

Thesis Title THE OXYGEN AFFINITY AND RED CELL 2,3-DPG IN
 BETA-THALASSEMIA / HB E PATIENTS

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ABSTRACT

The pathophysiology of oxygen transport were studied in 32 beta-thalassemia/Hb E (beta-thal/Hb E) patients (17 splenectomized and 15 non-splenectomized) by determination of red cells 2,3-DPG and oxygen equilibrium curves (OEC) in comparison with 30 normal individuals and other kinds of anemia . The results show definite abnormal response of beta-thal/Hb E red cells to the degree of anemia by inappropriate increase synthesis of 2,3-DPG and thus inappropriate increase of partial oxygen pressure at 50 % saturation (p50) . The observed values of 2,3-DPG and p50 are significantly lower than the expected values . The mean observed 2,3-DPG concentration was 14.95 ± 2.76 $\mu\text{mol/g Hb}$ and the mean expected value was 20.9 $\mu\text{mol/g Hb}$. The mean observed p50 was 29.14 ± 1.85 mm Hg and the mean expected value

was 38.8 mm Hg . Besides inappropriate increased red cells 2,3-DPG , the p50 was decreased by the effect of high percentage of Hb F content (36.05 %) in beta-thal/Hb E red cells . The study on Hb E both intact homozygous Hb E red cells and hemolysate showed no effect on p50 . Thus the pathophysiology of oxygen transport in beta-thal/Hb E red cells is abnormal in adaptation to the decline of hemoglobin concentration both in splenectomized and non-splenectomized patients .

The mechanism of inappropriate increased synthesis of 2,3-DPG is possibly due to the blockage of enzyme aldolase which sensitive to the thiol oxidation and heavy metal . This findings characterize only in beta-thal/Hb E and homozygous beta-thal . In alpha-thalassemia such as Hb H diseases , the low p50 is due to high oxygen affinity of Hb Bart's and Hb H . The red cells 2,3-DPG in alpha-thalassemia can increase appropriately with the degree of anemia . So , the severity of the clinical manifestation in beta-thal/Hb E patients correlates well with the degree of anemia and the extent of fuctional abnormal adaptation to the decline of Hb concentration .
