Time 14.30

HEREDITARY HYPODYSFIBRINOGENEMIA WITH DEFECTIVE RELEASE OF FIBRINOGENOPEPTIDE A (FIBRINOGEN FREIBURG) 0178

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A new autosomally inherited hypodysfibrinogenemia was recognized in four members in three different generations of a family. Only one patient had a major bleeding episode after trauma, the other affected members had ng history of excessive bleeding or thromboembolic disease. prohi The thrombin time and Reptilase time of plasma were greatly prolonged and partially corrected by the addition of calcium.Patient plasma pro-longed the thrombin time of normal plasma.Fibrinogen levels ranged strictly between 10 to 20 mg/100ml when measured as thrombin-clottable protein, whereas immunologically the fibrinogen levels were only slightly reduced. <u>.</u>

reduced. Functionally the major defect was impaired release of fibrinogenopeptide A upon incubation of the purified abnormal fibrinogen (94 % clottable protein)with thrombin and Reptilase. The abnormal fibrinogen showed a delayed polymerisation of its purified fibrin monomers. The described abnormal fibrinogen was indistinguishable from normal fibrinogen by polyacrylamide gel electrophoresis with and without sodium dodecyl sulfate. 0179 A NOVEL DYSFIBRI NOGENEMIA WITH ABNORMAL 7-CHAIN(FIBRI NOGEN NAGOYA). <u>Junki Takamatsu*, Kanji Ogata, Tadashi Kamiya, Katsuo Koie</u>, Ist Department of Internal Medicine, Nagoya University School of Medicine, Nagoya. <u>Takashi Takagi</u>, Biological Institute, Tohoku University, Sendai. <u>Sadaaki Iwanaga</u>, Biological Institute, op

14.45

0179

Kyushu University, Fukuoka, Japan.

Kyushu University, Fukuoka, Japan. Six individuals in 3 generations of Japanese family had prolonged thrombin clotting time but no history of hemorrhagic or thrombotic disease. Very low fibrinogen levels were but no history of hemorrhagic or thrombotic disease. Very low fibrinogen levels were obtained by thrombin clottable protein, while immunological procedures gave normal value of fibrinogen. The serum contained 40-80µg/ml of unclottable fibrinogen related antiger The patients' plasma had an inhibitory effect on the fibrin formation in normal plasma. Major defect of this fibrinogen was a delayed aggregation of fibrin monomers. ē On CM-chromatography (CM-52) of the S-carboxymethylated fibrinogen, three different On CM-chromatography (CM-52) of the S-carboxymethylated fibrinogen, three different r-chains, named r_x , r_x and r_x , were separated. They did not differ in their electrophoreties mobilities in SDS-PAGE, but were distinguishable in PAGE containing 8M urea. Moreover, the amino acid compositions and tryptic peptide mappings of each chain revealed a little difference from those of normal fibrinogen r chains, suggesting the difference in amin_acid substitution or oligosaccharide chain structure.

acid substitution or oligosaccharide chain structure. Based on these findings, we designated this fibrinogen as fibrinogen Nagoya; its possible identity without other dysfibrinogenemia has not been excluded.

15.00

0180 COMPARISON OF PLATELET AND PLASMA FIBRINOGEN FROM A SUBJECT WITH THE CONGENITAL ABNORMALITY, FIBRINOGEN PARIS I.

This (Jandrot-Perrus, M.W. Mosesson, M.H. Denninger and D. Ménaché. Service Central d'Immu-Μ. nologie et Hématologie, Hôpital Beaujon, Clichy, France.

Fibrinogen (ϕ) was obtained from normal platelets and plasma and compared with that from a subject with the congenital abnormality termed ϕ Paris I. The abnormality in this plasma ϕ is characterized by the presence of relatively large mutant γ chains (γ Paris I) that replace 50 % or more of the normal γ chain population and unlike γ chains do not form covalently linked dimers in the presence of f. XIIIa. Extracts of washed or washed trypsin-treated platelets, lysed in the presence of 8 M urea, were subjected to CM-cellulose chromatography. The ϕ containing peak from each sample was then concentrated and analyzed by SDS gel electrophoresis. All unreduced samples (5 % acrylamide) possessed a ϕ band migrating in the same position as intact ϕ . ϕ -containing slices from duplicate unstained gels of platelet samples were reduced with DTT and again subjected to SDS gent electrophoresis (10% acrylamide). All samples showed bands corresponding to the A , ${}_{\cal AB}$ and γ chains of normal plasma ϕ ; the band corresponding to the γ Paris I chain was not observed. Furthermore, there was no evidence of the presence of y Paris I chains in reduced specimens of a cross-linked fibrin clot from the γ Paris I subject's platelets. The fact that γ Paris I chains are absent from platelet ϕ Paris I subject, supports the conclusion that platelet and hepatic ϕ pools are separate pools and suggests that platelet and hepatic ϕ are not assembled from the same gene product.