

(SLL). Although it is relatively rare, appearing in 2% to 10% of CLL patients, RS often proves to be lethal due to its rapid progression and the scarcity of specific therapies. Venetoclax, a BCL2 inhibitor, has demonstrated efficacy in CLL but its role remains less explored in RS. Hence, there is a paucity of information regarding the direct employment of Venetoclax in the treatment regimen for RS. This study presents a case of Richter transformation being managed under treatment with Venetoclax. **Case report:** Case: A 51-year-old female patient, diagnosed with CLL with negative 17p deletion following investigations in 2015 due to autoimmune immune thrombocytopenia (ITP) and lymphocytosis, was given 6 cycles of FCR (fludarabine, cyclophosphamide, rituximab) due to steroid-resistant autoimmune thrombocytopenia, and complete response (CR) was achieved according to iwCLL criteria. After remission, the patient was monitored without treatment, and in 2020, full blood count, biochemical analysis, and peripheral smear were performed due to fatigue symptoms. The complete blood count showed leukocytes: 44600/mm³, lymphocytes: 39000/mm³, MCV: 86 fl, and hemoglobin: 9.5 g/dL. The patient, with no signs of hemolytic anemia, had no nutritional (Fe, B12, folate) deficiency, and normochromic normocytic anemia was detected. There were no mutations in the immunoglobulin heavy chain variable region (IGHV) genes. The patient, evaluated as relapsed stage 3 disease, was started on venetoclax-rituximab treatment. In the 11th month of the treatment, due to symptoms of fatigue, fever, night sweats, and weight loss, a bone marrow biopsy was performed after pancytopenia was observed, and a diagnosis of diffuse large B-cell lymphoma was made. Due to Richter transformation, DA-R-EPOCH (dose-adjusted rituximab, etoposide, prednisolone, vincristine, cyclophosphamide, doxorubicin) treatment was initiated. After 4 cycles of DA-R-EPOCH treatment, single-agent ibrutinib was started due to treatment-resistant disease and an ECOG performance score of 2. The patient, whose disease continued to progress under ibrutinib treatment, died from septic shock. **Conclusions:** This case underscores the complexities in treating Richter syndrome, particularly with venetoclax, and emphasizes the need for careful monitoring and understanding of potential transformations. The development of Richter transformation under venetoclax treatment highlights an area that requires further investigation and consideration in the management of CLL. Prospective studies and a comprehensive approach are vital to enhancing treatment strategies and improving outcomes for patients with this aggressive form of lymphoma.

<https://doi.org/10.1016/j.htct.2023.09.055>

Adult Hematology Abstract Categories

Chronic Myeloproliferative Diseases

PP 06

BIOMEDICAL ANALYSIS OF RED BLOOD CELLS IN POLYCYTHEMIA VERA, APPLICATION OF RAMAN SPECTROSCOPY

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Objective: Polycythemia vera (PV) is a chronic myeloproliferative neoplasm characterized by increased red blood cell mass. Excess erythrocytosis leads to elevated hematocrit, resulting in increased blood viscosity, a condition that promotes thrombosis. For years, red blood cells (RBCs) in PV were considered to be morphologically and functionally normal. This analysis aimed to check whether there are biochemical alterations in RBCs in PV that may be associated with thrombotic complications. **Methodology:** We included 5 patients with PV and 5 healthy individuals in the preliminary analysis of the biochemical properties of isolated RBCs focused on different forms of hemoglobin and heme. The analysis was conducted using Raman spectroscopy. **Results:** The results of the Raman spectra obtained from isolated RBCs suggest a larger contribution of ferrous heme iron in the sample of a patient with PV compared to a control sample. In the PV sample, a greater contribution of the high-spin heme iron, a molecular state typical for deoxyhemoglobin, was observed, which stays in line with higher ferrous content. The effect may indicate a weaker linkage of the protein with oxygen. **Conclusion:** Our analysis suggests the occurrence of biochemical alterations in RBCs in PV, together with RBC overproduction. Changes in the structure of hem and hemoglobin affect oxygen affinity. Our future study will focus on determining if described alterations in RBCs may contribute to the pathogenesis of thrombotic complications in PV.

<https://doi.org/10.1016/j.htct.2023.09.056>

PP 07

DISCONTINUATION OF TYROSINE KINASE INHIBITORS IN TUNISIAN CHRONIC MYELOID LEUKEMIA PATIENTS

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Objective: Some patients who achieve deep molecular remission (DMR) can successfully discontinue tyrosine kinase

inhibitors (TKI). TKI discontinuation in chronic phase CML is being implemented in the clinical routine. To investigate the outcome of the patients with chronic myeloid leukemia (CML) discontinued tyrosine kinase inhibitors (TKI) therapy **Case report:** TKI was prospectively discontinued in patients who were diagnosed with CML in the chronic phase treated with TKI for ≥ 5 years, and sustained molecular response 4.5 (MR4.5) for ≥ 2 years. Molecular relapse was defined as a single loss of major molecular response (MMR) (BCR-ABL1^{IS} $>0.1\%$). **Methodology:** Standard qRT-PCR techniques were performed to evaluate minimal residual disease (MRD) **Results:** Twenty-one patients with chronic-phase CML were enrolled. The median duration of TKI treatment before discontinuation was 117 months (49-177) months. The median follow-up time after TKI discontinuation was 20 months (range: 1-117 months). The estimated TFR rate was 62% and 47.6% at 12 and 24 months after discontinuation respectively. Five patients experienced loss of MMR within 7 months after TKI discontinuation. All relapsed patients promptly resumed TKI therapy and regained at least major molecular response. **Conclusion:** Our data on the Tunisian population may provide a basis for the safety and feasibility of TKI discontinuation particularly in CML patients who are in sustained deep molecular response with longer TKI treatment duration.

<https://doi.org/10.1016/j.htct.2023.09.057>

Adult Hematology Abstract Categories

Coagulation Diseases

PP 08

AZERBAIJAN EXPERIENCE OF HAEMOPHILIA CARE

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Objective: As the management of haemophilia is complex, it is essential that those with the disorder should have ready access to a range of services provided by a multidisciplinary team of specialists. There is a State Program aimed at solving this problem in Azerbaijan. The purpose of the study to learn complex epidemiological characteristics which are necessary for justification of strategy on treatment and prevention of haemophilia. **Methodology:** For planning of prophylactic treatment in Baku city, there was obtained the database of all patients (by sex, age, diagnosis, severity) registered in the city (625 persons). The main group consisting of 52 patients with severe and 40 patients with moderate haemophilia-A was formed. Different variant treatment of 162 patients was organized in HTC: chemical synovectomy with rifampicin (44); phonophoresis with refined naftalan oil (44); phonophoresis with hydrocortisone (28); electrophoresis with KJ (35). **Results:** 77.9% of patients observed in treatment and prophylaxis

facilities in Baku were men, 59% were diagnosed with haemophilia A, 18.8% with severe and 31.5% with moderate haemophilia. Prophylactic treatment reduces the average annual number of bleeding episodes by 2.2 times in severe haemophilia and 2.1 times in moderate haemophilia. The model of prophylactic treatment of hemophilia can be applied in the infusion model 2 or 3 times a week as far as possible. **Conclusion:** The role of physiotherapeutic methods of hemarthrosis treatment was assessed and positive results were obtained. Due to the prevalence of polymorbidity in patients with hemophilia the complexity of their observation and treatment and the participation of specialists from several specialties is necessary. As the duration of haemophilia is proportional to the frequency of its complications, starting the prophylactic treatment at the stage when patients are first diagnosed is recommended.

<https://doi.org/10.1016/j.htct.2023.09.058>

PP 09

CHARACTERIZATION AND MANAGEMENT OF PATIENTS WITH HEREDITARY FACTOR X DEFICIENCY: A RETROSPECTIVE SINGLE CENTER EXPERIENCE

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Objective: Factor X deficiency (FXd) is a rare coagulation disorder that can be either hereditary or acquired. **Case report:** We characterized patients with FXd and evaluated their bleeding patterns and treatment strategies. **Methodology:** This retrospective review includes patients with FXd managed at Ege University Medical Faculty Hospital Ege Adult Hemophilia and Thrombosis Center. We analyzed demographic characteristics, laboratory results, bleeding scores, and treatments of five patients with FXd (Table). Patient 1 was admitted for further evaluation of menometrorrhagia and prolonged postpartum bleeding. She required treatment following birth, tooth extraction, and fractional curettage during follow-up. Coagulation tests were run as a part of in vitro fertilization in patient 2 and were abnormal. Family history was significant for a history of thrombosis in her mother. Blood tests were positive for Prothrombin 20210 G/A heterozygous mutation and lupus anticoagulants. The patient has never had any bleeding episodes in the follow-up. Patient 3 has a history of menometrorrhagia, gingival bleeding, and prolonged bleeding after an abortion. The sister of the patient has FXd. In follow-up, she was treated for subcutaneous hematoma, gingival, and post-cesarean bleeding. Patient 4 presented for evaluation of menometrorrhagia. She was treated for polypectomy, two cesarean sections, tooth extraction, intermittent recurrent ecchymosis, and epistaxis. Patient 5 was diagnosed at age one and was referred to us for further management of his condition. His initial presentation was consistent with subdural hematoma. In the follow-up, he was treated for