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0676 STABILITY OVER TIME OF THE HETEROZYGOUS VWD PHENOTYPE

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A study has been made of 181 members of two large vWd kindred in the Carolinas. Analysis of the data from first testing showed that the trait, "affected"—meaning abnormal on one or more of 4 tests (BT, VIII:C, VIIIR:Ag, VIIIR:WF)—was transmitted as an autosomal dominant. There were 23 affected members in the smaller (418) kindred, and 57 affected in the larger (750) kindred. Penetrance of the vWd allele was reduced to 58%, and the "affected" phenotype was highly variable. When the tests were scored (+)= normal and (-)= abnormal, 11 of the 16 possible combinations of (+) and (-) were observed. Only 15% of those affected were of the classical phenotype, i.e. abnormal on all 4 tests.

A subsample (22 persons) of the larger kindred was re-tested twice, 1½ and 2 years after first testing. The "affected" phenotype was found to be very stable over time. In a total of 140 additional tests only 18 (12%) shifted from "abnormal" to "normal" or vice-versa; and only 5 of 22 persons (23%) shifted class; 2 shifting from "affected" to "non-affected", 2 from "non-affected" to "affected" and 1 from "affected" to "non-affected" to "affected".

The Prethrombotic State

Level 6 – Red Side

Discussion Group 12.00 – 12.45

?6-080 0677 THE VARIABILITY OF β THROMBOGLOBULIN AND PLATELET FACTOR 4 IN HEALTHY SUBJECTS

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Plasma β TG was measured by radioimmunoassay (RIA) in 202 healthy subjects (age range 12-103); 111 young (mean age 25.2), 34 middle aged (MA) (mean age 55.6) and 57 old (mean age 82.2). Their mean ± SE plasma β TG levels in ng/ml were 28.3 ± 1.5 (range 3-74), 31.9 ± 2.70 (range 7-65) and 49.99 ± 2.9 (range 14-95) respectively. Plasma β TG level was significantly raised in the old subjects compared to young or MA ($p \leqslant 0.0005$). Furthermore the ratio of plasma β TG to platelet concentration in whole blood (PC) was higher in the MA subjects compared to the young ($p \leqslant 0.009$). Plasma platelet factor 4 (PF4) was measured by RIA in 41 healthy subjects, 11 young and 30 old and correlated to plasma β TG. A significant correlation between the 2 proteins was found in the 2 groups ($r=0.8387$ in the young and $r=0.0602$ in the old subjects), indicating that both proteins are released in-vivo from the same pool and presumably at the same rate. The mean plasma PF4 level in ng/ml was 14.8 (range 6-48) in the young and 18.2 (range 7.7-50) in the old and the ratio of the plasma PF4 to PC was higher in the old subjects ($p < 0.04$). These results suggest that in-vivo platelet activation and "release reaction" are increased in old and MA subjects compared to young, presumably due to atherosclerotic vascular changes. This enhanced platelet activity may reflect a pre-thrombotic state.