



Bleeding score in Type 1 von Willebrand [vWD] patients using the condensed MCMDM-1 VWD Questionnaire



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Introduction and Objectives:

Excessive bleeding is a key component in patients with von Willebrand disease (vWD). However, reporting and interpretation being subjective led to the evolution of bleeding assessment tools. In 2008, the European Molecular and Clinical Markers for the Diagnosis and Management of type 1 VWD (MCMDM-1 vWD) questionnaire was established and independently validated. Our aim was to administer MCMDM-1 vWD bleeding score questionnaire to the Omani type 1 vWD patients and correlate it with the clinical phenotype.

Materials and Methods:

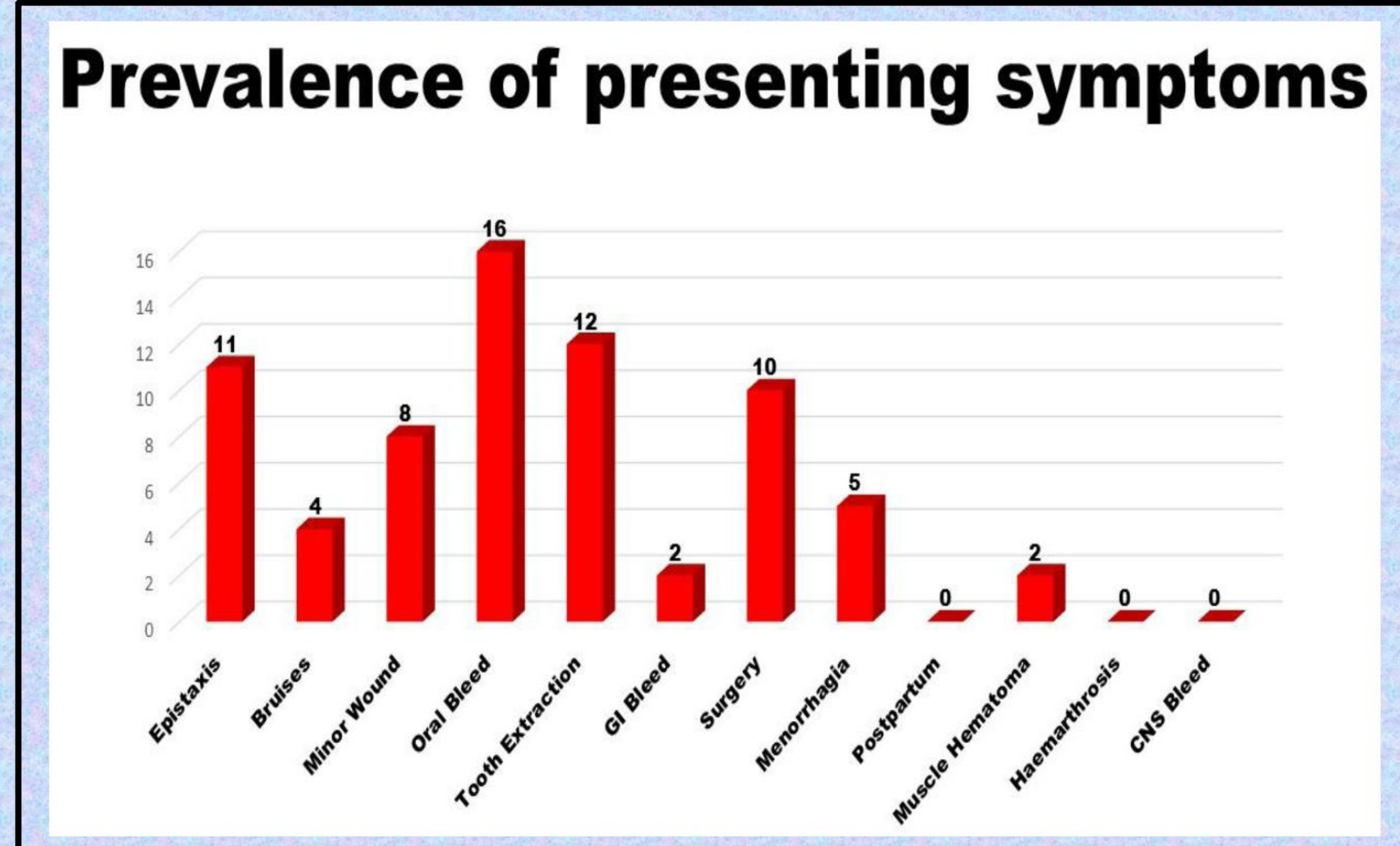
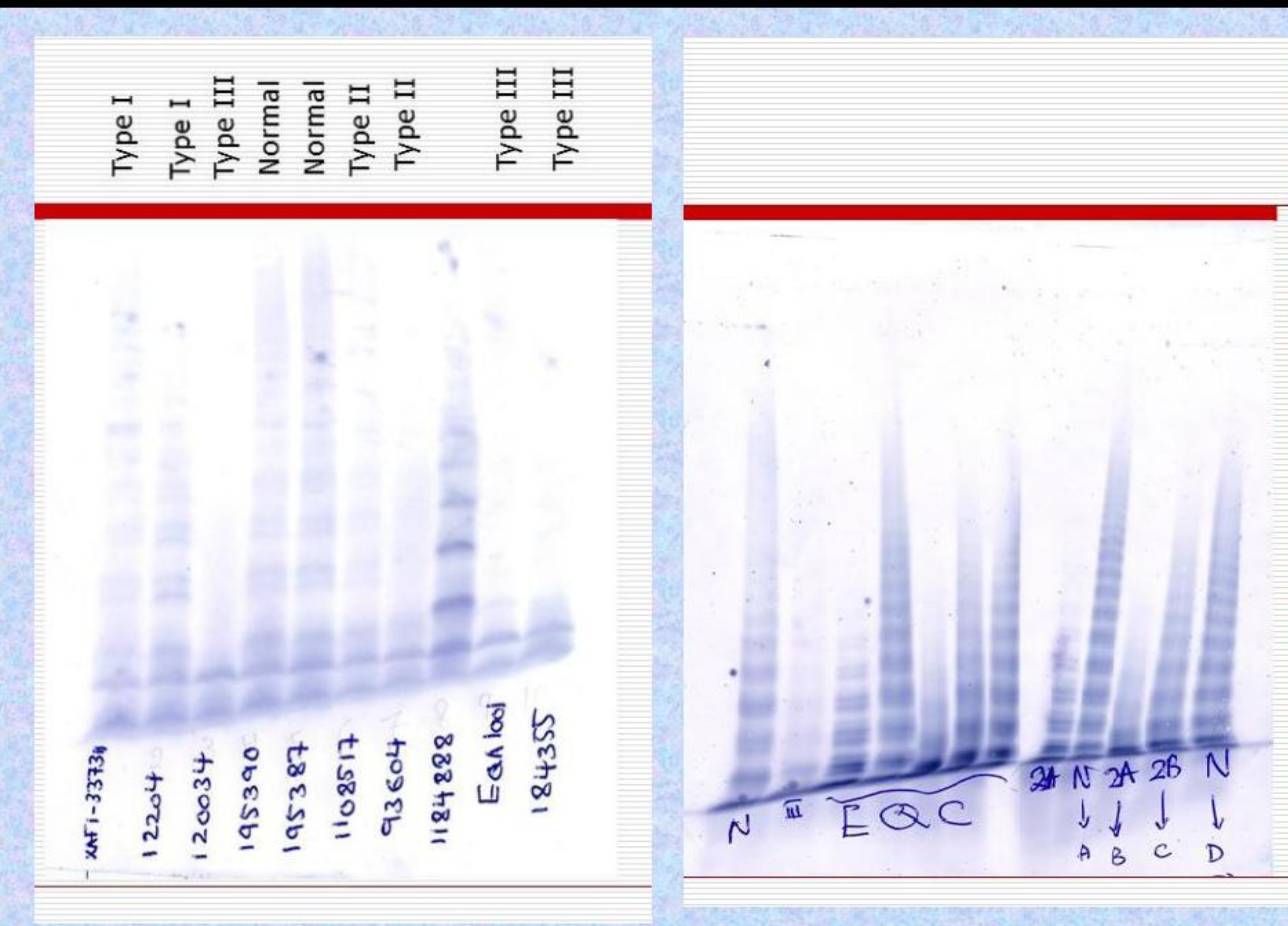
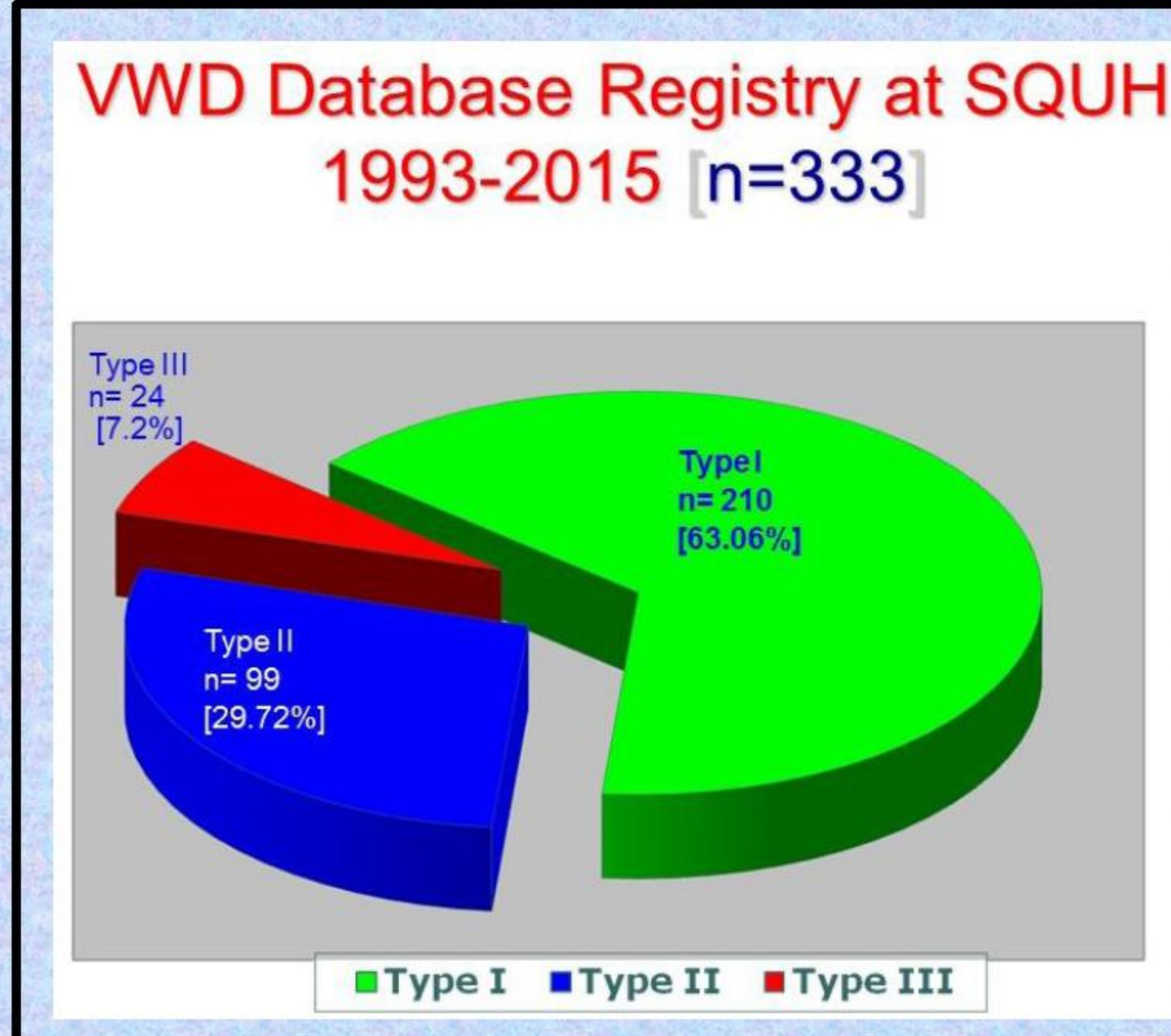
27 Type I vWD index cases diagnosed using the ISTH-SSC criteria were personally interviewed and administered the MCMDM-1 vWD bleeding score questionnaire. The bleeding score [BS] was calculated based on a history of bleeding from 12 different sites according to the standard MCMDM-1 vWD questionnaire.

Results:

The mean time to administer this questionnaire was 13 minutes with a range from 6 to 39 minutes. Overall, bleeding from the oral cavity was the most predominant symptom [59%], but amongst the females, menorrhagia was the most prevalent symptom [94%]. The prevalence of other symptoms were epistaxis [44%], tooth extraction [44%], bleeding from minor wounds [37%], cutaneous bleeds [33%], bleeding at surgery [33%], GI bleeds [7%] and hematoma [7%]. No patient had haemarthrosis or CNS bleeds. The mean BS in this cohort was 4.74 with a range from 0 to 17. 15% of the patients had a BS of 0, whereas, another 26% had a BS below 4. The BS was negatively correlated with FVIII:C levels, vWF:Ag, vWF:RiCoF and vWF:CBA respectively and the Pearson's correlation coefficient was respectively -0.11,-0.25,-0.29 and -0.28; $p > 0.05$.

Conclusions:

The MCMDM-1 vWD bleeding score questionnaire is designed to reflect the severity of bleeding amongst the vWD patients and our results demonstrates the inherent variability of this bleeding pattern with 41% below the BS reported in literature from the normal population [less than 4]. Furthermore, although the mean BS was abnormal in this cohort of vWD index cases, overall the BS did not correlate significantly with the surrogate laboratory parameters used for the diagnosis of vWD.



Summary

- Median bleeding score was 4 with an interquartile range between 1-7.
- Median time to administer the questionnaire was 12 minutes with IQR between 8-16 minutes.
- No patient presented with CNS bleeding or hemarthrosis.
- Correlation between the bleeding score of MCMDM-1 and ISTH-BAT questionnaire showed Spearman's rho of 0.780. [$p < 0.05$]

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