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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 dyserythropetic 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

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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

PP 56

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

cryoprecipitate was given instead. After the completion of the pre-pregnancy assessments, starting 3 months before the planned pregnancy and continuing for the whole pregnancy and for 3 months after birth, 25 units/kg plasma-derived concentrate at a dose of 25 units/kg was applied each time and every two weeks, and in cases where this could not be provided, the follow-up was continued by applying cryoprecipitate at a dose of 5 units / kg instead. During this whole process, FXIII levels ranged between 70% and 100%. The patient, who developed an abortion risk due to decidual bleeding in the first trimester, was hospitalized and an additional 25 units / kg plasma-derived FXIII concentrate was administered and a parenteral dose of 30 mg / kg tranexamic acid was applied until the signs of decidual bleeding disappeared. An additional 50 units/kg dose of plasma-derived FXIII concentrate was administered to the patient 30 minutes before birth who had a planned delivery by cesarean section at 38 weeks of gestation, and 30 mg/kg parenteral tranexamic acid was administered for 7 days following the delivery. FXIII level was detected 50% in the child of a healthy, 3500-g born boy. The patient and her baby, who are in the first year after birth, are followed up without any complications, and prophylactic plasma-derived FXIII concentrate or cryoprecipitate is administered to the patient once a month. **Discussion and Conclusion:** Inherited bleeding diathesis lead to an increased risk of bleeding and abortion in obstetric patients. Factor XIII deficiency is an extremely rare type among them. FXIII has a role in angiogenesis as well as hemostasis. Therefore, wound healing and tissue repair are impaired in Factor XIII deficiency. The risk of premature separation of the placenta, miscarriage especially in the first trimester, and postpartum uterine bleeding are increased in FXIII deficiency. Tranexamic acid can be used safely in obstetric patients with bleeding diathesis. It may be possible to ensure that patients with factor XIII deficiency have an uncomplicated pregnancy and delivery with regular follow-ups, regular prophylactic factor preparations, plasma replacements if they are not found, and in cases of bleeding, with additional doses of factor preparations or plasma replacement applications with tranexamic acid.

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EVALUATION OF THE FREQUENCY OF ARTERIAL AND VENOUS THROMBOSIS AND PREDISPOSING FACTORS IN PATIENTS USING ELTROMBOPAG

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Objective: Eltrombopag is a small molecule thrombopoietin-receptor agonist used orally for the treatment.There is a high risk of thrombosis associated with the use of Eltrombopag. Our aim in this study is evaluating the incidence of arterial and venous thrombosis in patients using Eltrombopag and followed up in our center with the diagnosis of ITP, MDS and aplastic anemia, and contributing to the literature with the data of Central Black Sea by retrospectively evaluating the predisposing factors. **Methodology:** In this study, the data of 144 patients who were treated with Eltrombopag with the diagnosis of ITP, MDS and aplastic anemia at Ondokuz Mayıs University Faculty of Medicine Hematology Clinic between 2009-2019 were analyzed retrospectively. The data of the patients were obtained retrospectively from the hospital management information system. **Results:** The study included 144 patients who treated with Eltrombopag. 66 (45.8%) of the patients were male and 78 (54.2%) were female. The mean age of the patients was 54.12 ± 20.08 years. 102 (70.8%) of the patient were diagnosed with ITP, 31 (21.5%) with aplastic anemia and 12 (7.6%) with MDS. Thrombosis was observed in 7 (4.9%) of 144 patients who treated with Eltrombopag. Venous thrombosis was found in 6 (4.2%) of these patients and arterial thrombosis was found in one patient (0.7%). **Conclusion:** Eltrombopag treatment poses a risk for thromboembolic events. The treatment process should be followed closely especially in patients with known risk factors for thrombosis.

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ETIOLOGY, TREATMENT AND FOLLOW-UP OF THROMBOSIS IN CHILDREN, ONE CENTER PROSPECTIVE TRIAL

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Objective: The aim of the study; To determine the frequency, etiological factors, treatment, long-term follow-up and recurrence rates of thrombosis in children. **Methodology:** Children with thrombosis in Ankara City Hospital between December 2018 and August 2021 were included. Patients were called or examined at 6–12-month intervals. **Results:** A total of 328 patients with a mean age of 6.9 were included. Catheter-related thrombosis was present in 52.7%.There were 14% arterial thrombosis and 59% venous thrombosis. Intracardiac thrombosis 16.2%, pulmonar thrombosis 2.4%, serebral thromboembolism %20 were detected.In the treatment, subcutaneous ondansetron (78.6%) was used mostly, but intravenous ondansetron was given in 6 patients and TPA was given 20 patients.In a mean follow-up of 15.8 months, 5 (1.5%) patients died due to thrombosis. **Conclusion:** Determining the etiological factors of patients with recurrence thrombosis is important for the duration of treatment in the follow-up.

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