

## POSTER SYMPOSIUM V

### Platelets: Hereditary Disorders.

ABSENCE OF FIBROBLAST-INDUCED FIBRIN CLOT RETRACTION IN A PATIENT WITH GLANZMANN'S THROMBASTHENIA AND ABNORMAL WOUND HEALING. G. de Gaetano, G. Remuzzi, G. Balconi and M.B. Donati. Istituto di Ricerche Farmacologiche 'Mario Negri', Milano, Italy, and Ospedale Maggiore, Bergamo, Italy.

Platelets and fibroblasts from normal individuals induce fibrin clot retraction (FCR). Absence of platelet-induced FCR is a characteristic abnormality of Glanzmann's thrombasthenia (GT) patients. The purpose of this study was to evaluate whether fibroblasts from a 16-year old girl with severe GT (Thromb. Haemost. 1976, 36, 286) would induce FCR.

Fibroblast culture was established from skin biopsy from the inner arm of the patient; fibroblasts were grown and FCR was evaluated as described previously (Europ. J. Cancer 1976, 12, 823). No clot retraction was observed in the samples containing GT fibroblasts during a 24-hour observation period. In control samples FCR started after about 3 hours and appeared to be completed within 12 hours (70-85%). Since fibrin-fibroblast interaction could play a role in normal tissue repair, the process of healing of the wound produced in this patient by the skin biopsy was followed. Neither haemorrhagic complications nor infections occurred during the first 10 days after the biopsy; at that time stitches were removed and the lips of the wound appeared to be well in line. However, abnormal tissue repair (lips swollen, out of the line and interwoven by recent granulation tissue) was evident 20 days later and persisted till the last outpatient control (6 months after biopsy).

These results would suggest that in GT the cellular abnormality, as indicated by absent FCR, is not restricted to platelets. Whether the abnormal platelet and fibroblast interaction with fibrin might be somehow related to defective wound healing following skin biopsy is not yet known.

MALONDIALDEHYDE (MDA) FORMATION IN PATIENTS WITH DEFECTIVE RELEASE REACTION. F.L. Pareti, L. Mannucci, A. Capitanio and D.C.B. Mills. Hemophilia and Thrombosis Center, Univ. of Milan, Italy and Specialized Center for Thrombosis Research, Temple University Hospital, Philadelphia, Pennsylvania, U.S.A.

MDA formation in platelet rich plasma (PRP) is considered to be an indicator of prosta-glandin endoperoxide production by the platelets, occurring during the release reaction. Six patients with a defect of the release reaction, storage pool deficiency (SPD), due to the lack of the storage pool of ADP, showed a normal amount of MDA formation in PRP after aggregation with collagen, thrombin (5U/ml) and after incubation with 10 mM N-ethyl-maleimide (NEM). The addition to PRP of 10  $\mu$ M ADP and 1  $\mu$ M serotonin (5HT), amounts comparable to those released during the release reaction, together with high concentrations of collagen, thrombin or NEM did not significantly enhance MDA formation. This suggests that MDA production is independent of released ADP and 5HT, and that the release induction mechanism is normal in these patients. Platelets from one patient with the "aspirin-like syndrome" did not produce MDA after stimulation with collagen, thrombin or NEM; neither did those from two normal donors after aspirin ingestion. The "aspirin-like syndrome" seems to be characterized by a specific defect in the release mechanism due to the inactivity of the cyclo-oxygenase, while the release mechanism is normal in SPD; the defect being due solely to the absence of releasable ADP.