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### RESEARCH ARTICLE

#### EVALUATION OF NOCTURNAL ENURESIS IN THALASSEMIC PATIENTS IN HEREDITARY BLOOD DISEASE CENTER AT KARBALA TEACHING HOSPITAL.

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#### Abstract

**Background:** Enuresis in thalassemic patients is one of psychosocial problem that presented to us in hereditary blood disease center at karbala teaching hospital.

**Aim:** To determine the type of enuresis, gender predominancy and associated certain predisposing factors in thalassemia major, thalassemia intermedia and sickle thalassemic patients in hereditary blood disease center at karbala teaching hospital.

**Method:** Across sectional study involved sixty-one patients with thalassemia and sickle thalassemia from 396 patients registered in hereditary blood disease center at karbala teaching hospital at age of 5-18 yrs and had enuresis. Medical history, physical examination, urinalysis and renal ultrasound were evaluated.

**Results:** The percentage of primary enuresis in thalassemic patients were 83.6%. The percentage of male patients were 67.2%. 75.4% had family history of enuresis. Just 13.1% had urinary tract infection and 98.4% with normal renal ultrasound.

**Conclusion:** The percentage of primary to secondary enuresis in thalassemic patient were similar to that of normal populations. The percentage of the males and family history of enuresis in enuretic thalassemic patients were more than that of normal population. After meticulous search we cannot find research concerned with enuretic thalassemic patients in Iraq.

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#### Introduction:-

Nocturnal enuresis refers to the occurrence of involuntary voiding at night after 5 yr. [1]

By 5 years of age, 80-85% are continent at night [2]. It may be primary or secondary; The primary mean nocturnal urinary control never achieved, estimated 75-90% of children with enuresis). Secondary 10-25%. [3]. The child was dry at night for at least a few months and then enuresis developed. In addition, 75% of children with enuresis are wet only at night, and 25% are incontinent day and night.

About 60% of children with nocturnal enuresis are boys. Family history is positive in 50% of cases. Primary nocturnal enuresis may be polygenetic. If one parent was enuretic, each child has a 44% risk of enuresis; if both parents were enuretic, each child has a 77%. Its frequency among adults is <1% [4]. Hemoglobinopathies abnormalities in 2 pairs of globin chains (tetramer). Over 1000 different mutations of the globin chains of the human hemoglobin molecule have been discovered [5]. Thalassemia refers to a spectrum of diseases characterized by reduced or absent production of one or more globin chains [6]. The thalassemias are inherited disorders of hemoglobin (Hb) synthesis. Their clinical severity widely varies, ranging from asymptomatic forms to severe. [7].

Classification of thalassemia: In clinical practice, the most important types affect either  $\alpha$ - or  $\beta$ -chain synthesis.

common forms of  $\beta$ -thalassemia are as follows:

1. Silent carrier  $\beta$ -thalassemia: Patients are asymptomatic
2.  $\beta$ -Thalassemia trait: Patients have mild anemia
3. Thalassemia intermedia: Patients have anemia of intermediate severity
4.  $\beta$ -thalassemia associated with  $\beta$ -chain structural variants: The most significant condition in this group of thalassemic syndromes is the Hb E/ $\beta$  thalassemia
5. Thalassemia major (Cooley anemia): This condition is characterized by transfusion-dependent anemia common forms of  $\alpha$ -thalassemia are as follows:

1. Silent carrier  $\alpha$ -thalassemia
2. Thalassemia trait: Characterized by mild anemia
3. Hb H disease: Represents  $\alpha$ -thalassemia intermedia
4. Thalassemia major: Results in the severe form. [8]

After meticulous searching we cannot find a published article concern with nocturnal enuresis in patients with thalassemia in hereditary blood disease centers, so we try to focus on this subject.

#### Patients and methods:-

396 hemoglobinopathies patients with thalassemia and sickle thalassemia from 5-18 years registered at hereditary blood disease center in Karbala teaching hospital for children from September 2016 to April 2017. From which 260 patients diagnosed as thalassemia major on regular blood transfusion every 2-4 weeks, 57 patients diagnosed as thalassemia intermedia, 79 patients diagnosed with sickle thalassemia syndrome. Only 61 patients were conducted in our cross-sectional study. Other types of hemoglobinopathies with or without diabetes and other chronic illness like heart failure, renal failure, diabetes insipidus were excluded from the study.

Detail history from all patients and their parents were taken including onset of voiding control, day and/or night wet, history of chronic disease (diabetes mellitus, heart failure, renal failure), diuretic drug ingestion, family history of enuresis. After routine physical examination; specific investigation for enuretic patients including general urine examination, urine culture and abdominal ultrasound for genitourinary system.

By using SPSS 21 (Statistical Package for the Social Sciences 21) the association between different types of thalassemia and enuresis evaluated by P value (significant when  $P < 0.05$ ).

#### Result:-

Primary enuresis in 51 (83.6%) of 61 patients, 44 (83.0%) in thalassemia major. Secondary enuresis was 10 (16.4%) of 61 patients, 9 (17%) thalassemia major. As shown in table I:

**Table-I:-** Primary and secondary enuresis in thalassemic and sickle thalassemia:

			Thalassemia		Sickle thalassemia	Total
			thalassemia major	thalassemia intermediate		
Enuresis	Primary	Count	44	2	5	51
		% within Thalassemia	83.0%	66.7%	100.0%	83.6%
	Secondary	Count	9	1	0	10
		% within Thalassemia	17.0%	33.3%	0.0%	16.4%
Total		Count	53	3	5	61
		% within Thalassemia	100.0%	100.0%	100.0%	100.0%

P value 0.4

Males in thalassemia major, thalassemia intermediate and Sickel thalassemia have enuresis were 41 (67.2%) and females were 20 (32.8%) as shown in table II:

**Table II:-**Gender of enuretic patient with thalassemia and sickle thalassemia

				Thalassemia		Sickle thalassemia	Total
				thalassemia major	thalassemia intermediate		
gender	Male	Count		34	3	4	41
		% within Thalassemia		64.2%	100.0%	80.0%	67.2%
	Female	Count		19	0	1	20
		% within Thalassemia		35.8%	0.0%	20.0%	32.8%
Total		Count		53	3	5	61
	% within Thalassemia	100.0%	100.0%	100.0%		100.0%	

P value= 0.3

Family history of enuresis was positive in 46(75.4%) from 61 thalassemic and sickle thalassemia patients as in table III.

**Table-III:-**Family history of enuresisin thalassemia and sickle thalassemia:

			Thalassemia		Sickle thalassemia	Total
			thalassemia major	thalassemia intermediate		
Family history	Positive	Count	43	1	2	46
		% within Thalassemia	81.1%	33.3%	40.0%	75.4%
	Negative	Count	10	2	3	15
		% within Thalassemia	18.9%	66.7%	60.0%	24.6%
Total		Count	53	3	5	61
		% within Thalassemia	100.0%	100.0%	100.0%	100.0%

P value =0.03

Urinalysis for urinary tract infection in thalassemic and sickle thalassemia patient reveal no significant finding 53(86.9%) as shown in table IV:

**Table-IV:-**significant finding for Urinary tract infection in enuretic patients with thalassemic and sickle thalassemia:

			Thalassemia		Sickle thalassemia	Total
			thalassemia major	thalassemia intermediate		
GUE	UTI	Count	8	0	0	8
		% within Thalassemia	15.1%	0.0%	0.0%	13.1%
	NoUTI	Count	45	3	5	53
		% within Thalassemia	84.9%	100.0%	100.0%	86.9%
Total		Count	53	3	5	61
		% within Thalassemia	100.0%	100.0%	100.0%	100.0%

P value= 0.5

Renal ultrasound was normal in 60(98.4%) of thalassemic and sickle thalassemia patients, as shown in table V.

**Table V:-**Renal ultrasound findings of thalassemia and sickle thalassemia patients:

			Thalassemia		Sickle thalassemia	Total
			thalassemia major	thalassemia intermediate		
Renal ultrasound	Normal	Count	52	3	5	60
		% within Thalassemia	98.1%	100.0%	100.0%	98.4%
	Abnormal	Count	1	0	0	1
		% within Thalassemia	1.9%	0.0%	0.0%	1.6%
Total		Count	53	3	5	61
		% within Thalassemia	100.0%	100.0%	100.0%	100.0%

P value = 0.9

### Discussion:-

In the current study, the result of Primary enuresis and secondary enuresis in thalassemia major, thalassemia intermediate and Sickle thalassemia were parallel to a cross sectional study performed among children (5-15 years old) visited the general pediatric out patient in Al-Imamain Al-Kadhmain Medical City in the capital Baghdad. In this study the prevalence of nocturnal enuresis was 29.5% (n=184), 84.8% (n=156) were of primary type and 15.2% (n=28) had secondary nocturnal enuresis [9].

Also this result goes with Robert M. Kliegman, et al, 2016 [4].

The result of enuretic males and female patient in thalassemic and sickle thalassemia, was similar to that of Alaa A. Selah, Atheer J. Al-Saffar in Iraqi JMS 2015, in which Males were significantly more enuretic than females (60.3% versus 39.7% and P= 0.02) [9].

Also this result goes with Robert M. Kliegman, et al, 2016, which reveals that Approximately 60% of children with nocturnal enuresis are boys. [4].

In Jamaica (the Caribbean) adopted the prospective interview method recorded a prevalence rate of 45 % among 8 years old sickle cell anemia patients; also noting a male predominance. [10].

The positive Family history of enuresis of thalassemic and sickle thalassemia patients goes with Ekinci O et al. who Find that family history of nocturnal enuresis and family problems were found to be more frequent in patients with thalassemia major. [11]. And Alaa A. Selah, Atheer J. Al-Saffar in Iraqi JMS 2015, in which positive family history of nocturnal enuresis (including both parents side and siblings), were significantly higher among enuretic children compared to non enuretic children [9].

Our result differs from that of Robert M. Kliegman, et al, 2016 which states that the Family history is positive in 50% of cases in general population. [4].

Urinalysis to evaluate urinary tract infection in thalassemic and sickle thalassemia patient reveal no significant finding and this not goes with Alaa A. Selah, Atheer J. Al-Saffar in Iraqi JMS 2015, and Mabiala Babela JR., et al. were children with positive history of urinary tract infection were significant. [9], [11].

However, this may be due to non-exclusion of confounding variables like diabetes mellitus, epilepsy and urinary tract infection these studies.

Renal ultrasound finding of thalassemic and sickle thalassemic patients seen in one patient (1.6%) (P value = 0.9) While Kovacevic L., et al. Study shows 12.54% of enuretic children and 5.38% of controls have abnormal finding (p = 0.004), the majority of these findings were clinically insignificant. [13].

### Recommendation:-

1. Thalassemic patients need evaluation of their psychosocial problems.
2. The health care provider in hereditary disease center should be focusing on enuresis as an additional psychosocial burden on thalassemic patients and their family.
3. We need multidisciplinary team for management of enuresis in hereditary blood disease center.
4. We hope to carry out Further studies regarding the incidence and prevalence of enuresis in thalassemic patients.

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