# Study of Exhaled Carbon Monoxide Concentration in Beta-Thalassemia and its Relation to Red Blood Cell Transfusion Therapy in Pediatrics

Thesis

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**Pediatrics**

By

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# دراسة نسبة غاز أول أكسيد الكربون فى هواء الزفير فى الأطفال المصابين بأنيميا بيتاثلاسيميا وعلاقته بنقل خلايا كرات الدم الحمراء

رسالة

توطئة للحصول على درجة الماجستير فى

طب الأطفال


مقدمة من

**الطبيب/ عبدالمحسن أحمد عبدالمحسن نايل**

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# SUMMARY

β-thalassemia is a major global public health probem. It is more prevalent in mediterranean countries. Population migration and intermarriage between different ethnic groups has introduced thalassemia in almost every country of the world.

Exhaled carbon monoxide is a relatively new non invasive marker in various systemic diseases.

The current study measured the level of exhaled carbon monoxide in β-thalassemia patients and its relation to red blood cell transfusion and chelation therapy.

The present study was carried on Fifty (50) Egyptian β-thalassemia patients: 23 (46%) females and 27 (54%) males with mean ages of 13.01±3.82 years. We measured ExCO immediately before RBC transfusion and after it by One day and One week. Thirty (30) age and sex-matched clinically healthy children were included as negative control.

***All studied patients were subjected to the following:***

Full medical history, thorough clinical examination, hemoglobin level and CO Analysis using CO analyzer (Pico+).


بسم الله الرحمن الرحيم
وفي أنفسكم
أفلا تبصرون
سورة الذاريات
الاية (٢١)
صدق الله العظيم


This work is dedicated to . . .

The memory of my mother, my elder sister

and to

My lovely son "Mido"

My father

My dear wife

My sister "Amal"

for being the light of my life.


*Thanks to* **ALLAH**, *Most Gracious, Most Merciful, who gives everything we have and gave me the power and patience to finish this work.*

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*Abdelmohsen*

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# INTRODUCTION

Beta-thalassemia (β-thalassemia) is a hereditary blood disorder characterized by reduced or absent beta globin chain synthesis, resulting in reduced hemoglobin in red blood cells (RBCs), decreased RBCs production and anemia ***(Galanello and Origa, 2010).***

There are three clinical and hematological conditions of increasing severity recognized in children, i.e., the β-thalassemia carrier state, thalassemia intermedia, and thalassemia major. The β-thalassemia carrier state, is clinically asymptomatic. Thalassemia major is a severe transfusion-dependent anemia. Thalassemia intermedia ranging in severity from the asymptomatic carrier state to the severe transfusion-dependent anemia ***(Cao and Galanello, 2010).***

β-thalassemia is prevalent in Mediterranean countries, the Middle East, Central Asia, India, Southern China, and the Far East as well as countries along the north coast of Africa. Population migration and intermarriage between different ethnic groups has introduced thalassemia in almost every country of the world. About 60,000 symptomatic individuals born annually, the great majority in the developing world. The total annual incidence of symptomatic individuals is estimated at 1 in 100,000 throughout the world ***(Galanello and Origa, 2010).***

Blood transfusions along with iron chelation therapy have been the mainstay of treatment for years ***(Singh et al., 2010).***

Human exhaled breath is a complex mixture of low molecular weight gases, containing a multitude of organic and inorganic molecules. Recent researches have focused on the promise that gases can serve as diagnostic indices of disease, including lung and airways diseases, and other systemic diseases ***(Ryter, 2010).***

Exhaled carbon monoxide (ExCO) may originate from the inspiration of ambient CO, as found in smoke and air pollution, but also from endogenous metabolic sources that include heme metabolism catalyzed by the heme oxygenase enzymes ***(Ryter and Sethi, 2007).***

The concentration of CO in end-tidal breath can be measured and used as an index for the rate of heme degradation, RBCs life span or severity of hemoglobinopathy in children ***(James et al., 2010).***

# AIM OF THE WORK

The aim of this work is to study exhaled carbon monoxide concentration in β-thalassemia and its relation to red blood cell transfusion therapy in children.

# THALASSEMIAS

The thalassemias are a group of inherited hematologic disorders caused by defects in the synthesis of one or more of the hemoglobin chains. Alpha thalassemia is caused by reduced or absent synthesis of alpha globin chains, and beta thalassemia is caused by reduced or absent synthesis of beta globin chains. Imbalances of globin chains cause hemolysis and impair erythropoiesis ***(Muncie and Campbell, 2009).***

## Historical Background:

In 1925, Thomas Cooley and Pearl Lee described a form of sever anemia, occurring in children of Italian origin and associated with splenomegaly and characteristic bone changes ***(Cooley and Lee, 1925).***

In the past 20 years, the two important forms of this disorder, alpha and beta thalassemia resulting from the defective synthesis of alpha and beta globin chains of hemoglobin, respectively, have become recognized as the most common monogenetic diseases in humans and globally it is estimated that there are 270 million carriers, of which 80 million are carriers of beta thalassemia ***(Cunningham et al., 2004 a).***