

STORAGE POOL DISEASE OF PLATELETS IN AN INFANT WITH THROMBOCYTOPENIC ABSENT RADII SYNDROME SIMULATING FANCONI ANEMIA. J. Zahavi, R. Gale and Z. Sacks. University of Tel-Aviv School of Medicine, Ichilov Hospital, Tel-Aviv, Israel.

This study reports the first example of storage pool disease, "aspirin like" variety, in a 4 month old infant with multiple congenital malformations including bilateral absent radii, thumbs and right kidney, dislocation of hip joints, equinovarus, convergent squint and hypospadias. Platelet count was decreased to $29,000/\text{mm}^3$ at birth and rose to $296,000/\text{mm}^3$ 4 months later. Studies of platelet function showed a slightly prolonged bleeding time (Ivy technic), decreased aggregation to collagen 0.003% and ADP 4 mM, total absence of aggregation to 1-epinephrine and decreased platelet factor 4 activity. Platelet serotonin content was slightly decreased. These abnormal findings were equally detected with the father's platelets, suggesting a genetic transmission of the platelet disorder. Platelet function of the mother, which is a first cousin of her husband, were normal. This study provides further evidence to the hereditary nature of thrombocytopenic absent radii (TAR) syndrome. The impaired platelet function seems to be an important and helpful test in the differential diagnosis of TAR syndrome and Fanconi anemia.

MORPHOLOGY OF BLOOD PLATELETS EXPOSED TO pH 5.3. J. Stewart and K. Davenport. SCOR Center for Thrombosis Research, Temple University Health Sciences Center, Philadelphia, Pennsylvania 19140 U.S.A.

Washed human platelets incubated at 37°C in citrate buffer of pH 5.3 released most of their stored compounds slowly ($t_{1/2}=60$ min). Addition of 2 mM NaF greatly increased the rate of release ($t_{1/2}=2$ min). Exposure of platelets to pH 5.3 caused three prominent ultrastructural changes: (1) extensive formation of long, narrow channels often parallel with the plasma membrane; (2) condensation of the densely staining component (DSC) (actin?) of the cytoplasm leaving large clear areas, the whole enclosed in an apparently intact plasma membrane; and, (3) aggregation and concentration of glycogen in these clear areas. Channel formation was immediate while the other changes developed over one hour. Most platelets contained a circular mass composed of fibrillar material (DSC) with or without granules. The ultrastructure of platelets exposed to 2 mM NaF at pH 5.3 for 3 min had the characteristics of platelets exposed to pH 5.3 alone for 60 min plus the "butterfly" appearance of platelets exposed to 10 mM NaF at pH 7.4 for 15 min. Glycogen was frequently located in bulbous protrusions. Exposure of platelets to pH 5.3 may cause a slow liberation of free intracellular Ca until the level is reached which induces secretion, while NaF may act by inducing a fast and specific rise in the free Ca level. The enhanced free [Ca] may cause the condensation and centralization of DSC, which in its course may force out the glycogen particles that were previously distributed in the cytoplasm.

METHOD FOR THE CONTROL OF FIBRINOLYTIC INHIBITORS. L. Badimón, C. A.-Villaverde, M. T. Borruel and F. G.-Valdecasas. Dept. Farmacología, Consejo Superior Investigaciones Científicas, Barcelona, Spain.

To determine easily and with precision the inhibitors of fibrinolysis, we have developed a method studying the critical Urokinase amount that can produce the fibrin plate lysis in 20 hours of incubation (37°C), by its addition to the fibrinogen solution when the fibrin plate is prepared. Placed on the coagulated fibrin layer samples (20 μl) of blood, plasma, serum or isolated inhibitors from euglobulins supernatant, we obtained circular areas of unlysed fibrin; plotting sample dilution, in logarithmic scale, against unlysed fibrin diameter, in decimal scale, a linear response is obtained. To give a standardization pattern we assayed a Trasylol curve that showed a linear response between 1 and 40 I.U./ml., so the results can be expressed in diameters of unlysed fibrin or in Trasylol units. As standard can also be used Iniprol in a range of 0.5 to 8 I.U./ml. or a pool of normal plasma, or whatever we have to evaluate in progressive dilutions. We have been contrasted this method with thromboelastographic and Blix ones, obtaining correlative results. This is an easy performance method that can be of clinical interest to study a great number of samples and to control the fibrinolytic agent treatment.