

the administration of the drug, 33% of the patients did not go to the hospital because they were afraid of the pandemic, 33% of them could not get treatment even though they went to the hospital, and 33% of them other reasons were reported. While 48% of the patients want an experienced health personnel to go to their home to perform their treatment, 52% do not want it, stating that they do not need it. None of the patients whose treatment was interrupted did not complain of bleeding during this period. **Conclusion:** It was seen that the patients experienced disruptions related to access to medication and treatment during the pandemic process. However, there were no major problems in this process, thanks to the help of their physicians and other institutions. It is important to emphasize the importance of treatment in hemophilia patients and to have easy communication with the center followed in order to overcome the pandemic process without complications.

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## LEUKEMIA

### OP 18

#### EVALUATION OF MRD-STATUS IN POST-INDUCTION PERIOD IN PEDIATRIC PATIENTS WITH ACUTE LYMPHOBLASTIC LEUKEMIA.

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**Objective:** The need to study the significance of minimal residual disease (MRD) at the induction therapy in patients with acute lymphoblastic leukemia (ALL) is beyond doubt. This has been confirmed by many years of work by many research groups. The role of MRD in the late stages of treatment and the impact of these values on patients survival requires research and discussion. **Aim:** To evaluate the influence of MRD-status in post-induction period on survival in patients with acute lymphoblastic leukemia. **Methodology:** From 2010 to 2022, 135 patients with primary B-ALL enrolled in ALL-IC BFM 2009 protocol. Median age was 5.4 year (range 1-17). Male was 62 (49,5%) and female 73 (54,1%). The diagnosis was based on WHO 2016 criteria. Stratification on prognostic risk groups was carried out according to protocol criteria. Prednisone response evaluated at day 8 of treatment. The 15th, 33th, and 78th (as post-induction) day response was assessed by bone marrow cytology and level of MRD by flow cytometry. **Results:** 5y-overall survival (OS) for patients with MRD-negative status on day 15 was 94,4±5,4% and 87,0±3,4% for MRD-positive (p=0,5). On day 33 patients with MRD-negative status achieved 5y-OS in 86,7±5,8% and 89,6±3,5% for MRD-positive (p=0,6). 5y-OS for patients with MRD-negative status on day 78 was 90.8±4.0%, MRD-positive - 90,4±6.5%. DFS for MRD-negative status was 88.5±4.5%, for MRD-positive - 66.3±11.8% (p=0,1). EFS for MRD-negative patients was 87.2±4,6% and for MRD-positive 66.3±11.8% (p=0,09). **Conclusion:**

We have found a tendency between MRD status on day 78 and the frequency of relapses in patients. At the moment, there are no reliable data on the effect of post-induction MRD status on survival. The assessment of MRD in the post-induction period has prognostic prospects and requires further study.

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## INHERITED BONE MARROW FAILURE DISEASES

### OP 19

#### GHOSAL HEMATODIAPHYSEAL DYSPLASIA (GHDD) DIAGNOSIS AND TREATMENT: CASE REPORT

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**Objective:** Ghosal hematodiaphyseal dysplasia syndrome (GHDD) is a rare autosomal recessive disorder characterized by increased bone density and regenerative corticosteroid-sensitive anemia. We describe GHDD in an 11-year old Azerbaijani boy with refractory anemia, mild thrombocytopenia and radiological metadiaphyseal dysplasia. The diagnosis was made based on clinical and laboratory examinations and genetic analysis. We have observed a significant improvement of anemia after administration of steroids. **Case report:** An 11-year-old boy with long-standing anemia, complained of fatigue, delayed physical development, and limited range of motion in the joint. Physical examination did not reveal LAP and hepatosplenomegaly. Among the dysmorphic craniofacial changes mentioned in the literature, has a tower-shaped skull, micrognathia, drooping ears, a long and wide philtrum, and a thin upper lip. Skeletal X-ray imaging showed fibrotic changes and varying degrees of osteopenia in the metaphysis of the long tubular bones. **Methodology:** The blood count: Hb 7.0 g/dl, HCT 24.5%, reticulocytes 5.6%, MCV 78fL, MCHC 28.6 g/dl, WBC count 6860/mm<sup>3</sup>, platelets 165000/mm<sup>3</sup>, ESR 75 mm/h, anisocytosis in erythrocytes and platelets were observed in a peripheral blood smear. Hemoglobin electrophoresis, iron studies, vitamin B12 and folic acid were normal. Coombs test was negative. Bone marrow examination showed hypoplasia in erythroid and megakaryocytic series and dysgranulocytopenia. **Results:** After detection of exon 12 ((p. Gly473Trp), rs149988492, CM215867) in the genetic panel analysis of anemia, steroid treatment at a dose of 1 mg/kg/day was started and anemia improved at 1-month follow-up (Hb level 6.8 g/dL to 11.9 g/dL), but mild thrombocytopenia was noted to persist. The clinically insignificant CRP elevation normalized during the treatment. **Conclusion:** GHDD should be

considered in patients with clinical and radiographic evidence of diaphyseal dysplasia as well as hematological abnormalities. In addition, bone dysplasia should be investigated in treatment-resistant hematological pathologies of unknown origin. Although GHDD is rare, clinicians should be informed that it responds well to steroid therapy.

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

OP 20

### COMPARISON OF THE QUALITY OF LIFE OF PATIENTS WITH A BETA-THALASSEMIA MAJOR, REGULARLY RECEIVING PARENTERAL AND ORAL CHELATORS

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**Objective:** Patients with  $\beta$ -thalassemia major ( $M\beta$ -th) are transfusion-dependent, which affects their quality of life. To maintain a safe level of iron in the body, patients with  $M\beta$ -th require adequate regular therapy with chelation drugs (CP). Currently, for the correction of iron overload in patients with  $M\beta$ -th, along with oral CP, parenteral CP continues to be used. However, oral and parenteral CP are perceived by patients ambiguously. Comparative assessment of the quality of life of transfusion-dependent children with  $M\beta$ -th receiving various CPs: parenteral deferoxamine and oral deferasirox. **Methodology:** For 2 years, a survey and clinical observation of 201 children with  $M\beta$ -th aged 2 to 18 years (boys 128, girls 73) was conducted. The control group consisted of apparently healthy children from preschool and school institutions ( $n=30$ ). Patients with  $M\beta$ -th underwent a quality of life study (PedsQL- Pediatrics Quality of Life Inventory, Generic Core Scales and PedsQLTM4.0) and a psychological examination. The survey was conducted after obtaining the informed consent of the parents of older children at the beginning and at the end of the study. Once a month, the necessary clinical and biochemical analyzes were carried out. Patients with  $M\beta$ -th regularly prescribed various CP regimens: deferoxamine subcutaneously; deferasirox, orally. **Results:** All studied patients with  $M\beta$ -th were divided into four age groups: group 1 - children under 4 years old according to parents ( $n=41$ ); group 2 - children 5-7 years old according to the assessment of children and parents separately ( $n=62$ ); group 3 - children 8-12 years old according to the assessment of children and parents separately ( $n=47$ ); Group 4 - children aged 13-18 years old according to the assessment of children and parents separately ( $n=51$ ). Each of the 4 groups of  $M\beta$ -th patients was divided into a subgroup taking only deferiprone and a subgroup taking only deferasirox. **Conclusion:** According to the Results of the survey, the indicators of the quality of life and the psychological state of children with  $M\beta$ -th receiving

parenteral and oral CP differed. So, in sick children with  $M\beta$ -th of different age groups, when taking parenteral CP in comparison with those taking oral CP, the quality of life was reduced, and the psychological state worsened significantly. This was especially impacted patients in the group of 8-13 years. In this group, there were more complex relationships with peers, parents, there was an increase in anxiety and aggressiveness, which is associated with the need for hours of use of the pump for subcutaneous injection of the drug, the presence of pathology that limits the use of oral chelators. In children of 4 different age groups, there is a significant difference in the values given by patients and their parents to the quality of life in patients receiving parenteral and enteral chelator therapy.

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## LYMPHOMAS

OP 21

### LABORATORY AND CLINICAL FEATURES OF TUMOR LYSIS SYNDROME IN CHILDREN WITH HIGH-GRADE NON-HODGKIN LYMPHOMA AND EVALUATION LONG-TERM RENAL FUNCTIONS IN SURVIVORS

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**Objective:** Tumor lysis syndrome (TLS) describes biochemical and clinical abnormalities resulting from spontaneous or treatment-induced necrosis of rapidly proliferating tumors such as Burkitt's lymphoma (BL). TLS can lead to complications like acute kidney injury (AKI) which can be fatal. In patients who had AKI in childhood, the frequency of kidney problems increases in later ages. Therefore, there is a need to examine long term kidney functions in patients with TLS. The purpose of our study is to investigate the laboratory and clinical features of tumor lysis syndrome in childhood non-Hodgkin lymphomas (NHL) and to reveal its impact on long term kidney function in survivors. **Methodology:** Our study was a single center retrospective study. 107 patients (0-18 years of age) admitted to our hospital between 1998-2020 years with a diagnosis of NHL and who received chemotherapy were included in the study. Clinical and laboratory characteristics of the patients at the time of diagnosis and within 14 days from the start of chemotherapy were examined. The presence of TLS and its laboratory and clinical features were examined according to the Cairo-Bishop criteria. The relationship between TLS and age, gender, histopathological subgroup, tumor stage, lactate dehydrogenase (LDH) level at presentation, bone marrow and kidney involvement were investigated. The presence of AKI was determined according to the Kidney Disease: Improving Global Outcomes criteria.