with ratio of 0.55. The PFA-100 showed elongation in 38 patients with sensitivity of 76%. PFA-100 had a sensitivity of above 80% for those with Ricof of 40-60%. There were 36 patients received intravenous Desmopressin at the dose of 0.15 µg/kg for primary haemostasis for surgery or procedure. The average dose was 13.1 mcg. All underwent the procedure and surgery without bleeding complication or any drug-related effect except 2 had hyponatraemia which was self-limiting. The post half dose desmopressin responses were only available in 16 patients with significant improvement in the FVIII (P = 0.016), vWF Ag (P = 0.0019) and Ricof (P = 0.0004). Nevertheless, the PFA-100 post desmopressin did not show any statistical significance in terms of therapeutic response. The diagnosis included mostly BUC-36%, followed by borderline vWF-24%, Type 1 vWD-20%, Platelet Function Defect-14%, Platelet Secretion Defect-2%, Macrothrombocytopenia-2% and haemophilia A carrier with borderline low vWF-2%.

Conclusion: The PFA-100 is a very useful screening test for patient with BUC complementing the ISTH-SSC Bleeding Assessment Tool in order to improve the clinical management. The efficacy of half dose desmopressin in this study which differs from the usual dose used for confirmed von Willebrand Disease in the standard practice should prompt clinicians to adopt such strategy in future.

MED-FP-028 (254) | Angiogenic alterations in VWD as a possible cause of epistaxis

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Objectives: The Ang/Tie system plays a major role in angiogenesis, where EC initiate proliferation, migration and vessel stabilization. Due to reduced levels or lack of Von Willebrand factor (VWF), not only hemostasis may be impaired, but also angiogenesis. As the Ang/Tie system is very sensitive, an imbalance of its members due to impaired levels of VWF may result in uncontrolled angiogenesis leading to angiodysplasia seen in Von Willebrand Disease (VWD), the most common inherited bleeding disorder. So far, the impact of VWF on the Ang/Tie system was examined in cell lines and mouse models. One of the most apparent symptoms of VWD is epistaxis, usually originating from a nasal plexus called Locus Kiesselbachi. In this study, we analyzed the Ang/Tie system in the Locus Kiesselbachi in a porcine VWD model to elucidate the possible pathomechanism of epistaxis in VWD.

Methods: Samples from fourteen pigs, representing type 1 (T1; n = 5), type 3 (T3; n = 3) VWD and a wild type (WT) control group (n = 6), respectively, were examined by immunohistochemistry (IHC) and RT-qPCR to detect the ligands angiopoietin (Ang) 1 and 2 and the tyrosine kinase receptors (Tie) 1 and 2. Expression levels were calculated using PECAM and PROCR as reference genes applying the ΔΔct method.

Results: TIE1 gene expression levels were significantly reduced from WT to T1 by 39% and to T3 by 58%. ANG1 also showed a clear tendency of lower expression in T3 individuals. For ANG2 and TIE2, a wider spread of expression levels was seen within each genotype and no significant differences between the genotypes were detected. In IHC the pattern of Ang1 in endothelial cells (EC; T1 = −37%; T3 = −33%) and vascular smooth muscle cells (VSMC; T1 = −36%; T3 = −31%) showed corresponding results.

Conclusion: We for the first time demonstrate profound alterations of the Ang/Tie system in the Locus Kiesselbachi in a VWD model. Thus, dysfunctional angiogenesis leading to angiodysplasia may contribute to the high frequency of epistaxis in VWD representing a part of its pathomechanism.

MED-MP-027 (644) | Spectrum of Von Willebrand disease in Southern India

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Introduction and Objective: Diagnosis of Von Willebrand (VWD) in low- and medium-income countries (LMIC) is challenging due to cost and lack of laboratory infrastructure. However, VWD continues to be one of the commonest bleeding disorders in these countries. The objective was to study the pattern of VWD and the utility of simple coagulation tests in the diagnosis in a tertiary centre in Southern India.

Patients and Methods: This was a retrospective study where data were collected from all patients who presented to a tertiary hospital in South India from January 2002 to March 2019. Diagnosis was confirmed by performing the following tests: bleeding time (BT), PFA200, APTT, Factor VIII, Ristocetin cofactor assay (vWF:RCo), Von Willebrand antigen (vWF:Ag) and Collagen binding assay (vWF:CBA) combined with history and parent's evaluation wherever applicable. Bleeding time was performed only by trained and experienced technical staff.

Results: There were a total of 445 cases with reduced levels of Von Willebrand Factor (vWF:Ag <50 u/dL). The distribution of cases is shown in Table below. Type 3 VWD was the most common subtype. 50% of type 1 was of the severe type with vWF:Ag <10. Bleeding time was prolonged in 79% of the total cases, all cases of type 3 VWD, 87% of severe type 1, 33% in mild Type 1, 60% in type 2 VWD and none of the cases with borderline levels of vWF:Ag. APTT was prolonged in 99.5% of type 3 VWD, 85.2% of severe type 1, 65.7% in mild type 1 and 49.5% of type 2 VWD. There was inverse correlation between BT and VWF:Ag (r = −0.52) & between BT and vWF:RCo (r = −0.66).

Conclusion: Diagnosis of VWD in resource-poor setting is challenging. Bleeding time though appears simple requires high level of skill and expertise. When performed under standardised conditions, BT is useful in the evaluation of VWD in LMIC.