

OSCILLATORY RESPONSE OF RISTOCETIN INDUCED-PLATELET AGGREGATION IN THROMBASTHENIA. J. Chediak, M. Telfer, B. Vander Laan, and I. Cohen. Division of Hematology, Department of Medicine, Michael Reese Hospital, University of Chicago, and Department of Biochemistry and Molecular Biology, Northwestern University, Evanston, Ill. U.S.A.

Since the introduction of ristocetin as an aggregating agent for the diagnosis of von Willebrand's disease, variable results have been reported on its effect on thrombasthenic platelets. The platelets from 10 thrombasthenic patients were subjected to aggregation with serially increasing concentrations of ristocetin. Unlike normal control platelets which showed normal irreversible aggregation using final concentrations of 1.25 mg/ml or higher, these platelets required increased concentrations of 1.8 and 2 mg/ml for maximal aggregation and intermediate concentrations (1.25 to 1.75 mg/ml) showed a cyclic oscillatory pattern of aggregation-disaggregation for 12-15 minutes, ending in partial disaggregation. If at this point creatine phosphate/creatin phosphokinase (CP/CPK) or ATP were added the oscillatory pattern was reinstated. Measurements of pH of the stirred platelet rich plasma (PRP) at the height and depth of oscillations changed only from 7.6 to 7.8 without correlation with the pattern. Incubation of the PRP with ADP prevented the ristocetin induced platelet aggregation; it was restored with the occurrence of oscillatory pattern upon addition of CP/CPK or ATP.

The oscillatory phenomenon of thrombasthenic platelets in response to ristocetin is a unique finding. The constancy of the pH at different stages of the oscillatory waves ruled out the possibility of a gross electrostatic effect. The membrane defect reported in thrombasthenic platelets may play a role in these findings.

CONSTITUTIONAL THROMBOCYTOPATHIA WITH ABNORMAL AGGREGATION DIFFERENT FROM STORAGE-POOL DISEASE AND ASPIRIN-LIKE SYNDROME. J. Conard, M. Samama, B. Vergaftig, C. Lecrubier, J. Breton-Gorius, F. Rendu, A. Bernadou, Hôtel-Dieu, Paris and Inserm U. 150, Paris - U.91 Créteil, France.

A thrombocytopathia associated with a lifelong hemorrhagic diathesis has been observed in a 18 years old woman. The bleeding time is prolonged. Platelet count and size, and clot retraction are normal. Adrenalin-induced aggregation is abolished and response to ADP and arachidonic acid are impaired. A storage-pool disease is unlikely since platelet ultrastructural aspects appear normal and the number of dense bodies is overnormal, that is confirmed by examination of platelets charged with mepacrine. The uptake of ^{14}C serotonin is not increased and it is in correlation with abnormal fluorescence of mepacrine-stained dense bodies. Contrasting with aspirin-like syndrome, the first phase aggregation is decreased and in vitro aspirin tolerance test abnormal. Finally, although platelets do not aggregate normally to arachidonic acid, production of Tromboxane A_2 and transferable platelet aggregating activity are present. Hence, the thrombocytopathia reported here could not be classified, but a congenital defect is suspected.

INFUSION OF CRYOPRECIPITATE PARTLY CORRECTS THE PROLONGED BLEEDING TIME IN PATIENTS WITH STORAGE POOL DEFICIENCY. S.M.Gerritsen, J.W.N.Akkerman and J.J.Sixma, Dept. of Haematology, University Hospital Utrecht, The Netherlands.

Four patients with Hermansky-Pudlak Syndrome (HPS)— storage pool deficiency, albinism and ceroid containing bone marrow macrophages — from one family, one unrelated HPS patient and one patient with storage pool deficiency (SPD) were alternately treated with cryoprecipitate from 16 donors or an equal amount of human albumin. Prior to infusion of cryoprecipitate template bleeding times were all longer than 20 min. Within 2 hrs after infusion bleeding times decreased 70% of initial values. This effect lasted for at least 6 hrs but had disappeared after 24 hrs. Infusion of albumin had no effect. Similar results were obtained in 2 additional cases of SPD that were treated with cryoprecipitate only. The abnormal platelet function tests and the biochemical abnormalities remained unchanged. Infusion of cryoprecipitate protected 4 SPD patients from bleeding during surgery. Infusion of cryoprecipitate may prevent bleeding in SPD patients. Its mechanism is still obscure.