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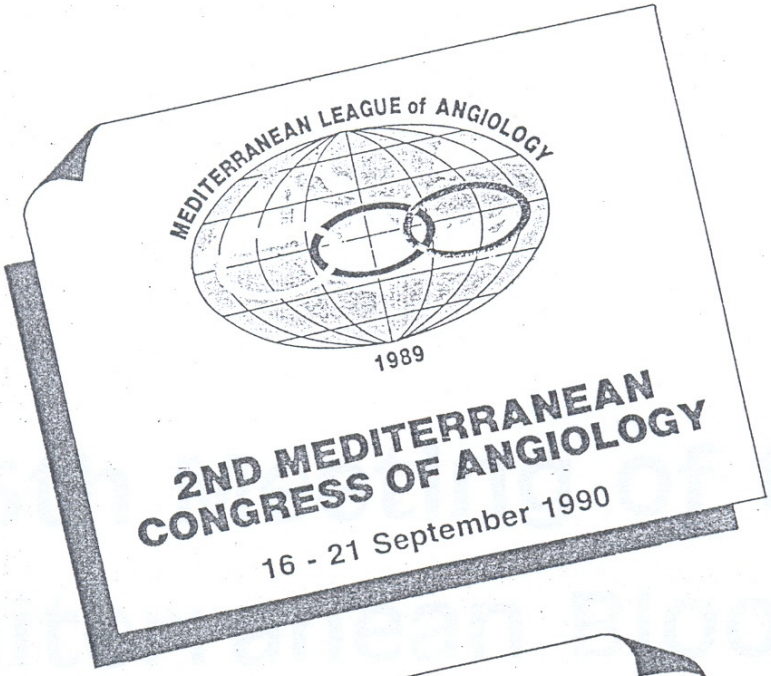

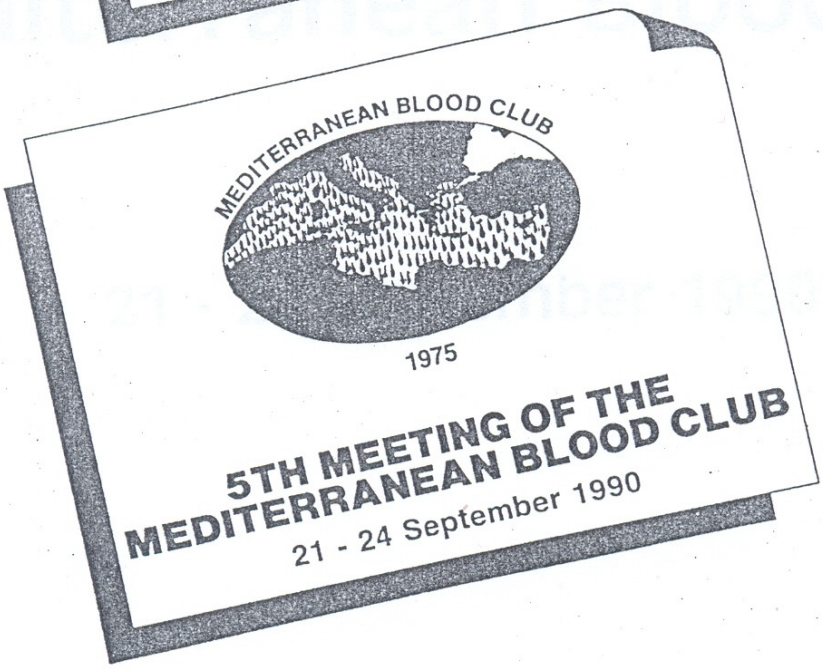

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MBC-1 (149)

EFFECTS OF ZINC SUPPLEMENTATION ON SOMATOMEDIN-C IN B-THALASSEMIA,

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Zinc deficiency is one of the factor which may contribute to the growth retardation in B-Thal patients. Zinc therapy promotes linear growth in these patients as we reported previously. Another factor related to linear growth is somatomedin-C (SM-C) and SM-C levels were found to be decreased in these patients. As well known, zinc is necessary for the synthesis of SM-C at the liver. So, it is logical to propose that zinc therapy promotes linear growth through SM-C. If this is true, it is expected that zinc supplementation will increase SM-C levels.

We conducted a survey in order to determine the effect of oral zinc supplementation on SM-C levels in twelve beta thalassemia cases, all with grovth retardation (below 3 percentile according to Turkish standards) were included in the study. Age range was 5.90 $\pm$ 11.67. Routine hematological tests, zinc and SM-C levels were determined prior to and after 12 months of zinc therapy with oral zinc sulfate (2 mg/kg of elementary zinc, daily). Zinc levels were determined with atomic absortion spectrophotometer; somatomedin-C levels were determined at the Reference Laboratories, Nichols Institute, Los Angeles, CA, U.S.A.

Plasma zinc levels, before and after 12 months of zinc therapy, were found to be 78.12 $\pm$ 18.12 and 104.33 $\pm$ 28.30 ug/dl respectively. The increase was significant ($P < 0.05$). Hair zinc levels before and after 12 months of zinc supplementation were found to be 150.35 $\pm$ 65.19 and 164.75 $\pm$ 101.62 ug/g respectively ($P > 0.05$). Normal plasma and hair zinc levels were 117.85 $\pm$ 5.82 ug/dl and 193.4 $\pm$ 53.1 ug/g respectively ($p < 0.0001$; $p < 0.05$).

SM-C levels were found to be below normal levels for age and sex. After 12 months of zinc therapy, there was no change in SM-C levels (0.36 $\pm$ 0.20 vs 0.25 $\pm$ 0.14 ; $p > 0.05$)

Although a significant relation between SM-C, zinc status and growth were reported in experimental studies; we were not able to increase SM-C levels in beta thalassemia patients after zinc supplementation. However during this period, expected linear growth was within normal limits. Hence, it was previously reported that despite an increase in growth velocity, SM-C levels may not show an increase. Or it may well be that zinc may effect linear growth from another pathway.

MBC-2 (150)

MOLECULAR BASIS OF BETA-THALASSEMIA IN TURKISH PC

(Experience of Pediatric Hematology and Oncology D of Ankara University)

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The molecular basis of Beta - Thalassemia in Turkish popu determined with hybridisation to synthetic oligonucleoti spesific to B-thalassemia mutations after using PCR techr

Twentyeight unrelated homozygous B-Thalassemia patier to 30 years, originating from different parts of Turkey we Of these six were in mild form.

Eight different mutations were found. The IVS I# 110 (G-A %); IVS II #1 (G-A) (10;17.85 %); IVS II #745 (C-G) (5;8.9 % AA) (4;7.14 %); FSC 8-9 (+G) (3;5.35 %); IVS I #6 (T-C) (1;1,8 (G-A) (2;3,57 %) and substitution at —30 (T-A) (2; 3,57 %) we ed. Six of them are still under investigation.

Of the 28 homozygous B-thalassemia patients 8 were fo homozygotes for IVS I# 110 ; 4 patients homozygotes for IV one for FSC 8; FSC 8-9; —30; IVS I #1; IVS II# 745 respecti patients were compound heterozygotes.

Mild disease was associated mostly with homozygosity fo (4 Cases).

Our results indicated that although the mutations thalassemics are heterogeneous, with the applicatio molecular techniques prenatal diagnosis can be performed