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D. KAISER and O.M. RENNERT*. University Children's Hospital, University of Berne, Berne, Switzerland.
Antiprotease activity in amniotic fluid in a family with $a_1$antitrypsin deficiency (a_1-AT).

We studied a family in which a 2½ years old child exhibited liver cirrhosis in association with a_1-AT deficiency of the PiZZ genotype. The diagnosis was established in the proband at the time her mother was again pregnant and in her 8th month of gestation. Antiprotease activity was measured by the trypsin inhibitory method of Eriksson and by immunodiffusion techniques. Amniotic fluid was obtained from the PiZ heterozygote prior to delivery by needle puncture of the amniotic membrane. This fluid was contrasted with term amniotic fluid from 5 PiMM women and 5 fluids obtained during the 16-18th weeks of gestation for diagnosis of other genetic diseases. The term control specimens had a trypsin inhibitory capacity = 0.031-0.106 mg/ml, first trimester fluids = 0.073-0.175 mg/ml. Immunodiffusion indicated that serum a_1-AT was present in amniotic fluid, concentration = 0.293 ± 0.078 mg/ml. The child from the pregnancy under study had hepatic disease and was a PiZZ homozygote. Her amniotic fluid had antitrypsin activity 10% of controls (0.006 mg/ml). The feasibility of antenatal diagnosis is suggested.

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Hereditary Transcobalamin II Deficiency: Clinical findings in a new family.

A second family with transcobalamin II (Tc II) deficiency was detected. Megaloblastic anemia in early life contrasted with normal vitamin B 12 serum level, but all B 12 was carried by alpha-1-globulin (Tc I), and the normal beta-binder (Tc II) was lacking. Family studies are compatible with autosomal recessive inheritance. Analogous findings of blocked cellular maturation in the intestine (leading to malnutrition) and in the lymphoid system (agammaglobulinemia) suggest that Tc II is of vital importance for rapidly proliferating cells. Complete correction of all disturbances after B 12 therapy in pharmacologic doses was linked with appearance of a new B 12-binding alpha-2-globulin (fetal binder ?).

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"Big chymotrypsin" from human leucocytes.

A chymotrypsin-like enzyme with substrate specificity resembling pancreatic α-chymotrypsin was isolated in small quantities from human polymorphonuclear leucocytes and compared with pancreatic chymotrypsin. The enzyme had activity against a variety of chymotrypsin substrates containing tyrosin residues. It was inhibited by DFP and by TPCK suggesting the presence of serine and histidine residues in the active center. However, the inhibition by TPCK was slower for the leucocyte enzyme than for pancreatic chymotrypsin. The behaviour of the enzyme in three different separation systems suggested that the molecular weight of the leucocyte enzyme and of its subunits obtained after treatment with DTT was about twice as great as for pancreatic chymotrypsin. The enzyme was localized in the cytosol and no association with the leucocyte granules could be demonstrated. The physiological role of "big leucocyte chymotrypsin" is at the present time unknown. It may be involved in phagocytosis or in the immunological response of the cell.

DFP=Diisopropylphosphofluoride
TPCK=L-1-tosylamide-2-phenylethychloromethylketone
DTT=Dithioerythrite

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Growth of disaggregated human adipocytes in Tissue Culture.

Recent investigations suggest that certain types of human obesity may be related to excessive multiplication of adipocytes in the last trimester of gestation and the first year of life. If true, such multiplication could well be modifiable. Adipose cellular multiplication at other ages has also been suggested. This study was undertaken to determine the growth rate, in vitro, of adipocytes from children of different ages. Using a modification of Rodbell's technique adipocytes were disaggregated from adipose tissues obtained from anterior abdominal wall at laparotomies. The isolated cells were grown in McCoy's medium supplemented with 20% fetal calf serum in 5% Co_2 and 95% balanced air at 37°C. The usually spherical adipocyte changed to a fibroblast appearance in several days and gradually lost intracellular lipids. Adipocytes from a 6 day old infant multiplied several fold, cells from a 4 month old multiplied fewer times while cells from children over 24 months have not been observed to multiply and tended to keep their intracellular lipids for longer periods. These findings suggest that the potential of human adipocytes to divide and multiply, at least in vitro, may be age dependent.