69 HETEROGENEITY OF IMMUNOLOGICAL ABNORMALITIES IN ATAXIA-TELANGIECTASIA(AT)

ELENA GALLI°, M.FIORILLI°, F.PANDOLFI°, G.RUSSO°, PAOLA BETTI°, STEFANIA BOSCARELLI°, LILIANA FIORE°, F.DIANZANI°°°, LUISA BUSINCO
Dpt. Pediatr., Dpt. Immunology°, Dpt. Virology°°, University of Rome, Italy

Peripheral blood mononuclear cells(PBMC) from 8 children with AT were evaluated with a panel of Monoclonal Antibodies(Mo.Ab)(OK-T,Ortho Diagnostic System)directed against T-cell subsets and for their in vitro functions in a pokeweed mitogen-induced immunoglobulin(Ig)biosynthesis assay.All the patients had a significantly reduced proportion of cells identified by Mo.Ab to a subpopulation of T lymphocytes with helper activity(OKT4)and produced low amounts or no IgA and IgG in vitro.Ig biosynthesis was increased by the addition of normal x-irradiated PBMC in one of 3 patients.Two patients had increased proportion of suppressor T cells (OKT8),and their cells were able to suppress Ig biosynthesis(IgM=91%;IgG=55%;IgA=80% of suppression)by PBMC untreated from normal donors.A great reduction or absence of γInterferon(IFN)production after induction with either Staphylococcal enterotoxin B or galactose-oxidase was showed,while normal values of αIFN were obtained.In one case we observed a low expression of Tac antigen and absent IL2 secretion.The percentage of HNK-1$^+$ mononuclear cells was normal as well as the NK lytic activity.

70 ADULT HEIGHT AND ITS PREDICTION IN PEDIATRIC PATIENTS WITH CHRONIC RENAL FAILURE (CRF)

G. GILLI*, K. SCHÄRER, O. MEHLS
University Children's Hospital, Heidelberg, and Florence Nightingale Hospital, Düsseldorf, FRG.

Earlier findings suggested that adult height (AH) of most pediatric patients treated by hemodialysis (HD) or transplantation (TP) is subnormal and that during treatment relative height is worthening (Proc.EDTA 18:329,1981). We have reexamined this problem with improved methodology in 41 pediatric pts followed longitudinally at different stages of CRF until AH (epiphyseal closure) was attained. In addition, AH was predicted by Tanner's method. At last observation 15 pts were treated by HD, 9 by TP and 17 conservatively (CT). AH was < 2 SD from normal mean in 1/13 male and 3/28 female pts when population specific standards were considered, but only 4 pts exceeded the mean. No significant differences were found between pts treated by HD, TP or CT nor between sexes. The mean age when AH was reached was 18.6 yrs in boys and 16.2 yrs in girls, i.e. close to normal means. An acceptable prediction of AH $\pm$ 2 cm was possible from the first available bone age in 60 % of all pts, with a mean prediction error of 2.7 cm. In conclusion, stunting is infrequent in pediatric pts with CRF who have reached AH. The method applied to predict AH allows a reliable prognosis of future growth in most children with CRF.