ongoing chemotherapy. Features similar to immune dysregulation in HLH also occur during pediatric acute leukemias. This immune dysregulation results unexpected cytopenias, fever, and splenomegaly in children with acute leukemia. We aim to analysis the pediatric acute leukemia pateints who had unexpected, prolonged cytopenias, and did not full-fill the HLH-2004 criteria set and received pulse methylprednisolone therapy up to three days **Methodology:** Data was analyzed retrospectively. The diagnosis of HLH was defined according to the HLH-2004 criteria set but two criterias (NK cell activity and sCD25 level) of HLH diagnosis were not studied due to lack of necessary equipment. Treatment response was defined as increasing neutrophil count above 500/mm3 in patients within the first seven days. **Results:** 12 patients received steroid for unexpected, prolonged cytopenias. Five or six of six criteria was not found. Four criteria in four, three criteria in five and two criteria in three patients was determined. All patients had cytopenia at least two of three lineages in peripheral blood, one of which was neutropenia. Hemophagocytosis in bone marrow sample was detected in eight patients. Ten patients (87%) recovered within the first seven days. Seven of nine thrombocytopenic patients recovered. **Conclusion:** In this report, the efficiency of short-term steroid treatment was demostrated in patients with unusual cytopenias who did not full-fill HLH criteria.

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EVALUATION OF VACCINATION RESPONSE IN CHILDREN AFTER TREATMENT FOR ACUTE LEUKEMIA

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Objective: Our study aims to evaluate the patients' immunity regarding childhood vaccination after leukemia treatment and determine the vaccines that require additional doses. **Methodology:** Sixty-six patients who were followed up with the diagnosis of ALL and AML between 2013 and 2016 were included in our study. The patient's gender, age at diagnosis, leukemia type, leukemia risk groups, vaccination status before chemotherapy (CT) and serologies of hepatitis A, hepatitis B, varicella, measles, rubella, mumps at the end of CT were recorded. **Results:** At the end of the treatment, loss of protective antibody response against hepatitis A (47.4%), hepatitis B (68.2%), varicella (64.2%), measles (45.5%), rubella (43.9%), and mumps (50%) vaccines were shown. Loss of protective antibodies against hepatitis A (66.7%), hepatitis B (100%), varicella (100%), measles (100%), rubella (91.7%), and mumps (91.7%) in high-risk ALL patients was higher than patients in standard-intermediate risk ALL. **Conclusion:** Loss of humoral immunity against hepatitis A, hepatitis B, varicella, MMR was shown in patients with leukemia at the end of the treatment. Due to the significant decrease in hepatitis B and MMR protective antibodies in the high-risk group, we recommend patients with leukemia who have completed chemotherapy to be vaccinated with hepatitis B vaccine three months and MMR vaccine six months after the treatment.

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PP 74

PONATINIB EXPERIENCE IN A PEDIATRIC CHRONIC MYELOID LEUKEMIA PATIENT

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Objective: Chronic myeloid leukemia (CML) is rarely seen in children. The development of myelofibrosis in CML is not uncommon and is associated with a poor prognosis. In cases unresponsive to treatment, tyrosine kinase mutation should be checked for drug resistance, second generation tyrosine kinase inhibitor (TKI) drugs (dasatinib/nilotinib) should be switched to and a suitable donor for bone marrow transplantation should be sought. Third-choice TKI can be used in children who are unresponsive to treatment and do not have a suitable donor. **Materials and Methods:** Experience of third-choice TKI(ponatinib) in a child with CML diagnosis due to unresponsiveness to treatment. **Results:** A 5.5-year-old female patient with no known disease was referred to us because of hepatosplenomegaly (liver 5 cm, spleen 10 cm). There was no laboratory disorder except for anemia (hgb 8.9 g/dL) and high LDH (1104 U/L). WBC was $11.1 \times 10^3/\mu$L neu $6.92 \times 103/\ \mu$L plt 304000/μL. Peripheral smear showed leukoerythroblastosis. Bone marrow biopsy result was evaluated as compatible with myelofibrosis and an increase in blast rate from 8% to 18% in the bone marrow. The patient was diagnosed CML accelerated phase with cytogenetic (46,XX,t(9:22)(q34;q11))and translocation (t(9:22)- p210,BCR/ABL positive) results and. Imatinib treatment was started at 400 mg/m^2.The copy number of BCR-ABL p210 checked before treatment was 72% IS. However, the patient developed febrile neutropenia, and imatinib dose reduction (< 200 mg/m^2) and interruption were required in the follow-up.Under imatinib treatment, BCR-ABL copy number was 16%IS at 1 month, 11%IS at 3 months, and 95%IS at 5 months. Due to the increase in the BCR-ABL copy number, nilotinib was switched to as a second-choice TKI(230 g/m2/dose, in 2 doses).No mutation could be detected in the c-ABL gene, which was examined for tyrosine kinase resistance. HLA groups were sent from the family and compatible donors were not found. Due to severe neutropenia in the follow-up, nilotinib could be continued at 50% dose. Under nilotinib treatment, the BCR-ABL copy number was 13% IS at 1 month, 10% IS at 2 months, and 31% IS at 3 months. The patient was started on ponatinib (18 mg/m^2/day)

as a third choice TKI. However, due to the deep neutropenia of the patient, it was possible to continue with a dose of 10 mg/m^2 from the 2nd week. With this dose, the neutrophil is around 0.8-1 $\times$ 10^3/μL. Under ponatinib treatment, BCR-ABL copy number was 6.6% IS at 1 month, 0.8% IS at 3 months, 0.09% at 5 months, and 0.05% at 6 months. No significant side effects were observed except neutropenia. **Conclusion:** There is no approved treatment in pediatric CML cases where the second choice TKI fails and there is no donor for transplantation. FDA approval for ponatinib in adult patients was obtained in December 2020. Ponatinib is a natural or mutant pan-BCR-ABL mutation inhibitor. It also inhibits VEGFR, FGFR,PDGFR,EPH and SRC kinases as well as KIT,RET,TIE2 and FLT3. The use of ponatinib should be evaluated by monitoring side effects/tolerance in pediatric cases where there is no other treatment option, and there is a need for studies on this subject.

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INHERITED BONE MARROW FAILURE DISEASES

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INVESTIGATION OF SALIVARY miR-9, miR-34a ve miR-196a LEVELS IN FANCONI ANEMIA AND ORAL SQUAMOUS CELL CARCINOMA PATIENTS

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Objective: Fanconi anemia (FA) is a rare bone marrow deficiency syndrome due to the DNA repair gene mutations, and Oral Squamous Cell Carcinoma (OSCC) is seen more frequently in FA patients than in the general population. The dysregulation of PI3K and Wnt signaling has been implicated in OSCC pathogenesis and abnormal expressions of miRNAs (a class of non-coding small regulatory RNAs) associated with these signaling pathways has been reported in OSHK patients. Salivary miRNAs are valuable biomarker candidates for OSCC development and prognosis. In this study, salivary levels of miR-9, miR-34a and miR-196a miRNAs related to PI3K and Wnt signaling pathways were examined in OSCC and FA patients and compared with the healthy control group. **Methodology:** Saliva samples were collected from 89 subjects including 25 OSCC patients, 24 FA patients and 40 healthy controls. Total RNA was isolated using Quick-RNA Miniprep Kit (Zymo Research) due to the kit instructions. cDNA was generated with miRCURY LNA miRNA PCR Assay (Qiagen, Hilden, Germany) and Quantitative real-time PCR was performed with miRCURY LNA SYBR Green PCR Kit (Qiagen, Hilden, Germany) according to the manufacturer's protocol. For the normalization of the expression levels of each miRNA, the mean expression U6 SnRNA was used as reference. The ΔΔCt value and the normalized miR-9, miR-34a and miR-196a salivary levels were calculated with Livak Method. **Results:** Our results showed that miR-9 and miR-34a levels in OSCC patients were significantly lower compared to healthy control groups (p= 0,01 and p= 0,012), and there was no significant difference in miR-196a levels (p> 0,05). In FA patients, miR-9 and miR-34 levels were lower than in control groups, likewise the OSCC patients (p =0,017 and p =0,014). There was no significant difference between miR-9, miR-34a, and miR-196a levels of FA patients and OSCC patients (p >0.05). **Conclusion:** According to our results, low levels of miR-9 and miR-34a in saliva are biomarker candidates that may be important for OSCC development. In FA patients, close follow-up of the levels of miR-9 and miR-34 would be appropriate considering OSCC development. Further studies are needed to confirm the potential of miR-9 and miR-34a as biomarkers for OSCC.

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A NOVEL MISSENSE MUTATION OUTSIDE DNAJ DOMAIN OF DNAJC21 IS ASSOCIATED WITH SHWACHMAN-DIAMOND SYNDROME

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Shwachman-Diamond Syndrome (SDS) and related bone marrow failure disorders are characterized by early onset pancytopenia with a hypocellular bone marrow, short stature, and pancreatic insufficiency, along with an increased risk for myeloid malignancies. Recently, several cases with an SDS-like syndrome have been reported to harbor mutations in the DNAJ domain of *DNAJC21*. Here, we report an intriguing case