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A STUDY OF B-THALASSEMIA IN PATIENTS REVIEWING THE THALASSEMIA CENTER IN AL-ZAHERA DAMASCUS, SYRIA

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ABSTRACT

Objective: This study was done to study B- thalassemia in patients reviewing the Thalassemia center in Damascus, Syria. **Materials and Methods:** This study was a retrospective study of the patients who reviewed the Thalassemia Center in Al-Zahera. This study included all cases from 20/2/2017 to 20/10/2017. **Results:** Most of the participants were males in 58 patients (58%). Most of the patients were younger than 5 years old with 47%. Splenectomy was done only in 9% of all patients, while 91% did not have it. Craniofacial features were present in 43 patients (43%). Multiple drug treatment was used in most of the cases 51%. **Conclusion:** This study highlights the need for large-scale epidemiologic research showing the prevalence and incidence of Thalassemia in Syria.

INTRODUCTION

Thalassemia is a well-known inherited hematologic disorder caused by a decrease or an absence of globin production.[1] Patients with thalassemia suffer from chronic hemolytic anemia and its sequelae. Thalassemia originates from varying genetic abnormalities that result in different clinical presentation. Non-transfusion dependent thalassemia (NTDT) or thalassemia intermedia (TI) is a milder form of thalassemia which does not require regular blood transfusion for survival. This group of thalassemia patients was recognized earlier as a TI but no consensus on diagnostic criteria has been reached due to high clinical variations ranging from asymptomatic to multi-organ involvement.[2-9] The terminology has been changed from TI to NTDT.[10] Generally patients with NTDT can maintain hemoglobin levels at 6–10 g/dl with occasional blood transfusions that may be required with fever, infection, or

pregnancy.[3,4,7,8,10] Complications of NTDT result from chronic hemolysis and tissue hypoxia, causing iron overload and problems in many organ systems.[5,6,8,11–20] According to the largest observational study on thalassemia intermedia (OPTIMAL CARE study; *n* = 584 TI patients), the three most common complications were osteoporosis, extramedullary hematopoiesis (EMH), and hypogonadism, respectively.[8]

Several complications that are associated with thalassemia intermedia are less frequently seen in thalassemia major, including EMH, leg ulcers, gallstones, and thrombophilia.[8] One of the most serious complications in NTDT is pulmonary hypertension which can be found in 11–50% of patients and leads to heart failure; the most common cause of death in NTDT patients.[3,4,6,8,11,13,14,16]

The proportion of patients classified by thalassemia type is changing due to advances in prenatal diagnoses and early detection. Higher numbers of NTDT patients are diagnosed and more fetuses with severe thalassemia are terminated.

Many previous studies aim to establish predictive factors for thalassemia complications and report that mechanisms for complications in thalassemia are multifactorial.[3,6,8,12,15,21–27]

In some countries, the prevalence of alpha-thalassemia is greater than that of beta-thalassemia which is different from the prevalence found in other regions.[25,26,28–30] The lack of studies and clear guidelines in this group can present a significant clinical challenge. This study aims to elucidate the prevalence of complications and identify predictive factors affecting complication of both alpha- and beta-NTDT patients.

MATERIALS AND METHODS

This study was a retrospective study of the patients who reviewed The Thalassemia in Al-Zahera center. This study included all cases from 20/2/2017 to 20/10/2017 who were diagnosed with B-thalassemia. All the data were collected only by the authors to ensure the privacy and all the names and personal information were blinded. Statistical analysis was done using SPSS 25.0.

RESULTS

Table 1: Variables of our study.

		N	%
Gender	Male	58	58
	Female	42	42
Age	<5	47	47
	6-10	23	23
	10-16	30	30
Treatment	One drug	49	49
	More than one	51	51
Splenectomy	Not Done	91	91.0
	Done	9	9.0
Craniofacial features	No	57	57.0
	Yes	43	43.0

Table 2: Ferritin levels in our study.

Ferritin levels before treatment			
		Frequency	**Percent**
Range ng/ml	1000-1500	1000-1500	24
	1500-2000	1500-2000	43
	>2000	>2000	33
Ferritin levels after treatment			
Range ng/ml	500-1000	13	13.0
	1000-1500	67	67.0
	1500-2000	20	20.0

Table 3: Hemoglobin levels in our study.

Hemoglobin levels before treatment			
		Frequency	**Percent**
Range g/dl	4-8	57	57.0
	8-10	30	30.0
	>10	13	13.0
Hemoglobin levels after treatment			
Range g/dl	4-8	54	54.0
	8-10	34	34.0
	>10	12	12.0

DISCUSSION

Most of the participants were males in 58 patients (58%) compared to 42 females (42%).

Regarding age of the participants, most of the patients were younger than 5 years old with 47%, while only 23% of the patients were between (6-10 years old). 30% were between 10-16 years old.

Ferritin values before treatment were between 1500-2000 ng/ml as the most common (43%), while values between 1000-1500 ng/ml were the least common (24%). 33% had ferritin levels higher than 2000 ng/ml.

Ferritin values after treatment were between 1000-1500 ng/ml as the most common (67%), while values between 500-1000 ng/ml were the least common (13%). 20% had ferritin levels between 1500-2000 ng/ml.

Hemoglobin values before treatment were between 4-8 g/dl as the most common (57%), while values more than 10 g/dl were the least common (13%). 30% had Hemoglobin levels between 8-10 g/dl. Hemoglobin values after treatment were between 4-8 g/dl as the most common (54%), while values more than 10 g/dl were the least common (12%). 34% had Hemoglobin levels between 8-10 g/dl. Regarding splenectomy, it was done only in 9% of all patients, while 91% did not have it. Craniofacial features were present in 43 patients (43%) compared to 57% with no changes. Multiple drug treatment was used in most of the cases 51%, while one drug treatment was used in 49% of all patients.

Compliance with Ethical Standards

Funding: This study was not funded by any institution.

Conflict of Interest: The authors of this study have no conflict of interests regarding the publication of this article.

Ethical approval: The names and personal details of the participants were blinded to ensure privacy.

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