

to retrospectively evaluation the patients diagnosed with LCH who were followed up and treated in our clinic. **Methodology:** The data of patients over the age of 18 years who were followed up and treated in Bozyaka Training and Research Hospital Hematology Clinic between 2015-2021 were scanned retrospectively from the hospital system. **Results:** Data of 6 patients were obtained. The mean age of the patients was 33.6. There was no difference between the genders. Pain was the reason for admission in 4 patients and was the most common symptom. While the most frequently involved system was the skeletal system with 5 patients, lung involvement was seen in 2 patients. Vinblastine and prednisolone combination therapy was given to 1 patient, who developed steroid-related avascular necrosis. One patient who was planned for combination treatment **Conclusion:** LCH is a rare disease especially seen in children and young adults. It can involve the skeletal system, lungs, and other organs. The prognosis is often good with excision of the lesion or systemic treatment.

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#### PP 52

##### PILOMATRIX CARCINOMA WAS BEYOND ANY EXPECTATIONS: A CASE REPORT OF CARCINOMA CLINICIAN SHOULD BE AWARE OF IT

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**Introduction:** Pilomatrix carcinoma is a rare cutaneous tumor derived from follicular matrix with about 150 reported cases and it is considered as locally aggressive tumor with a tendency to recur. In addition to lymph node metastases and poor prognosis, it is non-chemotherapy responding in systemic metastasis. **Method and result:** A 37-year-old Libyan man presented with two large, coalesced nodules in the face measuring about 3\*2 cm and 3\*3 cm, treated as a case of lipoma by surgical excision based on clinical diagnosis, reappearing of larger nodule 9\*6\*4 cm involving almost all the left cheek, surgical excision was done with histopathological features of pilomatrix carcinoma infiltrating to the subcutaneous adipose tissue and deep striated muscle with no clear margins. Conventional radiotherapy by electron beam was started using the linear accelerating machine, with total radiotherapy dose 60 gray (Gy) in 30 fraction for six weeks. No local recurrences, nor lymph node or systemic metastasis since June 2020. **Conclusion:** Pilomatrix carcinoma must always be considered in the differential diagnosis of nodular tumors of the head-and-neck due to high recurrence rate after simple excision. Furthermore, local radiotherapy post incomplete surgical excision is the best adjuvant therapy.

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#### PP 53

##### WHAT HAVE WE EXPERIENCED WITH COVID-19 IN PATIENTS WITH HEMATOLOGICAL DISORDERS?

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**Objective:** Patients with cancer are considered highly vulnerable to the COVID-19 disease. However, there are still few data in hematologic patients. Some small studies have shown a high mortality on patients with hematologic malignancies and COVID-19. In this study we aim to report a single center experience of the hematologic patient population with COVID-19 disease. **Methodology:** This single centre, retrospective, cohort study included a total of 111 adult patients (aged  $\geq 18$  years) with diagnosis of neoplastic and non-neoplastic hematologic diseases between March 2020 and August 2021. Ethics committee approval was obtained from the Istanbul University Istanbul Faculty of Medicine Clinical Research Ethics Committee. Categorical variables were compared using Pearson's Chi-square test. STATA16-MP was used for the statistical analysis. **Results:** A total of 111 patients (median age:55) with hematologic disease were diagnosed with COVID19. Ninety patients had neoplastic hematologic disorder. Forty-five patients were receiving anti-neoplastic treatment at the time of COVID19 diagnosis. A total of 21 patients (overall mortality rate:19%) died and 19 of them had neoplastic disorder. The malignancy mortality rate was estimated to be 21%. 45 of 90 cases were receiving chemotherapy. Ten of these 45 patients (22%) died due to COVID19 disease. **Conclusion:** In our study the majority of patients who died due to COVID-19 had hematological malignancies. The cytokine storm which affects the clinical outcome in COVID-19 may contribute to dismal prognosis in hematologic malignancies in which cytokine increase is a part of process. Most of the succumbed patients were relapsed refractory multiple line treated which may reflect the immune insufficiency. It seemed COVID-19 progress is mostly poor in hematologic malignancies compared otherwise healthy people.

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#### PP 54

##### A RARE CAUSE OF ANEMIA IN ADULTHOOD CONGENITAL DYSERYTHROPETIC ANEMIA

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**Objective:** Congenital dyserythropoietic anemia is a group of diseases characterized by ineffective erythropoiesis and multinuclear erythroblasts, mostly diagnosed in childhood. Although there are 3 main types, type II is the most common. We present our patient with congenital dyserythropoietic anemia, who was not diagnosed until the age of 49, to contribute to the literature. **Case report:** A 49-year-old male patient was admitted to our hospital with abdominal pain, weakness and yellowing of the eyes. His examinations revealed splenomegaly, cholelithiasis, anemia and hyperbilirubinemia. In the patient's anamnesis, he stated that he had jaundice and weakness since childhood, and that he knew that he had abdominal pain and spleen enlargement with advancing age. **Methodology:** Bone marrow biopsy was performed to the patient for a different diagnosis and cause. Binuclear erythroblasts were observed in the patient (fig. 1). As a result of the new generation sequencing performed on the patient who was evaluated as familial non-immune hemolytic anemia, c.1733T>C homozygous mutation in exon 15 of the SEC23B gene was detected and a diagnosis of congenital dyserythropoietic anemia type II was made. **Results:** Congenital dyserythropoietic anemias (CDA) represent a large group of diseases that mainly result in ineffective erythropoiesis. Morphological changes observed in the bone marrow over a long period of time were its main diagnostic features. Together with 3 main subtypes, they are examined in a total of 5 subtypes. CDA type II is most common. Clinically normal or slightly increased reticulocyte count is characterized by a variable degree of normocytic anemia. **Conclusion:** Diagnosing CDA cases: It is closely related to the clinician's ability to remember and access genetic tests, especially in advanced ages. Considering that access to genetic tests will increase in the future, many undiagnosed cases may come up. Although our treatment possibilities are limited in the current situation, future treatment methods are promising. However, studies are still needed to understand this disease and its mechanisms.

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#### QUALITY IMPROVEMENT / PATIENT SAFETY

PP 55

#### HEMATOLOGIC REFERENCE VALUES FOR HEALTHY ADULT SAUDIS

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**Background:** Laboratory hematological tests are widely used in clinical practice to assess health and disease conditions. Although, the International Federation of Clinical Chemistry and Laboratory Medicine (IFCC) and the Clinical and Laboratory Standards Institute (CLSI) recommended that reference

ranges should be established for each region, to the best of our knowledge, no study has described the reference values of routine hematological parameters in healthy Saudi adults. **Objectives:** To provide reference values of routine hematological parameters in Saudi adults according to age and gender. **Material and Methods:** A total of 827 adults potentially healthy Saudi participants with age ranging from 15 to 65 years were enrolled in this cross-sectional study from the central province of Saudi Arabia, Riyadh city. **Results:** The reference values of routine hematological parameters (full blood count, hemostatic profile, and biochemical tests of serum hematinic) according to gender were provided in detail (mean, SD, range, median, upper and lower limits) after exclusion of 157 due to various reasons. No difference in any hematological values were observed in relation to age. Current study hematological parameters' reference ranges were mostly different to the universal established ranges. **Conclusion:** This novel study provides the reference ranges of routine hematologic parameters for adult Saudi population for accurate assessments and appropriate management of routine clinical care, hence, to improve quality in health care.

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#### PEDIATRIC HEMATOLOGY ABSTRACT CATEGORIES

##### COAGULATION AND FIBRINOLYSIS DISORDERS

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#### A CASE REPORT WITH SEVERE CONGENITAL FACTOR XIII DEFICIENCY AND AN UNCOMPLICATED PREGNANCY AND BIRTH PROCESS

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**Introduction:** Factor XIII deficiency is an extremely rare type among bleeding diathesis. In factor XIII deficiency, normal results of coagulation screening tests are expected. It usually does not cause spontaneous bleeding. Apart from bleeding diathesis, it may cause delayed wound healing and recurrent spontaneous abortions in women. Here, we present a 32-year-old case with severe congenital factor XIII deficiency who had an uncomplicated pregnancy and birth with regular replacement therapies. **Case report:** A 32-year-old patient with severe congenital factor XIII deficiency, who had a history of spontaneous abortion at the 11th week of her first pregnancy, applied to our center with a request for childbirth. It was learned that the factor XIII levels of the patient could not be measured, that she was using plasma-derived FXIII concentrate at a dose of 25 units/kg every time once a month, and in cases where this could not be obtained, 5 units/kg