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Factors Affecting Compliance for Iron Chelating Agents in Thalassemic Patients in Basrah Province, Iraq

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Abstract
Objectives: To assess factors that affect patient's compliance for various type of chelating agents for thalassemic patients using direct questionnaires throughout 5 months period.
Methods: A prospective screening study done on patients with diagnosed transfusion-dependent β-thalassemia treated on different types of iron chelating agents. Thalassemia unit in AL-Zahraa teaching hospital. Evaluation of patient's preferability for chelation therapy, predictive values and reliability.
Results: In the current study, we found that 88 patients (60%) had accept the treatment in different degrees while the remaining number of patients (57 patients, 39.3%) had not accept their treatment.
Conclusion: In β-thalassemia compliance, monitoring using several methods is crucial as non-compliance is common and is associated with decreased survival. In addition, to have a good ideas about maintaining a reliable time for patient to continue on his chelating therapy.
Keywords: Compliance, iron-chelating agents, thalassemic patients, AL-Zahraa teaching hospital

Introduction

Beta-thalassemia is one of the commonest genetic disorders and consists of a reduction in the synthesis of the β globin chain of hemoglobin.[1] Although the disease spread worldwide, it is more prevalent among certain populations. Such as Italian, Greek, Indian and Chinese.[1,2] The incidence of thalassemia in its risk groups is 10–20/1000 births. Hemoglobin A (adult hemoglobin, HbA) is a tetrameric molecule which contains 2 α chains and 2 β chains. Hemoglobin F (fetal hemoglobin, HbF) consists of 2 α chains and 2 γ chains and is the predominant Hb during prenatal life and shortly after birth. However, immediately after birth, HbF starts to decrease gradually to replace by adult hemoglobin. β thalassemia major becomes symptomatic from 3 to 12 months of age, as HbA fails to replace the declining levels of HbF.[2] As the production of chains is deficient, the bone marrow tries to overcome the defect by synthesizing an excess of α chains, which associate and form insoluble aggregates within newly formed red cells.[2] This is injurious to the immature red cell and ultimately results in cell lysis. As the bone marrow fails to correct the resulting peripheral anemia, extra medullary organs such as the liver,[3,4] spleen and lymph nodes become involved in erythropoiesis, leading to enlargement of the organs. At least 91 mutations and several deletional mutations have been identified within or around the β-globin chain gene located on chromosome 11, all affecting the expression of the β-globin chain gene.[2] Different levels of expression are associated with different clinical pictures of the disease. B-thalassemia major defined as β-thalassemia that requires regular transfusions to sustain life. In 1925, Thomas Cooley, a Detroit pediatrician, described similarities in the appearance and clinical course of the disease in four children of Greek and Italian immigrants.[5] The advanced clinical picture that he described, consisting of severe anemia, hepatosplenomegaly, growth retardation and bone deformities, no longer seen at present in North America due to regular transfusion programs.[2] The aim of regular transfusions is to maintain the hemoglobin at a level that prevents hypoxia and ineffective erythropoiesis.[6,7] Transfusions prevent the consequences of chronic hypoxia, they help these patients to have a normal appearance and to prolong survival, and they have created a new problem iron overload.[8-10]

Methods

Patients with diagnosed transfusion-dependent β-thalassemia were approached regarding possible participation in a randomized prospective study of iron chelators Desferoxamine (DFO) and Exjade (Deferasirox). All patients were take one of these two drugs and will followed for a study period of five months. This study meant to evaluate the factors that affect the compliance for these drugs in β-thalassemia patients.

Patient Population

The number of patients in the current study was 145.

Outcome Measures

The primary end-point of chelation therapy is a reduction in the body iron burden. The standard test used worldwide for assessment of iron overload is serum ferritin.

Results

In the current study, 145 patients were studied (80 patients 55.2% were males, while 65 patients 44.8% were females), (Figure 1). The youngest patient was two years and six months (2.5 years) old, while the oldest patient was (16 years) old. The mean age for the patients was 9.7 years. In the current study, we found that 68% of patients live in rural areas, while the remaining 32% of patients were live in urban areas. In the current study, the number of patients were taking oral chelating therapy (Exjad) was 116 patients (80%), while the number of patients taking injectable chelating therapy (Desferoxamine) was 29 patients (20%), (Figure 2). In the current study, we found that 42 patients (29%) had very accepted to their treatment and they are on regular way of treatment, 34 patients (23.4%) had not accept their treatment and they are on irregular

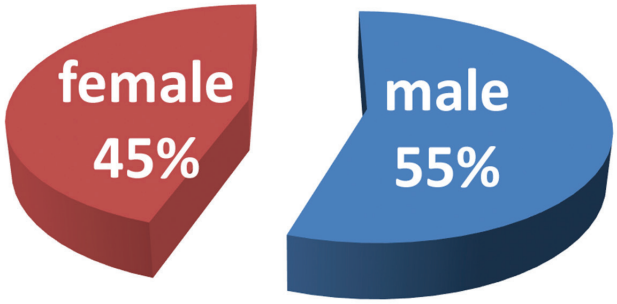

Fig. 1 **Gender of patients.**

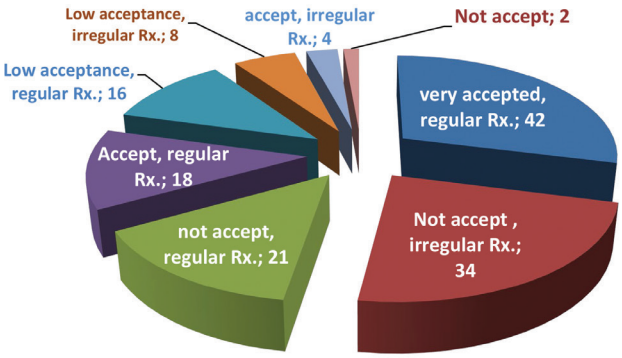

Fig. 3 **Compliance to therapy.**

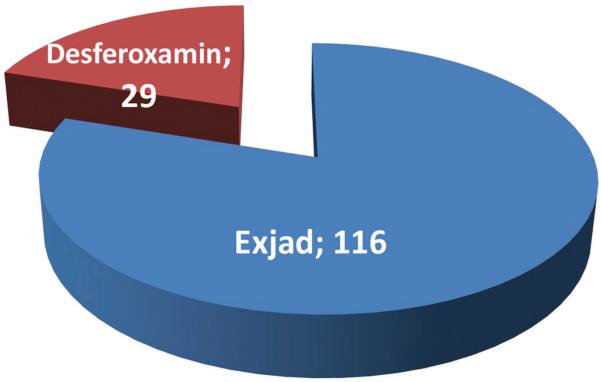

Fig. 2 **Type of chelating therapy taken.**

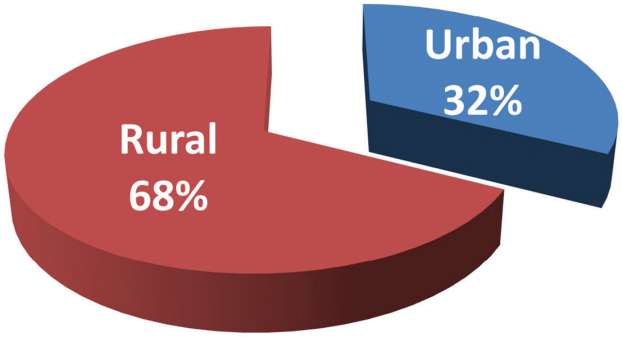

Fig. 4 **Residence of patients.**

way of treatment. 21 patients (14.4%) had not accepted their treatment and they were take their medication on regular way. 18 patients (12.4%) had accept their treatment and they take their medication regularly. 16 patients (11%) had low acceptance to their treatment and they were taking their medication regularly. 8 patients (5.5%) had low acceptance to their medication and they were taking their treatment on irregular way, 4 patients (2.7%) were accept their treatment and they take their medication on irregular way, while 2 patients (1.3%) of the total number of patients had not accepted the treatment, (Figure 3). In the current study, the majority of patients live in a rural areas (98 patients; 67.5%) while 47 patients (32.5%) live in an urban areas, (Figure 4). In the current study, we found that 88 patients (60%) had accept the treatment in different degrees while the remaining number of patients (57 patients, 39.3%) had not accept their treatment. The causes of not accepting their treatment are; nausea and vomiting in 23 patients (15.8%), abdominal pain in 15 patients (10.3%), Abnormal taste in 10 patients (6.8%). Interference with the time of their breakfast with their families in 10 patients (6.8%), Pain and swelling at the site of injection in 7 patients (4.8%). Better reduction in the serum ferritin so they leave Exjad and shift to Desferoxamin in 7 patients (4.8%). Neglection to take their medication in 6 patients (4.1%), Allergic reaction to Exjad occur in 4 patients (2.7%). Interference with the child activity in 2 patients (1.3%), Cyanosis occurred in 2 patients (1.3%). Finally, the child was refractory to his treatment in 2 patients (1.3%).

Discussion

In the current study, the males represent 55% of patients, while the females represent 45%. These was near the results of study done by Shimmaa Mostafa and Maha Abd Elaziz,[8] where the males represent 66% while the females were 34%. And compared with the study done by Way Seah Lee et al.[9] in which the males were 52.5% and females were 47.5%. The mean age for the patients in the current studay was 9.7 years that is near the result of Way Seah Lee et al.[9] which is 9 years. In the current study we found that the residence of 67.5% of the patients was in the rural areas, while the remaining 32.5% live in an urban areas, this was compared with the results obtained from Shimmaa Mostafa and Maha Abd Elaziz[8] which found that 40% of patients live in an urban areas while 60% of the patients were living in a rural areas. In the current study, the compliance was 60% that was near the compliance in the study made by Dr. Elena Pope which was 60.1 ± 11. However, it was lower than the results of study done by Way Seah Lee et al.[9] which was 81%.

Conclusion

In β-thalassemia compliance, monitoring using several methods is crucial as non-compliance is common and is associated with decreased survival. In addition, to have a good ideas about maintaining a reliable time for patient to continue on his chelating therapy.

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