + Vinkristin. Before the start of maintenance therapy, all patients with leukemia confirmed the preservation of the previously achieved remission, and the functional and organic state of the liver. With positive results, the continuation of leukemia treatment began. And when the symptoms of toxic hepatitis were detected, the severity was determined. According to this indicator, 3 forms of flow were issued: light, medium-heavy and heavy forms. The tactics of conducting therapy of each form were chosen by us. **Results:** Of 51 patients, 42 had toxic hepatitis (82%). It was mild in 12 patients (23.5%), moderate in 26 children (50.9%), and severe in 4 children (7.8%). In the mild form of hepatitis, patients were prescribed intravenous administration of Riboksin + Aevit (orally) for 10-14 days, or alternatively, per os Ursobil + Aevit. This combination made it possible to restore all clinical and laboratory parameters in patients within 14-21 days, and at the same time, without interrupting chemotherapy. Moderate and severe forms of hepatitis occurred mainly during the period of reinduction (54.7%). The administration of intravenous ademetionine (Heptral) in the form of monotherapy for 8-12 days allowed continuous reinduction courses. Following him, the administration of an oral combination of Ursobil + Lipoic acid + Aevit for 14-21 days allowed to preserve the long-term effect. In severe hepatitis, chemotherapy was suspended, and patients were prescribed intravenous ademetionine (Heptral) in combination with oral Ursobil + Aevit for 10-14 days, and along with this detoxification therapy was carried out in parallel. Such treatment gave an improvement in clinical and laboratory parameters. Subsequently, intensive therapy was suspended, and chemotherapy was started accompanied by

Ursobil + Aevit + Lipoic acid for the next two weeks. This choice of therapy allowed us to preserve the restored indicators for a long time. There were no deaths or severe complications from toxic hepatitis in any case.

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#### HEMOGLOBINOPATHIES (SICKLE CELL DISEASE, THALASSEMIA ETC...)

PP 30

#### RARE UNSTABLE ALPHA GLOBIN VARIANT HB TAYBE (HBA1:C.118\_120DELACC) WITH HBA2 POLY A MUTATIONS, CAUSES TO HEMOLYTIC ANEMIA IN TWO CASES FROM AZERBAIJAN

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**Objective:** Sequence variants are usually silent and rarer in  $\alpha$ -globin, some may lead to an unstable protein with hemolytic or thalassemic phenotype. The most common HBA variant is ConstantSpring which is leading to an unstable elongated protein chain. Others may be due to ins/del in the  $\alpha$ -globin, one of them Hb-Taybe caused deletion of Thr residue at codon 40 of the HBA1. We report, for the first time, 2 cases with hemolytic anemia due to the presence of Hb-Taybe in *trans* with HBA2 poly-A mutations. **Methodology:** Patients were managed in the Thalassemia Unit of the National Hematology Center. Detailed pedigrees are drawn, medical recordings are reviewed and peripheral blood samples are collected. Sanger sequencing was performed with *in house* designed primers (HBA1, NM\_000558.5 and HBA2, NM\_000517.6) on genome analyser (ABI3500). Deletion and duplication analysis is performed by MLPA. **Results:** P-1: A 4-yr-old male who was diagnosed at the age of 1 yr with congenital hemolytic anemia. He presented with jaundice, the blood film showed moderate hypochromia and anisopoikilocytosis. He was transfusion dependent. P-2: A 45-yr-old female who was diagnosed at the age of 5 yr with congenital hemolytic anemia. She underwent splenectomy at the age of 32 yr due to moderate anemia. After splenectomy she was not transfusion dependent. Relevant clinical and laboratory data is presented at table. **Conclusion:** The clinical presentation is variable from mild hemolytic anemia to regular transfusion requirement. One patient had to have splenectomy in order to ameliorate the transfusion requirement. This study supports the requirement of  $\alpha$ -globin gene analyses, and a careful evaluation of cases with hemolytic anemia, particularly in populations where thalassemias are endemic, in order to avoid missing any of the rare globin variants and to offer accurate genetic counseling.

	P-1	P-2
Age at diagnosis (yr)	1	5
Consanguinity of parents	-	-
Spleen size below the left costal margin (cm)	3	underwent splenectomy
Cholelithiasis	+	-
Hb (g/dL)	6.1	8.4
RBC ( $10^6/\mu\text{l}$ )	2.93	3.01
MCV (fL)	76.8	108
MCH (pg)	20.8	27.9
Hb A2 (%)	1.7	1.7
Hb F (%)	6	0.1
Serum Iron ( $\mu\text{g/dL}$ )	113	149.9
Serum Ferritin (ng/ml)	115.8	1623
LDH (units/L)	-	554
Total bilirubin (mg/dl)	1.91	2.93
Direct Bilirubin (mg/dl)	0.62	0.76

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#### PP 31

#### HBH DISEASE AND SYSTEMIC LUPUS ERYTHEMATOSUS

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**Objective:** The association between thalassemia and systemic lupus erythematosus (SLE) is very rare. There are many articles in the literature showing that patients diagnosed with SLE with Beta-Thalassemia have a more severe hemolytic picture. The combination of Alpha thalassemia and SLE was first reported in an article published on January 30, 2021, by the staff of Guangzhou Hospital in the People's Republic of China. Our report is about combination of HbH disease and SLE, too. **Case report:** A 31-year-old female patient with HbH disease who had been irregularly monitored by a hematologist for 12 years received a blood transfusion for the first time during her 4th pregnancy and has not seen a hematologist since. At 12 weeks of gestation (7th pregnancy), a severe hemolytic anemic clinic was observed and erythrocyte mass transfusion was initiated. However, as different types of allergic reactions were observed during and after hemotransfusions autoimmune tests were held. **Methodology:** As a result, Direct Antiglobulin Test (DAT), Anti Nuclear Antibody (ANA), and anti-dsDNA positive, complement C3 levels were found below standard. The diagnosis of SLE was confirmed based on the fact that the patient's previous 6 pregnancies resulted in miscarriages and stillbirth. At a later stage, as a result of detailed instrumental and laboratory examinations, she was diagnosed with Lupus nephritis and steroid treatment was started under the control of a nephrologist. **Results:** Unit erythrocyte mass was transfused during cholecystectomy in this patient who was taken to the hospital with seizure pain in the right subcostal area that suddenly began at 22 weeks of gestation. 24-week pregnancy was ceased due to

intrauterine growth retardation. In the next month of follow-up, during the hospitalization 7 units of washed erythrocyte mass were transfused to the patient who was brought to the hospital with severe anemia after positive Covid-19 PCR analysis. **Conclusion:** In case published about the first patient with HbH disease and SLE it was reported an increase in the severity of anemia and the maintenance of Hb value in the range of 9.0-10.0 g / dl with steroid. According to our researches there were found similarities between the outcomes of these two studies. Studies suggest that SLE patients with severe hemolytic clinics in regions with a high prevalence of thalassemia should be investigated for hemoglobinopathies.

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#### TRANSFUSION MEDICINE / APHERESIS / CELL PROCESSING

#### PP 32

#### EVALUATION OF CLINICAL AND LABORATORY FINDINGS OF THERAPEUTIC PLASMAPHERESIS IN CHILDREN

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**Objective:** Therapeutic plasmapheresis is an extracorporeal treatment method. The abnormal component of the patient's plasma is removed from the blood and replaced with the remaining blood components with a selected replacement fluid. We aimed to evaluate the demographic characteristics, procedure indications, procedure methods, differences between pre- and post-procedure laboratory parameters, and procedure-related complications of pediatric patients who underwent therapeutic plasma exchange (TPE). **Methodology:** Pediatric patients who underwent therapeutic plasmapheresis in Adana City Training and Research Hospital between 2018-2021 were included in our study. In this period, the number of pediatric patients who underwent therapeutic plasmapheresis was 61, and the total number of procedures was 238. The data of the patients were obtained from the files of the apheresis unit and the hospital registry system by retrospective analysis. Statistical analysis of the study was made with the SPSS v20 program. **Results:** 25 patients were female, 36 patients were male. Youngest patient was 6 months old and eldest was 17 years old. Patients weight range was between 5 and 104 kilograms. 191 of the procedures were TPE, 47 of them were lipid apheresis. The most common indications were hepatic failure, familial hyperlipidemia, neurological disorders, hematological disorders, sepsis with MODS and