

Relation between nutritional status and inherited disorders of hemoglobin (thalassemia) in Egyptian children

Heba SA¹; Baseem AZ² and Abdelhamid SE³

^{1,2}PhD Nutrition, ³ MD in pediatrics

1Clinical nutrition Dep. NNI, 2-Cairo University Hospitals, 3-Hid of Ped Dep. Banha University

ABSTRACT

The thalassemias are a group of inherited disorders of hemoglobin, first reported independently from the United States and Italy in 1925. The word 'thalassemia', derived from Greek roots for 'the sea' and 'blood', was invented under the mistaken belief that these disorders were confined to the Mediterranean region. The thalassemia's are the commonest genetic disorders in humans and represent an increasing public health problem in the tropical countries in which they occur at a high frequency. A retrospective design was used in the study. This design according to the available literature is considered to be appropriate as it allows the researcher to assess the dependent variable in the present (thalassemia) and then links this factors that affecting occurring on the sample, socioeconomic and physical parameters. The total sample size was 100children (50Boy) and (50 Girl). Their ages ranged 6-12 years. The study showed that the changes in physical, physiological, anthropometrics measurement, laboratory investigation of blood and behavioral affected by the nutritional status of them.

Key word: *Thalassemia, diet, children*

INTRODUCTION

The thalassemias are the commonest genetic disorders in humans and present an increasing public health problem in the tropical countries in which they occur at a high frequency (**Weatherall and Clegg, 2000**).

Thalassemia is an inherited disorder of autosomal recessive gene disorder caused by impaired synthesis of one or more globin chains. The impairment alters production of hemoglobin (Hb). The term "thalassemia" is derived from the Greek words "Thalassa" (sea) and "Haema" (blood). The clinical manifestations are diverse, ranging from absence of symptoms to profound fatal anemia in utero, or, if untreated, in early childhood (**Giardina and Forget, 2008**).

The α -thalassemia is most prevalent in Asian and African populations. Persons of Mediterranean and African descent have the highest incidence of β -thalassemia. Other abnormalities of

hemoglobin also occur with increased frequency in these populations: therefore, thalassemia may coexist with other disorders of hemoglobin such as the sickle cell syndromes, hemoglobin E (Hb-E), or hemoglobin C (Hb-C) (**Weatherall, 1987**).

Thalassemia is a heterogeneous inherited disorder of hemoglobin synthesis due to mutations of the globin gene, leading to various degrees of quantitative defect in globin production and reduced synthesis or complete absence of one or more of the globin chains, resulting in ineffective erythropoietin and anemia.

Beta-thalassemia major (BTM) is usually presented at 4 - 6 months of life, due to the protective effect of high hemoglobin F concentration at birth that slowly declines through the first year of life. Manifestations are those of anemia (pallor, lethargy...etc), failure to thrive and organomegaly. Patients presenting later will have signs of extra medullary hematopoiesis;

(frontal bossing of the skull, hepatosplenomegaly, thinning of long bones cortices, widening of medullary and diploic spaces; resulting in bossing of skull, prominence of the upper incisors and wide separation of orbits) (**Kesse-Adu and Howard, 2013**).

The thalassemia syndrome is classified according to which of the globin chains, α or β , is affected. These are 2 major groups α - and β -thalassemia (**Giardina and Forget, 2008**).

Of interest, when compared to age and gender matched controls, the children with thalassemia had similar dietary intake at baseline despite marked growth and body fat deficits. After increasing caloric intake by 30–50% over an 8-week period, they observed significant improvements in weight; fat stores, albumin and insulin-like growth factor 1 (IGF-1) levels compared to the non-supplemented thalassemia groups. The improvement in IGF-1 following nutritional therapy supports the opinion that components of the growth failure in

thalassemias are related to nutritional deficiency. Unfortunately, the majority of the published work in this area has summarized observations or interventions in relatively small numbers of subjects primarily conducted outside of North America in low profile journals, therefore has received little attention (**Walter et al., 2006**).

Borgna-Pignatti (2007) reported that thalassemic children had increased energy expenditure and protein turn over, while they had the average energy intake lower than the recommended daily dietary allowances. These patients also had multiple deficiencies of vitamins and minerals such as vitamin A, vitamin E, foliate, vitamin B12, and zinc.

Therefore, thalassemic children are at risk of energy and nutrient deficiencies. The results of malnutrition can affect the maturation and growth of the children. Previous studies showed that thalassemic patients had growth failure, delayed puberty and underweight. Nutritional statuses in

thalassemia patients should thus be evaluated (**Claster *et al.*, 2007**).

In addition to the potential increased requirement for total calories, others have shown that the requirements for specific nutrients may be increased in patients with thalassemia. The clearest example of this is with regard to foliate metabolism. Folate, an essential nutrient required for normal erythropoietin activity, has been shown recently to be readily catabolized by ferritin. Thus, in patients with thalassemia with hyperactive erythropoiesis, confounded by iron overload, foliate requirements are increased and deficiencies are commonly reported (**Ozdem *et al.*, 2008**).

Optimal nutritional status is imperative for achieving the genetic potential for growth and pubertal development in children, and poor growth and delayed puberty is considered a red flag to any pediatrician leading to exploration of possible nutrient specific or global nutritional deficiencies. In the adult

patient, altered nutritional status may adversely affect immune function, oxidative stress, bone health, and/or may limit the effectiveness of some medications (**AAPCN, 2008**).

In sufficient antioxidant intake has been associated with oxidative stress. In the early 1990s, a group in Thailand was able to reduce circulating glutathione peroxidase in young patients with thalassemia through supplementation with 200 mg of vitamin E for 4–8 weeks (**Vogiatzi *et al.*, 2009**).

Thongkijpreecha *et al.*, (2011) assessed the nutritional status in patients with thalassemia intermediate. The results of anthropometric measurements in this study indicated that patients with thalassemia intermediate had growth impairment. Although the subjective global assessment (SGA) and biochemical parameters appeared normal in these patients, their dietary intakes were apparently inappropriate. Therefore, the nutritional assessment and appropriate nutritional interventions should be incorporated into

therapeutic plans for these patients to improve growth status and clinical outcomes.

One nutrient studied widely in patients with thalassemia over the past few decades is zinc, an essential trace element found in the diet that cohabitates along with iron. Dietary survey studies suggest that decreased dietary zinc intake is limiting in a small percentage of patients with thalassemia. Increased urinary excretion and elevated requirements are more likely the reason for depressed circulating plasma zinc levels (**Fung *et al.*, 2012**).

Fung (2016) focused the relationship between nutritional status and three commence morbidities in patients with thalassemia. There is a vital need for studies to explore the optimal dose of calcium and vitamin D, which minimizes hypercalcemia while improving bone outcomes. Moreover, there is a paucity of research focused on the ultimate goal of reducing fracture risk. Future research could consider nutrient

supplementation cocktails for health outcomes in both patients with transfusion dependent thalassemia and in non-transfused thalassemia patients. Until these data are gathered, it is suggested that both pediatric and adult patients be monitored frequently and nutritional deficiencies corrected when observed in order to improve the overall health and quality of life in patients with thalassemia.

So, this study carried out to investigate the constellation between different nutritional, biochemical, socioeconomically factors and thalassemia.

Design:

A retrospective design was used in the study. This design according to the available literature is considered to be appropriate as it allows the researcher to assess the dependent variable in the present (thalassemia) and then links this factors that affecting occurring on the sample, laboratory and research physical health.

- **Sampling:**

The total sample size was 100 children (50 Boys) and (50 Girls). Their ages ranged between 6-12years.

- **Setting of the study:**

This study was conducted at Banha university hospital.

- **Period of study:**

The present study started in December 2016 and ended in January 2018.

Instrumentation

Demographic data

Demographic data including age, sex, educational level of parents and their income were estimated.

24 - Hour Nutrient Recall Method

Estimates of the adequacy of nutrients intake were based on data of individual dietary intake. The individual Intake component of the survey consisted of a 24-hour recall method. The format of the form used

to record food intake of specific foods and dietary supplements. The sample asked to recall everything that they ate within the last 24-hour or the previous day. Description of all foods and averages consumed, including food obtained away from home and the exact eaten quantities (**Rasanen, 1979**).

Assessment of Nutrient intake from food consumption data

The daily food intake had been assessed for each subject from the data collected by the 24-hour recall method for seven days. Portion size had been converted into equivalent weights. Kinds and amounts of the consumed food per meal had been tabulated. Nutrient values were derived from standard reference tables (**Food and Nutrition Board (2008) and NNI Food Composition Table in Cairo (2006)**).

For each food calculation were made of the contribution of food energy, protein, carbohydrate, fat, iron,

calcium, phosphorus, sodium, potassium, zinc, magnesium, vitamin "A, D, E, C, B complex" and foliate. The nutritive value of the diet was then compared with the calculated total of the recommended dietary allowances (WHO, 2010) appropriate for the individuals in the study. Estimation of the adequacy of nutrient intake was based on data on individual dietary intakes.

Anthropometric measurements ***Height***

It was measured using the Raven Monometer, with direct reading of height. Then, the following categories of height status were determined according to World Health Organization (WHO, 2007).

Weight

It was recorded using platform scale; the scale was standardized by known weight before the survey in each

studied site and corrected according to WHO (2007).

Assessment of Body Mass Index for age: from 5-19 years old, the Z score body mass index was used for males. The following categories of weight status were determined according to WHO Z score body mass index released by WHO, (2007).

Over weight >+1 SD

Obesity >+2SD

Thinness <-2SD

Sever thinness <-3SD

The other tested anthropometric measurements as arm circumference, triceps skin fold thickness and arm muscle circumference were recorded by the methods of (Jelliffe, 1966).

Laboratory investigation

Calorimetric determination of all blood parameters was carried according to Drabkin, (1949).

Statistical Analysis

The first step done in order to analyze the collected data was to tabulate all the raw values for each variable, and for girls and boys children. Correlation coefficients between all the variables of this study were obtained and their significances were also identified by **Artimage and Berry (1987)**. Statistical Package for Social Science, Computer Software, IBM, SPSS Ver. 16.0 in 2008, SPSS Company, London, UK.

Results and discussion

Table (1) shows the intercorrelation between social variables for children with thalassemia. With respect to family member, it correlated significantly with pocket money ($P<0.05$). Father's income, it correlated significantly positive with mother's income, other income, total income and the pocket money ($P<0.01$). Also positive correlation was found between mother's income and

other income, total income and the pocket money ($P<0.01$). As for other income, it correlated significantly positive with total income and pocket money ($P<0.01$), while there are a negative correlation coefficient other between the income and the hours of watching Television ($P<0.05$).

Furthermore total income, it correlated significantly positive with the pocket money ($P<0.01$) and a negative correlation was found between the pocket money and the hours of watching T.V ($P<0.05$). Sleeping hours, it correlated significantly positive with hours of watching television ($P<0.01$). This finding was consistent with **Ali et al., (2015)** who observed that thalassemia patients had significantly higher scores in the social domain.

The results of table (2) discussed the correlation

coefficient between social variables and nutrients intake. With regard to age in years it correlated significantly positive with vitamin "D" and vitamin "B6" ($P < 0.01$) ($r = 0.238$ and 0.271) respectively, calories, total fat and niacin ($P < 0.05$) ($r = 0.177$, 0.178 and 0.190) respectively, father's income, it correlated significantly positive with calories, total protein, total fat, calcium, phosphorus, and niacin ($P < 0.01$) ($r = 0.266$, 0.344 , 0.318 , 0.318 , 0.241 and 0.327) respectively. With regard to mother's income, it correlated significantly positive with calories, total protein, total fat, carbohydrates, calcium, vitamin "B1" and niacin ($P < 0.01$) ($r = 0.455$, 0.402 , 0.392 , 0.382 , 0.492 , 0.453 and 0.508) respectively. With respected to other income, it correlated significantly positive with calories, total protein, total fat, calcium, phosphorus, vitamin "A" and

niacin ($P < 0.01$) ($r = 0.355$, 0.431 , 0.422 , 0.433 , 0.317 , 0.293 and 0.405) respectively. Also, It was found a correlated significantly positive with carbohydrates and vitamin "B6" ($P < 0.05$) ($r = 0.187$ and 0.187) respectively, also between total income and calories, total protein, total fat, calcium, phosphorus, vitamin "A", and niacin ($P < 0.01$) ($r = 0.379$, 0.446 , 0.438 , 0.438 , 0.337 , 0.258 , and 0.402) respectively. Total income had a positive correlation with carbohydrates and vitamin "B6" ($P < 0.05$) ($r = 0.215$ and 0.204) respectively. Sleeping hours, it correlated significant positive with carbohydrates, total iron, magnesium, vitamin "B1", vitamin "B2" and vitamin "B12" ($P < 0.01$) ($r = 0.298$, 0.271 , 0.301 , 0.252 , 0.256 and 0.313) respectively. Also there are a correlation significant positive between hours watching T.V and calories, carbohydrates, phosphorus, total iron,

magnesium, vitamin "E", vitamin "B1", vitamin "B2", vitamin "B6", and vitamin "B12" ($P < 0.01$) ($r=0.248, 0.331, 0.308, 0.478, 0.472, 0.300, 0.367, 0.367, 0.241, 0.293$) respectively.

A negative correlation was found between family member, and calories ($P < 0.05$) ($r = 0.142$), also among vitamin "C", vitamin "B6" with family member ($P < 0.01$) ($r = 0.252$ and 0.025) respectively. The majority of the intake of these nutrients was at the low satisfied point and this due to the low level of nutrient intake and the low level of social variables (*Kaplowitz et al., 2001; Grunbaum, 2004 and Thoits, 2010*). Complementary and alternative medicine (CAM) as some foods is frequently being used in thalassemia patients to ensure their sense of well-being and help them overcome the complications of their

illnesses (**Bordbar et al., 2018**).

Data presented in table (3) shows the intercorrelation between anthropometric measurements. It could be noticed that a positive correlation coefficient between weight, height, body mass index, arm circumference and triceps skin fold thickness ($P < 0.01$). ($r=0.492, 0.805, 0.689$ and 0.553 respects). With respect to height, it correlated significantly positive with arm circumference ($P < 0.05$). Body mass index, it correlated significantly positive with arm circumference triceps and skin fold thickness ($P < 0.01$). Arm circumference correlated significantly positive with triceps skin fold thickness and arm muscle circumference ($P < 0.01$). As for there is a negative correlation was found between triceps skin fold thickness and arm muscle circumference ($P < 0.01$).

Table (4) explained the correlation between anthropometric measurements and social variables. With respect to weight, it correlated significantly positive with age in years, father's income, other income, pocket money amount, total income, sleeping hours and hours of watching TV ($P < 0.01$). It can be noticed that the highest correlated significantly positive with other income and pocket money amount ($P < 0.05$), negatively with hours of watching TV. As for body mass index correlated significantly positive with age in years, sleeping hours, hours of watching TV ($P < 0.01$) and father's income, pocket money and total income ($P < 0.05$). Also arm circumference it correlated significantly positive with pocket money and sleeping hours ($P < 0.05$) and there are correlated significantly positive with hours of watching TV. Also triceps skin fold thickness it

correlated significantly positive with family member, hours of watching TV ($P < 0.05$) and sleeping hours ($P < 0.01$) while correlated negatively with age in years ($P < 0.05$). A positive correlation coefficient was observed between arm muscle circumference and age in years ($P < 0.01$) and negatively with family member ($P < 0.05$) and sleeping hours ($P < 0.01$). These results were harmony with search done by **Ali et al., (2015)**

Data of table (5) show the correlation coefficient it was observed a positive correlation coefficient between K calories and weight, body mass index, arm circumference, ($P < 0.01$) and arm muscle circumference ($P < 0.05$). Also total protein, it correlated significantly positive with weight, body mass index, arm circumference and triceps skin

fold thickness ($P < 0.01$) and correlated positive with height ($P < 0.05$). Carbohydrates intake, it correlated significantly positive with weight, arm circumference, body mass index and triceps skin fold thickness ($P < 0.01$). In addition to calcium intake, it correlated significantly positive with height, weight ($P < 0.01$) and correlation with arm circumference ($P < 0.05$). Also phosphorus intake, it correlated significantly positive with weight, body mass index and arm circumference ($P < 0.01$). While vitamin "D" intake, it correlated significantly positive with body mass index ($P < 0.01$). Also vitamin "C" intake, it correlated with arm muscle circumference ($P < 0.05$) and negatively with triceps skin fold thickness ($P < 0.01$). Vitamin "B1" and weight, arm circumference, body mass index and triceps skin fold thickness ($P < 0.01$). Vitamin "B2" intake, with

weight, body mass index ($P < 0.05$) and arm circumference and triceps skin fold thickness ($P < 0.01$). Vitamin "B6" and weight, body mass index and arm circumference ($P < 0.05$) and arm muscle circumference ($P < 0.01$). Folate intake, with weight ($P < 0.05$) and body mass index, arm circumference and triceps skin fold thickness ($P < 0.01$) and negatively with height, ($P < 0.05$). While vitamin "B12" intake, with weight, body mass index and triceps skin fold thickness ($P < 0.01$) and correlation negative with height ($P < 0.01$), arm muscle circumference ($P < 0.05$). Finally niacin intake with weight and body mass index, arm circumference ($P < 0.01$) and triceps skin fold thickness ($P < 0.05$).

Table (6) explains the mean and SD for the whole sample with respects to laboratory investigation

comparisons between girls and boys groups. Although the mean value of hemoglobin level for girls group was lower than boys groups, ($P < 0.05$). Thalassemia is an inherited disorder of autosomal recessive gene disorder caused by impaired synthesis of one or more globin chains. The impairment alters production of hemoglobin (Hb). So, the clinical manifestations are diverse, ranging from absence of symptoms to profound fatal anemia in utero, or, if untreated, in early childhood (**Giardina and Forget, 2008**)

Table (7) shows the correlation coefficient between laboratory investigation and social variables. A positive correlation was found between all blood tested parameters (hemoglobin, hematocrit, ferritin and erythrocytes levels) and father's income, other income, total income ($P < 0.01$) and family member ($P < 0.05$). While, it correlated negatively with sleeping hours ($P < 0.05$).

Data of table (8) explained the correlation coefficients between laboratory investigation and nutrients intake. With respect to all blood tested parameters (hemoglobin, hematocrit, ferritin and erythrocytes levels) correlated significantly negatively with k. calories, total protein, Carbohydrates, phosphorus, vitamin "B1", Vitamin "B2", niacin ($P < 0.01$) and total fat, ($P < 0.05$).

Pyridoxine B6 is the name given to three related compounds found in many foods, particularly in meat, liver, fish and some vegetables. It functions in many enzyme systems, especially those concerned in protein synthesis.

There is no correlation was found between vitamin "C" and laboratory investigation, this mean ascorbic acid can't aid the absorption of iron from the intestine in case thalassemia patients.

Finally there are negative correlation coefficients observed between all blood tested parameters (hemoglobin, hematocrit, ferritin and erythrocytes levels) and some nutrient intake. This was also recorded for total iron. The reason for this is not explained by low iron intake, which is actually high in boys group and satisfied in girls groups. This means that reasons than low iron intake were responsible for low hemoglobin level in blood serum, such as losses with sweat, high-impact contact with higher activity during childhood and adolescence, sport anemia, drinking tea (tending to be of concentrated tea drink) right after feeding and other unknown factor such as possible internal parasites (even if healed) causing low iron absorption. This result is in comparable with the results obtained by **(Kail and Cavanaugh 2010)** and the

results obtained by **(Jamie 2008)**.

The result of table (9) show the correlation coefficients between laboratory investigation and anthropometric measurements all blood tested parameters (hemoglobin, hematocrit, ferritin and erythrocytes levels) correlated negative with weight, arm circumference and triceps skin fold thickness ($P < 0.01$).

CONCLUSION

Study concluded that Body mass index, it correlated significantly positive with arm circumference triceps and skin fold thickness. There was observed a positive correlation coefficient between macronutrients and micronutrients with weight, body mass index, arm circumference, and arm muscle circumference. There are negative correlation

coefficients observed between all blood tested parameters and some nutrient intake. The changes in physical, physiological and behavioral affected by the nutritional status.

SaudiMed J 2015; 36(5): 575e579.

Artimage GY and Berry WG (1987):

Statistical Methods 7th Ed. Ames, Iowa Stata University Press, 39-63.

References

AAPCN, American Academy of Pediatrics, Committee on Nutrition (2008):

Pediatric Nutrition Handbook, R.E. Kleinman, Eds., 6th ed. American Academy of Pediatrics. Elk Grove Village, IL.

Ali SS; Tarawah AM; Al-Hawsawi ZM, Zolaly MA and Turkustani W(2015):

Comprehensive patient care improves quality of life in transfusion dependent patients with b-thalassemia.

Bordbar M; Pasalar M; Safaei S, kamfiroozi R; Zareifar S; zekavat O and Haghpanah S (2018):

Complementary and alternative medicine use in thalassemia patients in Shiraz, southern Iran: A cross-sectional study, Journal of Traditional and Complementary Medicine 8, 141-146

Borgna-Pignatti C (2007):

Modern treatment of thalassemia intermedia. GJH 138:291-304.

Claster S; Wood JC and Noetzli, L (2007):

Nutritional deficiencies in iron overloaded patients with hemoglobinopathies. *Am. J. Hematol.*, 84: 344–348.

Drabkin D (1949):

The standardization of hemoglobin measurements. *Am. J. Med. Sci.*, 21(7): 710.

Food and Nutrition Board (2008):

A Report of the Panel on Macronutrients, Subcommittees on Upper Reference Levels of Nutrients and Interpretation and Uses of Dietary Reference Intakes, and the Standing Committee on the Scientific Evaluation of Dietary Reference Intakes. Dietary Reference Intakes for Energy, Carbohydrate, Fiber, Fat, Fatty Acids, Cholesterol, Protein, and Amino Acids (Macronutrients). THE NATIONAL ACADEMIES PRESS, Washington, D.C. ISBN0-309-08537-3.

Fung EB (2016):

The importance of nutrition for health in patients with transfusion-dependent thalassemia. *Annals of the New York Academy of Sciences*, 1368: 40-48.

Fung EB; Xu Y and Trachtenberg F (2012):

Inadequate dietary intake in patients with thalassemia. *J. Acad. Nutr. Diet.*, 112:980–990.

Giardina PJ and Forget, B.G. (2008):

Thalassemia syndromes, In: Hoffman, R.; Benz, E.J. and Shattil, S.S. *Hematology: Basic Principles and Practice*. 5th ed. Philadelphia, Pa: Elsevier Churchill Livingstone; chap 41.

Grunbaum BG (2004):

Thalassemia syndromes. In: Hoffman, R.; Benz, E.J. and Shattil, S.S. *Hematology: Basic Principles and Practice*. 5th ed. Philadelphia, Pa:

Elsevier Churchill Livingstone;
chap 41.

Jamie S (2008):

Nutrition in adolescence. In Mahan, L.K. and Escott-Stump, S Krause's Food, Nutrition, and Diet Therapy (12th ed). Philadelphia: W.B. Saunders Harcourt Brace.

Jelliffe DB (1966):

The assessment of the nutritional status of the community .Geneva Switzerland World Health Organization 1966.(World Health Organization Monograph Series No. 53) 271 p.

Kail and Cavanaugh (2010):

Human Development: A Life-Span View, 5th Edition Belmont, CA: Wadsworth Publishing.

Kaplowitz K; Karydis I, Karagiorga-Lagana M, Nounopoulos C, and Tolis G (2001):

Basal and stimulated levels of growth hormone, insulin-like growth factor-I (IGF-I), IGF-I binding and IGF-binding proteins in beta-thalassemia major. J Pediatr. Endocrinol. Metab, 17(1), p.p. 19-29.

Kesse-Adu R and Howard J (2013):

Inherited anaemias: sickle cell and thalassaemia. Medicine, 41, 4: 219-24.

NNI (National Nutrition Institute) (2006):

National Nutrition Institute Food Composition Table, Cairo,A.R.E., 2006.

Ozdem S; Kupesiz Aand Yesilipek A (2008):

Plasma homocysteine levels in patients with β -thalassemia major. Scand. J. Clin. Lab. Invest., 68: 134–139.

Rasanen L (1979):

Nutrition survey of Finnish rural children. VI Methodological study

comparing the 24-hour recall and the dietary history interview. *American Journal of Clinical Nutrition* 32: 2560-2567.6

Thoits P; Kangsadalampai O; Pongtanakul B and Kulwara M (2010):

Nutritional Status in Patients with Thalassemia Intermedia. *J. Hematol. Transfus. Med.*, 21: 167-76.

Thongkijpreecha P; Kangsadalampai O; Pongtanakul B and Kulwara M (2011):

Nutritional Status in Patients with Thalassemia Intermedia. *J. Hematol. Transfus. Med.*, 21: 167-76. *Transfusion and deferoxamine. Haematologica*, 89, p.p. 1187 - 1193.

Vogiatzi MG; Macklin EA and Trachtenberg FL (2009):

Differences in the prevalence of growth, endocrine and vitamin D abnormalities among the various thalassemia syndromes in North America.

Br. J. Haematol., 146: 546–556.

Walter PB; Fung EB and Killilea DW (2006):

Oxidative stress and inflammation in iron-overloaded patients with thalassaemia or sickle cell disease. *Br. J. Haematol.*, 135: 254–263.

Weatherall DJ and Clegg JB (2000):

The thalassemys In: Stamatoyannopoulos G-Perlmutter RM, Marjerus PW and Varmus H (eds) *Molecular Basis of Blood Diseases*, 3rd edn. Philadelphia: WB Saunders (in press)

Weatherall DJ (1987):

Fortnightly review: The thalassaemias *BMJ* 314 p. 1675.

WHO (2007):

Onis, M; Onyango, A.W; Borghi, E; Siyam, A; Nishda,

C and Siekmann. J (2007):
Development of a WHO
growth reference for school –
age children

and Managing, Report of a
WHO Con- sultation.

WHO (2010):

World Health Organization,
“Blood disease. Preventing

Table (1): The Intercorrelation between social variables for children with thalassemia

Variable	Age in years	Rooms number	Family members	Father's income	Mother's income	Other income	Total income	Pocket money amount	Sleeping hours
RsN	0.024								
FMs	0.006	0.312**							
F'sI	0.060	0.313**	0.112						
M'sI	0.193	-0.110	0.127	0.888**					
OI	0.045	0.302**	0.121	0.887**	0.725**				
TI	0.074	0.247**	0.073	0.879**	0.718**	0.944**			
PM	-0.020	0.216*	0.086	0.875**	0.716**	0.942**	0.726**		
SH	0.057	-0.035	0.094	-0.082	0.049	-0.005	-0.027	-0.19	
HWTV	0.146	-0.030	-0.092	-0.142	-0.081	-0.183*	-0.140	-0.187*	0.449**

* P < 0.05 ** P < 0.01

RsN=Rooms number FMs=Family members F'sI=Father's income

M'sI= Mother's income OI= other income TI= Total income

PMA= Pocket money amount SH= Sleeping hours

HWTV =Hours of watching TV

Table (2): The correlation coefficient between social variables and nutrients intake

Variable	AY	RN	FM	FI	MI	OI	TI	PM	SH	HWTV
Cal	0.18*	0.09	-0.14	0.27**	0.46**	0.36**	0.38**	0.36**	0.21*	0.25**
T. Protein	0.02	0.13	-0.08	0.34**	0.40**	0.43**	0.45**	0.42**	0.15	0.16
T. Fat	0.18*	0.11	-0.17	0.32**	0.39**	0.42**	0.44**	0.43**	0.06	0.12
Carb.	0.17	0.02	-0.11	0.14	0.38**	0.19*	0.22*	0.21*	0.3**	0.33**
Ca	0.04	0.17	-0.1	0.32**	0.49**	0.43**	0.44**	0.33**	0.06	-0.01
Ph.	0.091	0.18*	-0.02	0.24**	0.38**	0.32**	0.34**	0.33**	0.17	0.31**
T. Iron	0.07	0.07	0.05	0.10	0.19	0.11	0.14	0.06	0.27**	0.48**
Mg	0.08	0.03	0.11	-0.04	0.27*	-0.07	-0.04	-0.03	0.30**	0.47**
Vit. "A"	-0.01	0.06	0.00	0.16	0.33*	0.29**	0.26**	0.25**	0.12	0.14
Vit. "D"	0.24**	-0.09	0.07	0.00	-0.2	0.01	0.03	-0.04	0.16	0.09
Vit. "E"	0.10	-0.10	-0.12	-0.12	0.27	-0.15	-0.13	-0.19*	-0.03	0.30**
Vit. "C"	0.15	-0.07	-0.25**	-0.01	0.24	0.06	0.05	-0.04	-0.041	-0.03
Vit."B1"	0.05	0.06	0.05	0.14	0.45**	0.14	0.15	0.07	0.25**	0.37**
Vit."B2"	0.03	0.08	0.03	-0.004	0.21	0.003	0.01	0.07	0.26**	0.37**
Vit."B6"	0.27**	-0.05	-0.36**	0.15	0.24	0.19*	0.20*	0.29**	-0.15	0.24**
Vit."B12"	0.07	-0.16	-0.04	-0.12	0.06	-0.14	-0.12	-0.04	0.31**	0.29**
Folate	0.10	-0.06	0.06	-0.16	-0.01	0.27**	0.23**	-0.27**	0.36**	0.54**
Niacin	0.19*	0.10	-0.03	0.33**	0.51**	0.41**	0.40**	0.38**	0.19*	0.17

* P < 0.05 ** P < 0.01

AY= Age in years RN= Rooms number FM= Family members FI= Father's income
 MI= Mother's income OI= other income TI= Total income PM= Pocket money
 SH= Sleeping hours HWTV= Hours W. TV

Table (3): The interrelation between anthropometric measurements

Variable	Weight	Height	BMI	Arm Circumference	Triceps Skin Fold Thickness
Height	0.49**				
Body Mass Index	0.81**	-0.13			
Arm Circumference	0.689**	0.197*	0.696**		
Triceps Skin Fold Thickness	0.553**	0.093	0.479**	0.283**	
Arm Muscle Circumference	0.124	0.154	0.051	0.557**	-0.612**

*P<0.05, p<0.01**

Table (4): The correlation coefficient between anthropometric measurements and social variables

Variables	Weight	Height	BMI	Arm Circumference	Triceps Skin Fold Thickness	Arm Muscle Circumference
AY	0.28**	-0.03	0.29**	0.15	-0.21*	0.26**
RN	0.02	0.13	-0.05	0.06	0.07	-0.03
FM	0.02	-0.01	0.01	-0.04	0.20*	-0.20*
FI	0.27**	0.09	0.21*	0.07	0.13	-0.06
MI	0.22	0.11	0.16	0.12	-0.01	0.06
OI	0.28**	0.21*	0.17	0.11	0.15	-0.06
PM	0.27**	0.21*	0.17*	0.19*	0.07	0.08
TI	0.28**	0.18	0.17*	0.1	0.4	-0.05
SH	0.4**	-0.03	0.44**	0.21*	0.56**	-0.33**
HW TV	0.3**	-0.24**	0.41**	0.39**	0.19*	0.12

* P < 0.05 ** P < 0.01

AY= Age in years RN= Rooms number FM= Family members FI= Father's income
 MI= Mother's income OI= other income TI= Total income PM= Pocket money
 SH= Sleeping hours HWTV= Hours W. TV

Table (5): The correlation coefficient between anthropometric measurements and nutrient intake

Variables	Weight	Height	BMI	Arm Circumference	Triceps Skin Fold Thickness	Arm Muscle Circumference
Calories	0.57**	0.09	0.55**	0.54***	0.16	0.2*
Total protein	0.48**	0.20*	0.4**	0.4**	0.35**	0.09
Total fat	0.39**	0.16	0.38**	0.38**	-0.003	0.22*
Carbohydrate	0.46**	-0.05	0.46**	0.48**	0.44**	0.18
Calcium	0.24**	0.24**	0.12	0.18*	0.07	0.12
Phosphorus	0.34**	0.11	0.31**	0.38**	0.14	0.16
Total iron	0.36**	-0.03	0.32**	0.35**	0.38**	-0.005
Magnesium	0.33**	0.01	0.34**	0.32**	0.15	0.17
Vitamin "A"	-0.17	-0.03	-0.17	0.02	0.06	0.01
Vitamin "D"	0.17	-0.14	0.27**	0.15	0.03	0.09
Vitamin "E"	-0.04	0.04	-0.05	0.09	-0.15	0.21*
Vitamin "C"	-0.1	0.09	-0.16	-0.04	-0.23**	0.17*
Vitamin "B1"	0.3**	0.01	0.30**	0.31**	0.31**	0.02
Vitamin "B2"	0.19*	0.07	0.19*	0.32**	0.32**	-0.04
Vitamin "B6"	0.19*	0.005	0.3*	0.21*	-0.18	0.32**
Vitamin "B12"	0.4**	-0.3**	0.44**	0.13	0.4**	-0.22*
Folate	0.20*	-0.2*	0.31**	0.28**	0.24**	-0.02
Niacin	0.34**	0.14	0.33**	0.33**	0.2*	0.09

*P<0.05**p<0.01***p<0.001

Table (6): The mean and standard (SD) deviations for the whole sample with respects to laboratory investigation comparisons between female and male groups

Variables	Girls(50)		Boys(50)		P
	Mean	SD	Mean	SD	
Hemoglobin	8.94	1.44	9.06	2.098	0.044*
Hematocrit	18	4.023	16.45	1.26	0.05
Erythrocytes	2.64	0.003	2.55	0.083	0.007
Ferritin	14.35	0.091	16.97	0.043	0.004

Table (7): The correlation coefficient between laboratory investigation and social variables

Variables	Hemoglobin	Hematocrit	Erythrocytes	Ferritin
Age in years	-0.036	-0.031	-0.034	-0.033
Rooms number	0.171	0.123	0.120	0.121
Family members	0.327*	0.325*	0.324*	0.319*
Father's Income	0.528**	0.521**	0.531**	0.530**
Mother's Income	0.287	0.281	0.251	0.241
Other income	0.452**	0.453**	0.422**	0.462**
Total income	0.396**	0.391**	0.381**	0.385**
Pocket money amount	0.003	0.001	0.002	0.001
Sleeping hours	-0.279*	-0.253*	-0.263*	-0.260*
Hours of watching TV	-0.093	-0.089	-0.079	-0.083

*P<0.05**p<0.01

Table (8): The correlation coefficients between laboratory investigation and nutrients intake

Variables	Hemoglobin	Hematocrit	Erythrocytes	Ferritin
Calories	-0.436**	-0.426**	-0.431**	-0.437**
Total protein	-0.443**	-0.465**	-0.455**	-0.446**
Total fat	-0.298*	-0.301*	-0.311*	-0.308*
Carbohydrate	-0.431**	-0.440**	-0.434**	-0.430**
Calcium	-0.088	-0.079	-0.029	-0.059
Phosphorus	-0.433**	-0.429**	-0.439**	-0.449**
Total iron	-0.05	-0.04	-0.06	-0.03
Magnesium	-0.187	-0.107	-0.177	-0.167
Vitamin "A"	0.017	0.097	0.087	0.047
Vitamin "D"	0.043	0.023	0.033	0.054
Vitamin "E"	-0.029	-0.053	-0.035	-0.042
Vitamin "C"	0.011	0.021	0.019	0.031
Vitamin "B1"	-.0428**	-.0426**	-0.427**	-0.425**
Vitamin "B2"	-0.470**	-0.474**	-0.475**	-0.473**
Vitamin "B6"	-0.290*	-0.291*	-0.289*	-0.293*
Vitamin "B12"	-0.122	-0.123	-0.147	-0.157
Folate	-0.22	-0.34	-0.12	-0.06
Niacin	-0.533**	-.534**	-0.530**	-0.531**

* P < 0.05 ** P < 0.01

Table (9): The correlation coefficients between laboratory investigation and anthropometric measurements

Variable	Hemoglobin	Hematocrit	Erythrocytes	Ferritin
Height	-0.365**	-0.345**	-0.367**	-0.369**
Body Mass Index	-0.093	-0.087	-0.043	-0.073
Arm Circumference	-0.255	-0.245	-0.266	-0.246
Triceps Skin Fold Thickness	-0.452**	-0.462**	-0.446**	-0.466**
Arm Muscle Circumference	-0.565**	-0.555**	-0.579**	-0.566**
Variable	-0.17	-0.10	-0.16	-0.12

* P < 0.05 ** P < 0.01

العلاقة بين الحالة التغذوية والاضطرابات الموروثة من الهيموجلوبين (الثلاسيميا) عند الأطفال المصريين

هبة سعيد عبد الحليم^١ و باسم علي زوين^٢ و عبد الحميد صلاح الهمشري^٣

١- قسم التغذية الاكلينيكيه – المعهد القومي للتغذية

٢- مستشفيات جامعه القاهرة

٣- قسم طب الأطفال – كلية الطب – جامعه بنها

الملخص العربي

الثلاسيميا عبارة عن مجموعة من الاضطرابات الوراثية للهيموجلوبين ، تم التعرف عليها لأول مره عن طريق الولايات المتحدة وإيطاليا في عام ١٩٢٥ . تم اختراع كلمة "الثلاسيميا" المشتقة من الجنور اليونانية لـ "البحر" و "الدم" ، تحت الاعتقاد الخاطئ بأن كانت هذه الاضطرابات تقتصر على منطقة البحر الأبيض المتوسط. إن الثلاسيميا هي أكثر الأمراض الوراثية شيوعاً لدى البشر ، وهي تمثل مشكلة متزايدة في مجال الصحة العامة في البلدان الاستوائية التي تحدث فيها بمعدل عالٍ. تم تصميم هذه الدراسة علي أن تكون بأثر رجعي. ويعتبر هذا التصميم وفقاً للأدبيات المتاحة مناسباً لأنه يسمح للباحث بتقييم المتغير التابع في الحاضر (الثلاسيميا) ثم يربط هذه العوامل التي تؤثر على العيانات والعوامل الاجتماعية والاقتصادية والمادية. الحجم الإجمالي للعينة كان ١٠٠ طفلاً (٥٠ ولد و ٥٠ بنت). تراوحت أعمارهم بين ٦-١٢ سنة. وأظهرت الدراسة أن التغيرات في الفيزيائية والفيزيولوجية والسلوكية تتأثر بالحالة التغذوية لهم. كما تأثرت تحاليل الدم والمقاييس الجسمية بالقيمة الغذائية لنظامهم الغذائي.

الكلمات المفتاحية: الثلاسيميا- الغذاء – الأطفال