

Study Tanner staging of β -thalassemic patients attending thalassemic Center in Ibn Al-Atheer Teaching Pediatric Hospital

By

Dr.Kahtan Adnan Abdullah^{*1}; Dr. Mohammed Ahmed Jassim Alogaidi^{*2}; Dr.Raed Jabbar Hussain^{*3}

^{*1} Senior pediatrician F.I.B.M.S,JAR pediatric teaching hospital Suliamania; ^{*2 & *3} FIBMS Pediatrics, Al-Elwiya pediatric teaching hospital, Baghdad- Alrusafa/Health Directorate, Ministry of Health and Environment, Baghdad, Iraq;

Abstract:

Purpose: determine any relationship between tanner staging of the patients and transfusion program, iron overload and chelation therapy and study tanner staging of β -thalassemic patients attending Thalassemic Center in Ibn Al-Atheer Teaching Pediatric Hospital. **Patients and Methods:** A descriptive-analytic study (case series study) was done on β -thalassemic patients attending Thalassemia Center in Ibn Al-Atheer Teaching Hospital in Mosul, during the period from the 1st of January to the 30th of June 2019. Sixty patients with β -thalassemia, 45 of them are β -thalassemia major cases and 15 are β -thalassemia intermedia cases. The patients of the major type were sub-classified into 3 groups according to their ages: Group 1, Patients from the age of ≥ 13 years; Group 2, Patients from the age of 14 years to 16 years and Group 3, Patients from the age of more than 17 years. **Results:** current study showed male (77.78, 66.67) % more than female (22.22; 33.33) % in both types of thalassemia (Major and Intermedia) respectively. so the most of the patients in this study live in urban areas (58.33%) and (41.67%) in rural areas, mean age at diagnosis of thalassemia major was 7.16 months, Delayed tanner staging was found in 64.44% of patients with thalassemia major, while in thalassemia intermedia only 33.33% were considered to be on a delayed stage. as well as 4(80%) of Tanner Stage (II) for patients in age group (≥ 13) years compare to 20% for stage (III), so 50% for both stage (II and III) respectively in age group (14-16) years. **Conclusion:** Two-third of the patients with thalassemia major had delayed puberty (64.44%), while one-third of the patients with thalassemia intermedia had delayed pubertal development (33.33%).

keywords: Tanner staging; age group; β -thalassemic patients; Ibn Al-Atheer Teaching Pediatric Hospital

Introduction:

Thalassemia is a congenital blood disease that informs of a defect in the production of a globin chain, result in absence production of β -globin chain completely (major-thalassemia), or a partial reduction (intermediate thalassemia) ⁽¹⁾, Thalassemia syndromes are the most common genetic disorder on a world, wide bases, which is inherited on autosomal recessive bases, characterized by reduced rate of production of one or more of the globin chains of hemoglobin. β -Thalassemia was first described by Cooley in 1925⁽²⁾.

Thalassemia is prevalent in Mediterranean countries, the Middle East, central Asia, India, Southern China and the Far East as well as countries along the north coast of Africa and in South America. The highest carrier frequency is reported in Cyprus (14%), Sardinia (10.3%) and South East Asia ⁽³⁾.

Thalassemia has been classified into three main types, which include beta-thalassemia minor, beta-thalassemia intermedia, and beta-thalassemia major (homozygous condition). Based on severity, the thalassemia intermedia and thalassemia major (TM) are further classified into transfusion-dependent thalassemia (TDT) and non-transfusion-dependent thalassemia (NTDT) respectively. The spectrum of severity ranges from mild anemia to moderate and severe anemia. Its clinical features include severe hemolytic anemia, bone abnormalities, and

hepatosplenomegaly (HSM) ^(4,5). **Aim of current study:** To determine any relationship between tanner staging of the patients and transfusion program, iron overload and chelation therapy and study tanner staging of β -thalassemic patients attending Thalassemic Center in Ibn Al-Atheer Teaching Pediatric Hospital.

Patients and Methods:

Anemia is usually severe with low hemoglobin to be less than 5.5 g/dl, unless blood transfusions are given with hypochromic microcytic anemia, many bizarre, fragmented RBC, poikilocytosis and target cells are present. Hemoglobin electrophoresis in β -Thalassemia major (β^0/β^0) show: HbF > 90%, HbA₂ is increased (normal 2 – 3 %), HbA is absent.

A descriptive-analytic study (case series study) was done on β -thalassemic patients attending Thalassemia Center in Ibn Al-Atheer Teaching Hospital in Mosul, during the period from the 1st of January to the 30th of June 2019.

Sixty patients with β -thalassemia, 45 of them are β -thalassemia major cases and 15 are β -thalassemia intermedia cases. The patients of the major type were sub-classified into 3 groups according to their ages: Group 1, Patients from the age of ≥ 13 years; Group 2, Patients from the age of 14 years to 16 years and Group 3, Patients from the age of more than 17 years.

The following information were collected from the patients:

Name, age, gender, residence, age of diagnosis of thalassemia, type of thalassemia, number of blood transfusions per year, age of starting desferal therapy, number of days per week patients receiving desferal, complications of thalassemia, and family history of delayed puberty.

In the physical examination the patients were assessed for their tanner staging according to the following criteria: For male: Pubic hair, penis, testes, axillary hair, and facial hair. For female: Pubic hair, breasts,

axillary hair, menarche. In addition, the weight and height of the patients were recorded.

The laboratory investigations done for the patients include: Serum iron; Total Iron Binding Capacity (TIBC); Transferrin Saturation, The bone age of all patients was estimated by a radiologist through X-ray. Results were analyzed using t- test to find the *P* value.

Results and discussion:

Table (1): Distribution of thalassemia patients according to Gender

Types of thalassemia	Gender			
	Male		Female	
	No.	%	No.	%
Major	35	77.78	10	22.22
Intermedia	10	66.67	5	33.33
Total	45	75	15	25

Gender distribution of the patients with thalassemia is shown in table (1), which appearance male (77.78, 66.67) % more than female (22.22; 33.33) % in both types of thalassemia (Major and Intermedia) respectively.

Table (2): Correlated between types of thalassemia and residence.

Type of Thalassemia	Residence			
	Urban areas		Rural areas	
	No.	%	No.	%
Major	24	53.33	21	46.67
Intermedia	11	73.33	4	26.67
Total	35	58.33	25	41.67

Table -2 showed most of the patients in this study live in urban areas (58.33%) and (41.67%) in rural areas, while another study performed by Al-Haji Ahmed (1992) shows that 55.2% of the patients were from rural areas and 44.8% from urban areas. This difference may be due to better compliance of urban families ⁽⁶⁾.

Table (3): Correlated between types of thalassemia and Age of Diagnosis.

Type of Thalassemia	Age of Diagnosis	
	Mean	Range
Thalassemia Major	7.16 (months)	3 – 18 (months)
Thalassemia Intermedia	3.87 (years)	1 - 13 (years)

The mean age at diagnosis of thalassemia major was 7.16 months, which was much lower than the age at diagnosis in Saudi Arabia (18 ± 0.5 months). It appears that earlier presentation of the disease may reflect the severity of the disease in our country. (Table -3).

Table (4): correlation ship between tanner staging of the patients with thalassemia major and age groups.

Age Group		Tanner Stage				
		I	II	III	IV	V
≥ 13	No.	12	2	0	0	0
	%	(85.71%)	(14.29%)	0	0	0
14 – 16	No.	3	8	1	0	0
	%	(25.00%)	(66.67%)	(8.33%)	0	0
17 \leq	No.	7	5	4	2	1
	%	(36.84%)	(26.32)	(21.05)	(10.53%)	(5.26%)
Total		22	15	5	2	1

Delayed tanner staging was found in 64.44% of patients with thalassemia major, while in thalassemia intermedia only 33.33% were considered to be on a delayed stage. Also, Yesilipek M.A. in Turkey in 1993 found that 74.50% of his patients had delayed puberty⁽⁷⁾.

Table (5): correlation ship between tanner staging of the patients with thalassemia intermedia with age groups.

Age Group (years)		Tanner Stage				
		I	II	III	IV	V
≥ 13	No.	0	4	1	0	0
	%	0	(80%)	(20%)	0	0
14 – 16	No.	0	1	1	0	0
	%	0	(50%)	(50%)	0	0
17 ≤	No.	0	0	0	4	4
	%	0	0	0	(50%)	(50%)
Total		0	5	2	4	4

Results in table -5 showed 4(80%) of Tanner Stage (II) for patients in age group (≥ 13) years compare to 20% for stage (III), so 50% for both stage (II and III) respectively in age group (14-16) years. The results were compared with those obtained in 12 normal subjects of whom six were prepubertal (mean (SD) age 11 (0-6) years; bone age 10-1 (0 8) years), and six were in Tanner's stage 3-4 (mean) age 13-8 (0-8) years; bone age 13-6 (0-4) years⁽⁸⁾.

Conclusion: Two-third of the patients with thalassemia major had delayed puberty (64.44%), while one- third of the patients with thalassemia intermedia had delayed pubertal development (33.33%).

References:

1) Michael, R . ; Debaun,E. ; Vichinsky, E R Behram, R M Kliegman and H B Jensen, Nelson textbook of pediatrics. 2350-2353, 2016.

2) William W. Hay, Anthony R. Hayward, Judith M. Sondheimer: *Current Pediatric Diagnosis and Treatment*, 14^h Ed. 1997; 737 – 38.

3) TAHER A.T., CAPPELLINI M.D. and MUSALLAM K.M.: Recent advances and treatment challenges in patients with non-transfusion-dependent thalassemia. *Blood reviews*, 26: S1-2, 2012.

4) Needs T, Gonzalez-Mosquera LF, Lynch DT. StatPearls [Internet]. StatPearls Publishing; Treasure Island (FL): May 15, 2021. Beta Thalassemia. [[PubMed](#)]

5) Viprakasit V, Ekwattanakit S. Clinical Classification, Screening and Diagnosis for Thalassemia. *Hematol Oncol Clin North Am.* 2018 Apr;32(2):193-211. [[PubMed](#)].

6) Al-Haji Ahmad FF: *Hemoglobinopathies in Mosul*, M.Sc Thesis, University of Mosul, Iraq, 1992.

7) Yesilipek MA, Bircan I, Oygur N, Ertug H, Yegin O, Guven AG: *Growth and Genderual Maturation in Children with Thalassemia Major*. Akdeniz University Medical School, Antalya, Turkey. *Hematologica*, 1993; Jan-Feb; 78(1): 30 – 3.

8) DE SANCTIS.V. ; VULLO,C.; KATZ,M; WONKE, t B; TANAS; t B,; B BAGNI, t.Gonadal function in patients with thalassaemia major, *J Clin Pathol* 1988;41:133-137.