DDAVP : A NEW PHARMACOLOGIC APPROACH TO THE MANAGEMENT OF HEMOPHILIA AND VON WILLEBRAND'S DISEASE. P.M.Mannucci, Hemophilia and Thrombosis Ctr, Univ. of Milano, Italy

I-deamino - 8 D - arginine (DDAVP) infusion causes a marked increase of F VIII - related properties in patients with moderate and mild hemophilia (H) and von Willebrand's disease (VWD) The possibility was evaluated that such an autologous F VIII response might be hemostatically effective, allowing patients (pts) to undergo surgery without blood products. 0.3 μ/Kg of DDAVP given before dental surgery and repeated in the early post-operative period were followed by a 2 - to 3 fold rise of F VIII coagulant activity (VIII : C) in 4 pts with moderate and mild H. In two, there was no abnormal bleeding after dental extraction, whereas plasma concentrates became necessary to control oozing in the remaining 2 pts. The adoption of higher dosage (0.4-0.5 μ/Kg) in pts with higher starting VIII : C (9% or more) was followed by a more marked response (4 to 6 fold). Hence, VIII : C levels up to 100% of average normal could be achieved and dental extractions as well as major surgical procedures (such as cholecystectomy, thoracotomy and two tonsillectomies) were carried out successfully in 6 pts with mild H and 2 with VWD. The mean halfdisappearance time of autologous VIII : C was 9.4 hours (range 7.5-11.6). Plasma and urine osmolality showed no consistent variation. DDAVP appears a promising pharmacologic alternative to plasma concentrates in the management of a number of pts with H and VWD.

HEMOPHILIA A IN A GIRL WITH DELETION OF A PART OF THE LONG ARM OF ONE X CHROMOSOME. M. Samama, Ch. Perrotez, R. Houissa, A. Hafsia, J. Seger. Departments of hematology Hôtel-Dieu and Hospital Saint-Antoine, Paris, France and Hospital Aziza Othmana, Tunis, Tunisia.

A severe hemorragic diathesis has been discovered in a 10 years old girl whose brother has severe hemophilia with classical haemarthrosis. The factor VIII biological activity is less than 1 % in both of them with an almost normal factor VIII related antigen. The bleeding time was normal. There was no consanguinity and paternity was not disproved by extensive blood grouping tests. Cytogenetics studies showed a deletion of a part of the long arm of one X chromosome. The formula of the caryotype is 46, X, del X (q. 212). The sex chromatin showed the presence of a phenotypic female. The X chromatin was smaller than normal. This is the first case to our knowledge of true female haemophilia A due to a deletion of one X chromosome.

RELATION OF GROWTH HORMONE AND VON WILLEBRAND FACTOR ACTIVITY. K.E. Sarji, H.B. Schraibman, J.H. Levine, M. Barnes, R.M.G. Nair, J. Sagel and J.A. Colwell. V.A. Hospital and Dept. of Med., Med. Univ. of S.C., Charleston, S.C.

Growth hormone (GH) has been implicated in the pathogenesis of diabetic angiopathy although the exact mechanism remains unknown. von Willebrand factor activity (vWF) is significantly higher in diabetic than in normal plasma ($p<.001$). In view of this, we have investigated the relationship of radioimmunoassayable GH and vWF activity, as measured in a bioassay using washed normal human platelets and ristocetin. Samples were obtained after an overnight fast, during oral glucose tolerance tests, and during sleep. A significant positive correlation was seen between GH over a range of 0.46 to 12.2 ng/ml and vWF activity over a range of 1 to 470% ($p<.01$). vWF activity was suppressed by oral glucose in normals and diabetics, with the amount of suppression related to the degree of glucose intolerance. Maximal suppression of vWF activity was coincident with maximal GH suppression. Samples from 4 normal subjects drawn through an indwelling catheter at 30-minute intervals during sleep showed peaks of plasma GH and vWF activity with GH peaks preceding vWF peaks by an hour or less. Two patients with panhypopituitarism and one patient with isolated GH deficiency were similarly studied. GH levels were low in these patients, but vWF levels were normal; both GH and vWF showed little sleep-related change.

A single intramuscular injection of GH produced a marked increase of vWF activity in all 3 patients within 30 minutes and a later increase after 5-6 hours. We conclude that a regulator of glucose metabolism, possibly GH, is involved in regulation of vWF activity.