MEASUREMENT OF THE FREE ERYTHROCYTE PROTOPORPHYRIN IN CHILDREN WITH MICROCYTOSIS

THESIS

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BY

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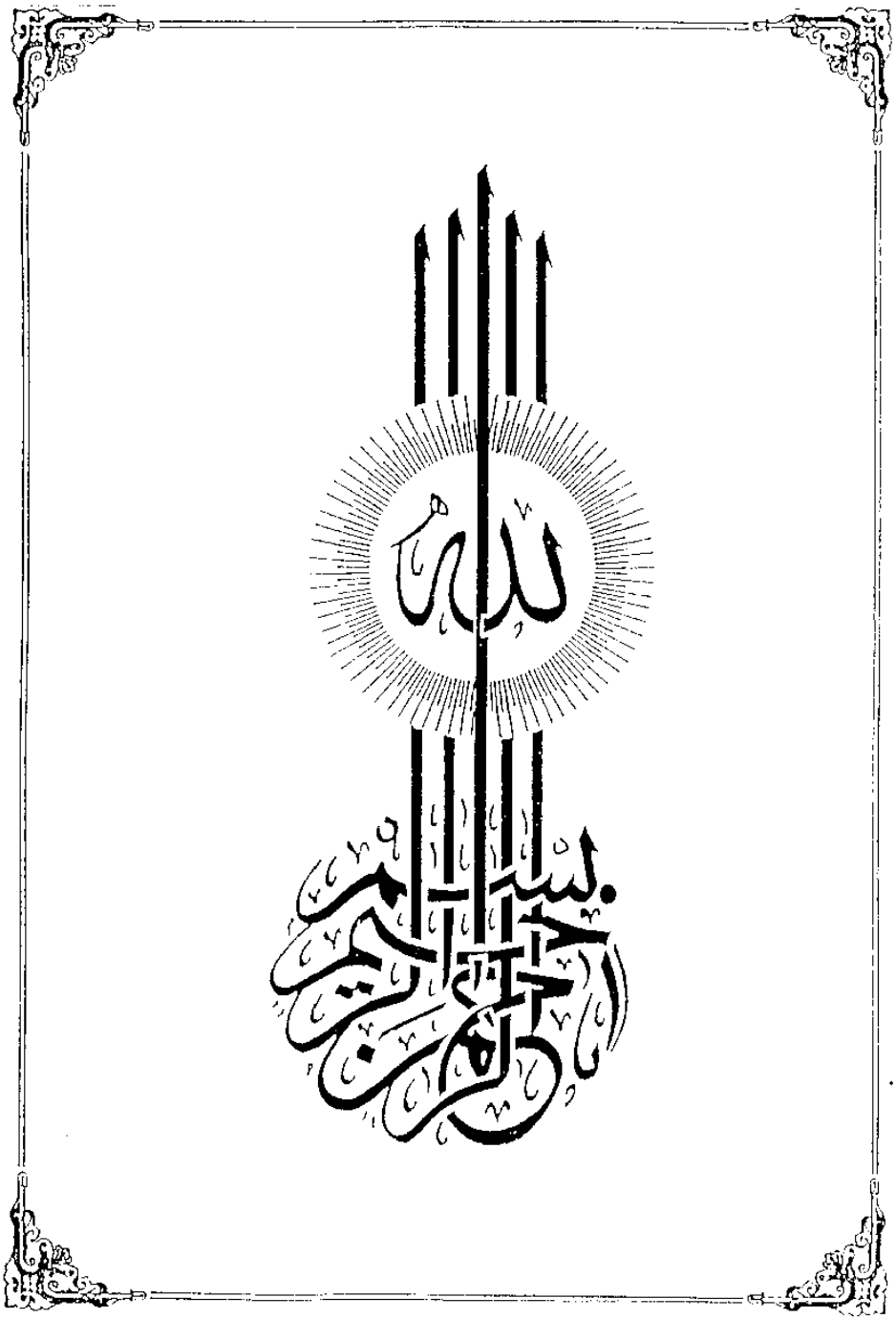
IN THE NAME OF GOD



TO MY PARENTS AND MY DAUGHTER,
FOR THEIR CONTINUOUS ENCOURAGEMENT AND LOVE

Aknowledgement

ACKNOWLEDGEMENT

I wish to express my appreciation and gratitude to ***Professor A.S. Khalifa,*** *Professor of pediatrics, to whom I am deeply indebted for his helpful supervision.*

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INTRODUCTION AND AIM OF WORK

Thalassaemia syndromes and iron deficiency are considered to be the commonest haematologic disorders that pose major public health problems in many parts of the world, particularly in the underprivileged communities. Reports throughout the world support the high prevalence of iron deficiency anaemia in young children, especially in the developing countries (Bothwell et al., 1979); meanwhile, heterozygous B-thalassaemia is considered as the most common type of the thalassaemias (Pierce et al., 1977). In Egypt, Sabry et al. (1973) found that its incidence was 1.3%.

B-thalassaemia trait mimicks iron deficiency in the presence of both microcytosis and suppressed haemoglobin synthesis (Bainton Finch, 1964; Nathan, 1972; and Ward, 1979), and it is usually not possible to make a distinction between both disorders on examination of the blood smear.

For identification of iron deficiency, it was found that determination of the plasma ferritin concentration, which closely parallels body iron reserves, is the most

valuable criterion (Jacobs et al., 1972; Lipschitz, et al., 1974) on the other hand, globin synthetic rate measurement provides the most definitive method to diagnose heterozygous B-thalassaemia (Beris et al., 1980 and Congote, 1981). But since these procedures of diagnosis are not suitable for general use in the routine clinical laboratory, a simple, accurate and precise method is needed to differentiate between iron deficiency as a cause of anaemia and B-thalassaemia trait.

Dagg, Goldberg and Lockhead (1966) found that the estimation of free erythrocyte protoporphyrin (FEP), together with percentage saturation of the total iron binding capacity, allowed an accurate diagnosis of latent iron deficiency as supported by Langer et al., (1972) and Van Eijk et al. (1974).

Meanwhile, Pearson, O'Brien and McIntosh (1973) have demonstrated that the electronic measurement of mean corpuscular volume (MCV) was a reliable screening test for thalassaemia trait among adults; but this was not suitable procedure in children due to the age related changes occurring in MCV during childhood, as

described by Berman et al. (1980), and because of the incidence of microcytosis caused by iron deficiency.

AIM OF WORK:

The aim of this work is to measure the free erythrocyte protoporphyrin (FEP) simultaneously with the determination of the mean corpuscular volume (MCV) in children with microcytosis, since heterozygous B-thalassaemia and iron deficiency anaemia are known to be common haematological disorders among children in the region.

An attempt to evaluate the usefulness of such test in the differential diagnosis between both conditions will be the matter of this study.

Review of Literature

1- HAEMOGLOBIN

Molecular Structure of Haemoglobin:

Haemoglobin is one of the most important biological constituents of the body. It has been known for over a century that human haemoglobin is a heterogenous intra-erythrocytic respiratory protein of a molecular weight 64 000 daltons, and it is thought to be ellipsoid in shape with a central cavity (Ingram et al., 1956).

The basic structure of haemoglobin consists of two alpha and two non alpha polypeptide chains of globin, colourless protein, on the surface of which are attached four small prosthetic groups of haem molecules that impart to haemoglobin its red colour and its unique oxygen carrying properties (Granick, 1949) (Figure 1).

GLOBIN

In normal adult haemoglobin (A), there are two alpha-chains made up of 141 amino-acids each, and two Beta-chains each of which are 146 amino-acids long (Rhine-Smith et al., 1957).

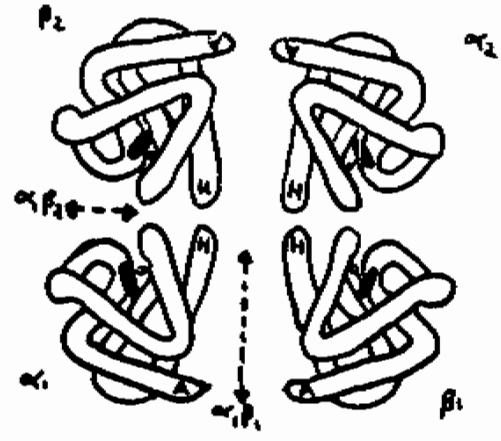

Figure (1): The arrangement of the 4 polypeptide chains of haemoglobin.

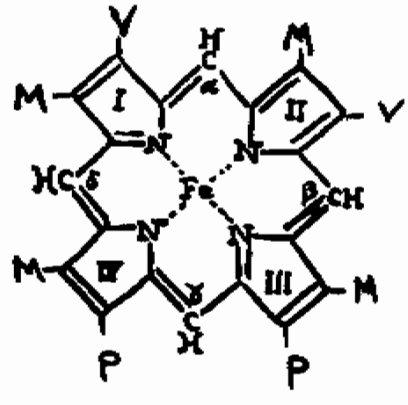

Figure (2): Fe protoporphyrin.

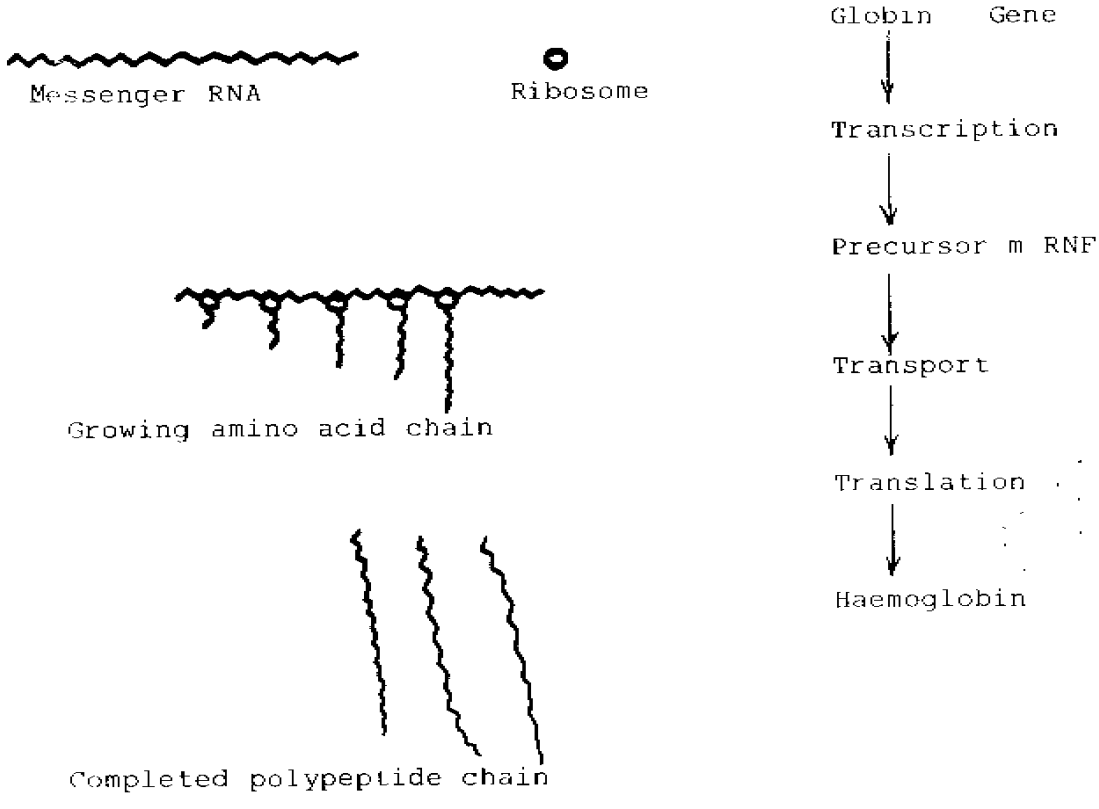

Figure (3): Messenger RNA carries the coding for the sequence of amino acids required to build globin. A ribosome moves to mRNA, and the appropriate a.a. on its specific transfer RNA is brought. Further a.a. are joined on to form the polypeptide globin chain.