ages of 1-7.The first line therapy consists of intravenous immunoglobulin, anti-D immunoglobulin or corticosteroids. Second-line treatment options are immunosuppressive therapy, Rituximab.Thrombopoietin receptor agonists are used, which increase platelet production in the bone marrow. Our case report on a child with refractory chronic ITP, who failed the first and second line therapy.

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PP 63

ESSENTIAL THROMBOCYTOSIS IN CHILDREN

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Objective: Sporadic essential thrombocytosis is a very rare disease in the childhood age group and its frequency has been reported as 1/1,000,000. WHO 2008 essential thrombocytosis diagnostic criteria; high platelet count for more than one year (>450 × 109/l), exclusion of reactive or secondary causes of thrombocytosis (iron deficiency, megaloblastic anemia, acute phase reactants, trauma, operation), no family history of myeloproliferative neoplasm and thrombocytosis, and WHO It can be summarized as the absence of myeloid neoplasm criteria. **Methodology:** In this study, seven cases diagnosed as sporadic essential thrombocytosis in our Pediatric Hematology clinic are presented. Six of the patients were girls and one was a boy. The median age at presentation was 13 years (the youngest 5 months, the oldest 15 years old). Application complaints: Headache, vertigo and tinnitus in adolescent children were not present in young children, they were detected incidentally. Thrombus was not detected in any patient. The median platelet count at diagnosis was 1442 × 109/l (range 963- 2438). **Results:** An increase in megakaryocytes was detected in bone marrow aspiration, no cytogenetic anomalies were found. Jak-2 (V617F) mutation was detected in one case and CALR mutation in two cases. No MPL (W515L) mutation was found in any case. In one case with a CALR mutation, a known type 2 mutation was detected, and in the other a new, previously unidentified mutation was detected. In the other four cases, no clonality was detected. Three cases with mutations and two cases with no mutations are being followed up with hydroxyurea therapy. The other two cases are using low-dose aspirin. Follow-up periods range from six months to nine years. No complications developed. **Conclusion:** Thrombocytosis is a common problem in childhood. Reactive and secondary causes are usually identified. Essential thrombocytosis is a diagnosis that should be considered after excluding other causes. Mutation studies should be performed in pediatric patients who meet the WHO 2008 criteria. While Jak-2 (V617F), CALR and MPL (W515L) mutations are seen in 90% of cases in adults, these three mutations are only seen in 25% of the childhood age group. The high number of cases with no mutations indicates that new candidate genes should be sought and studied.

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RED BLOOD CELL DISORDERS

PP 64

GARDNER DIAMOND SYNDROME: A CASE REPORT

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Case report: Gardner Diamond Syndrome (GDS) is a rare autoimmune disorder also known as autoerythrocyte sensitization syndrome represented with skin lesions. These lesions mostly occur after a triggering factor. The pathophysiology of the disease is not completely understood yet. In this case report, the characteristic features of GDS is presented; furthermore, our aim is to emphasize the effect of emotional stress during the disease.

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IMMUNODEFICIENCIES / NEUTROPHIL DISEASES

PP 65

MYELOPEROXIDASE DEFICIENCY: SINGLE CENTER EXPERIENCE

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Objective: Myeloperoxidase (MPO) is a hemoprotein expressed in azurophilic granules of neutrophils and lysosomes of monocytes. It is caused by mutations in the MPO gene on chromosome 17 and is estimated to affect 1:2000-4000 people. It is the most common inherited defect of phagocytes. Microbial killing is impaired in patients with MPO deficiency, but most patients are asymptomatic, except for diabetic patients. In this article, we aimed to present our patients diagnosed with primary MPO deficiency. **Methodology:** During the investigation for the etiology of neutropenia in the hematology department of our hospital, patients who were diagnosed with MPO deficiency were examined. In the evaluation of the patients, it was observed that the neutrophil count in the hemogram printout and the counted neutrophil count in the peripheral smear were inconsistent. We performed MPO staining with FCM from the peripheral blood samples of the patients and we found that the neutrophils were MPO negative. **Results:** A 1-day-old male patient has no additional disease (c.608A>C H mutation). C.578G>C mutation was detected in the follow-up due to ANA+ in a 6.5-year-old female patient. A c.2031-2A>C mutation was found in the 18-year-old patient who was being followed up with the diagnosis of Granulamatous Polyangiitis and his sister. A c.493del mutation was detected in an 11-year-old patient who was diagnosed with ITP 5 years ago. The noval mutation was detected in the patient followed up with the diagnosis of

retinoblastoma. **Conclusion:** MPO deficiency may occur primarily as well as secondary. A number of point germ line mutations cause primary MPO deficiency. Most patients asymptomatic without an increase in infection. Severe infectious complications were not observed in any of our patients. We wanted to emphasize that MPO deficiency should also be kept in mind in patients whose neutropenia etiology was investigated.

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LEUKEMIA

PP 66

CHARACTERISTICS AND OUTCOME OF T(8;21)-POSITIVE CHILDHOOD ACUTE MYELOID LEUKEMIA: A SINGLE INSTITUTION'S EXPERIENCE

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Objective: Compared with other cytogenetic acute myeloid leukemia (AML) groups, patients with core-binding factor AML (CBF-AML) are considered as a favorable AML risk group based on their high remission rate and survival probabilities. However, up to 30-40% of these patients can still relapse after standard intensive induction and consolidation chemotherapy. **Methodology:** From 2004 to 2020, 147 AML patients reviewed. Ten of 147 patients were followed up with t(8;21) chromosomal anomaly. The t(8;21)(q22;q22) was detected by reverse transcription polymerase chain reaction (RT-PCR) and/or floresan in situ hibridizasyon (FISH). We analyzed patients' demographic data: sex, white blood cell count at diagnosis, central nervous system status, additional cytogenetic anomaly and recurrence rates, stem cell transplant status and survival rates. **Results:** Two of 10 patients were female. The median age was 10 years (3-17 years). Median followup was 36 months (2-114 months). The mean white blood cell count of 10 patients was 21.5 ($\times$ 109/l) at diagnosis. One out of 10 patients had granulocytic sarcoma and 2 had central nervous system involvement. Additional cytogenetic anomalies were detected in 90% of the patients, of which 2 relapsed and 3 died. One patient received hematopoietic stem cell transplantation and died because of HSCT complications. **Conclusion:** Recent studies show that CBF-AML includes different groups with different clinical outcomes. We found that 50% of our patients achieved complete remission and 50% experienced relapsed disease or death. After we were able to monitor the t(8;21) level with RT-PCR, we diagnosed relapsed disease in 1 patient with additional cytogenetic anomaly. RT-PCR is essential for optimal handling of these patients to predict patients' relapse risk and to detect minimal residual disease.

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PP 67

BK-VIRUS ASSOCIATED HAEMORRHAGIC CYSTITIS CONCOMITANT WITH CHEMOTHERAPY IN AN ADOLESCENT GIRL WITH ACUTE LYMPHOBLASTIC LEUKEMIA

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Case report: Haemorrhagic cystitis (HC) is characterized by focal or diffuse haemorrhagic and inflammatory changes of the bladder mucosa. Polyoma BK virus (BKV) infection is an important underlying condition that provokes hematopoietic stem cell transplantation (HSCT)-related HC. Although commonly reported in transplant recipients, BKV associated HC, and tubulointerstitial nephritis rarely occurs in paediatric acute lymphoblastic leukemia (ALL) patients receiving chemotherapy. A 15-year-old girl diagnosed with T cell ALL, receiving high-risk chemotherapy protocol, complained about dysuria and lower abdominal pain with macroscopic haematuria. Her complaints started under meropenem, teicoplanin, amikacin, and caspofungin treatment due to neutropenic fever with severe mucositis. There wasn't any bacterial growth in the urine or blood culture. PCR analysis detected 2,2 $\times$ 109 copies/mL of BKV in urine. The antibiotics other than ciprofloxacin were discontinued. Her complaints are alleviated day by day. She did not experience any urinary symptoms or haematuria, and the BKV copy number declined to 3,3 $\times$ 107 copies/mL during follow-up.Contributing factors of BKV associated HC are highly relevant in HSCT recipients. However, patients receiving intensive chemotherapy may have similar conditions. A predisposing and potential manageable factor such as BKV should be searched in paediatric haematology practice.

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PP 68

A CASE OF METHOTREXATE-INDUCED PHOTOSENTIVITY REACTION

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Case report: Methotrexate is an essential drug effectively used in acute lymphoblastic leukemia. Doses above 500 mg/m2 are defined as high-dose methotrexate (HDMTX). Since HDMTX is known to cause serious morbidity, it is given with a standard