Poster Board P5-044

## 0739 AN INHIBITOR TO FACTOR VIII ANTIGEN PRESENTING AS ACQUIRED VON WILLEBRAND DISEASE

K.M. McGrath*, C.A. Johnson and J.J. Stuart, Department of Medicine, Bowman Gray School of Medicine, Winston-Salem, North Carolina, U.S.A. and Department of Human Metaboli & Clinical Biochemistry, University of Sheffield, Sheffield, England.

A patient with clinical and laboratory evidence of acquired von Willebrand disease is described. He presented with the recent onset of spontaneous hemorrhage and demonstrated a prolonged bleeding time, reduced factor VIII:C, and undetectable factor VIII:AG and factor VIIIR:WF. Following transfusion of cryoprecipitate, there was a smaller than expected immediate increase in factor VIII:C, factor VIIIR:WF and factor VIIIR:AG, with rapid return to baseline levels, and no secondary rise in factor VIII:C. An inhibitor could be demonstrated in the patient's plasma which markedly decreased the level of factor VIIIR:AG in normal plasma while only weakly decreasing the activity of factor VIIIR:WF. Activity against factor VIII:C was demonstrable in vitro using a concentrated inhibitor preparation. The inhibitor was contained in the IgG fraction of plasma and lacked precipitating properties. This inhibitor, which demonstrates a specificity not previously seen in spontaneous anti-factor VIII antibodies, provides further evidence for the separate identity of the antigen and von Willebrand factor sites of the factor VIII molecule. It also indicates that inhibition of ristocetin-induced aggregation of platelets is not the most sensitive method for the detection of an inhibitor in all cases of acquired von Willebrand disease.

P5-045

## 0740 COAGULATION DISORDERS IN FULMINANT HEPATITIS TREATED BY DIALYSIS

M.MORFINI*, F.MARTINELLI, S.CINOTTI, S.BONCINELLI, F.MAZZOTTA, PACI P., S.BANDINI, F.TONELLI, F.CORDOPATRI, A.MORETTINI, F.PACINI and P.ROSSI FERRINI, Department of Haematology and Liver Unit, Florence, Italy.

A young man, 28 years old, was admitted to Infectious Division with transaminases over 2500 U/ml, reduced cortical activity and decerebrate posturing. A prolonged APTT, Prothrombin below 15%, very low levels of Factor II, V, VII (5-20%), AT III by S-2238 (17%), Antiplasmin (20%) and Plasminogen (5%) by S-2251, Prekallikrein by S-2302 (5%), Fibrinogen (70mg%) but very high VIII AHF (4.5U/ml), VIII AGN (4.0U/ml) and VIII VWF (3.8U/ml) were recorded. After daily dialysis sessions with polyacrilonitrile membrane (RP 6) a marked improvement was observed. The patient awoke while prothrombin and platelet recovery took place, fibrinogen, VIII AHF and Factor V rose over normal value to 600mg%, 7.5U/ml and 3U/ml rispectively. Antiplasmin, Plasminogen, PKK showed a slow but costant improvement. Unfortunatly a venous thrombosis and sepsis set in during the 2nd day after dialysis, with a rapid decrease of platelets. Heparin infusion 1 mg/kg b.w. was infused every 6 hrs. After 15day platelet returned to normal value and VIII AHF to initial level but it was still higher (4U/ml) than normal value after 2 mounths from recovery.

P5-046

## 0741 HOMOLOGOUS ANTIBODIES TO FACTOR IX SHORTEN THE BOVINE THROMBOPLASTIN COAGULATION TIME OF HUMAN PLASMA

K.H. Örstavik, Institute of Medical Genetics, University of Oslo, Norway

We have previously demonstrated that neutralization of factor IX in normal plasma by heterologous antisera shortens the one stage prothrombin time determined with bovine thromboplastin. In this study we demonstrate a similar effect of a homologous antibody. Addition of 1/5 volume of plasma from a patient with hemophilia B- and an acquired inhibitor to factor IX (titre 5 U/ml) gave a shortening of the prothrombin time of plasma from 7 normal persons (mean 34 sec) compared to the prothrombin time determined after addition of 1/5 volume of control plasma from a patient with hemophilia B- and no acquired inhibitor (mean 39 sec). Addition of inhibitor plasma had no effect on the prothrombin time of plasma from 6 patients with hemophilia B-. Complexes between factor IX and the human inhibitor could be demonstrated both before and after the coagulation with bovine thromboplastin. These complexes were demonstrated as a factor IX antigen with a reduced electrophoretic mobility in crossed immunoelectrophoresis against a rabbit antiserum to factor IX. The results indicate that normal factor IX loses the ability to act as an inhibitor in the coagulation with bovine thromboplastin after having formed a soluble complex with a homologous antibody.