PLATELETS, ENDOTHELIAL CELLS AND VON WILLEBRAND FACTOR. Z.M. Ruggeri. Hemophilia and Thrombosis Centre. University of Milano and Policlinico Hospital, Milano, Italy.

In plasma and platelets (Pl) from 10 patients (pts) with "severe" recessive von Willebrand's disease (vWD) factor VIII-related antigen (VIIIR:AG) was unmeasurable in 7 and extremely low in 3 using a very sensitive immunoradiometric assay (IRMA); ristocetin cofactor (VIIIR:RCo) was always unmeasurable and VIII:AG was not detectable on immunofluorescence on endothelial cells (EC) of the vessel wall. In 12 pts with "classical", dominant vWD characterized by reduced plasma level of VIIIR:AG and VIIIR:RCo, normal values were found in Pl and the mobility of Pl VIIIR:AG on crossed immunoelectrophoresis was not different from that of normal controls; VIIIR:AG was normal on EC. In 7 pts showing a faster electrophoretic mobility of plasma VIIIR:AG (variant: vWD), the same abnormality was found in Pl. VIIIR:AG concentration was normal both in plasma and in Pl when measured by electroimmunodiffusion, whereas IRMA gave lower values and VIIIR:RCo was decreased; VIIIR:AG was normal on EC. These findings show that "severe" vWD is the expression of a marked reduction of factor VIII synthesis fully expressed in EC, Pl and plasma. In "classical" vWD the plasma defects are not reflected in the cellular compartments, suggesting a defective transfer from EC and Pl to plasma. Patients with "variant" vWD are the expression of a qualitative alteration of the factor VIII molecule, functionally defective both in Pl and in plasma.

FACTOR VIII ASSOCIATED ANTIGEN, PLATELET RETENTION AND PLATELET AGGREGATION IN PATIENTS WITH VASCULAR DISEASES. I. Scharrer and U. Pander, Dept. of Angiology, Center of Internal Medicine, University of Frankfurt/ Main, F.R.G.

Factor VIII associated Antigen (F.VIII ASS AG) or Factor VIII related protein (FVIIIRP) was found in the vessel wall by several investigators. Breddin and co-workers described enhanced platelet aggregation in patients with vascular diseases. Therefore we investigated the level of F. VIIIassAG in patients with vascular diseases and the relation of it to platelet aggregation and platelet retention. The following methods were used: Quantitative immunelectrophoresis in antiserum containing agarosegel according to Laurell, platelet retention test by application of the adeplat-S-pump system and platelet aggregation test III according to Breddin. We compared the results of 50 healthy persons and 53 patients with vascular diseases. We found a significant correlation of F.VIIIassAG to the age of 50 healthy males. The Spearman rang correlation coefficient was r= 0.4188. We could also demonstrate a relation of the increased level of F.assAG^{S} to different vascular diseases. The patients with myocardial infarction (18), with peripheral arterial occlusion (18), with cardiac valve replacement (9) and with peripheral venous thrombosis (8) showed a distinctly increased F.assAG. We found neither a correlation of F.VIIIassAG to the aggregation nor to the retention.

SYMPOSIA SESSION

II Hemophilic Arthropathy: Current Concepts in Management