weight loss of 6-7 kg.Lumbar and whole spinal MRI revealed changes in the intensity of the medullary signal, mild decrease in the height of L2-L3 and T10 . EMG revealed polyneuropathy.PET showed a moderate uptake of FDG in the localization of the bone marrow. The spleen was enlarged-size-157 mm. **Methodology:** Laboratory findings: hemogram-WBC-11.15 × 103/$\mu$L, Hgb-15g / dL, HCT-48%, PLT-604 × 103/$\mu$L. Bone marrow biopsy, imprint, aspiration revealed moderately hipercellular bone marrow with increasing in all 3 series, groupings in megakaryocytes, containing limited (3-4%) kappa monoclonal plasma cell population; moderately increasing reticulin fibers (grade 1 according to WHO). Karyotype 46, XY; multiple myeloma FISH panel: translocation 4; 14 and translocation 11; 14 (+). JAK2V617F-50.48% (+). **Results:** The key point in the diagnosis was trilineage hyperplasia of the bone marrow,because the reticulin fibrosis may occurs in 20% of PV cases. Thus, the patient was diagnosed with LPV. Due to the detection of plasma cells in the bone marrow (3-4%), kappa light chains, with the diagnosis of LPV, the diagnosis of MGUS was established.The patient was prescribed ASS 100 mg per os, Hydrea at a dose of 500 mg every other day. For MGUS, the "wait and watch" tactic was chosen. **Conclusion:** In the diagnosis of LPV, along with molecular genetic research, trepanobiopsy of the bone marrow plays a leading role. The possibility of a combination of myeloproliferative and other diseases should not be ruled out.

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## THE OUTCOME OF FATHERHOOD IN PATIENTS WITH PHILADELPHIA NEGATIVE MYELOPROLIFERATIVE NEOPLASMS, A SINGLE INSTITUTION EXPERIENCE

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**Objective:** The aim of this retrospective study is to evaluate fertility in the Philadelphia-negative MPN male patients and the effect of treatment received on male fertility and the outcome. **Methodology:** This is a single-center, mixed-design study (retrospective + phone interviews) conducted within the National Center for Cancer Care and Research. **Results:** 120 patients were interviewed, only 19 patients (15.7%) had met the inclusion criteria. The majority of patients had lost follow-up or cannot be contacted, and 29.1% of patients had their families completed by the time of diagnosis. The treatment received includes hydroxyurea, interferon, and ruxolitinib. The mode of delivery was normal vaginal delivery in 68% of the pregnancies. The total number of conceptions was 27; three stillbirths were reported. **Conclusion:** The data showed that most MPN male patients on treatment had their offspring born normally with no delivery complications, no reported congenital anomaly or growth retardation, and no report of MPN-related cancers. Though, further studies with a larger sample size are required to fully understand the effect of medications on the outcome of fatherhood in Philadelphia negative MPN patients.

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PP 17

## CONCOMITANT JAK2 AND BCR-ABL1 IN PATIENTS WITH CHRONIC MYELOID LEUKEMIA CLINICAL IMPACT AND RESPONSE TO THERAPY: A SYSTEMATIC REVIEW

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**Objective:** The aim of this review is to assess patients with chronic myeloid leukemia with concomitant JA2 positive for - their characteristics - response to treatment **Methodology:** We searched the English literature (Google Scholar, PubMed, and SCOPUS) for studies, reviews, case series, and case reports of patients with chronic myeloid leukemia who had JAK2 mutation. Inclusion criteria: were the presence of JAK2 mutation in CML patients with BCR-ABL1 rearrangement and, secondly, age ≥18yrs. The search included all articles published up to 20th April 2021. **Results:** A total of 25 patients met our criteria of the search. Twenty-two patients were diagnosed in the chronic phase, 2 patients in the accelerated phase, and one patient transformed to the blast phase. More females n=16 and 10 males. The mean age at the time of diagnosis was 61.3 years. 9 patients had to be switch to second-line therapy. Age and gender distribution and the presence of splenomegaly or organomegaly are almost the same. Males were slightly more than females. **Conclusion:** It is difficult to conclude that multi-kinase inhibitors are superior to imatinib in treating CML with concomitant JAK2 mutation. But the result of the reported cases showed that multi-kinase inhibitors are more likely to be successful in achieving remission and loss of JAK2 mutation. However, it is difficult to generalize the result without further studies due to the few numbers of patients and the unusual association.

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*COAGULATION DISEASES*

PP 18

## DOUBLE HETEROZYGOTIC FV DEFECT WITH HETEROZYGOTIC FV LEIDEN MUTATION AND FV DEFICIENCY IN THROMBOSIS

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**Objective:** FV Leiden mutation causes activated protein C (APC) resistance and causes an increase in thrombin level.

Although moderate bleeding is seen in severe factor V deficiency, less than 1% of patients experience bleeding. Cases in which thrombosis is prominent in the presence FV Leiden mutation and FV deficiency have been reported. Here, we present a patient with FV deficiency with FV Leiden heterozygous mutation in the etiology of recurrent abortion. **Case report:** A 41-year-old female patient who applied to her primary care physician with bilateral lumbar pain upon finding INR: 1.43 (0.8-1.2) and APTT: 37.6 seconds (25-36.5), the patient was recommended to apply to our out patient clinic. The patient who described two spontaneous abortions (at the age of 25, the first in the 2nd trimester and the other in the 3rd trimester), also had a history of ecchymosis in the extremities caused by minor trauma at intervals. **Methodology:** PT, INR and APTT returned to normal with the mixing test performed on the patient (12.1 sEC, 1.03 and 28.6 sec, respectively).Afterwards, FV, which is one of the factors in the common pathway of coagulation, was found low in the examination repeated twice (12.3% and 10.2%) (N: 62-139%). The APCR studied twice in screening for thrombophilia was 1.4 and 2.4 (N: 2.61-3.32) Protein C, protein S, antithrombin III levels were within normal limits, LAC and APA were negative. **Results:** According to this result, FV Leiden heterozygous mutation was detected in the genetic thrombophilia panel. Also the patient had FV deficiency . **Conclusion:** Authors termed the coexistence of heterozygous FV Leiden mutation and type1 FV deficiency as pseudohomozygous FV Leiden mutation. In our and other studies, we concluded that thrombosis was clinically significant, where as bleeding was rare and mild. We think that prolonged PT and APTT results in patients with a history of thrombosis with FV Leidenmutation are also stimulating in evaluating FV activity.

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PP 19

## BLEEDING MANAGEMENT DURING DELIVERY AND POSTPARTUM PERIOD IN GLANZMANN THROMBASTHENIA: EXPERIENCES FROM TWO CASES

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**Objective:** Glanzmann thrombasthenia (GT) is a hereditary bleeding disorder. The platelets lack $\alpha$IIb$\beta$3integrin and fail to aggregate. Pregnancy can also lead to isoantibody formation when fetal cells with $\beta$3integrins pass into the circulation of a mother lacking them; a consequence is neonatal thrombocytopenia and a high risk of mortality. We here present our experience with two GT patients, in which rFVIIa was our choice for bleeding prophylaxis and/or control during delivery and postpartum period. **Case report:** Case 1: A 28-year-old woman with GT was hospitalized. She was on 38th gestational week (GW). Vaginal delivery was completed with rFVIIa prophylaxis. Postpartum 5th day rFVIIa stopped. The patient discharged with a minimal vaginal bleeding. Postpartum10th day, she developed severe bleeding. GT seemed to be the only related factor. rFVIIa restarted with tranexamic acid and misoprostol. Two apheresis units of platelets were transfused. That time, rFVIIa continued 7 days. **Methodology:** Case 2: A 26-year-old woman with GT developed hematuria on 30th GW. No urinary system pathology was found. With. rFVIIa treatment, hematuria was ceased. On 39th GW, during labor she developed massive bleeding. As urgent management, 8 random units of platelet and 5 units of packed red blood cell were transfused with local vaginal compress. rFVIIa treatment was initiated. On 10th days of rFIIa with minimal vaginal bleeding the patient was discharged from the hospital. **Results:** In both of the patients, no major neonatal bleeding problem was experienced. **Conclusion:** GT patients are at risk for heavy bleeding during labor, deliver or postpartum. Platelet transfusion is simple and easy option for bleeding control. In alloimmunized patients pooled platelet should be used. The use of rFVIIa appears to be safe and relatively effective.

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## CASE REPORT OF MARGINAL ZONE LYMPHOMA DETECTED WHILE INVESTIGATING ETIOLOGY FOR HEMOSTASIS DISORDER

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**Case report:** In this article, we wanted to present our case in which we detected SMZL during examining for defects in coagulation tests and correlated the PT and aPTT elevation with the development of inhibitors against coagulation factors related to this disease. The PT and aPTT values of the patient diagnosed with MZL did not improve in the mixing test, and no other etiology was found. With the second course of chemotherapy, the patient's values improved.

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## ACQUIRED FACTOR XIII DEFICIENCY WITH RUNX1 MUTATION, A REPORT OF TWO CASES TREATED WITH FACTOR XIII CONCENTRATE

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