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INCREASED THROMBOTIC TENDENCY IN A FAMILY WITH HEREDITARY ANGIONEUROTIC EDEMA (HANE). W. Kirschstein, U. Hoffmann, S. Simianer, E. Dempfle, C.Kortsik, D.Heene. I. Med. Klinik, Klinikum Mannheim, University Heidelberg, D 6800 Mannheim, FRG.

Biochemical hallmark of HANE is a reduction of C1-inhibitor. We observed a family with type II disease (non-functional protein), in which 3 of 6 affected members had arterial thromboembolic events at young ages. For evaluation of alterations in the hemostatic system analysis included: fibrinogen, FVII, FVIII, FIX, FXI, FXII, prekallikrein PK, antithrombin III ATIII, protein C PC, α_2 antiplasmin α_2AP, α_2 macroglobulin α_2 MG, plasminogen activator inhibitor PAI, plasminogen PG, euglobulin clot lysis time ECLT and tissue plasminogen activator tPA at baseline and after venous occlusion. The results are shown in part in the table:

	FVII %	FVIII %	FIX %	FXI %	FXII %	PK E/ml	PC %	ECLT min base	ECLT min cuff	tPA ng/ml base	tPA ng/ml cuff
1.	117	115	180	125	98	1,06	153	356	258	9,7	5,2
2.	228	246	170	105	95	1,11	115	375	343	24,2	44,8
3.	96	280	150	130	110	0,70	93	-	-	4,8	-
4.	174	160	135	160	200	1,21	181	360	181	12,4	27,8
5.	134	124	170	150	102	1,26	181	370	388	16,4	16,0

There is evidence of nearly no response to venous occlusion in 2 and a diminished response in 1 out of 4 patients.
We conclude, that the increased thrombotic tendency in this family is related to the increased potential of prephase coagulation factors and impaired fibrinolytic response to venous occlusion concomitantly with the reduction of the main inhibitor of the contact activation system.

PRETHROMBOTIC STATE

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URINARY FIBRINOPEPTIDE A IN A HEALTHY POPULATION AND IN PATIENTS WITH PERIPHERAL VASCULAR DISEASE. J. Dawes (1), O. Drummond (1) and N. Goodfield (2). MRC/SNBTS Blood Components Assay Group, Edinburgh (1) and Royal Infirmary, Edinburgh, UK (2).

Urinary fibrinopeptide A (FpA) concentrations may be a useful clinical marker of the activation of coagulation. They are not susceptible to false positives resulting from ex vivo activation, and sampling is noninvasive.
Individual urine voidings were collected from cohorts of 30 healthy subjects in each age decade from birth to 70 years, and a further 24 between 70 and 100 years. Below the age of 70 the urinary FpA concentration was 1.72 $\pm$ 0.76 ng/ml, and there was no effect of age or sex. Within this population, 4% of samples contained FpA concentrations above the upper limit of normal (mean + 2.5 SD); intensive investigation of one case failed to reveal any renal or coagulative disorder, though the urinary FpA levels remained high (8.4 - 14.2 ng/ml). Above 70 years old, 29% of urinary FpA concentrations exceeded the upper limit of normal established on a younger population. Thus, urinary FpA does increase with advanced age, but this may well result from occult disease.
Sampling of every urine voiding over 48h in 3 healthy individuals established that there is no diurnal pattern either in urinary FpA concentration or in rate of FpA excretion.
Urinary FpA was unaffected by the phase of the menstrual cycle.
Urine samples from patients with peripheral vascular disease were assayed, and 24% contained elevated concentrations of FpA.
Urinary FpA is probably a valuable marker of low grade activation of coagulation, particularly in chronic conditions where the assay of plasma samples is frequently uninformative.

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INTEREST OF THE RATIO OF INCREASE OF β-THROMBOGLOBULIN ($\Delta^+ \beta$TG) AND OF FIBRINOPEPTIDE A (Δ^+ FPA) FOR DIAGNOSIS AND TREATMENT OF THROMBO-EMBOLIC DISEASES.

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The interest of release of TG and FPA for the diagnosis and treatment of prethrombotic and thrombotic disorders is well known.
The ratio of increase of these two parameters ($\Delta^+ \beta$TG/Δ^+ FPA) seems to bring some additional informations.
This ratio, normally about 1, increases in isolated or preponderant platelet activation and decreases when platelet activation plays a minor role than the plasmatic factors. A more logical choice of therapeutics and a better control of its effectiveness are so possible.
This study includes 91 cases of established thrombosis (20 arterial and 71 venous) and 272 cases of prethrombotic disorders (58 Raynaud syndromes, 54 cases of venous insufficiency, 60 of hip prosthesis, 40 of coronary by-pass, 60 of valvular replacement). The ratio $^+$ TG/ $^+$ FPA was calculated before, during and after efficacious or inefficacious treatment. In the cases of established thrombosis, our results confirm the leading role of platelets in the development of arterial thrombosis. The cases of venous thrombosis may be divided in two groups : simple venous thrombosis when the plasmatic factors play a leading role and complicated or recurrent venous thrombosis where the platelets play an equivalent or even a greater role.
In the cases of prethrombotic states, the role of the platelets which is important on the arterial side is generally far from negligeable on the venous side. In cases of valvular replacement or of coronary by-pass the modification of the ratio lead us very frequently to modify our prophylactic therapeutics.