PREVALENCE OF BETA THALASSEMIA IN A SAMPLE OF SCHOOL AGE CHILDREN IN SULAIMANI CITY

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ABSTRACT

Background

Beta thalassemia is a major health problem in Sulaimani. The disease is usually diagnosed early but puts a huge burden on the health sector due, as well as a high morbidity and mortality of the Patient. The carrier state is mostly under diagnosed.

Objectives

To determine the prevalence of Beta thalassemia trait in school aged children in Sulaimani city.

Materials and Methods

A prospective cross-sectional study, undertook in 2 months peroid in 2017 in Sulaimani city, in which a total of 300 students were randomly taken from five primary school. A complete blood count was done, the serum hemoglobin electrophoresis and iron studies were done for individuals with anemia (PCV < 36%) or microcytosis (MCV < 76 fL).

Results

Out of 300 subjects 43 (14.3%) had anemia (including 17 (5.7%) had iron deficiency anemia), 13 (4.3%) had Beta thalassemia (major and minor).

Conclusion

The high prevalence rate of Beta thalassemia carrier state necessitates strengthening the preventive program, and neonatal screening to control and prevent cases of thalassemia major.

Keywords: *Beta thalassemia, Anemia, Prevention, Sulaimani.*

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INTRODUCTION

Hemoglobinopathies are the most common single gene disorders worldwide with a considerable frequency in certain areas particularly Mediterranean and Middle Eastern countries, including Iraq. Hemoglobinopathies include structural variants (such as Hb S, Hb C, Hb E), and the thalassemias [1, 2].

As a consequence of globalization and decreases in neonatal and childhood mortality, the severe α and β thalassemias have become worldwide clinical problems [3-5].

The thalassemias are a heterogenous group of inherited red blood cell disorders causing a quantative decrease in globin chain production leading to ineffective erythropoiesis and hemolysis. Seven percent of the world's population are thalassemia carriers. The phenotype is variable, ranging from an asymptomatic state with no anemia, to severe microcytic anemia with transfusion dependence [6,7].

If alpha chains are affected alpha thalassemia results, if beta chains are affected the result is beta thalassemia. The clinical effects depend upon both the type and extent of gene mutations and upon whether inherited from one or both parents [7]. Clinically there are four alpha-thalassemia syndromes: silent carrier, alpha-thalassemia trait, HbH disease, and hydrops fetalis syndrome [8].

Beta-globin gene mutations causing beta-thalassemia involve both coding and noncoding sequences. Thalassemia major is homozygous or compound heterozygous for β° or β^{+} genes (beta-globin mutations allowing no [0] or some [+] global chain production), whereas thalassemia minor represents heterozygous disease [9].

Beta thalassemia major and intermedia are defined based on RBC transfusion requirements, those with major disease require more than eight transfusions per year (usually every 3 to 4 weeks), whereas those with intermedia require fewer than eight transfusions per year. The indications for transfusion therapy include severe anemia and prevention of subsequent complications, growth failure, and significant extramedullary hemopoiesis, which should be prevented for a variety of reasons [6].

Stem cell transplantation can cure transfusion dependence in thalassemia major, but its availability is limited by donor availability. Gene therapy using autologus human stem cells (HSCs) holds promise as corrective therapy in those without suitable donor [6].

Beta- thalassemia major is an important health problem in Sulaimani province, with more than 600 registered cases in a population over 1.5 million. The high carrier rate and the frequency of consanguineous marriages, necessitate strengthening the effective prevention program [10].

In a number of populations at risk (Greek, Turkish, Cypriots, Italians) have controlled the disease at the population level by programs based on carrier screening, genetic consulting and prenatal diagnosis [11].

Aim of study is to determine the prevalence of Beta thalassemia trait in school aged children in Sulaimani city.

SUBJECTS AND METHOD

This cross-sectional study was carried out from April 15th 2017 to June 15th 2017 at primary schools inside Sulaimani city. Permission to conduct the study in these schools was taken from Sulaimani General Directorate for Education. By a stratified random sampling taking in consideration age and sex, five schools were selected. Three hundred pupils (150 boys and 150 girls) from about 1900 pupils aged from 8- 11 years were included in this study.

Two weeks before starting, schools were visited and the consent letter was sent for the parents for taking permission of them to take 5 ml blood sample from each pupil. At the day of visit (5 ml) of blood was withdrawn by venipuncture by an experienced nurse in pediatric blood sampling from those pupils whom we had their consent paper signed by their parents (there was 15% non response), (1 mL) of the blood was put in EDT A(ethylene diamine tetra acetic acid) tube for a CBC(complete blood count) test, another (1 mL) EDTA sample was put aside for Hb electrophoresis, and (3 mL) as clot sample for serum ferritin , serum iron and iron binding capacity.

The child's name and age, the date of sampling, the identification number, and the name of the school were recorded in the notebook. And the code number of each pupil recorded on his or her own tube. In the laboratory, a complete blood count was performed on the uncoagulated blood by an Orpheee mythic 18 automated machine using CBC line kit.

After the results of the (CBC)s, all cases with anemia (Hb <12gm/dL) [12], and all cases with microcytosis (MCV <76fL) [12] were then sent for Hb High-performance liquid chromatography using (Bio-Rad D-10 Hemoglobin Analyzer) and the serum sample was sent for iron profile using (spectrophotometric method by fully automated Roche Cobas C311 chemistry analyzer) and transferrin saturation(TS) of the anemic pupils was calculated by dividing the concentration of serum iron (SI) by the total iron binding capacity (TIBC). Criteria for IDA was considered in anemic pupils with transferrin saturation <16% [12].

The statistical analysis was done manually, and the charts done using Microsoft Excel.

RESULTS

In this study the total number of participants was 300 pupils (150 boys and 150 girls) with their ages ranging from 8 to 11 years, the median age was 9.5 years.

The prevalence of Beta thalassemia minor among school age children was (3.6%), 11 from 300, while thalassemia major/intermedia accounted for (0.7%), 2 cases only from 300, (Figure 1).

And among the thalassemia cases, 7 (54%) had anemia (PCV <36%), 6 (46%) had normal PCV but low MCV (<76 fL), (Figure 2).

Anemia (PCV<36%) was found in 43 (14.3%) pupils, 17 of them (5.7%) had Iron deficiency anemia, 7 (2.3%) had beta thalassemia trait or beta thalassemia intermedia (Figure 3). The other 19 pupils didn't meet the diagnostic criteria for neither thalassemia nor iron deficiency. The prevalence of iron deficiency anemia as defined by transferrin saturation less than (16%), accounted for 39.5% of the anemic cases.

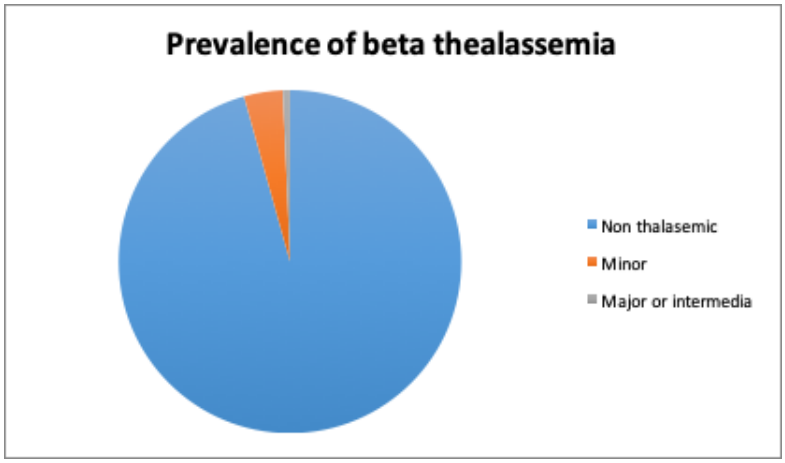

Figure 1. Shows the prevalence of Beta thalassemia

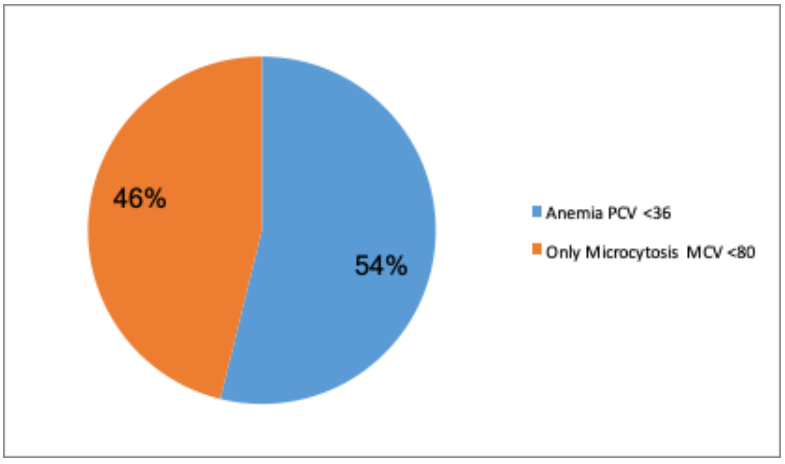

Figure 2. Shows the percentage of anemic cases with thalassemia, versus thalassemia cases who only had microcytosis with normal PCV.

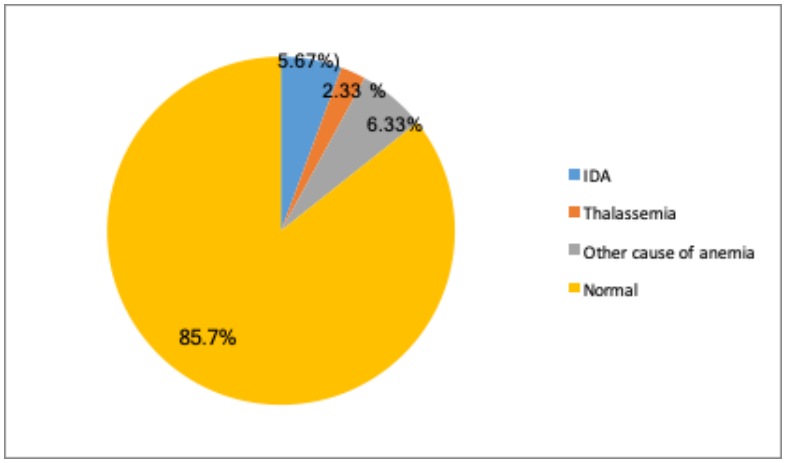

Figure 3. The percentage of anemic subjects in our studied group, and the cause of anemia. Note: As we mentioned before not all cases with beta thalassemia minor had anemia.

Table 1. Prevalence of anemia and microcytosis in the studied sample.

Variable	No	%
PCV		
>36	43	14.3
≥ 36	257	85.7
Total	300	100.0
MCV		
>76	66	22
≥80	234	78
Total	300	100.0

PCV<36%, MCV<76 are abnormal level in this age group

Table 2. Prevalence of anemia, Iron deficiency anemia, thalassemia minor, thalassemia major and other cases of anemia in the study.

Variable	No	%
Normal results	257	85.7
Anemia	43	14.3
Iron deficiency anemia	17	5.7
Thalassemia minor	11	3.7
Thalassemia major	2	0.7
Other anemias	19	6.3
Total	300	100

DISCUSSION

This is the first study done in Sulaimani city to estimate prevalence of Beta Thalassemia syndrome and carrier state among other causes of anemia in school age children. It differs from the study by Jalal S D et.al which was done on adults attending the premarital health centre at Sulaimani in 2007 and had a much bigger sample [10].

As because of the big number of carriers and the high frequency of consanguineous marriages which can range in between (25%-70%) [10-13], beta thalassemia is a big burden on the health department in Sulaimani in particular, and Iraq and the other countries in the neighbourhood.

Our study showed that beta thalassemia is a common health problem as we found the prevalence of it (3.7%) among school age children. Also anemia in general is common at (14.3%) of the participants.

Our study showed consistent results with other studies in Iraq, like 4.6% in Basra [14], 4.4% in Baghdad [15], 3.7% in Dohuk [16], and the previous study in Sulaimani which was done on married couples which showed 4.14% carrier rate [10] .

This was a lower rate than a study in Mosul 6.7% [17], and in Erbil 6.94% [18], but the Mosul study was conducted on patients attending 3 major hospitals in the city.

Also a similar results were found in the neighbouring Iran 4-8% [19], 3.1% in Jordan [20], 3% in Saudi Arabia [21], and lower numbers in Turkey 1.6% and 2% in two different studies in Ankara and Adiyaman respectively [22, 23].

Although some other Mediterranean countries had higher rates of carrier beta thalassemia, like 9% in Egypt [24], 7.77% in Libya [25] and 7.4% in Greece [26].

In conclusion the study documented high prevalence rates of Beta thalassemia in Sulaimani like most the other parts of Iraq and neighbouring countries. This necessitates strengthening the established preventive program, and perform neonatal screening, also by education of documented cases, counselling and prenatal diagnosis for their offspring.

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