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Blood, 1 August 2002, Vol. 100, No. 3, pp. 778-785

CLINICAL OBSERVATIONS, INTERVENTIONS,
AND THERAPEUTIC TRIALS

von Willebrand factor cleaving protease (ADAMTS13) is deficient in recurrent and familial thrombotic thrombocytopenic purpura and hemolytic uremic syndrome

Giuseppe Remuzzi, Miriam Galbusera, Marina Noris, Maria Teresa Canciani, Erica Daina, Elena Bresin, Silvia Contaretti, Jessica Caprioli, Sara Gamba, Piero Ruggerenti, Norberto Perico, and Pier Mannuccio Mannucci for the Italian Registry of Recurrent and Familial HUS/TTP

From the Mario Negri Institute for Pharmacological Research, Clinical Research Center for Rare Diseases, Aldo e Cele Daccò, Villa Camozzi-Ranica; Unit of Nephrology and Dialysis, Azienda Ospedaliera, Ospedali Riuniti di Bergamo; and Angelo Bianchi Bonomi Hemophilia and Thrombosis Center, IRCCS Maggiore Hospital and University of Milan, Italy.

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Whether measurement of ADAMTS13 activity may enable physicians to distinguish thrombotic thrombocytopenic purpura (TTP) from hemolytic uremic syndrome (HUS) is still a controversial issue. Our aim was to clarify whether patients with normal or deficient ADAMTS13 activity could be distinguished in terms of disease manifestations and multimeric patterns of plasma von Willebrand factor (VWF). ADAMTS13 activity, VWF antigen, and multimeric pattern were evaluated in patients with recurrent and familial TTP (n = 20) and HUS (n = 29). Results of the collagen-binding assay of ADAMTS13 activity were confirmed in selected samples by testing the capacity of plasma to cleave recombinant VWF A1-A2-A3. Most patients with TTP had complete or partial deficiency of ADAMTS13 activity during the acute phase, and in some the defect persisted at remission. However, complete ADAMTS13 deficiency was also found in 5 of 9 patients with HUS during the acute phase and in 5 patients during remission. HUS patients with ADAMTS13 deficiency could not be distinguished clinically from those with normal ADAMTS13. In a subgroup of patients with TTP or HUS, the ADAMTS13 defect was inherited, as documented by half-normal levels of ADAMTS13 in their asymptomatic parents, consistent with the heterozygous carrier state. In patients with TTP and HUS there was indirect evidence of increased VWF fragmentation, and this occurred also in patients with ADAMTS13 deficiency. In conclusion, deficient ADAMTS13 activity does not distinguish TTP from HUS, at least in the recurrent and familial

forms, and it is not the only determinant of VWF abnormalities in these conditions.

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