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The Weight Assessment and its Relation to Hemophilia Joint Health Score and Quality of Life in Hemophilic Patients

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Abstract

Background:*Hemophilia is characterized by spontaneous and provoked mucocutaneous, joint, muscles, gastrointestinal, and central nervous system bleeding, leading to major morbidity and even mortality of safe plasma - derrived and recombinant coagulation factor, people with hemophilia are now living longer with fewer bleeding and infections complications. The evaluation of hemophilia care is possible by assessing physical, emo- tional, mental, social, and behavioral components of well-being and func- tion as perceived by the patients and observers .Health-related quality of life (HRQoL) is considered one of the most relevant health outcome measures in medicine. Our study aims to* assess *weight of hemophilic children and correlate the weight to Hemophilia Joint* htlaeH*ScoreandAlsoQuality-of-lifeinhemophilicpatients .****Methods:****case control study was conducted at Pediatric Department of Faculty of Medicine in Zagazig University between October 2019 and March 2020.This study included 66 patients already diagnosed as hemophilia. These patients were classified into two groups; Group I;33 hemophilic patients with arthropathy. GroupII;33 hemophilic patients without arthropathy.*

Both sex had been included in the study are subjected to the following: *Full history taking including personal history. Thorough clinical examination: Assessmentof anthropometric measurements Quality of life using Hamo - Qol questionnaire****Results:****The obesity (85-95precentile) was found more in hemophilia with arthropathy[33.3%] than in hemophilia without arthropathy [10.3%].Patients with hemophilia in our study had been found 100%[p=1] Unsatisified about quality of life in both groups with H.A.&without H.A.****Conclusion:*** *All hemophilic patients received treatment on-demand only, No prophylaxis treatment. Regarding to H.A. our study reported that severity of H.A. Correlate positively with (age (years), weight , BMI , waist (cm), Global Giat Score and with HJHS total Score). In contrast , it correlate negatively with factor plasma level . All hemophilic patients (with or without hemoarthropathy) suffer from unsatisified quality of life (100%).*

Key words: *Weight Hemophilia Joint Health Score- Quality of Life*

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I. Introduction:

Hemophilia is characterized by spontaneous and provoked mucocutaneous, joint, muscles, gastrointestinal, and central nervous system bleeding, leading to major morbidity and even mortality of safe plasma - derrived and recombinant coagulation factor, people with hemophilia are now living longer with fewer bleeding and infections complications[(1)].

Obesity is a major contributing factor to the global burden of chronic disease and disability . According to the who, obesity rates have increased at least threefold since1980 in parts of North America , the Pacific Islands, Australia and China. [(2)].

quality of life significantly improved in the last 35 years. Recombinant factor concentrates further increased the safety of the products, eliminating the risk of virus and prion contamination. Primary pro- phylaxis reduced joint bleeds and development of target joint and chronic arthropathy, but inevitable continuous medical support is a real economical burden for the health system.

The evaluation of hemophilia care is possible by assessing physical, emo- tional, mental, social, and behavioral components of well-being and func- tion as perceived by the patients and observers .Health-related quality of life (HRQoL) is considered one of the most relevant health outcome measures in medicine. Since 1990 more than 20 studies for assessment of HRQoL in hemophilia were published [(3, 4)].

Our study aims to assess weight of hemophilic children and correlate the weight to Hemophilia Joint htlaeHScoreandAlsoQuality-of-lifeinhemophilicpatients .

II. Patients and Methods

A. Technical design:

- **Study design:** case control study.
- **Sitting:** This study had been conducted at Pediatric Department of Faculty of Medicine in Zagazig University between October 2019 and March 2020.

Sample size: Using epi-info, power 80% and CI 95%, assuming that mean ± SD of feelings in health-related quality of life in group I was 16.64 ± 20.4 and 30.9 ± 20.3 in group II. So, the total sample had been 66 children.

Subjects:

This study included 66 patients already diagnosed as hemophilia.

These patients were classified into two groups;

- Group I;33 hemophilic patients with arthropathy.
- GroupII;33 hemophilic patients without arthropathy.
- Level of clotting factor VIII or IX less than 10% in both groups.

Inclusion Criteria:

- Known patients diagnosed hemphilia A or B.

- ❖ Age up to 18 years.
- ❖ Both sex had been included.

Exclusion Criteria:

- ❖ Patients older than 18 years.
- ❖ Patients with congenital anomalies.
- ❖ Patients with other bleeding disorders as defect in factors V, VII, I, II, or combined deficiency of factor V and VIII (F5F8D) patients with hemophilic arthropathy combined with other hematological diseases.
- ❖ Patient with other bone and joint diseases.

B. Operational design:

Both sex had been included in the study are subjected to the following:

1) Full history taking including personal history.

2) Thorough clinical examination:

- Assessment of vital signs and color.
- Assessment of any bone deformity, pain or muscle weakness.
- Hemophilia Joint Healt Score (HJHS).

3) Assessmentof anthropometric measurements on WHO growth chart (Weight/age, height/age and BMI/age).

4) Drugs in hemophilic patients.

5) Nutritional assessment.(by assess Weight/age, height/age and BMI/age,M.A.C.,Mid-thigh circumference,Waist circumference).

C. Quality of life using Hamo - Qol questionnaire.

Administrative design:

1) Approval will be obtained fromZagazig University Research Board (IRB Institutional).

2) An official permission was obtained from Hematology unit ,Zagazig University Hospitals.

3) An official permission was obtained from theInstitutional Research.

4) Verbal consent from patients and their parents.

Statistical Analysis

The data were coded, entered and processed on computer using SPSS (version 18).The results were represented in tabular and diagrammatic forms then interpreted.

Mean, standard deviation, range, frequency, and percentage were use as descriptive statistics.

The following test was done:

❖ **Chi-Square testX^2** was used to test the association variables for categorical data.

❖ **Student's t-test** was used to assess the statistical significance of the difference between two population means in a study involving independent samples.

❖ **Student's paired t-test** was used to assess the statistical significance of the difference between two population means in a study involving paired samples.

❖ **ANOVA (F test)** For normally quantitative variables, to compare between more than two groups, and Post Hoc test (LSD) for pairwise comparisons

❖ **r→Pearson'sProduct correlation coefficient:** it evaluates the linear association between 2 quantitative variables (one is the independent var.X, and the other is the dependent var., Y).value of "r" ranges from -1 to 1

0= no linear correlation

1= perfect positive correlation

-1 = perfect negative correlation

Positive= increase in the independent variable leads to increase in the dependent variable

Negative = increase in the independent variable leads to decrease in the dependent variable.

P value was considered significant as the following:

* P > 0.05: Non significant

* P ≤ 0.05: Significant

III. Results:

Table (1): Comparison between Hemophilia with arthropathy group and Hemophilia without arthropathy group regarding demographic data.

			Hemophilia with arthropathy group	Hemophilia without arthropathy group	t. test	P. value
Age (years)	**Mean ± SD**		10.68 ± 4.28	7.17 ± 4.12	3.376	0.001
Sex	**Female**	**No.**	0	1	X^2 1.015	0.314
		%	.0%	3.0%		
	Male	**No.**	33	32		
		%	100.0%	97.0%		

There was no statistically significant difference between Hemophilia with arthropathy group and Hemophilia without arthropathy group regarding Sex

There were statistically significant increase in Age (years) among Hemophilia with arthropathy group than Hemophilia without arthropathy group

Table (2): Comparison between Hemophilia with arthropathy group and Hemophilia without arthropathy group regarding BMI and BMI percentile.

			Hemophilia with arthropathy group	Hemophilia without arthropathy group	X^2	P. value
BMI	**Mean ± SD**		23.30 ± 3.03	19.29 ± 3.23	**t. test** 1.305	0.019
BMI percentile	**stunted (<5th percentile)**	**No.**	1	4	2.412	0.120
		%	3.0%	13.8%		
	severe under weight (5th -10 percentile)	**No.**	4	2	0.482	0.488
		%	12.1%	6.9%		
	under weight (10th - 25 percentile)	**No.**	2	2	0.018	0.894
		%	6.1%	6.9%		
	normal (25th -75 percentile)	**No.**	12	9	0.196	0.658
		%	36.4%	31.0%		
	overweight (75th -85 percentile)	**No.**	2	2	.018a	0.894
		%	6.1%	6.9%		
	obese (85th -95 percentile)	**No.**	11	3	4.666	0.031
		%	33.3%	10.3%		
	morbid obesity (> 95 percentile)	**No.**	1	6	4.806	0.028
		%	3.0%	20.7%		

There were statistically significant increase in BMI among Hemophilia with arthropathy group than Hemophilia without arthropathy group

There is statistically significant increase in obese among Hemophilia with arthropathy group than Hemophilia without arthropathy group

There is statistically significant increase in morbid obesity among Hemophilia without arthropathy group than Hemophilia with arthropathy group.

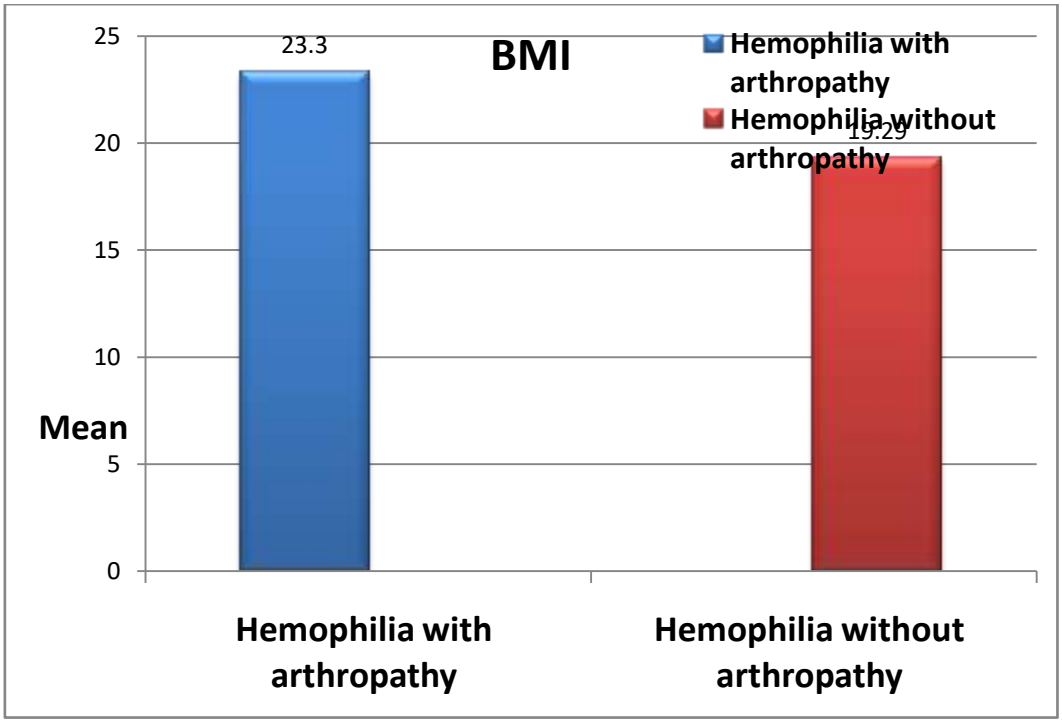

Figure (1): Comparison between Hemophilia with arthropathy group and Hemophilia without arthropathy group regarding BMI.

This figure shows that there is increase in mean\BMI in group of arthropathy than without arthropathy.

Table (3): Comparison between Hemophilia with arthropathy group and Hemophilia without arthropathy group regarding M .A .C (cm), Waist (cm) and hip (cm).

		Hemophilia with arthropathy group	Hemophilia without arthropathy group	t. test	P. value
M .A .C (cm)	**Mean ± SD**	23.92 ± 3.17	21.06± 3.38	3.545	0.001
Waist (cm)	**Mean ± SD**	64.74 ± 9.29	57.30 ± 11.11	2.950	0.004
hip (cm)	**Mean ± SD**	36.88 ± 5.79	32.12 ± 6.29	3.197	0.002

There were statistically significant increase in M .A .C (cm), Waist (cm) and hip (cm) among Hemophilia with arthropathy group than Hemophilia without arthropathy group

Table (4): Comparison between Hemophilia with arthropathy group and Hemophilia without arthropathy group regarding QOL.

			Hemophilia with arthropathy group	Hemophilia without arthropathy group	X^2	P. value
QOL	satasified	No.	0	0	0	1
		%	0%	0%		
	Un satasified	No.	33	33		
		%	100%	100%		

There was no statistically significant difference between Hemophilia with arthropathy group and Hemophilia without arthropathy group regarding QOL due to maternal depression.

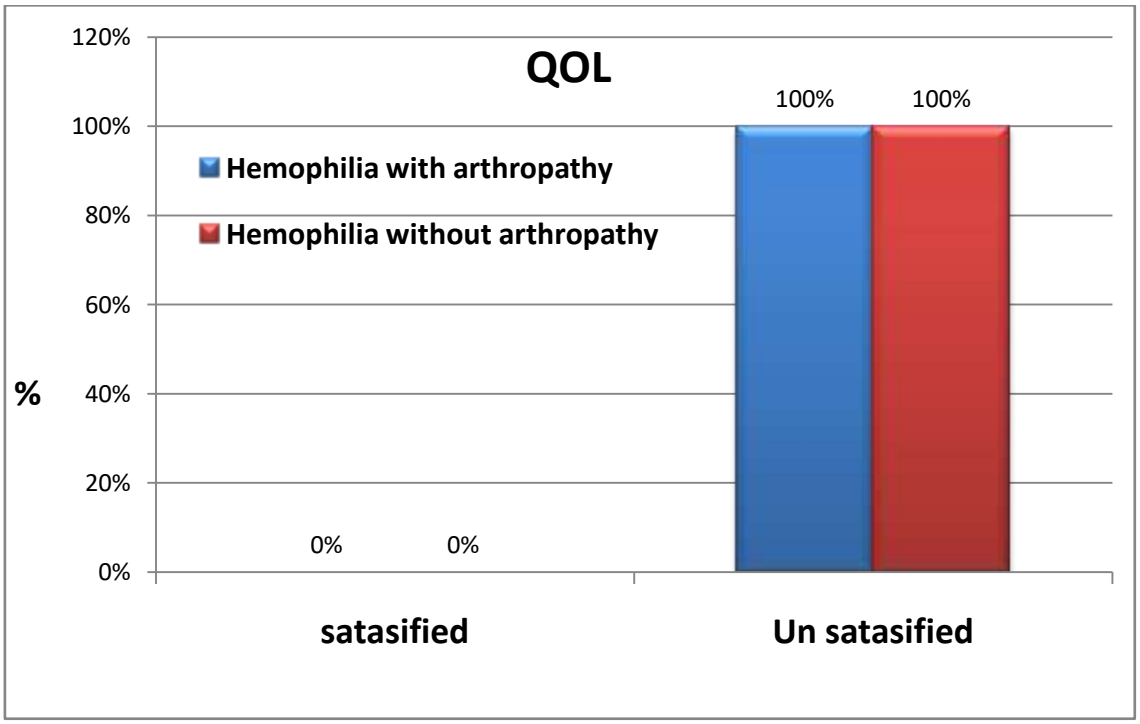

Figure (2): Comparison between Hemophilia with arthropathy group and Hemophilia without arthropathy group regarding QOL.

IV. Discussion

In our study ,We found that there was statistically significant increase in BMI among hemopilia with arthropathy group than hemophilia without arthropathy group.

This agreed with **Majumdar et al,**[5]that reported that there is positive correlation between BMI and risk of bleeding as well as resulting arthropathy and obesity accelerate development of hemophilic a .yhtaporhtra

In our study,We found that there was significant positive correlation between HJHS total score and ages yeaby**BLADEN et al.,**[6] who reported that older children had lower odds of having zero HJHS

And this also agreed by **Hilliard et al,**[7]several studies that reported apositive relationship between age and HJHS and Lithuanian study **saulyte et al.,**[8]observed higher joint score in older .nerdlihc

In our study, we found that there is significant increase in unsatasification of quality of life in hemophilic stneitapinbothgroupswitharthropathyandwithout arthropathygroups (by100%).

These results were nearly agreed with **Zhanget al.,**[9] who reported that hemophilia decrease htlaeh related ytilauqof efil.

Moreover,deergasihtby **Murtadha et al.,**[10]that confirmed that severity of hemophilia adversely affects ytilauq of life.

This also mentioned by **Miners et al.,**[11]**, and Tantawy et al.,**[12]**,**and**Mercan et al.,**[13] who said hemophilia adversely affect quality of ,efillimitspatientsactivitiesandhandersthierllives .

Our study showing no stastically significant difference in joint score and start of treatment and this agreed by **Sha Liu et al.**[14]that said no significant difference in mean joint score was found between patients receiving on demand therapy and those receiving on demand to low dose prophylactic therapy

In our study, we found that there is no statistically difference between hemophilia with arthropathy group and without arthropathy regarding on prophylaxis or on demand.in our study both groups on demand not prophylactic treatment.AS well AS**Hemophilia Federation of America** [15]mentioned that on demand is another treatment option for individual with hemophilia .patient can inject clotting factor on need to treat specific episodes .factor replacement is injected following an injury to provide clotting factor and promote normal healing .

On contrast to our study,Amulti centerEuropianstudy in 903 patients reported that patients treated with prophylaxis had significantly higher score in general health and mental health [16].

THE WHO ,has designated prophylaxis as therapy of choice for all children with severe hemophilia compared with treatment in case of bleeding only (on demand ,prophylaxis is associated with increased clotting factor consumption especially in childhood [17, 18 , 19].

A 2017 literature review concluded that individuals using prophylaxis had signifigantly better average health related quality of life than those who received on demand factor [20].

Prophylaxis had served as a whole marks of treatment for individual with hemophilia as a standard treatment in developed countries[21, 22, 23].

In our study limitations are both higher cost and limited availability of clotting factor concentrates. AS clotting factor concentrates are expensive, issues of cost have hampered the introuduction of prophylaxis in developing countries.

V. Conclusion:

All hemophilic patients received treatment on-demand only, No prophylaxis treatment. Regarding to H.A. our study reported that severity of H.A. Correlate positively with (age (years), weight, BMI, waist (cm), Global Giat Score and with HJHS total Score). In contrast, it correlates negatively with factor plasma level. All hemophilic patients (with or without hemoarthropathy) suffer fromunsatisified quality of life (100%).

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