(28.7%) hemophilia B (HB). The clinical severity of hemophilia A ranged between mild (10, 10.3%), moderate (2, 2.1%) and severe (83, 85.6%), while for hemophilia B (mild 13 (33.3%), moderate 2 (5.1%) and severe 24 (61.5%) respectively. There were 76 (55.9%) had chronic joint disability. Factor inhibitors with different titers were detected in 24 (24.7%) of HA and only 2 (5.1%) of HB. Out of the whole cohort 136 had been tested for causative variants, 17 (12.5%) were positive for inv-22 and 4 (2.9%) for inv-1, while all negative HA were selected for analysis by next generation sequencing. We are reporting 3 cases of females with severe forms of hemophilia.We are reporting different mutations that was consistent in group of tested members of same family /trip. We confirmed as previously reported high frequency of inv 22 and we found 7 novel mutation out 12 detected variants for HA and one novel mutation out of 13 detected variants for HB. **Conclusion:** These results will enrich the spectrum of variants and enlarge the factor VIII and factor IX proteins database in the Saudi Arabian population. Establishing a molecular genetic based tests for fast, easy, and cost effective reliable mutation screening that can also be applied in the future for prenatal and pre-implantation genetic diagnosis.

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PP05

SERUM LEVEL OF VASCULAR CELL ADHESION MOLECULE AND P SELECTIN AS THROMBOPHILIC RISK FACTOR FOR EARLY VASCULAR ACCESS THROMBOTIC OCCLUSION IN HEMODIALYSIS PATIENTS

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Objective: Hemodialysis (HD) vascular access (VA) failure is the dominant cause of morbidity and the major cost of care for ESRD patients. The aim of the present work is to assess the serum level of vascular cell adhesion molecule and P-selectin in HD patients as markers for early thrombotic VA event. **Methodology:** 90 CKD patients divided into three groups: group I: 10 subjects apparently health, group II: 10 Patients with CKD stage IV-V on medical treatment and group III: 70 Patients with CKD stage V on HD with recent VA insertion divided into 2 subgroups: Subgroup III a: 57 patients with AV shunt and subgroup III b: 13 patients with permanent catheter. Laboratory investigations done (blood urea, serum creatinine, eGFR, CBC, PT, and INR), imaging and serum VCAM 1, P selectin before and 6 months after HD **Results:** There was positive connection between VCAM and P selectin and dialysis with statistics in form of p value (<0.001). Markers level pre dialysis and after 6 months of dialysis revealed that range of p selectin and VCAM1 level after 6 months are higher than pre dialysis level. **Conclusion:** Detection of elevated serum level of circulating sVCAM-1 and s P-selectin could be useful in the prediction of native AVF and permanent catheter thrombosis in chronic HD patients. The association between sVCAM-1 and s P-selectin and thrombosis in HD patients increases the evidence of the role of adhesion molecules in VA thrombosis

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PP06

SUCCESSFUL MANAGEMENT OF SEVERE CONGENITAL FACTOR X DEFICIENCY DURING PREGNANCY AND LABOR WITH PCC IN TWO SISTERS

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Introduction: Factor X (FX) deficiency is an autosomal recessive disorder caused by quantitative or qualitative defects in the FX protein. FX deficiency has an estimated worldwide prevalence of one in 1000000. (1) Pregnancy in women with congenital FX deficiency has been associated with adverse fetal outcomes (abortion and preterm labor) (2,11). We report two cases of successful pregnancy with factor X deficiency. **Case 1:** A 29-year-old woman with congenital factor X deficiency and prior abortion on prophylaxis PCC every 4 weeks. She was treated with PCC 25unit/kg twice weekly during the pregnancy course. At week 32 of pregnancy, she presented with labor pain. Lab showed PT 20.7 PTT 52.5 INR1.5 Fibrinogen 3.8 Hb13.8 platelet 195 WBCs 7.6 factor X 0.15. She was given PCC 25 units/kg until a level of 0.4 was achieved. She delivered a healthy, 1.9 kg baby by normal vaginal delivery. The estimated blood loss was 150 ml. She then received FX 15 units/kg for 3 days postpartum to maintain FX level >30% and INR <1.5. No episodes of abnormal bleeding were observed during pregnancy, labor or postpartum. **Case 2:** A 36-years-old woman with congenital factor X deficiency and two prior abortions, on prophylaxis PCC every 4 weeks. She received prophylaxis PCC 25units/kg twice weekly during the course of this pregnancy. At week 38 of pregnancy, she delivered a healthy 3.2 kg baby by cesarean section (CS) after failing labor induction. Lab showed PT 23.7 PTT 50.4 INR1.7 Fibrinogen 2.3 CBC was normal.FX 0.13. She was given PCC 25 units/kg until a level of 0.4 was achieved. The estimated blood loss was 500 ml. She then received FX 15 units/kg for 7 days postpartum to maintain FX level >30% and INR <1.5. She was discharged on tranexamixc acid. No episodes of abnormal bleeding were observed during pregnancy CS or post-partum. **Discussion:** Although FX activity increases during normal pregnancy, levels usually remain insufficient for hemostasis at delivery in women with severe FXD (4,5,6). FX replacement therapy with PCC or FX concentrate may be required to treat or prevent bleeding in FXD.Therefore, a therapeutic dose of PCC 20–30 iu/kg is expected to increase plasma FX activity by 0.4–0.6 iu/ml. Further infusions at 1- to 2-d intervals may be required if sustained treatment is necessary (3). There are reports of FX replacement with PCC during pregnancy in women with previous adverse pregnancy outcomes (7,8) and FX

replacement during labor with PCC or FFP, but with different regimens [9]. Our patients were treated with PCC prophylaxis during pregnancy and 25 units/kg during labor. No bleeding nor thrombosis was seen in both cases. The British guidelines recommend PCC 20–40 iu/kg during the third trimester for women with history of bleeding and with FX activity <03 iu/ml with the goal of achieving FX activity >04 iu/ml. They also recommend, to consider further PCC 10–20 iu/kg once daily to maintain FX activity >03 iu/ml for at least 3 days post-partum. [10] **Conclusion:** Prophylactic PCC resulted in excellent hemostasis in two of our patients, including one that delivered by C-section.

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LYMPHOMA

PP 07

PREVENTION CAN BE THE BEST TOOL FOR ADULT T-CELL LEUKEMIA. UPDATED T-CELL BRAZIL PROJECT

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Objective: T-cell Brazil project started in April 2017 an ambispective study focusing to collecting epidemiological and clinical data from the most frequent subtypes of PTCL, among them the ATL. As of July 2022 T-cell Brazil database contained 81 (16%) ATL out of 520 registered cases. Our goals are to describe demographic and clinical features, analyze the overall and progression-free survival (OS and PFS), and try to identify factors that could influence outcome. **Methodology:** Brazilian Registry using REDcap Platform by Vanderbilt realized descriptive and bivariate analyses, then it was applied Kaplan-Meier method and log-rank test to obtain survival estimates, and besides that, it was used the Cox Regression to identify any factor that could influence the OS and PFS. **Results:** The median age was 52 years (24-91); 32 (39%) male; the majority of clinical subtypes were 52% lymphoma type; 81% received chemotherapy. The best response assessment after first-line treatment was: progression or no response in 31%; 26% complete response; 21% partial response, 21% not available (NA) due to death or on treatment; 34% of patients were alive and the 24-month OS and PFS was 33% and 21%, respectively. As predictors for PFS and OS were B symptom and elevated LDH values. **Conclusion:** This study, even recognizing a limited sample size, highlights the poor prognosis associated with ATL, mainly acute and lymphoma type, with high mortality rates. Hence, apparently, a good shot, it would be one of the bases for the prevention of ATL to establish a disease entity of "chronic active HTLV-1 infection" that defines high-risk carriers for ATL development, and then, enables preventive intervention.

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PP08

AN UNUSUAL OCULAR LYMPHOMA, PRIMARY INTRAVITREAL LYMPHOMA DIAGNOSED INCIDENTALLY

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Objective: Ocular lymphoma involvement can be either secondary during systemic lymphoma or primary. Diagnosis can be troublesome due to insidious disease onset. Uveitis is the main differential diagnosis. The prognosis is poor. **Case report:** A 62-year-old male patient was evaluated during a periodical check-up for hypertensive retinopathy. The unexpected good vision quality with severe left vitreous infiltration and not associated macular edema contributed to malignancy suspicion. A diagnostic procedure was performed bilaterally. Both of the vitreal tissue revealed atypical lymphoid cells with B-Cell phenotype. Cranial MRI, PET-CT, and CSF analysis documented the case as primary vitreoretinal lymphoma (VRL). **Methodology:** First-line treatment was with intravitreal methotrexate (MTX). After 10 courses, high-dose cytarabine-based treatment was given as consolidation. Considering high recurrence rates, stem cells were mobilized and cryopreserved for future use for autologous stem cell transplantation (ASCT). **Results:** Follow-up was 3 monthly. After 10 months of remission period, retinal disease relapse was spotted. After 5 cycles bilateral intravitreal