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ASSESSING COMPLIANCE TO THERAPY IN A SAMPLE OF PATIENTS WITH THALASSEMIA IN AL-ZAHERA CENTER

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ABSTRACT

Objective: This study was done to study the compliance of patients with B- thalassemia to treatment in 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 non-compliant participants were younger than 5 years old, while in compliant patients, 46.2% of patients were younger than 5 years old, also 46.2% of the patients were between (6-10 years old). Most of the non-compliant participants were males, while in compliant patients females were more common. In non-compliant patients, Splenectomy was not done in most of the cases while it was not done at all in all compliant patients. Cranial features were found in 47.1% and 15.4% of non-compliant and compliant patients, respectively. **Conclusion:** This study highlights the need for large-scale epidemiologic research showing the prevalence and incidence of Thalassemia and the compliance to treatment 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. We studied the patients' compliance to treatment and considered all those who kept reviewing the center as compliant. 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: Age Distribution based on the Compliance to treatment in our study.

Age			
Compliance		**Frequency**	**Percent**
Non-Compliant	<5	41	47.1
	6-10	17	19.5
	10-16	29	33.3
	Total	87	100.0
Compliant	<5	6	46.2
	6-10	6	46.2
	10-16	1	7.7
	Total	13	100.0

Table 2: Gender Distribution based on the Compliance to treatment in our study.

Gender			
Compliance		**Frequency**	**Percent**
Non-Compliant	Male	53	60.9
	Female	34	39.1
	Total	87	100.0
Compliant	Male	5	38.5
	Female	8	61.5
	Total	13	100.0

Table 3: Ferritin levels based on the Compliance to treatment in our study.

Ferritin level before treatment			
Compliance	**Ferritin level**	**Frequency**	**Percent**
Non-Compliant	1000-1500	20	23
	1500-2000	42	48.3
	>2000	25	28.7
	Total	87	100
Compliant	1000-1500	4	30.8
	1500-2000	1	7.7
	>2000	8	61.5
	Total	13	100
Ferritin level after treatment			
Compliance	**Ferritin level**	**Frequency**	**Percent**
Non-Compliant	500-1000	4	6
	1000-1500	63	71
	1500-2000	20	23
	Total	87	100
Compliant	500-1000	9	70
	1000-1500	3	23
	1500-2000	1	7
	Total	13	100

Table 4: Hemoglobin levels based on the Compliance to treatment in our study.

Hemoglobin levels before treatment			
Compliance	**Hemoglobin levels**	**Frequency**	**Percent**
Non-Compliant	4-8	48	55.2
	8-10	26	29.9
	>10	13	14.9
	Total	87	100.0
Compliant	4-8	9	69.2
	8-10	4	30.8
	Total	13	100.0
Hemoglobin levels after treatment			
Compliance	**Hemoglobin levels**	**Frequency**	**Percent**
Non-Compliant	4-8	51	58.6
	8-10	34	39.1
	>10	2	2.3
	Total	87	100.0
Compliant	4-8	3	23.1
	>10	10	76.9
	Total	13	100.0

Table 5: Splenectomy rates based on the Compliance to treatment in our study.

Splenectomy rates			
Compliance	**Splenectomy**	**Frequency**	**Percent**
Non-Compliant	Not done	78	89.7
	Done	9	10.3
	Total	87	100.0
Compliant	Not done	13	100.0

Table 6: Cranial features rates based on the Compliance to treatment in our study.

Cranial features			
Compliance		**Frequency**	**Percent**
Non-Compliant	No	46	52.9
	Yes	41	47.1
	Total	87	100.0
Compliant	No	11	84.6
	Yes	2	15.4
	Total	13	100.0

DISCUSSION

Regarding the age of the participants, most of the non-compliant participants were younger than 5 years old with 47.2%, while only 19.5% of the patients were between (6-10 years old). 33.3 % were between 10-16 years old. While in compliant patients, 46.2% of patients were younger than 5 years old, also 46.2% of the patients were between (6-10 years old). Only one patient (7.6% of compliant patients) was between 10-16 years old. (Table 1).

Most of the non-compliant participants were males in 53 patients (60.9% of non-compliant patients) compared to 34 females (39.1% of non-compliant patients). While in compliant patients females were more common in 61.9% (of compliant patients) compared to males in 38.5% (of compliant patients). (Table 2).

Ferritin values before treatment in non-compliant patients were between 1500-2000 ng/ml as the most common (48.3%), while values between 1000-1500 ng/ml were the least common (23%). 28.7% had ferritin levels higher than 2000 ng/ml. While, in compliant patients 61.5% had ferritin levels higher than 2000 ng/ml, 30.8% were between 1000-1500 ng/ml and one patients (7.7%) was between 1500-2000 ng/ml. (Table 3).

Ferritin values after treatment in non-compliant patients were between 1000-1500 ng/ml as the most common (71%), while values between 500-1000 ng/ml were the least common (6%). 23% had ferritin levels between 1500-2000 ng/ml. While, in compliant patients 70% had ferritin levels between 500-1000 ng/ml, 23% were between 1000-1500 ng/ml and one patients (7%) was between 1500-2000 ng/ml. (Table 3).

In non-compliant patients, Hemoglobin values before treatment were between 4-8 g/dl as the most common (55.2%), while values more than 10 g/dl were the least common (14.9%). 29.9% had Hemoglobin levels between 8-10 g/dl. While after treatment, Hemoglobin values were between 4-8 g/dl as the most common (58.6%), while values more than 10 g/dl were the least common (2.3%). 39.1% had Hemoglobin levels between 8-10 g/dl. In compliant patients, Hemoglobin values before treatment were between 4-8 g/dl as the most common (69.2%) and 30.8% had Hemoglobin levels between 8-10 g/dl. While after treatment, Hemoglobin values were more than 10 g/dl as the most common (76.9%), while values between 4-8 g/dl were 23.1%. (Table 4).

In non-compliant patients, splenectomy was not done in most of the cases 89.7%, while it was not done in all compliant patients. Splenectomy was only done in 10.3% of non-compliant patients. (Table 5).

Cranial features were found in 47.1% and 15.4% of non-compliant and compliant patients, respectively. (Table 6).

Compliance with Ethical Standards

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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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