IDENTIFICATION AND SOME PROPERTIES OF HEMOGLOBIN QUEENS [α 34 (B15) LEU → ARG] IN A THAI SUBJECT

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A THESIS SUBMITTED IN PARTIAL FULFILMENT OF
THE REQUIREMENTS FOR THE DEGREE OF
MASTER OF SCIENCE

IN

BIOCHEMISTRY

FACULTY OF GRADUATE STUDIES

MAHIDOL UNIVERSITY

1986

ABSTRACT

A twenty-four year old healthy Thai male of Chinese ancestry with normal hematologic profile was found to have an abnormal hemoglobin (about 12% of total hemoglobin) moving slower to the anode than Hb A on electrophoresis in starch gels and on cellulose acetate membranes in Tris-EDTA borate buffer, pH 8.6 and pH 9.1 respectively. Another faint abnormal band moving slower to the anode than Hb A_2 was also detected on starch gel electrophoresis so that the abnormality in the propositus was believed to be an α chain variant. Electrophoresis of globin chains on cellulose acetate in acid-urea-mercaptoethanol buffer, pH 6.5 and on Triton-X 100-acid-urea polyacrylamide gels indicated that the abnormal globin chain moved faster to the cathode than normal α chain. Preparative isolation of the abnormal hemoglobin was performed by DEAE-cellulose column chromatography and the abnormal α chain was purified by CM-cellulose column chromatography. A fingerprint of the tryptic peptides of the abnormal α chain from the propositus showed that $\alpha T_p V$ was lost but two new peptides were present, which had amino acid compositions indicating the replacement of a leucyl residue in position 34 by an arginyl residue. This amino acid interchange in the α_1 β_1 contact was first found in 1979 by Tatsis and named Hb Queens [α 34 (B15) Leu → Arg]. The whole hemolysate of the propositus showed normal heat stability and isopropanol stability, but the isolated abnormal

hemoglobin was somewhat less heat stable than Hb A. Red blood cells showed normal morphology and osmotic fragility, and were negative for the inclusion body test.