

STORAGE POOL DEFICIENCY IN NEONATAL PLATELETS. Marie J. Stuart. Dept. of Peds., SUNY, Upstate Med. Ctr., Syracuse, N.Y.

It is recognized that newborns exhibit a transient thrombocytopathy. Some investigators have described this as an "aspirin-like" defect, while others have ascribed it to a "storage-pool" abnormality. In an attempt to resolve this discrepancy platelets from 10 normal newborns, 10 normal adults and 10 aspirin treated adults were studied. None of the mothers of the newborn infants had ingested drugs known to interfere with platelet function. Platelet aggregations with ADP (3uM), epinephrine (5uM) and collagen demonstrated abnormal 2nd phase aggregation in both the newborns and in the aspirin treated adults. When mixtures of equal volumes of platelets from newborn and aspirin treated controls were tested, normal, irreversible aggregation was observed. Mutual correction was not observed on preincubation of newborn platelets with aspirin prior to mixing. When platelet malonaldehyde production was measured as an index of prostaglandin synthesis, in the presence of thrombin or NEM, no statistically different differences were observed between normal adults and newborn platelets. Malonaldehyde production was markedly reduced in the aspirin treated controls. Storage-pool deficient platelets, when combined with aspirinized platelets, prove mutually corrective in aggregation studies since storage pool deficient platelets overcome the abnormality in prostaglandin synthesis of the aspirin treated cells. These latter cells in turn provide the necessary storage-pool nucleotides to cause mutual correction and irreversible aggregation of the mixture. Our findings indicate that newborn platelets have a "storage-pool" defect.

GIANT (BERNARD-SOULIER) PLATELETS ARE NORMAL SIZED IN CIRCULATION. M.M. Froimovic, John G. Milton, Department of Physiology, McGill University, Montreal, Canada, and J.P. Caen, Department of Hemostasis and Experimental Thrombosis, Hôpital Saint-Louis, Paris, France.

Bernard-Soulier Syndrome (BSS) is a platelet disorder for which one of the characteristics is the appearance of "giant" platelets on peripheral blood smears. A comparison is made between the shape distributions of platelets obtained from normal (9) and BSS (3) donors. Geometric parameters are evaluated from a cinematographic analysis of freely rotating glutaraldehyde-hardened and unfixed platelets and of platelets on blood smear. On blood smear, normal platelets have a mean diameter of  $1.8 \pm 0.2 \mu\text{m}$ , where 10% have a diameter greater than  $2.5 \mu\text{m}$  (Weiss, et al (1974). *Am. J. Med.* 57: 920). On the other hand, BSS platelets are significantly larger, having a mean diameter of  $3.3 \pm 0.7 \mu\text{m}$ , with 80% having a diameter greater than  $2.5 \mu\text{m}$ . These observations are consistent with all published reports concerning BSS platelets and suggest donors homozygous for this disorder (Bithell, et al (1972) *N.Y. Acad. Sci.* 201: 145). In contrast, the freely circulating disc-form of the BSS platelet is essentially indistinguishable from a normal platelet: mean diameter and thickness of  $3.2 \pm 0.3 \mu\text{m}$ , and  $1.4 \pm 0.4 \mu\text{m}$  respectively, as compared with  $3.2 \pm 0.3 \mu\text{m}$  and  $1.1 \pm 0.4 \mu\text{m}$  for normal platelets.

It is concluded that the giant size of BSS platelets results from abnormal behaviour of these platelets during the preparation of the blood smear.

DEFECTIVE RISTOCETIN AGGREGATION NOT ATTRIBUTABLE TO A VON WILLEBRAND OR BERNARD-SOULIER TYPE DEFECT. J.L. Wautier, A.T. Nurden, H. Michel and J.P. Caen. Hôpital Lariboisière and Hôpital Saint-Louis, Paris, France

In von Willebrand's disease the absence of platelet aggregation by ristocetin has been correlated with abnormalities in the von Willebrand factor (VIII<sub>W</sub>), while in the Bernard-Soulier syndrome (BSS) a platelet membrane defect involving surface glycoproteins has been reported. During a 3 year period an absence of platelet aggregation induced by ristocetin was observed in a 53 year old caucasian man with enlarged spleen, normal platelet count and eosinophilic leukaemia. The platelets react normally with collagen and ADP but the aggregation induced by thrombin and bovine VIII<sub>W</sub> were reduced. The patient's platelets were well agglutinated by an antibody reacting with a component absent in the BSS. The bleeding time, VIII levels, platelet adhesion to subendothelium (rabbit aorta) were normal. No abnormalities were detected in the surface glycoproteins as studied by SDS polyacrylamide gel electrophoresis however the platelet sialic acid content was slightly reduced. It is concluded that an abnormality additional to those previously described may be the course of the defective ristocetin induced platelet aggregation in this patient.