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VON WILLEBRAND DISEASE: A SINGLE INSTITUTION EXPERIENCE

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BACKGROUND/AIMS: Von Willebrand disease (VWD) has been found to be the most common inherited bleeding disorder in Caucasians, with a prevalence of 1%, yet it has not been well recognized in Taiwan and Asian countries. This study aims to identify patients with VWD by clinical manifestation and laboratory tests in Taiwan.

METHODS: VWD was detected by a panel of laboratory tests, including bleeding time, aPTT, factor VIII activity assay, Von Willebrand antigen (VWF:Ag) and ristocetin cofactor activity (VWF:RCo), and platelet function analyzer (PFA) test. VWF multimer analysis was performed by western blot to confirm the disease subtype.

RESULTS: From October, 2003 to July, 2006, 40 out of 371 suspected patients (10.7%) from 32 families were identified to have VWD. These included 12 men and 28 women. Their median age was 29 years, covering a range of 4 to 58 years. Three patients were found to have an acquired type of VWD, another 3 had comorbidities (2 with hypothyroidism and 1 with essential thrombocytopenia)and 37 patients had an inherited variant of VWD. The most frequently encountered reasons for which the VWD panel studies were done were family inheritance detection (20%), easy bruising (18%), excessive bleeding after dental extraction or procedures (13%), and iron deficiency anemia (13%). Menorrhagia accounted for 79% in female patients. By laboratory examinations, the mean value of VWF:Ag and VWF:RCo was 47.9 ± 21.6 % and 31.7 ± 16.5%, respectively. The sensitivity of bleeding time, aPTT, factor VIII activity, VWF:Ag, VWF:RCo and PFA test to detect VWD was 47%, 34%, 35%, 53%, 85% and 81%, respectively. Of 19 patients with VWF multimer analysis, 18 patients revealed to have type I VWD, only 1 patient had type IIA.

DISCUSSION/CONCLUSIONS: Our study demonstrated that VWD is not an uncommon inherited bleeding disease in Taiwan. Case detection was difficultand only 1/5th proportion of patients was found by family inheritance study. VWF:RCo and PFA test are the more sensitive tests for the VWD identification.

Key words: Von Willebrand disease; VWF: antigen; Ristocetin cofactor activity.