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JONGKOL SANKOTE : ABNORMAL IRON DEPOSITS IN THALASSEMIC RED CELLS. THESIS ADVISOR : RUCHANEEKORN KALPRAVIDH Ph.D., PENSRI POOTRAKUL M.D. 169 p. ISBN 974-662-672-8

This study explores whether deferiprone at a low dose of 50 mg/kg/day is effective in chelating RBC membrane free Fe in long term administration for patients with β-thalassemia (β-thal) in Thailand. The ferrozine reaction for RBC membrane free Fe in Thai β-thal and α-thal patients was examined. The study revealed that the RBC membrane free Fe in various genotypes of 100 thalassemic patients was higher than that in normal subjects. Normal subjects showed undetectable amounts but the mean free Fe level on RBC membranes of β-thal was 35.86±31.57 nmole Fe/mg protein, higher than on α-thal which was 5.69±5.88 nmole Fe/mg protein. The RBC membrane free Fe in nonsplenectomized β-thal/Hb E was 18.63±22.44 nmole Fe/mg protein, and was markedly high in splenectomized β-thal/Hb E, 51.12±31.25 nmole Fe/mg protein. In α-thal 1/HbCS, RBC membrane free Fe was 9.42±6.32 nmole Fe/mg protein while α-thal 1/α-thal 2 showed lower levels of 2.13±2.00 nmole Fe/mg protein. An increase in amount of RBC membrane free iron was related to the decrease in hemoglobin concentration, an increase in transferrin iron saturation, serum ferritin, and RBC ferritin.

The subjects include 12 patients of both sexes with β-thal diseases (10 β-thal/Hb E, 2 β-thal homozygotes), age 19-46 years who are non-transfusion dependent or irregularly transfused. They received deferiprone 50 mg/kg/day for a period of 17-84 weeks except 3 cases who received 25 mg/kg/day. One of the latter subjects had the dose increased to 50 mg/kg/day after 24 weeks of administration. Ten cases were evaluable. Their RBC membrane free Fe at initial of trial ranged from 17.70-116.94 nmole Fe/mg protein. After treatment, the RBC membrane free Fe decreased from 67.98±36.05 nmole Fe/mg protein to 5.61±5.71 nmole Fe/mg protein with the range from undetectable amounts to 14.68 nmole Fe/mg protein. The drug was then withheld afterward. The RBC membrane free Fe disappeared with deferiprone treatment in 3 patients. The serum ferritin levels also decreased to normal or near normal level after deferiprone administration, including an increase in urinary iron excretion and slight decrease in RBC ferritin. The patients treated with deferiprone required less or no transfusion during the treatment. There were no serious side effects related to deferiprone except gastrointestinal symptom which disappeared within 2 weeks after administration. Therefore, deferiprone is effective in chelating iron in non-transfusion dependent thalassemia.