T. Samori, M. Yatabe, M. Ukita, M. Fujimaki and K. Fukutake (Department of Clinical Pathology, Tokyo Medical College, 7–1, 6-Chome, Nishi-Shinjuku, Shinjuku-Ku, Tokyo, Japan): **A New Type of Congenital Dysfibrinogenemia (Fibrinogen Tokyo) with Defective Stabilization of Fibrin Polymers.** **(45)**

A new type of congenital dysfibrinogenemia characterized by abnormal stabilization of fibrin polymers under the normal concentration of plasma factor XIII and normal thrombin time has been discovered in Tokyo.

The molecular abnormality of this abnormal fibrinogen are shown by an anodal immunoelectrophoretic mobility and an abnormal pattern of D fragment in fibrinogen degradation products by plasmin digest on immunoelectrophoresis and crossimmunoelectrophoresis.

However, the plasma fibrinogen level of this case measures always in the normal range, when immunological methods or thrombin dependent methods are used, and the decreased level of plasma factor XIII is indicated by the use of assay methods based on clotlysis.

Andrei Z. Budzynski, Victor J. Marder, Doris Ménaché and Marie-Claude Guillin (Specialized Center of Research in Thrombosis, Temple University, Philadelphia, Pennsylvania U.S.A. and Service Central d'Immunologie et Hematologie, Hôpital Beaujon, Clichy, France): **Structural Abnormality in the Gamma Polypeptide Chain of Paris I Fibrinogen.** **(46)**

Congenital abnormal fibrinogen Paris I has an abnormal γ chain which is probably the cause of impaired polyperization of fibrin monomers and an inability to crosslink in the presence of Factor XIII. SDS-polyacrylamide gel electrophoresis demonstrated that the $A\alpha$ and $B\beta$ chains of Paris I fibrinogen are normal. However, only 1/3 of the γ chains are normal, the remaining 2/3 migrating as a heavier species between the $B\beta$ and γ chains. The conversion of Paris I fibrinogen into crosslinked fibrin in the presence of thrombin, calcium and Factor XIII is accompanied by the disappearance of the normal γ chain and the formation of γ chain dimers, but the abnormal γ Paris I chain did not disappear. There was only partial disappearance of the α chains, with approximately 2/3 not participating in the crosslinking reaction. It would appear that 2/3 of the fibrinogen molecules in the plasma of the propositus have abnormal γ Paris I chains, which are approximately 2500 daltons heavier than the normal γ chains. Plasmic digestion of Paris I fibrinogen results in the formation of an abnormal Fragment D with a heavier γ chain remnant. Degradation of Paris I fibrin yields a normal Fragment D-D and an abnormal Fragment D. The data indicate that the structural defect in γ Paris I chain occurs in the carboxy-terminal region, which can account for both the defective polymerization and imperfect crosslinking.

D. Shepro, H. B. Hechtman and F. A. Belamarich (Boston University, Departments of Biology and Surgery, Boston MA. 02215, U.S.A.): **Endothelium and Serotonin Transport.** **(47)**

The transport of serotonin (5-HT) by endothelial cells (EC) was measured in isolated blood vessels, freshly harvested cells and in the isolated dog lung. In addition cultured bovine aortic EC were used to measure 5-HT transport when they reached confluency (7 days). The EC in all preparations removed ^{14}C-5-HT even in the presence of iproniazid (5×10^{-4} M). In freshly isolated EC and cultured EC imipramine (10^{-4} M) reduced uptake. Metabolic inhibitors, cold (4° C), ouabain (10^{-5} M) also reduced uptake suggesting that 5-HT uptake may be coupled to an active transport mechanism. Six analogues of 5-HT only reduce uptake in freshly isolated EC. Lungs that showed variable degrees of pulmonary insufficiency did not remove 5-HT despite significant platelet entrapment. These results indicate that: (1) 5-HT transport is a function of all EC and not unique to the pulmonary circulation; (2) uptake of 5-HT by EC is coupled to an active transport mechanism; (3) a model using cultured EC to measure 5-HT transport is comparable to other systems, e.g. platelets and brain; (4) EC damage results in a loss of 5-HT uptake by the lung.