(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