HEMATOLOGY, TRANSFUSION AND CELL THERAPY

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ORAL PRESENTATIONS

Adult Hematology Abstract Categories

Chronic Myeloproliferative Diseases
OP 01

THE RELATIONSHIP BETWEEN POLICYTEMIA VERA AND METABOLIC SYNDROME: THE SINGLE CENTER EXPERIENCE

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Objective: Polycythemia vera (PV) is the most common myeloproliferative neoplasm. It is known that while the amount of substances such as malonyl-dialdehyde, which are known as oxidative stress markers, increases in PV and metabolic syndrome (MS), antioxidant molecules decrease. There are very few studies investigating the clinical relationship between PV and MS. In our study, we determined the incidence of MS in patients diagnosed with PV in our center and investigated the relationship between MS and PV. **Methodology:** Forty patients with PV were included in the study. The study included non-smoker patients over the age of 18 who were followed up in our center and diagnosed with polycythemia vera according to the diagnostic criteria specified by the World Health Organization in 2016, by examining bone marrow aspiration biopsy and JAK mutation. The diagnosis of metabolic syndrome was made according to the criteria set by the International Diabetes Association. **Results:** Of the 40 patients included in the study, 23 (57.5%) were diagnosed with MS. Gender, age, HbA1c, fasting blood glucose, hemoglobin, ferritin, triglyceride, HDL, systolic and diastolic blood pressures, waist circumference measurements of PV patients with MS were compared with PV patients without MS. HbA1c, glucose, Triglyceride, blood pressure, values showed a statistically significant difference between the groups diagnosed with MS and PV. **Conclusion:** The incidence of MS in our country is 32.9%. In our study, the incidence of MS in patients with PV was found to be higher than the Türkiye average. Oxidative stress seems to be important in the etiology of the two diseases, so our study shows that it is important for the clinician to be careful in patients diagnosed with PV and MS. Although there seems to be a relationship between PV and MS in our study, the data need to be confirmed by studies with a higher number of patients.

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Adult Hematology Abstract Categories

Lymphoma
OP 02

THE OUTCOME OF PERIPHERAL T-CELL LYMPHOMA PATIENTS FAILING FIRST-LINE THERAPY, FROM PROSPECTIVE COHORT OF T-CELL BRAZIL PROJECT

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Objective: In Brazil, the National Institute of Cancer estimates for the years 2023-2025 about 12,040 new cases of NHL, about 1,444 of peripheral T-cell lymphomas (PTCLs). T-cell Brazil project is an ambispective study inserting new diagnosis from January 2015 to December 2022. Our goal was to explore a prospective cohort (PC), April 2017-December 2022, analyzing primary refractory and relapse (R/R) PTCLs pts to explore bad factors for overall survival (OS). **Methodology:** PC enrolled 461 pts who received 1ª treatment line. Descriptive analyses, Kaplan-Meier method, Log-Rank test to compare groups and Cox Regression to identify risk factor for OS using IBM-SPSS software v.24. **Results:** It was identified 171 (37%) pts, 71% refractory and 29% relapsed. Median mo. from treatment to R/R was 6 mo. (1-49). Overall, 42% received 2nd line treatment and these 11% had to bone marrow transplantation. After a median 17 months (0-51) of follow up, 64% pts had died, and 74% due to lymphoma, 17% infections, 9% toxicities. Refractory pts (HR=2.51, $P<0.0001$), IPI=2-4 (HR=3.19, $P<0.0001$) and >1 extranodal site (HR=1.76, $P=0.01$) were associated with a higher risk of death in a Cox Regression. **Conclusion:** This study confirms outcomes for patients treated according to standards treatment. No difference was found in OS with respect to histology. Results confirm that peripheral T-cell lymphomas patients had dismal outcome after relapse or progression, besides of higher IPI and more than one extranodal site at diagnosis. However, HCT as salvage can possibly prolong life as some studies already indicated.

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OP 03

IBRUTİNİB-OBİNUTUZUMAB COMBİNATİON THERAPY İN THE TREATMENT OF RELAPSED NODAL MARGİNAL ZONE LYMPHOMA: A CASE STUDY

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Background: Marginal Zone Lymphoma (MZL) is a type of non-Hodgkin lymphoma (NHL) originating from B-lymphocytes. It is characterized as a slow-growing or indolent lymphoma and is considered a rare disease. The report focuses on a case of MZL diagnosed in childhood, which relapsed after initial treatment and subsequently went into remission following ibrutinib-obinutuzumab treatment. **Case Report:** In 2010, a 9-year-old girl with no previously known systemic illnesses was diagnosed with stage 4B nodal marginal zone lymphoma outside a pediatric center. Initially, she achieved remission following treatment with rituximab-bendamustine but experienced a relapse in 2012. Subsequent to lymph node excision and Methotrexate, Ifosfamide, Etoposide, and Dexamethasone (MIED) therapy, all conducted outside the pediatric center, she received an autologous stem cell transplant in 2013. Five years after the transplantation, she applied to our center when she was 18 years old, exhibiting widespread lymphadenopathy and suffering a relapse of stage 4B nodal MZL. Treatment with ibrutinib-obinutuzumab was commenced, leading to a full response after six cycles, without any adverse effects. Maintenance therapy with ibrutinib was initiated to avert further recurrence. **Conclusion:** The treatment of relapsed nodal MZL continues to be challenging. In patients who have previously received repeated cytotoxic chemotherapy, the combination of ibrutinib-obinutuzumab may be an effective and safe option to avoid cumulative toxicity of chemotherapy. Further studies with more cases in R/R nodal MZL will contribute to the management of the disease.

Keywords:
Marginal Zone Lymphoma (MZL)
Non-Hodgkin lymphoma (NHL),Ibrutinib-Obinutuzumab
Relapsed Nodal MZL
Lymphadenopathy

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Adult Hematology Abstract Categories

Myeloma

OP 04

ISATUXIMAB PLUS CARFILZOMIB AND DEXAMETHASONE VERSUS CARFILZOMIB AND DEXAMETHASONE IN PATIENTS WITH RELAPSED MULTIPLE MYELOMA (IKEMA): FINAL OVERALL SURVIVAL ANALYSIS

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Objective: Isatuximab (Isa, anti-CD38 monoclonal antibody) is approved in combination with carfilzomib (K) and dexamethasone (d), for relapsed multiple myeloma (MM) patients (pts) after ≥1 prior therapy. Final progression free survival (PFS) analysis after 2 years showed mPFS of 35.65 mo (Isa-Kd) vs 19.15 mo (Kd). Here, we report the final overall survival (OS) from IKEMA planned 3 years after the primary PFS analysis. **Methodology:** Pts with 1–3 prior lines of therapy were randomized 3:2 to receive Isa-Kd (n=179) or Kd (n=123). Treatment (tx) was given until progressive disease, unacceptable toxicity, or pt wish. Safety was assessed in all treated pts. **Results:** As of 7 Feb 2023, 23.5% (Isa-Kd) and 5.7% (Kd) pts were on tx. Median follow-up: 56.61 mo. OS benefit was more in Isa-Kd pts (mOS was NR; [95% CI: 52.172–NR] vs 50.6 mo [95% CI: 38.932–NR]; HR: 0.855; nominal one-sided p=0.1836). Isa-Kd had longer TTNT vs Kd (median 43.99 vs 25.0 mo; nominal one-sided p=0.0002), as was PFS2 (median 47.18 vs 32.36 mo; nominal one-sided p=0.0035). The safety profiles were comparable to interim and final PFS analyses. Grade ≥3 TEAEs: 84.2% (Isa-Kd) vs 73.0% (Kd). **Conclusion:** This final OS analysis shows a meaningful trend for OS benefit with Isa-Kd vs Kd despite subsequent tx with anti-CD38 agents, introduction of tx with novel mechanism of action among further therapies, and the COVID-19 pandemic. Improvements in TTNT and PSF2 were observed and sustained PFS benefit still observed at PFS2. The Isa-Kd safety profile was consistent with previous analyses, supporting it as a standard-of-care therapy for relapsed MM pts.

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OP 05

REAL-WORLD (RW) TREATMENT PATTERNS AND OUTCOMES IN PATIENTS (PTS) WITH RELAPSED/REFRACTORY MULTIPLE MYELOMA (RRMM) WITH AT LEAST ONE PRIOR THERAPY IN TURKEY

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Objective: Data on RW treatment patterns and outcomes in RRMM Pts who received at least one prior line of therapy (LoT) are lacking outside the US and Europe. This study evaluated RW clinical characteristics, treatment patterns, and outcomes among Turkish Pts who received at least one prior MM-specific therapy. **Methodology:** This retrospective chart review included RRMM Pts who had received at least one prior LoT and initiated a second-line (2L) or third-line (3L) MM-specific treatment regimen between 01-Jan-2015 and 31-Dec-2020. Patients' demographics and clinical characteristics, treatment patterns, and overall survival (OS) were evaluated. **Results:** Of the 107 RRMM Pts initiating 2L treatment, 91.6% experienced symptomatic disease [prominent symptoms: anemia (71.0%); bone lesions (53.3%)]. Table 1 presents other clinical and demographic characteristics. Bortezomib (BOR)-based regimens were most used in first-line (1L) regardless of stem-cell transplant (SCT) status (SCT induction: 68.7%; non-SCT: 79.5%), and lenalidomide (LEN-based regimens were used as 1L maintenance (40.3%). LEN-free regimens were used in 58.1% (2L) and 35.6% (3L) of Pts, with DVd (29.5%) and DRd (19.5%) being the most utilized regimens in 2L and 3L, respectively (Fig. 1). In total, 53.1% were LEN-retreated and 30.8% were LEN-refractory. The median (interquartile range) duration of treatment on 2L [7.0 (6.0, 10.5) months] and 3L [7.1 (6.0, 14.0) months] was short (Table 2). After 2L and 3L initiation, 57.9% and 25.6% of Pts had disease progression; median OS was 10.4 and 12.8 months, respectively (Table 3). **Conclusion:** BOR-based regimens were commonly utilized in 1L. LEN-based regimens were used as maintenance therapy in 1L and as retreatment in RRMM Pts. Newer therapies (Daratumumab- or Carfilzomib-based regimens) were utilized in 2L and 3L. The short duration of therapy, high disease progression rate, high LEN retreatment, and refractoriness rates indicate the need for new LEN-free regimens for treating RRMM Pts in Turkey.

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OP 06

URETERAL AMYLOİDOSİS: A CASE REPORT OF SUCCESSFUL MANAGEMENT WİTH SURGERY, RADİATİON, AND CHEMOTHERAPY

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Background: Ureteral amyloidosis is a unique and infrequent form of amyloidosis characterized by the deposition of amyloid proteins within the ureters. These tubes, responsible for transporting urine from the kidneys to the bladder, can become obstructed due to this protein accumulation, potentially leading to renal complications. We are presenting a case ureteral amyloidosis. **Case Report:** A 48-year-old male with no known prior medical conditions presented with a three-month history of right-sided pain, frequent and painful urination, reduced urine output, and hematuria. Blood tests showed a hemoglobin level of 12.8 g/dL and MCV of 73. Urinalysis revealed pyuria and hematuria. An upright abdominal X-ray indicated hydronephrosis, and an abdominal CT scan

detected grade 2 hydronephrosis on the right side and a suspicious mass in a 1 cm segment of the distal right ureter outside the bladder. The mass was excised, and histological examination with crystal violet and Congo red staining showed a strong positive reaction for amyloid. Immunohistochemical analysis confirmed the diagnosis of lambda light chain amyloidoma. Systemic screening for amyloid deposition was negative except for the ureter. Nine months post-operation, the patient returned with recurrent pain and oliguria. A CT scan revealed a mass at the excision site, consistent with lambda light chain amyloidoma. Considering it a recurrent disease, the patient underwent intensity-modulated radiation therapy (IMRT) with a total dose of 20 Gy in 10 fractions of 2 Gy each. Two months post-radiation, with recurring symptoms, the patient received four cycles of bortezomib-dexamethasone treatment. Post-treatment, the patient's symptoms improved, and CT imaging showed the disappearance of the mass lesion. **Conclusion:** Ureteral amyloidosis, though rare, can present with significant clinical symptoms. Early detection and a combination of surgical and medical interventions, as demonstrated in this case, can lead to symptom resolution and improved patient outcomes.

Keywords:
Amyloidosis
Ureteral Amyloidosis
Bortezomib
Surgery
Radiotherapy

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Adult Hematology Abstract Categories

Platelet Diseases
OP 07

THE ROLE OF ADAMTS13 ACTIVITY LEVELS ON DISEASE EXACERBATION OR RELAPSE IN PATIENTS WITH IMMUNE-MEDIATED THROMBOTIC THROMBOCYTOPENIC PURPURA: POST HOC ANALYSIS OF THE PHASE 3 HERCULES AND POST-HERCULES STUDIES

Johanna KREMER HOVINGA [1], Javier DE LA RUBIA [2], Katerina PAVENSKI [3], Ara METJIAN [4], Paul KNÖBL [5], Flora PEYVANDI [6], Spero CATALAND [7], Paul COPPO [8], Umer KHAN [9], Laurel A. MENAPACE [10], Ana PAULA MARQUES [11], Sriya GUNAWARDENA [10], Marie SCULLY [12]

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Objective: The management of exacerbations and disease relapse is important for patients with immune-mediated thrombotic thrombocytopenic purpura (iTTP). Severe ADAMTS13 (a disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13) deficiency during clinical remission is associated with risk of relapse and may guide prophylactic immune-modulatory therapy. We evaluated ADAMTS13 activity as a potential biomarker of exacerbation or relapse risk in the HERCULES and post-HERCULES studies. **Methodology:** This is a post hoc analysis of integrated data from the modified intent-to-treat (mITT) population of the Phase 3 HERCULES trial (NCT02553317) comparing caplacizumab and placebo (both plus standard-of-care treatment) in patients (pts) with iTTP and the 3-year follow-up post-HERCULES study (NCT02878603). ADAMTS13 activity was determined at baseline, weekly during treatment (post-TPE) and twice during follow-up. Recurrence risk was assessed according to ADAMTS13 activity, using TTP adverse event codes. **Results:** 49/144 (34%) pts in the HERCULES mITT had a recurrence during HERCULES or post-HERCULES. 140/144 pts had follow-up data after treatment end. Of these, 39 pts (28%) had a recurrence after treatment end; mean [SD] ADAMTS13 activity was 20.5% (28.7) in pts with recurrence vs 54.0% (34.9) in pts without; [P<0.0001]). ADAMTS13 activity was <20% at treatment end in 69.2% (27/39) and 27.1% (26/96) pts with/without recurrence (P<0.0001). Similar trends were seen across both treatment groups (Table). **Conclusion:** Regardless of the treatment received (caplacizumab or placebo), lower ADAMTS13 activity levels at end of treatment were associated with a higher risk of recurrence in the HERCULES and post-HERCULES studies. These data highlight the predictive value of ADAMTS13 levels on the risk of recurrence and may assist clinical decision-making in the treatment of iTTP. This content was first presented at ASH 2022 (abstract #2493).

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OP 08

EVALUATION OF VITAMIN D STATUS IN ADULT PATIENTS WITH IMMUNE THROMBOCYTOPENIA

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Objective: 25-OH-vitamin D has been demonstrated to have immunomodulatory effects in addition to maintaining calcium and bone homeostasis. Numerous autoimmune diseases have been linked to a deficiency in this nutrient. Immune cells can metabolize vitamin D and express the vitamin D nuclear receptor. In this study, we aimed to examine the relationship between vitamin D levels and adult patients newly diagnosed with ITP. **Methodology:** The methodology used for this investigation was retrospective. Our primary outcomes were the relationships between 25(OH)D value and platelet count as well as the clinical manifestations of ITP at the time of diagnosis and 25(OH)D value. We also looked at how the various factors and 25(OH)D levels correlated. **Results:** When the vitamin D levels of the patients included in the study were evaluated at the time of diagnosis of ITP; 15 (25%) had vitamin D sufficiency, 15 (25%) had vitamin D insufficiency, 30 (50%) had vitamin D deficiency. There was no statistically significant difference between the median ages of the patients in all 3 groups. In the group with sufficient vitamin D level, male gender was observed more than female gender (p:0.001). **Conclusion:** When we compared the vitamin D levels of the patients according to their response to first-line treatment, no significant difference was found in terms of vitamin D levels in patients who did not respond to treatment, who responded partially, and who responded completely (p:0.32). Similarly, no significant difference was found between response to second-line treatment and vitamin D levels (p:0.16). There was no statistically significant difference in vitamin D between relapsed and non-relapsed

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OP 09

CHANGES IN MUCOSA-ASSOCIATED INVARIANT T CELLS (MAIT), ASSOCIATED CYTOKINES, AND MR-1+ CELL NUMBER AND PHENOTYPE IN THE PERIPHERAL BLOOD OF PEDIATRIC ITP PATIENTS WITH AND WITHOUT ELTROMBOPAG THERAPY

Ahmet Eken [1], Metin Çil [2], Zehra Busra Azizoglu [1], Ramazan Üzen [1], Nazly Najat ASAAD [1,5], Sahin CALIK [1], Koray DORTERLER [3], Enes Mehmet Turkoglu [1], Yunus Emre DOĞAN [3], Ebru Yılmaz [3], Alper Ozcan [3], Musa Karakükçü [3], Goksel Leblebisatan [4], Ekrem Ünal [3]

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Objective: Immune thrombocytopenia (ITP) is an autoimmune disease characterized by thrombocytopenia caused by the formation of antibodies against platelets. Mucosa-associated invariant T cells (MAIT), a subset of unconventional T cells present in the blood and mucosa, are activated in an MR-1-mediated manner, respond to certain infections and cytokines and produce various effector cytokines. **Case report:** In this study, changes in blood MAIT cells were investigated in pediatric ITP patients who received and did not receive Eltrombopag. Twenty healthy volunteers (n:20), 60 untreated, and 16 treated patients (with Eltrombopag) were included in the study. **Methodology:** PBMCs isolated using the Ficoll-Hypaque density gradient were stained with appropriate surface markers and subjected to flow cytometric analysis. In addition, intracellular cytokine staining was performed to measure the level of IFN-γ, IL17A, IL-22, TNF-α cytokines after PMA/Ionomycin stimulation, and all data were analyzed using FlowJo and GraphPad 8. **Results:** Independent of Eltrombopag treatment, MAIT cell absolute counts were decreased in ITP patients. CD45RO levels of the $CD8^{+}$MAIT subtype increased, $\alpha\beta^{+}$ T cells decreased, and $\gamma\delta^{+}$ T cell frequency increased in ITP patients. In patients, the frequency of MAIT cell-derived IFN-γ and TNF-α decreased, MR-1 expression, which is responsible for MAIT cell activation in the $CD3^{-}$fraction, increased, and this level decreased to the levels in healthy controls in individuals receiving Eltrombopag treatment. **Conclusion:** The low HLA-DR levels seen in CD3- cells in ITP patients reached the levels of healthy controls in the group receiving Eltrombopag. These results show that the number and activation status of MAIT cells in ITP patients change and Eltrombopag treatment modulates MAIT cell activity.

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Adult Hematology Abstract Categories

Stem Cell Transplant
OP 10

LONG-TERM OUTCOMES OF ALLOGENEIC STEM CELL TRANSPLANTATION FOR RELAPSED/REFRACTORY HODGKIN AND NON-HODGKIN LYMPHOMA: MULTI-CENTER EXPERINCE FROM TURKEY

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Objective: In this multicenter retrospective study, we evaluated the efficiency on survival and safety of allogeneic hematopoietic stem cell transplantation (allo-HSCT) in patients with relapse/refractory (R/R) Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL). **Methodology:** A total of 110 patients with R/R HL or NHL who underwent allo-HSCT were evaluated between July 2007 and October 2022 in 7 adult stem cell transplantation centers. The primary endpoints of this study were progression-free survival (PFS), graft versus host disease-free, relapse-free survival (GRFS) and overall survival (OS) after the allo-SCT **Results:** Forty-one (37.3%) of total patients were diagnosed with HL, 69 (62.7%) were NHL. The median age at the time of transplantation was 39,5 years (16-67) and 66 (60%) of them male. The mean follow-up time was 67,5±8.1 months and the rates of 5-years OS, PFS, and GRFS were 38.4%, 59.3% and 49.5% respectively. In multivariate analysis, OS was significantly impacted by both conditioning regimen type and acute GvHD degree. Myeloablative conditioning regimen and grade 3-4 acute GvHD had a statistically significant negative effect on OS (HR: 1.74, 95% CI: 1.02-2.98, p=.042, and HR: 2.03, 95% CI: 1.12-3.68, p=.019, respectively). Mismatch unrelated donor (HR: 3.91, 95% CI: 1.58-9.67, p=.003) and CMV reactivation (HR: 1.99, 95% CI: 1.11-3.58, p=.020) were statistically significant negative effect on GRFS. **Conclusion:** According to our results, PFS, OS, and GRFS are not impacted by the disease subtype. However, the transplantation results are affected by the conditioning regimens, donor type, acute GVHD status, and CMV reactivation

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Adult Hematology Abstract Categories

Other Diseases
OP 11

INCREASED CAROTID INTIMA MEDIA THICKNESS AS AN INDICATOR OF INCREASED CARDIOVASCULAR RISK IN PATIENTS WITH PRIMARY FAMILIAL ERYTHROCYTOSIS

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Objective: Erythrocytosis is a group of disorders frequently encountered in haematology practice. Erythrocytosis (polycythemia) is considered to be an elevated haemoglobin (Hb) and/or haematocrit ratio (Hct) in peripheral blood. This ratio is defined as an Hb value >16.5 g/dL in males and >16.0 g/dL in females and an Hct value >49% in males or >48% in females. Erythrocytosis is basically divided into primary and secondary according to EPO (Erythropoietin) level. Both groups are divided into hereditary and acquired forms. EPO level is normal in the primary form. Primary Familial Erythrocytosis (PFE) form often includes EPO mutations (germline mutations). Mutations in EPO receptors result in increased erythrocyte production despite physiological EPO levels. It is inherited and often has a family history of early cardiovascular and cerebrovascular disease events. Primary acquired polycythaemia is Polycythaemia Vera, which includes (somatic mutations; clonal) (JAK2 mutations). Here JAK mutations have mutations of the JAK2V617F or Exon 12 region. It is a chronic myeloproliferative disease involving the bone marrow with the risk of leukaemia and myelofibrosis. The basic rule in secondary causes is increased EPO levels. Secondary inherited type includes germline mutations (VHL, EGLN1, EPAS) and methaemoglobinaemia. Acquired secondary polycythaemia is mainly due to hypoxic causes. In this group of patients, lung, cardiac, endocrine, high altitude and renal transplantation are the main causes. In the approach to polycythaemic patients in haematology outpatient clinics, patients are followed up with intermittent phlebotomies unless the patient has P Vera, normal EPO and JAK mutation. There is no common follow-up and treatment integrity for this group of patients including our study. Although PFE does not have the risk of haematological malignancy, cardiac and cerebral events at an early age are common in family members in the anamnesis of patients. In line with this result, we wanted to evaluate the possible cardiovascular risk in patients in the PFE group and measured carotid intima-media thickness (CIMT) with high-resolution B-mode carotid

ultrasonography, which is known to be a suitable method for detecting subclinical atherosclerosis. Our study was supported by TUBITAK with 1002 programme code and 215S524 project number. Increased CIMT is an indicator of atherosclerosis and increased risk of cardiovascular disease. In our study, we found that CIMT measurements were increased in PFE patients compared to the control group. With this result, we think that subclinical atherosclerosis is increased in these patients. Our aim is to ensure that increased cardiovascular risk in this group of patients and their family members should be taken into consideration and examined more closely. **Methodology:** The study included 64 polycystic patients admitted to Namık Kemal University Medical Faculty Haematology outpatient clinic. Hb levels above 16.5 g/dL in males and 16 g/dL in females were considered polycythaemic. Patients with normal EPO levels and JAK2 analyses (-) were considered as PFE. As a control group, 29 healthy subjects with normal Hb levels were included in the study. Patients with high EPO levels and JAK2 analyses (+), known malignancy and active infection were excluded from the study. CIMT measurements were performed in the supine position with their heads tilted backwards after resting for 15 min. The right and left carotid arteries were imaged by an experienced cardiologist using a high-resolution B-mode ultrasound device (GE Vivid S5: General Electric VingMed Systems, Horten, Norway) with a 12L-RS broadband linear transducer. Right and left common carotid arteries were visualised in the longitudinal plane. The measurements were made manually by determining a 1cm segment 2 cm below the carotid bulb. 3 measurements were averaged. Carotid plaques were not included in the measurement. **Results:** IMTs of the patients were determined as follows. Both CIMT were found to be higher in the patient group. Significant carotid intima media thickness was found in the patient groups compared to the control group. This difference was detected in both carotid arteries. **Conclusion:** Cardiovascular and cerebrovascular events are common in family members of PFE patients, especially with male predominance and sudden death occurring at a young age. Although PFE patients have increased cardiovascular risks, they are often not followed up closely enough from a cardiac point of view in outpatient clinics. Mutations defining PFE are not frequently used in clinical practice. These mutations are mostly found in the 8th exon of the EPO receptor gene. However, since the frequently defined mutation cannot be demonstrated in many cases, the term idiopathic familial polycythaemia is used instead of PFE in some sources. Studies have shown that cardiac load will increase due to increased viscosity as a result of increased erythrocyte mass and endothelial dysfunction will occur due to increased shear stress in the endothelium. An increase in CIMT is an early indicator of subclinical atherosclerosis. As a result of our study, we found that the increase in CIMT, which is an indicator of increased cardiovascular risk, was significantly and statistically significantly increased in the patient group compared to the control group in B mode ultrasond measurements. PFE patients require combined follow-up in haematology and cardiology outpatient clinics. We believe that family investigations are important for the protection of future generations. We think that it is important to screen family members in PFE patients beyond defining a possible risk of cardiovascular disease only in the patient himself/herself in order to prevent complications that may occur in the future and for preventive medicine.

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OP 12

OUTCOME OF APLASTIC ANEMIA ACCORDING TO DISEASE SEVERITY

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Objective Background: Aplastic anemia is pancytopenia with a hypocellular bone marrow [<25 % (or 25 to 50 % if <30 % of residual cells are hematopoietic)] due to failure of the bone marrow in the absence of marrow fibrosis or abnormal infiltrates. For therapeutic guide, the disease is classified into moderately severe, severe and very severe aplastic anemia depending on the degree of cytopenia .Accordingly, patients with severe or very severe forms are started on therapy urgently while patients suffering from non-severe AA are treated conservatively with as needed PRBCs, platelets and growth factors support. Allogenic Hematopoietic stem cell transplantation is the standard of care for young patients with severe AA. **Aims:** Survival following allogenic Hematopoietic stem cell transplantation or immunosuppressive therapy were compared in aplastic anemia according to severity and the prognostic factors related with survival identified. **Methodology:** This is a retrospective study of 156 patients with AA. The outcome of these patients were first analyzed according to the first-line treatment received (SCT vs. IST with no subsequent transplant). The outcome was further stratified based on their risk stratification into moderate, severe, and very severe. Patient's characteristics were summarized using frequencies with percentages for categorical variables and medians with interquartile ranges for continuous data. Probabilities of OS and EFS were summarized using Kaplan-Meier estimator. Survival curves were compared using log-rank test. P-value< 0.05 was considered significant. Analysis was conducted using RStudio 2022.07.2 Build 576 ©ℓ 2009-2022 RStudio, PBC. **Results:** A 156 patients, 92 (59%) were treated with SCT and 64 (41.0%) with IST. 24(15.4%) patients were moderately severe AA, 56 (35.9%) severe AA and 76 (48.7%) very severe AA. Overall survival was 83.7 % in the allogenic hematopoietic stem cell transplantation and 78.8 % in patients given immunosuppressive therapy front-line group (P=0.4).In both group overall survival was 97 % for moderately severe AA, 82 % for severe AA and 77 % for very severe AA. In the allo-SCT cohort, under multivariate analysis, Overall survival for moderately severe, severe and very severe aplastic

anemia was 66.0%, 81.4% and 86.3 % respectively (P=0.5). While, in IST group OS for moderately severe, severe and very severe aplastic anemia was 93.8%, 86.6% and 56.1 % respectively (P=0.005). Age of 20 years or under positively affected overall survival in allogenic hematopoietic stem cell transplantation group, whereas age more than 20 years negatively affected overall survival in this group. The factors influencing the overall survival were use of allo-SCT, an age under 20-years-and moderately severe AA. **Conclusion:** Aplastic anemia in adolescents has a very good outcome. If a matched sibling donor is available, Hematopoietic stem cell transplantation is the first choice treatment. If such a donor is not available, immunosuppressive treatment may still be an acceptable second choice also because, in case of failure, hematopoietic stem cell transplantation is a very good rescue option. Use of SCT, age of < 20 years in sever AA and IST in non-severe AA were favorably associated to OS. Therefore, younger age SAA patients, with HLA-matched donors, may benefit more from allo-SCT.

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OP 13

GLOBAL RESEARCH PATTERNS ON BLOOD DONOR DEFERRAL: AN ANALYSIS OF THEMES, TRENDS, AND INFLUENCE

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Background: Blood banking relies heavily on deferral policies for safety. Recognizing current academic themes can highlight research opportunities, encourage collaboration, ensure funding, understand audience interests, steer public sentiment, and inspire productive competition, thereby prompting impactful studies. **Materials and Methods:** We analyzed 1034 blood deferral papers from Web of Science and Scopus, focusing on publication count, uniqueness, timeline, and themes like Men who have Sex with Men (MSM), HIV, COVID-19, anemia, and machine learning. We also assessed the global distribution of these studies to understand prevalence and associations with geography, demographics, and economic factors. Results and Conclusions: The study uncovered 1037 articles; MSM (107), HIV (234), Anemia (201), COVID-19 (40), and machine learning (59). Most papers were from the US, UK, Canada, reflecting their robust research capabilities. The US led in HIV and anemia studies, with India significantly contributing to anemia research. India led in COVID-19 studies, with substantial participation from the US. Machine learning research primarily came from the US and India, with significant Chinese contributions. The trending literature on blood deferral underscores the need to comprehend evolving blood banking dynamics. Machine learning, with its transformative capacity, is a prime research area. These insights could guide future studies and policymaking, maintaining blood safety.

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OP 14

UNUSUAL PRESENTATİON OF RHABDOMYOSARCOMA WİTH BONE MARROW INVOLVEMENT AND CERVİCAL MASS: A 17-YEAR-OLD FEMALE CASE REPORT

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Background: Rhabdomyosarcoma (RMS) is a rare type of cancer that originates in the skeletal muscle cells. It's most commonly found in children but can occur at any age. The cancer is characterized by the presence of cells that resemble immature muscle cells, and it can grow and spread rapidly. Rhabdomyosarcoma (RMS) is a rare cancer that originates in skeletal muscle cells and can be found in various parts of the body, including the head and neck, genitourinary tract, extremities, and other less common areas such as the trunk and retroperitoneum. Bone marrow infiltration in Rhabdomyosarcoma (RMS) is a relatively rare occurrence. We are presenting a case Rhabdomyosarcoma with Bone Marrow Involvement and cervical Mass. **Case Report:** A 17-year-old female patient with no known previous illnesses presented to an external center with complaints of coughing, difficulty swallowing, weight loss, and fatigue that had begun a month prior. During a physical examination, a 2 cm mass was observed in the left cervical region, along with an enlarged appearance of the thyroid gland. Complete blood count revealed hemoglobin at 10.6 g/dL, leukocytes at 1000 mm3, neutrophils at 200 mm3, and platelets at 70000 mm3, leading to a referral to a hematology clinic. Upon repeated observation of pancytopenia, early myeloid precursors were seen in a peripheral smear. Due to a high suspicion of lymphoma, a bone marrow biopsy was performed, revealing widespread mononuclear cell infiltration. Immunohistochemical analysis showed desmin(+), myogenin (+), and Ki67 80% positivity, leading to a diagnosis of rhabdomyosarcoma. A PET-CT scan to determine the extent of the

disease revealed multiple involvements in the bone and lungs. Treatment with Vincristine, doxorubicin, cyclophosphamide, and dexamethasone was initiated, resulting in a significant regression of the masses and an improvement in the cytopenia picture. **Conclusion:** The presence of RMS in the bone marrow can complicate both the diagnosis and treatment of the disease. It may require additional diagnostic procedures, such as bone marrow biopsy, to confirm the presence of RMS cells. Treatment may also need to be more aggressive, approaches. Bone marrow involvement in RMS is considered a more advanced stage of the disease and may be associated with a more challenging prognosis. Early detection and tailored treatment are crucial for managing RMS with bone marrow infiltration.

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OP 15

NEW MOLECULAR TARGETS IN CANCER CELL BIOENERGETIC PATHWAYS

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Research Supervisor: L.Y. Grivtsova, PhD in medical science, PhD in biology science, Head of Laboratory Medicine Department, Head of Clinical Immunology Laboratory of A.F. Tsyba MRSC, a branch of the Federal State Budgetary Institution "NMRC of Radiology" of the Ministry of Health of the Russian Federation Over the last ten years, the ideas of molecular oncology about the energy metabolism of malignant cells have changed dramatically, and new molecular mechanisms in the cascade pathways of cancer bioenergetics are being searched for. Numerous data show that the emergence and development of tumors are closely related to the metabolism of iron ions (Fe). Inorganic substrates, namely iron ions involved in the metabolic processes of the tumor cell, have received limited attention in the world literature to date. Our research group has put forward and is developing the concept of «Energy metaplasia of cancer cells», i.e. acquisition of an additional autotrophic way of energy production (respiratory reactions involving iron ions) in the process of oncogenesis. Proof of the hypothesis opens prospects for explaining some issues of oncogenesis and a new approach to the treatment of cancer. The aim of the study: to investigate and obtain evidence for the existence of respiratory (chemosynthetic) reactions involving iron ions as a way to obtain energy in cancer cells. The studies were conducted on the basis of the «Center of Cell Technologies», Samara city, Russia, under the guidance of specialists from A.F. Tsyba MRSC, Obninsk city, Russia. All experiments were conducted in vitro using HeLa cell line (cervical carcinoma) and human mesenchymal stromal cell line (MSC) culture as a control. The proof-of-concept study was carried out in 3 stages. The 1st stage was analytical review, the 2nd stage - study of energy metabolism by extracellular flux analysis on the SeaHorseXFp apparatus (USA), the 3rd stage - bioinformatic study on search in the human genome for homolog genes responsible for chemosynthetic reactions using blastp and exonerate programs. As a result of the analytical review of works on the evolution of the way of energy production by plant and animal cells, a possible chemosynthetic reaction in cancer cells - oxidation of iron ions (Fe^{+2} - Fe^{+3}+E) was revealed. As a result of 50 performed protocols on SeaHorseXFp cell metabolism analyzer we found suppression of two classical pathways of energy production - oxidative phosphorylation (by 54,2%) and glycolysis (by 85,4%) in malignant HeLa culture in contrast to normal index in MSC cell culture. As a result of bioinformatic study, 6 proteins and 11 domains related to iron metabolism were found in the human genome, which are highly similar in sequence to the genes responsible for chemosynthetic reactions involving iron ions in iron bacteria. Thus, respiratory chemosynthetic reactions involving iron ions are possible in malignant cells, which allows the cancer cell to change its energy phenotype and acquire an additional autotrophic way of energy production, allowing it to acquire the properties of uncontrolled growth and metastatic spread. This molecular cascade requires additional study and is of interest as a target for the development of targeted antitumor drugs.

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Pediatric Hematology Abstract Categories Coagulation and Fibrinolysis Disorders
OP 16

THE COEXİSTENCE OF NOVEL MUTATİONS OF FX, DİMETHYLGLYCİNE DEHYDROGENASE GENES WİTH FAMİLİAL EPİSODİC PAİN SYNDROME: A CASE REPORT

Hatice Mine Çakmak [1]

[1] *Duzce Unıversıty*

Objective: Congenital Factor X (FX) deficiency is an autosomal recessive disorder with variable clinical severity associated with heterozygosis or homozygosis inheritance. Genetic mutations are located on the glutamic acid domain on exon two and the catalytic site of FX on exons 7, 8. Missense mutations of specific patients or families are reported. Severe forms of the disease result from homozygosis or compound heterozygosis genetic mutations. In this case report, we aim to write a rare cause of epistaxis and novel mutations of FX and DMDGH (**Dimethylglycine Dehydrogenase)** deficiencies, and he and his family are the second with TRAP1-related FEPS1 (familial episodic pain syndrome) in the World. **Case report:** A 6-year-old boy, born in Turkey, with no known

chronic medical condition, was admitted to the pediatric hematology-oncology policlinic with epistaxis lasting for approximately 10 minutes and repeating daily. The family history revealed prolonged bleeding episodes in his father, uncle, aunt, and uncles' sister. The patient denied mucosal bleeding, spontaneous bruising, or prolonged bleeding after dental extraction. The physical examination included; weight: 22 kg (50-75 p), height: 127 cm (>95 p), and regular systemic features. In addition, prolonged PT (21.1s) and normal aPTT levels were found. In his family, prolonged PT was also detected in his father (16.9s) and sister (13.7s). Factor and coagulation levels and their normal ranges consistent with age are given in Table 1. In the clinic exom study, NM_000504.4:c. 785 G>A p.Gly262Asp heterozygosis mutation on the F10 gene has a nonsynonymous_SNV effect, causing Factor X deficiency (Table 1). This mutation is a new change undefined in the Clinvar database with a DANN score of 0.988. According to the ACMG rules (PP3, PM2, PP2), this mutation is pathogenic (Figure 1). Clinic exom study revealed other mutations (Table 2) associated with the case's clinic features. For example, the patient had a fish odor and muscle tiredness associated with dimethylglycine dehydrogenase deficiency. In addition, the patient suffered from episodic pain syndrome (upper body pain after cold, physical stress, and fasting) and frequent fevers that may be associated with Immunodeficiency and TNFRS13B mutation. Episodic pain syndrome was common in the patient's father's family (uncle, uncle's sister, aunt, grandfather, aunt, and aunt's three children) (Figure 2). However, the diagnosis of immune deficiency is not defined in his family. Family segregation mutation analyses are under study. **Results:** Factor X deficiency is a rare coagulation disorder. The clinic severity differs according to the genetic mutations generally localized to the glutamic domain exon 2 [(1)]. Gokcebay et al. represented an infant with a homogenous FX gene mutation in Exon2 (Gly51Arg) with an FX serum level of 0.03 U/ml. This infant had umbilical cord bleeding and cephalic hematoma and received fresh frozen plasma and activated prothrombin complex concentrate (aPCC). PT (INR) was elevated only[(5)]. Nagaya et al. reported Four heterozygous mutations [p.Gly154Arg, p.Val236Met, p.Gly263Val, and p.Arg387Cys] and a compound heterozygous FX gene mutation (p.Gly406Ser and p.Val424Phe) were identified [(6)]. Another case report showed that a heterozygous nonsense mutation in the F10 gene led to prolonged vaginal bleeding after polypectomy [(7)]. In neonates, FX levels <10% may cause severe bleeding like CNS, gastrointestinal, hematomas, and hemarthroses. In addition, severe deficiency may cause epistaxis and menorrhagia. The results of the EN-RBD study showed a variable target level of 10% to 20% up to 40% to prevent bleeding. In addition to fresh frozen plasma, Apcc, FIX/FX, and FX concentrates are available to treat FX deficiency. Doses of treatment and schedules differ according to the surgery preparation, prophylaxis, or bleeding. Tranexamic acid is preferred for menorrhagia, nosebleeds, presurgery, and surgery to prevent excessive factor administration (8,9). Our study showed mild FX deficiency with a nucleotide protein change of NM_000504.4:c.785 G>A p. Gly262Asp, and a novel mutation of FX deficiency. Recurrent epistaxis episodes were controlled with a nasal tampon. We plan to administer tranexamic acid for uncontrolled nasal bleeding. In trauma and surgery, fresh frozen plasma and concentrates are the treatment options. Familial non-inflammatory pain syndromes (FEPS) are divided into three groups; only one reported family has TRAP1-related FEPS1 syndrome. The predisposing factors for FEPS3 in children are cold, fatigue, hunger, and the rainy season. Pain mainly occurs in the afternoon or at night; paroxysmal pain lasts for tens of minutes, then relieves, and then starts again after a short interval. Unlike FEPS3(cardiac ion channel disease and congenital myotonia), caused by the SCN11A mutation with pain in the distal limbs, TRAP1-related FEPS1 syndrome has pain symptoms in the upper body with autonomic symptoms. Currently, there is no specific drug for treating familial paroxysmal pain syndromes. The primary treatment is to use analgesics, keep warm, and avoid cold environments. Here, we report the second family with TRAP1-related FEPS1 syndrome. The pain is in the upper body, similar to his family [(10)]. Dimethylglisin dehydrogenase deficiency, as fish odor syndrome, is a likely benign condition with mild muscle involvement [(11)]. We report a novel variant consistent with the clinical situation, a heterozygotic stop gain, NM_013391.3:c.972 G>A p.Trp324 mutation, defined as pathogenic/VUS/likely benign. **Conclusion:** This is the first report of F10, DMGDH novel mutations, and the coexistence of TRPA1 (clinic was compatible) and TNFRSF13B with a need for investigation for immunodeficiencies in a child. In conclusion, this is the first patient with two novel mutations for FX and DMDGH deficiencies, and his family is the second with TRAP1-related FEPS1 syndrome in the World.

Laboratory Values (normal ranges)	Patient	Father	Mother	Sister (7 years old)
PT (s)	21.1 (10.1-12.1)	16.9 (11-14)	13 (11-14)	13.7 (10-12.1)
APTT (s)	32.6 (26-36)	27.8 (27-40)	28.7 (27-40)	26.8 (26-36)
Factor VII (%)	75.3 (65-180)	97.7 (61-127)	97.7 (65-180)	69.4 (61-127)
Factor X (%)	**20.3** (88-94)	**65.9** (70-150)	78.5 (70-150)	**69.5** (88-94)

Gen	Nucleotide Protein Change	Zygosis	dbSNP	Effect	Variant	Classification
F10	NM_000504.4: c.785 G>A p. Gly262Asp	het	-	nonsynonymous-SNV	-	Disease (Dominance, OMIM#) Factor X Deficiency (OR; 227600)
DMGDH	NM_000504.4: c.785 G>A p. Gly262Asp	het	rs139044238	Stop gain	Pathogenic/ VUS/Likely benign	Dimethylglisin dehydrogenase deficiency (OR;605850)
TRPA1	NM_007332.3: c.2564 A>G p.Asn855Ser	het	rs398123010	nonsynonymous-SNV	Pathogenic	Familial episodic pain syndrome (OD;615040)
TNFRSF13B	NM_012452.3: c.198 C>A p. Cys66	het	rs144718007	Stop gain	Pathogenic	Tumor Necrosis Factor Receptor Superfamily Member 13B;604907

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Pediatric Hematology Abstract Categories

Red Blood Cell Disorders
OP 17

ASSESSMENT OF VITAMIN B12 AND HOMOCYSTEINE LEVELS IN PREGNANT WOMEN ADMITTED FOR DELIVERY AND CORD BLOOD SAMPLES OF THEIR NEWBORN BABIES: A MULTICENTER STUDY

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Objective: Vitamin B12, an essential micronutrient, plays a vital role in various physiological processes, particularly during pregnancy and fetal development. The growing popularity of vegetarian diets and socioeconomic barriers to consuming animal-based products contributes to Vitamin B12 deficiency becoming a global issue. Understanding the B12 status in pregnant women and its potential impact on newborns is of utmost significance as it can have far-reaching implications for both maternal and infant health. This research aims to investigate the vitamin B12 and homocysteine levels in pregnant women admitted for delivery and analyze corresponding cord blood samples from their newborn babies. **Methodology:** This prospective study was conducted in three tertiary care hospitals and included pregnant women aged ≥16 years admitted for delivery and their newborns ≥34 weeks. The demographic data and the results of complete blood counts performed within the previous 24 hours before birth were recorded. The levels of vitamin B12 and homocysteine were measured in blood samples and cord blood samples taken from pregnant women and their newborns, respectively. The study parameters were compared between the two groups based on the mothers' and babies' homocysteine and B12 levels. **Results:** The study included 615 Turkish and 217 foreign pregnant women. Anemia affected 36% of pregnant, with a higher frequency in mothers with B12 deficiency. The mean B12 level in pregnant women was 157±75.3 pg/ml, with 14.8% having elevated homocysteine levels. The levels of B12 and homocysteine of the newborns were 234.7±13.2 pg/ml and 9.13±5.75 mol/L, respectively. Vitamin B12 deficiency was found in 48.9% of newborns, while homocysteine levels were slightly elevated or elevated in 19.1%; both findings were significantly more common in babies born to B12-deficient mothers. **Conclusion:** In our study, vitamin B12 deficiency was significant in pregnant mothers and their neonates, with a substantial connection to cord blood homocysteine levels. Further study is needed to determine the impact of this deficit on mother and newborn health. Implementing approaches to timely detecting Vitamin B12 deficiency and, if necessary, providing adequate Vitamin B12 supplementation during pregnancy could play a pivotal role in enhancing the health and well-being of both the mother and the child.

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OP 18

A GHOSAL HEMATODIAPHYSEAL DYSPLASIA CASE; EXCELLENT RESPONSE TO NON-STEROIDAL ANTI-INFLAMATORY DRUG TREATMENT

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Objective: Ghosal hematodiaphyseal dysplasia (GHDD) is a very rare autosomal recessive disease caused by prostaglandin metabolism disturbances due to biallelic mutations on chromosome 7q33-34 which lead to decrease in thromboxane synthase function. Previously long-term corticosteroid was the only treatment for GHDD. Currently, non-steroidal anti-inflammatory drugs (NSAIDs) as a targeted therapy are preferred alternatively. Here, a genetically confirmed GHDD case responsive to ibuprofen is presented. **Case report:** A 9-year-old girl presented to our clinic with severe normocytic anemia, swelling, and pain in her lower limbs. In physical and radiologic examination metadiaphyseal dysplasia was diagnosed. The diagnosis of GHDD was confirmed with genetic analysis. The patient was treated with ibuprofen (30 mg/kg/day) with excellent response to both pain and hematologic parameters in 15 days period. **Conclusion:** Ghosal

hematodiaphyseal dysplasia is a very rare disease. The patients manifest with metadiaphyseal dysplasia, severe anemia, chronic fatigue, and inflammation. Previously long-term corticosteroid was the only treatment for GHDD with multiple significant long-term complication risks. NSAIDs, in this case, ibuprofen, are the current and new treatment options with relatively safe side effect profiles. But only a few cases with short-term follow-up were reported in the literature.

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OP 19

THE SIGNIFICANCE OF NEXT-GENERATION SEQUENCING IN NON-IMMUNE HEMOLYTIC ANEMIAS AMONG NORMOCHROMIC-NORMOCYTIC ANEMIAS

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Objective: Next-generation sequencing studies increased the exact diagnosis of unexplained normochromic-normocytic anemias and other anemias. Targeted next-generation sequencing studies allow the diagnosis of cytoskeleton defects, atypical cases, and some enzyme deficiencies. We aimed to compare the children with non-immune hemolytic anemia (n=13), and the others without non-immune hemolytic anemia (n=19) in the means of demographics, diagnosis, detected mutations, and laboratories. **Methodology:** In this study, the children who were examined in the Pediatric Hematology-Oncology Clinic of Duzce University School of Medicine and had unexplained anemia (n=29) underwent next-generation studies. The demographics, laboratory values, and genetic findings of two groups (non-immun hemolytic anemia and the others) were compared. We also found two novel mutations, one with hereditary spherocytosis and one with hereditary elliptocytosis. Mean, standard deviation, median minimum, maximum, frequency and ratio values were used in descriptive statistics of the data. The distribution of variables was measured with the Kolmogorov-Simirnov test. Independent sample t test and mann-whitney u test were used to analyze quantitative independent data. The chi-square test was used to analyze qualitative independent data. SPSS 28. 0 program was used in the analysis Results **Conclusion:** The demographics and the laboratory results are explained in Table 1. Comparing the non-immune hemolytic anemia patients (n=13) with the others (n=19), we found that membrane disorders rates, identified mutations associated with anemia, mean cell volume, mean cell hemoglobin, thrombocyte, reticulocyte, and absolute reticulocyte levels were higher, hemoglobin and erythrocyte levels were lower in the non-immun lower in the non-immune hemolytic anemia group (Table 2). The novel mutations are shown

Table 1) Demographics of the patients with unexplained anemia

		Min-Max.	Median	Mean.±s.d./n-%
Age (years)		0.1 - 17.0	0.1	5.3 ± 4.8
Age at onset of symptoms (years)		0.0 - 17.0	0.0	2.1 ± 4.2
Gender	Girl			9 6.8%
	Boy			23 17.4%
Nonimmune Hemolytic Anemia (+)	(+)			13 9.8%
	(-)			19 14.4%
Nonimmune Hemolytic Anemia (+)	(-)			22 16.7%
	(+)			10 7.6%
Identified Anemia Mutation	(-)			29 22.0%
	(+)			3 2.3%
Other defined mutations	(-)			23 17.4%
	(+)			9 6.8%
Enzyme Deficiency	(-)			27 20.5%
	(+)			5 3.8%
Erythrocyte count (10^9)		1.7 - 4.6	1.7	3.3 ± 0.9
Hct (%)		14.7 - 38.0	14.7	27.9 ± 6.3
Hb (g/dl)		4.7 - 27.0	4.7	9.9 ± 3.7
MCV (fL)		14.7 - 111.0	14.7	84.3 ± 15.8
MCH (pg)		22.0 - 97.0	22.0	30.6 ± 12.6
MCHC (g/dl)		30.0 - 36.1	30.0	32.9 ± 1.5
RDW (%)		10.9 - 21.7	10.9	15.1 ± 2.6
Thrombocyte count (x10^3/ml)		94.0 - 567.0	94.0	354.1 ± 112.2
Reticulocyte (%)		0.1 - 23.7	0.1	3.9 ± 5.7
Adjusted reticulocyte count (%)		0.0 - 12.7	0.0	2.8 ± 3.3
Transfusion rates (/y.l)		0.0 - 4.0	0.0	0.6 ± 1.3
Total bilirubin (mg/dl)		0.0 - 14.0	0.0	4.0 ± 4.9
İndirect bilirübin (mg/dl)		0.0 - 13.2	0.0	2.8 ± 4.1
Ferritin (ng/ml)		14.5 - 1392.0	14.5	192.0 ± 315.8

Abbreviations: Hct: Hematocrite, Hb: Hemoglobin, MCV: Mean cell volüme, MCHC: mean cell hemoglobin concentration, RDW: red cell distribution width

Table 2) Comparing the children with versus without non-immune hemolytic anemia

		non-immune hemolytic anemia (+)		non-immune hemolytic anemia (-)		p	
		Mean.±SD/n-%	Median	Mean.±SD/n-%	Median		
Age (years)		5.1 ± 6.2	1.5	5.4 ± 3.9	5.0	0.247	m
Age at onset of symptoms		2.4 ± 5.7	0.0	1.9 ± 3.0	0.0	0.544	m
Gender	Female	6 46.2%		3 15.8%		0.061	X^2
	Male	7 53.8%		16 84.2%			
Immun hemolytic anemia	(-)	6 46.2%		16 84.2%		**0.023**	X^2
	(+)	7 53.8%		3 15.8%			
Identified anemia mutation	(-)	10 76.9%		19 100%		**0.028**	X^2
	(+)	3 23.1%		0 0.0%			
Other defined mutation	(-)	9 69.2%		14 73.7%		0.783	X^2
	(+)	4 30.8%		5 26.3%			
	(+)	3 23.1%		2 10.5%			
Erythrocyte (10^2/ml)		2.9 ± 0.8	3.2	3.5 ± 0.9	3.7	**0.030**	m
Hct (%)		26.2 ± 6.4	28.0	29.0 ± 6.1	29.8	0.234	m
Hb (g/dl)		8.6 ± 2.1	9.2	10.8 ± 4.3	10.2	0.058	m
MCV (fL)		90.8 ± 9.5	90.0	79.9 ± 17.9	82.0	**0.021**	m
MCH (pg)		35.1 ± 18.9	30.6	27.6 ± 3.2	27.0	**0.030**	m
MCHC (g/dl)		33.0 ± 1.7	32.6	32.9 ± 1.4	32.6	0.835	t
RDW (%)		15.4 ± 3.5	14.0	15.0 ± 1.8	15.0	0.673	m
Thrombocyte (x10^3/ml)		412 ± 104	384	314 ± 102	314	**0.013**	t
Reticulocyte (%)		6.6 ± 7.1	3.2	1.1 ± 0.6	1.2	**0.001**	m
Adjusted reticulocyte count (%)		4.4 ± 4.0	2.3	0.9 ± 0.4	1.0	**0.001**	m
Transfusion rates (/year)		0.9 ± 1.6	0.0	0.2 ± 0.6	0.0	0.150	m
Total bilirübin (mg/dl)		4.8 ± 4.7	3.3	3.2 ± 5.1	0.4	0.124	m
İndirect bilirübin (mg/dl)		4.0 ± 4.6	2.3	1.5 ± 3.0	0.2	0.065	m
Ferritin (ng/ml)		295 ± 186	236	144 ± 356	23	**0.005**	m

Abbreviations: Hct: Hematocrite, Hb: Hemoglobin, MCV: Mean cell volüme, MCHC: mean cell hemoglobin concentration, RDW: red cell distribution width, ^{t}t test / ^{m} Mann-whitney u test / $^{X^2}$ Ki-kare test

Patient	Gen	Nucleotide Protein Change	Zygosis	dbSNP	Effect	Variant Classification	Disease (Dominance, OMIM#)
1	ANK1	NM_001142446.2:c.747 C>G p.Tyr249*	het	-	stop gain	-	Sferocytosis type 1; AD:18900
1	SPTB	NM_001024858.3:c.4891 C>T p.Arg1631Cys	het	rs372503030	nonsynonymous-SNV	VUS	Spectrin Beta Erythrocytic; 182870)
2	SPTB	NM_001024858.3:c.4 355C>T p.Ala1452Val	het	rs768609633	nonsynonymous-SNV	VUS	Spectrin Beta Eritrocytic; 182870

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Pediatric Hematology Abstract Categories

Immunodeficiencies / Neutrophil Diseases
OP 20

A NEXT-GENERATION SEQUENCING TEST FOR CONGENITAL NEUTROPENIA IN PEDIATRIC PATIENTS

Ayça Koca Yozgat [1], Fatma Burçin Kurtipek [1], Zeliha Güzelküçük [1], Dilek Kaçar [1], Turan Bayhan [2], Namık Yaşar Özbek [1], Neşe Yaralı [2]

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Objective: Congenital neutropenia (CN) is a rare inherited hematological disease and its phenotypic, histologic and molecular aspects are heterogeneous. Congenital neutropenia can manifest as isolated neutropenia or neutropenia with extra-hematopoietic abnormalities, immunodeficiency or metabolic diseases and results in recurrent, life-threatening bacterial infections. Mutations in more than 20 genes have been demonstrated to cause CN, some of which cause complex phenotypes. **Case report:** Usually caused by ELANE mutations although mutations in other genes like HAX-1, G6PC3, and GF11 have also been reported. Identifying the causative mutation aids in the diagnosis and ruling out other secondary causes of neutropenia. In this study we aimed to identify the molecular defects in CN patients by next generation sequencing (NGS). **Methodology:** Next generation sequencing test was performed on peripheral blood specimens of 17 patients diagnosed with congenital or chronic neutropenia and bone marrow failure and hematological malignancy ruled out from January 2021- June 2023. The genes in the NGS panel were; LAMTOR2, CLPB, HAX1, USB1, SBDS, JAGN1, TAZ, ELANE, G6PC3, WAS, CXCR4, GFI1, VPS45, VPS13B. **Results:** The median age at presentation of neutropenia was 28.9 months. Mean neutrophil count at diagnosis was 380± 259/mm3. Bone marrow aspiration was performed in ten patients and myeloid maturation arrest was observed five. Granulocyte colony stimulating factor was given for seven patients and all had a response. In the NGS panel TAZ mutation was detected in one patient compatible with Barth Syndrome and VPS13 double heterozygous mutation was detected in one patient compatible with Cohen Syndrome. **Conclusion:** Considering the heterogeneity of CN in terms of genotypes and phenotypes expanded next generation sequencing panel would be necessary. The early onset of the disease, clinical severity and associated high risk of malignant transformation in CN strongly suggests the need for early diagnosis and therapeutic intervention.

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Pediatric Hematology Abstract Categories

Leukemia
OP 21

BK-VIRUS INFECTIONS IN PEDIATRIC LEUKEMIA PATIENTS DURING LEUKEMIA TREATMENT

Dilek Kaçar [1], Zeliha Güzelküçük [1], Ayça Koca Yozgat [1], Melek Işık [1], Neşe Yaralı [1]

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Objective: Polyoma BK virus (BKV) infection/reactivation is an important underlying condition that provokes hemorrhagic cystitis (HC) in hematopoietic stem cell transplantation (HSCT) recipients. However, BKV associated infections can rarely occur in acute leukemia patients without receiving HSCT. Here we present 12 pediatric acute leukemia patients with BKV infection during leukemia treatment. **Methodology:** Between September 2019 and July 2023, in Ankara Bilkent City Hospital, BKV by quantitative polymerase chain reaction (PCR) detected in the urine of 12 pediatric leukemia patients who had not got HSCT but receiving intensive chemotherapy. The clinical characteristics of these infections were retrospectively evaluated. **Results:** Ten of the 12 patients had acute lymphoblastic leukemia (ALL). Seven of the 10 ALL cases were T cell ALL. Ten of the patients were male and 10 of the patients' age were 10 years and older. Eleven patients experienced HC and one patient had epididymitis. The copy number of BKV varied between 470 to 1.3 trillion /mL. Seven patients had got treatment ranging from hydration, ciprofloxacin to bladder irrigation. Except a refractory T cell ALL patient, all of the patients had clinical improvement. **Conclusion:** Although it is a major complication of HSCT and solid organ transplantation, BK virus infection can also occur in pediatric acute leukemia patients during treatment. As in HSCT recipients, male gender and older age seems as risk factors in leukemia patients. Due to complete loss of virus specific T lymphocytes, T cell ALL patients may be more prone BK virus activation.

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OP 22

MOLECULAR CHARACTERISTICS AND TREATMENT RESPONSE TO COG ALL PROTOCOL IN CHILDREN; A 16-YEAR SINGLE-CENTER STUDY

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Objective: Acute Lymphoblastic Lekumia is the most prevalent cancer type in children. While most data from Turkey primarily focuses on BFM protocols, our study uniquely concentrates on the COG ALL protocols. In this study we aimed to analyze the molecular characteristics and outcomes of the patients diagnosed with ALL who were treated at the Pediatric Hematology and Oncology department of Acıbadem Mehmet Ali Aydınlar University Faculty of Medicine, Altunizade Hospital. **Methodology:** We have reported all the cases that have achieved complete treatment of ALL. Patient risks were assessed by diagnosis, demographics, and clinical settings, followed by protocol selection. Analysis of risk stratification involved immunophenotyping, and genetic characteristics. **Results:** 46 patient participated in the study. Standard risk, high risk and very high risk was observed in 60.5%, 27.9% and 11.6% patients respectively for B ALL and all T-ALL patients were admitted to the high risk group. 28.3% had negative prognostic genetic mutations. At the end of the induction therapy (At the 29th day), 80.4 % of the patients had MRD level below 0,1%. Mean survival time was 71,6 months. 4,3% of patients had bone marrow relapse, and after second-line treatment, are now relapse free. **Conclusion:** This study assesses the pediatric ALL patients treated with COG protocols from Turkey in a single center over a span of 16 years. Follow up processes and therapy responses taking into consideration their demographical characteristics, clinical attributes, genetic profiles, complications and outcomes. COG ALL Protocols in Turkey are being used only by the COG international corresponding members and are as promising as the BFM protocols.

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Pediatric Hematology Abstract Categories

Inherited Bone Marrow Failure Diseases
OP 23

INVESTIGATION OF SALİVARY MIR-9 AND SERUM CIP2A LEVELS IN FANCONI ANEMIA PATIENTS AT HIGH RISK OF DEVELOPING ORAL SQUAMOUS CELL CARCINOMA

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Objective: Fanconi anemia (FA) is a rare bone marrow failure syndrome caused by mutations in DNA repair genes, and the risk of developing Oral Squamous Cell Carcinoma (OSCC) in FA patients is higher than in the normal population and is seen at younger ages. mi-RNAs and proteins associated with signaling pathways such as PI3K and Wnt, which play a role in cancer pathogenesis, are important biomarker candidates for OSCC development. Tumor suppressor miR-9 have been reported that abnormally expressed in many different cancers and OSCC. Cancerous inhibitor of protein phosphatase 2A (CIP2A) is a characterized human oncoprotein that has been studied in the most of human malignancies. Squamous cell cancers frequently develop in FA patients. Therefore, in this study, we aimed to evaluate the salivary miR-9 and serum CIP2A levels of our FA patients who are likely to develop cancer, and to evaluate them in terms of the risk of developing OSCC and compared them with the healthy control group. **Methodology:** Saliva and serum samples were collected from 25 OSHK patients, 24 FA patients and 40 healthy volunteers, and total RNA was isolated from saliva samples and quantitative Real-Time PCR was performed with the miRCURY LNA miRNA PCR Assay (Qiagen, Hilden, Germany). miR-9 saliva levels were normalized and calculated by the Livak Method. ELISA (Bioassay Technology Laboratory, Shanghai, PRC) method was used to measure serum CIP2A levels. **Results:** According to our data, salivary miR-9 levels of both OSCC and FA patients were lower than healthy controls (p=0,01 and p=0,017). In OSCC patients, miR-9 level was related to lymph node metastasis (p=0,04). Serum CIP2A levels in OSCC patients and were higher than in healthy controls (p<0,001). **Conclusion:** Our findings indicate that miR-9 and CIP2A may be remarkable biomarkers in the development of OSCC. Since FA patients have a high risk of developing OSCC, close follow-up of the physical examination findings of miR-9 and CIP2A levels can be beneficial.

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Pediatric Hematology Abstract Categories

Stem Cell Transplantation
OP 24

BUSULFAN-CYCLOPHOSPHAMIDE OR TREOSULFAN-FLUDARABINE-THIOTEPA-BASED MYELOABLATIVE CONDITIONING FOR CHILDREN WITH THALASSEMIA MAJOR, SINGLE CENTRE EXPERIENCE

Nihan Bayram [1], Yontem Yaman [1], Isik Odaman Al [1], Kursat Ozdilli [1], Murat Elli [1], Sema Anak [1]

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Objective: Hemoglobinopaties are the most common genetic disorder worldwide. Patients with transfusion-dependent thalassemia major (TDT) are deficient in β-globulin chain production, resulting in ineffective erytropoiesis and hemolysis. Consequently, patients with TDT suffer from primary and secondary iron overload, leading to severe organ dysfunction. In despite of significant improvements in supportive care, especially monitoring and treatment of iron overload and its complications, organ dysfunction progresses in adulthood, resulting in significant morbidity and mortality. Allogeneic haematopoietic stem cell transplantation (HSCT) is the current standart of care for patients with thalassemia major, except clinical trials on gene therapy and gene editing as alternative curative options. Despite improvements in supportive care, blood transfusions and organ damage from iron overload situation before HSCT, predict worse outcome. Recent studies have reported a rate of graft rejection of 8 to 12 % in pediatric patients with TDT undergoing HSCT. Furthermore, the role of conditioning regimen in the outcome has been extensively investigated. Busulfan, treosulfan, fludarabine, thiotepa, cyclophosphamide are common agents of the conditioning regimen for HSCT. Busulfan is an alkylating agent that is mainly eliminated through the liver. Busulfan is associated with sinusoidal obstruction syndrome, pulmonary toxicty, seizures, chronic gonadal dysfunction, and late mortality. Treosulfan is the prodrug of L-epoxybutane, a water-soluble, bifunctional alkylating agent. Treosulfan-containing regimens achieve a high rate of stable donor engraftment, reduced transplant-related mortality and low rate of GVHD. Therefore, treosulfan has been considered to replace busulfan in conditioning regimens in patients with TDT. Experience with treosulfan-based conditioning in pediatric patients is more limited than studies in adult series. However, the data has promising results. Thus, here were reported a retrospective study of patients with TDT undergoing HSCT, in which we compared those with busulfan and those with treosulfan in their conditioning regimen. **Methodology:** We retrospectively evaluated all the consecutive cases of pediatric patients underwent allogenic HSCT and busulfan-based or treosulfan-based conditioning regimens between 2015 and 2021 at Istanbul Medipol University Pediatric Bone Marrow Transplantation Unit. 47 patients were included to the study. Patients between 0 and 18 years of age that unerwent allogenic HSCT for TDT with a treosulfan or busulfan base conditioning regimen during the period of the study were included. In our center, Busulfan-Cyclophosphamide was the conditioning regimen between 2015-2017. Busulfan dose was adjusted according to patient's weight (3-15 kg: 5,11mg/kg/d; 15-25 kg: 4,9mg/kg/d; 25-50 kg: 4,1mg/kg/d; 50-75 kg: 3,3mg/kg/d; 75-100 kg: 2,7mg/kg/dd), and then recalibrated according to AUC. Cyclophosphamide dose was 50mg/kg/d, 4 days. We started using treosulfan-based conditioning in patients with any risk factor for busulfan toxicity in 2018. Conditioning regimen is; treosulfan 10-12-14 g/m^2/d (based on age) 3 days, fludarabine 40 mg/m^2/d 4 days, thiotepa 10mg/kg/d 1 day. GVHD prophylaxis was administered as ATG, methotrexate and cyclosporine. Prophylaxis of venoocclusive disease (VOD) with defibrotide was administered whether the patient had a risk factor or not. **Results:** A total of 47 patients undergoing 49 allogenic HSCT were included: 32 HSCT (65%) with busulfan and 17 (35%) with treosulfan based conditionings. Median age was 7,16 years (2,15-15,9), with no significant difference between the busulfan and treosulfan cohorts (7,9; 7,15). There were 22 (47%) girls and 25 (53%) boys. In the total study population, an HSCT was received from a matched sibling donor (MSD) by 31 patients (65%) and from a 10/10 matched unrelated donor (MUD) by 14 patients (29%). One patient had an 6/6 matched mother and one patient had a 6/6 matched father. There was a significant difference between busulfan and treosulfan cohorts: An HSCT was performed with a MSD by 26 patients (86%) in the BU-Cy group versus 5 (33%) in the TREO-FLU group. The stem cell source was bone marrow (BM) for 75% (n=37) of transplantations and peripheral blood stem cells (PBSC) for 22% (n=11). In one transplantation, both BM and PBSC were used. There was a significant difference between the groups: BM in 87% of transplantations for BU-Cy group; and 47% of transplantations in TREO-FLU group. Thirteen patients experienced acute graft versus host disease (GVHD): 8 patient with skin GVHD (17%: 5 in BU-Cy group, 15%; 3 in TREO-FLU group, 15%), 3 patients with gastrointestinal (GIS) GVDH (6%: 1 in BU-Cy group, 3%; 2 in TREO-FLU group, 11%), 2 patienst with both skin and GIS GVHD (4%, both of two were in the BU-Cy group). However, there were significant differences in donor types and stem cell sources between two groups. There are 3 patients following-up with chronic GVHD: 2 with bronchiolitis obliterans (1 in BU-Cy group and 1 in TREO-FLU group) and 1 patient with ocular GVHD (in BU-Cy group) Ten patients had VOD and all of them were in BU-Cy group (21% of whole population, 30% of BU-Cy group) .Four of 10 patients were followed-up in intensive care unit, and 3 of them had seizures therewithal. We did not have mortality due to VOD. In the total study population, primer engraftment failure number was 3 (6%: all in BU-Cy). We performed second HSCT in 2 of 3, and 1 of 3 died. Number of secondary graft rejection was 2 (4%: 1 in BU-Cy, 1 inTREO-FLU). Their bone marrow turned into TDT with normal series of granulocytes and platelets and parents did not prefer the second transplantation. Number of prolonged isolated thrombocytopenia was 2 (4%: both in BU-Cy): One had platelet recovery with eltrombopag treatment and the other died due to severe GIS GVHD. The median follow-up of all patients was 6 years (2-7 years). OS was 93,75% in the BU-Cy group and 100% in the TREO-FLU group. We had 2 transplant-related mortality: One patient was 15-year-old boy, underwent BU-Cy based allogenic HSCT

from his MSD. He had primer engraftment failure with aplasic bone marrow. The other was 12-year-old boy, underwent BU-Cy based allogenic HSCT from his MSD. He had severe GIS GVHD and prolonged isolated thrombocytopenia. **Conclusion:** Despite busulfan based conditionings used to be more common approach in pediatric patients underwent allogenic HSCT for TDT, treosulfan-based conditioning is gaining acceptance. Our retrospective study confirms the efficiacy and safety of both agents. Treosulfan, fludarabine and thiotepa seem to be appropriate for minimizing the risk of complications, particularly for VOD.

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OP 25

EFFECT OF GRAFT VERSUS HOST DISEASE PROPHYLAXIS ON THE LEUKEMIA FREE SURVIVAL IN PEDIATRIC PATIENTS WHO HEMATOPOETIC STEM CELL TRANSPLANTED FOR LEUKEMIA

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Objective: Hematopoietic stem cell transplantation (HSCT) is an important treatment modality for leukemia, the most common childhood malignancy. Graft versus host disease, one of the most important complication of transplantation, is the most important cause of morbidity and mortality. In our study, we aimed to show the effect of methotrexate doses given in transplants due to leukemia, the development of acute or chronic GVHD, on leukemia-free survival. **Methodology:** Patients who underwent HSCT due to leukemia, between April 2010-October 2020 at a pediatric transplantation unit were included in the study. Methotrexate doses given to patients; were grouped as 10mg/m^2 on day 1,3,6; 10mg/m^2 on day 1,3, 5mg/m^2 on day 6; 10mg/m^2 on day 1, 3; 10mg/m^2 on day 1 and 5 mg/m^2 on day 3,6; 10 mg/m^2 on day 1 and also 5 mg/m^2 on day 1. The effects of these groups on event-free and overall survival were evaluated. **Results:** Recurrence was not observed in 72 of 93 patients evaluated in the ALL group (77.4%). The conditioning regimens were considered TBI-Busulfan-based regimens. No significant difference was observed in terms of LFS. The absence of aGVHD in the ALL patient group significantly prolongs LFS, when evaluated according to CR1-2-3 groups, CR2 significantly extended the LFS time. Effect of GVHD prophylaxis on LFS was evaluated no significant effect of methotrexate dose on LFS was observed. **Conclusion:** The most important factor affecting leukemia-free survival is the state of remission. The longest duration of LFS was detected in CR1. The effect of methotrexate dose as GVHD prophylaxis has not been determined. There was no consensus in the studies on methotrexate doses in the literature. It is necessary to study with a larger cohort.

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Pediatric Oncology Abstract Categories

Rare Tumours and Histiocytosis

OP 26

LANGERHANS CELL HISTIOCYTOSIS IN TURKISH CHILDREN; 30 YEARS OF EXPERIENCE FROM A SINGLE CENTER

Selma CAKMAKCI [1], Arzu YAZAL ERDEM [1], Derya OZYORUK [1], Neriman SARI [1], Seda SAHIN [1], Meriç KAYMAK CIHAN [2], Suna Emir [3], İnci ERGURHAN ILHAN [1]

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Objective: Langerhans-Cell Histiocytosis, the most common histiocytic disorder, is characterized by inflammatory lesions with infiltrating CD1a+/CD207+ pathologic dendritic cells. The extent of disease is highly variable, from single lesion disease to life-threatening disseminated multisystem disease. We aimed to determine the demographic characteristics and the clinical outcomes of children with LCH. **Methodology:** The files of 81 patients diagnosed with LCH in Ankara Oncology Hospital, Dışkapı Children's Hospital and Ankara City Hospital between 1993 and 2023 were retrospectively analyzed. Data collected from the files included characteristics, age, sex, symptoms, physical examination findings, site of involvement, laboratory findings at diagnosis, procedure applied, treatment type used, and outcome. **Results:** The median age was 5 (0.1-17) with a median follow-up of 3 years (0.1-14) (Table1). The most common complaint was a bone lesion-related symptom; swelling (31%), pain (19%). Surgery was the only treatment in 19, chemotherapy in 22, radiotherapy in 1, surgery + chemotherapy in 35 (43%). Vinblastine + prednisolone was most commonly (36%) used. A patient with BRAF600VE was treated with vemurafenib. Recurrence was detected in 13 (16%) patients. Three patients died (3.7%) with refractory disease. **Conclusion:** Bone and skin were the most frequently involved systems in our study. Prognostic factors affecting event-free survival (EFS) were multisystem disease (5-year EFS 62% versus 87%, p=0.01) and hematologic system involvement (5-year EFS 42% versus 82%, p=0.02). Consistent with the literature, our overall survival (OS) rate was found to be high (5-year OS 95%). Patients with single-system disease had excellent survival (100%).

	No (n=81)	%
Median age at diagnosis (range)	5 (0,1-17 years)	
Age distribution		
≤24 ay	22	27
>24 ay	59	73
Sex		
Male	55	68
Female	26	32
Staging		
Single-system disease	57	70
Multisystem disease	24	30
Sites of involvement		
Bone isolated	38	47

Bone multiple	28	35
Skin	18	22
Lymph node	13	16
Lung	13	16
Liver	8	10
Hematologic	6	7
CNS/neurodegenerative	6	7
Diabetes İnsipidus	6	7
GIS	2	3
Chemotherapy protocol		
DAL-HX 83	31	38
LCH-III	19	24
LCH-IV	8	10

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Pediatric Oncology Abstract Categories

Supportive Care and Palliative Care
OP 27

EVALUATION OF VIRAL RESPIRATORY TRACT INFECTIONS IN PEDIATRIC HEMATOLOGY-ONCOLOGY PATIENTS BEFORE COVID-19 PANDEMY

Deniz Tugcu [1], Leyla Valıyeva [1], Sifa Sahın [1], Rumeysa Tuna [1], Mustafa Bılıcı [1], Ayşegül Unuvar [1], Serap Karaman [1], Gülşah Tanyıldız [1], Selda Hancerli [2], Sevim Mese [3], Ali Agacfıdan [3], Ayper Somer [2], Zeynep Karakas [1]

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Objective: Respiratory viruses are an important cause of morbidity and mortality in pediatric hematology oncology patients. We aimed to determine the infection rate, clinical and epidemiological characteristics of respiratory viruses in pediatric patients with hemato-oncological malignancy, aplastic anemia and congenital neutropenia and to show how these viruses affect the primary disease course and treatment. **Methodology:** Between August 2015 and December 2018, 97 patients aged between 5 months and 215 months who were admitted to Istanbul University, Istanbul Faculty of Medicine, Department of Pediatric Haematology-Oncology with acute respiratory tract infection findings and diagnosed with Haemato-Oncological Malignancy, Congenital Neutropenia, Aplastic Anaemia and who had viral respiratory panel were retrospectively analysed. In the viral respiratory panel test, nasal swab samples of the patients were evaluated by RT-multiplex PCR method. SPSS (Statistical Package for the Social Sciences) 22.0 programme was used for statistical analyses **Results:** A total of 97 patients, 52 males (53.6%) and 45 females (46.4%), aged between 5 months and 215 months (78.81±60.17 months, median 60 months) were included in the study. The most common viral respiratoty panel (VRP) positivity was observed between 5 months and 208 months and the mean age was 85.49±61.73 months (median=81 months). Although 44.3% (n=43) of the patients presented in winter and 23.7% (n=23) in autumn, VRP positivity was more common in patients presenting in spring (n=43, 70%) and winter (n=22, 51.2%) seasons. When the VRP results of the patients were analysed; 50.5% (n=49) were positive; 39.2% (n=38) were monoinfection, 11.3% (n=11) were co-infection) and 49.5% (n=48) were negative. When we looked at the VRP results, rhinovirus (hRV) was the most common virus with a frequency of 22.4% (n=11). Other viruses were Respiratory Synsititial Virus (RSV) A/B (14.2% n=7), Parainfluenza (14.2% n=7), Influenza (8.2% n=4), Coronavirus (8.2% n=4), Metapneumovirus (2.1% n=1), Mycoplasma pneumonia (6.1% n=3). Among the co-infections seen in a total of 11 patients, hRV and RSV A/B were the most common viruses accompanying other viruses with a rate of 63.6% (n=7). Among a total of 67 patients who were in various stages of CT and whose treatment was completed, the most common VRP positivity was seen in patients in the induction phase with a rate of 28.3% (n=19). Of the 12 patients with co-infection, 5 (41.6%) were in the induction phase. Cough (n=59 60.8%) and fever (n=47 48.5%) were the most common presenting complaints, accompanied by wheezing (n=17 17 17.5%), respiratory distress (n=11 11.3%), diarrhoea/vomiting (n=9 9.3%) and muscle pain (n=9 9.3%). VRP was positive in 43.9% of patients presenting with fever. The most common hRV virus was found most frequently in spring and winter seasons. Viral respiratory infection positivity was most frequently seen in ALL (n=16 33.3%), second most frequently in Hodgkin's Lymphoma (n=5 10.5%) and Neuroblastoma (n=5 10.5%). Among the patients, upper respiratory tract infection (URTI) (74.2%, n=72) was more common than lower respiratory tract infection (LRTI) (25.8%, n=25). The rate of LRTI in co-infections (28.0%, n=14) was higher than the rate of URTI (6.9%, n=5) and was statistically significant (p=0.021). When hemogram and biochemistry results were analysed, although neutropenia (50.5%) and lymphopenia (50.5%) were observed at a high rate in patients with positive VRI, they were not statistically significant when compared with VRP positivity. Of the patients with VRP positivity (50.5% n=49), 34.6% (n=17) required hospitalisation due to viral respiratory infection. Of the patients included in the study, 4 patients need intensive care unit due to bacterial pneumonia (Mycoplasma pneumonia and Pneumocystis jireovici), bleeding into a mass (hepatoblastoma) and pericardial effusion (peripheric T cell lymphoma). In 7 patients whose chemotherapy duration was prolonged, the duration of treatment prolongation ranged between 4 and 60 days (mean 19.29±20.69 and median 10 days). No VRI-related mortality was observed among the patients during the follow-up period. **Conclusion:** Identification of respiratory viruses in pediatric hematology oncology patients contributes to the management of their primary disease.

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Pediatric Oncology Abstract Categories Survivorship and Late side effects
OP 28

LONG-TERM EVALUATİON RESULTS OF OUR PATİENTS DİAGNOSED WİTH NASOPHARYNGEAL CARCİNOMA: A SİNGLE CENTER EXPERİENCE

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Objective INTRODUCTION: Nasopharyngeal carcinoma represents less than 1% of all childhood cancers. It is most common between 10-20 years with male predominance. Patients most often come with complaints of sizeable cervical lymph nodes, epistaxis, and headache. Since it is unsuitable for surgery due to its anatomical localization, the diagnosis can be made with tru-cut or lymph node excisional biopsy. The most common type of undifferentiated is seen in childhood. It is the type most closely associated with the Epstein-Barr virus. The mainstay of treatment is radiotherapy and chemotherapy. Survival rates are over 75%. However, surviving patients have to cope with the side effects of long-term radiotherapy and chemotherapy. Therefore, studies are ongoing to reduce treatment-related toxicities. **OBJECTIVE:** In this study, our aim was to examine the demographic data, long-term survival results, and late treatment-related side effects of our patients with nasopharyngeal carcinoma. **Methodology:** The data of patients diagnosed with nasopharyngeal carcinoma who were treated at Uludag University Faculty of Medicine, Department of Pediatric Oncology were analyzed retrospectively. **Results:** Twenty-four cases admitted to the pediatric oncology outpatient clinic between 2003 and 2023 were included in the study. The female/male ratio of the cases was 11/13. The mean age at diagnosis was 14.4±1.7. The most common complaints at admission were cervical lymphadenopathy and headache. Two of the cases were sibling cases diagnosed in different years. One patient was diagnosed with papillary adenocarcinoma and received only surgical treatment. Other 23 cases were diagnosed as non-keratinized undifferentiated carcinoma and received radiotherapy and chemotherapy. The patients were given a protocol consisting of bleomycin, epirubicin, and cisplatin. ICE protocol consisting of ifosfamide, etoposide, and carboplatin was given to relapsed cases. Radiotherapy was given after 3 cycles of chemotherapy. In the follow-up of the cases, one case died due to refractory disease, three cases due to relapse, and one case due to sepsis. The cases were followed up for an average of 5.8 years (min: 1 year, max: 19 years). The survival of our cases was 79.2% in the 20-year follow-up. The most common long-term side effects developing secondary to treatment were xerostomia (n=12), dysphagia (n=12), and malnutrition developing secondary to these. Six cases had hypothyroidism and one case had hyperthyroidism. Fibrosis secondary to radiotherapy was seen in 10/24 patients. Apart from these side effects, seven cases had hearing loss, recurrent otitis, and nascent speech. **Conclusion:** Nasopharyngeal carcinoma is a rare tumor in childhood. Because it is rare, treatment approaches have been created based on adult patients. Depending on the doses of radiotherapy, and chemotherapy taken at an early age, many side effects that reduce the quality of life are seen in patients who live for a long time. Studies are needed to reduce these side effects. We wanted to contribute to the literature by publishing the long-term results of our cases.

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