

Quality of life & self-esteem in Hemophilic children and adolescents

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Abstract

Background: The present study aimed to assess of quality of life and self esteem and to discover the relation of both to the clinical and therapeutic parameters in children with hemophilia. To achieve this target, 100 hemophilic children were included in the study. They had a mean age of 10.09 ± 4.44 . They were subjected to careful history taking and thorough clinical examination. In addition, they were subjected QOL and self esteem assessment. It was shown that 48% of the studied children are obese, the most commonly encountered manifestation was target joint 75.0% followed by gum bleeding 72%, circumcision bleeding 69.0%, epistaxis 54.0%, hemarthrosis 50.0% muscle hematoma 49.0%, dental bleeding 45%, and limited joint movement 33.0%. Regarding the QOL, the present study found that it is clear that the studied children had generally poor quality of life domain. This is manifested by the mean total QOL score which is only 50.9, comparison between individual and total QOL scores in the studied age groups had revealed that children of the middle age had significantly better QOL scores when compared with the other two groups. The relatively poor QOL scores in the present study is explained by the higher frequency of joint problems which had detrimental effects on the studied children physical health and other QOL domains.

Also patients with joint problems including hemarthrosis, target joint and limited joint movement had significantly worse QOL score when compared with patients without, assessing patients self- esteem revealed significantly lower Coppersmith self- esteem inventory in patients when compared with controls, obese children had significantly lower self esteem scores when compared with children with normal weight. Finally, we showed a statistically significant inverse correlation between self esteem and QOL

جودة الحياة وتقدير الذات لدى الأطفال والمراهقين المصابين بالهيموفيليا

الخلفية: جودة الحياة في مرضى الناعوري (سيولة الدم) في مصر تحتاج إلى جهود مضيئة من فرق الرعاية المتكاملة للأطفال من أخصائيي الدم والأطباء النفسيين من أجل تقييم صحيح وتحسين جودة الحياة. ومن بين العوامل التي تسهم بشكل كبير في الحالة العاطفية للطفل الناعوري احترام الذات. لذلك، فإن الدراسة الحالية تهدف إلى تقييم نوعية الحياة وتقدير الذات واكتشاف العلاقة بين كل من المعايير السريرية والعلاجية في هؤلاء الأطفال. لتحقيق هذا، تم إدراج 100 طفل الناعوري كان لديهم متوسط عمر 10.09 ± 4.44 . كما تعرضوا إلى تناول تاريخي دقيق وفحص سريري شامل. بالإضافة إلى ذلك، أنهم تعرضوا إلى اختبار جودة الحياة وتقدير الذات. تبين أن 48% من الأطفال يعانون من السمنة المفرطة. حيث كان المظهر الأكثر شيوعاً الذي يواجهه هو المفصل المستهدف 75.0%، يليه نزيف اللثة 72%، ثم نزيف الختان 69.0%، ثم الرعاف 54.0%، ثم تسمى المفصل 50.0% وكتل دموية في العضلات 49.0%، نزيف الأسنان 45.0%، وحركة المفاصل محدودة 45.0%. ونزيف اللثة 33.0%. في دراستنا، كان الأطفال الذين لديهم مستوى اجتماعي واقتصادي أعلى لديهم مجموع أفضل لجودة الحياة بالمقارنة مع الذين لديهم مستوى اجتماعي واقتصادي متوسط ومنخفض وأيضاً، كان المرضى الذين لديهم مستوى اجتماعي واقتصادي متوسط كانوا أصحاب جودة حياة أفضل من المرضى الذين يعانون من انخفاض المستوى الاجتماعي والاقتصادي. أيضاً المرضى الذين يعانون من مشاكل المفاصل- بما في ذلك تسمى المفصل، وحركة المفاصل المحدودة- لديهم نتيجة أسوأ بكثير لجودة حياة. الأطفال البناء أقل بكثير تسجيلات تقدير الذات بالمقارنة مع الأطفال ذوي الوزن الطبيعي.

الاستنتاجات: الأطفال الناعوري لديهم نوعية حياة رديئة وانخفاض تقدير الذات مقارنة مع أقرانهم الأصحاء، المستوى الاجتماعي والاقتصادي، والسمنة، وتأثير المفاصل، والعلاج تحت الطلب هي المحددات الرئيسية لجودة الحياة وتقدير الذات في هؤلاء المرضى، جودة الحياة ترتبط بشكل ملحوظ مع تقدير الذات.

التوصيات: ينصح بدراسة وطنية أكبر لتحديد التأثير الدقيق للناعوري على جودة الحياة، واحترام الذات والمتغيرات النفسية الأخرى، ويجب أن يكون التشاور النفسي عنصراً أساسياً في إدارة أطفال الناعوري، ويجب أن تشمل مراكز العلاج فريق متكامل من أطباء الأطفال وأمراض الدم والأطباء النفسيين والأخصائيين الاجتماعيين، والتثقيف الصحي للعائلات والمدرسين للتعامل السليم مع الأطفال وتوفير بيئة آمنة، المزيد من الجهود الرسمية لتوفير العلاج عوضاً عن نقل الدم المتكرر، المزيد من الجهود الغير رسمية لتكوين روابط ومجموعات لدعم الأطفال المرضى.

Introduction:

Hemophilia is a hereditary (sex- linked, recessive) bleeding disorder in which a decreased level of clotting factor activity in the blood may lead to spontaneous hemorrhages in muscles and joints. As the gene which causes this disorder is located on the X- chromosome, hemophilia predominantly affects male (Bottos et al., 2007).

The worldwide incidence of hemophilia A is approximately 1 case per 5000 male individuals, with approximately one third of affected individuals not having a family history. The prevalence of hemophilia A varies with the reporting country, with a range of 5.4 to 14.5 cases per 100.000 male individuals (Konkle et al., 2009).

Hemophilias are chronic diseases and there is no cure for hemophilia A or B. Concentration of the missing factor should be raised with intravenous infusion to prevent or stop bleeding. Bleeding might occur into the joints or muscles and is very painful. Considering the above mentioned problems, it is increasingly important that providers are able and can consider other aspects of this disease including the aspects of mental health (Ghanizadeh and Baligh-Jahromi, 2009).

These health aspects as an important component of the management have increasingly gained recognition in recent years (Globe et al., 2002). However, there are only a limited number of studies that had focused on lifestyle and quality- of- life issues (Naraine et al., 2002). Hemophilic subjects show a lower level of self- esteem compared with their healthy counterparts. However, there are no differences between the hemophilic subjects and their healthy counterparts in depression (Canclini et al., 2003).

Aim:

1. Assessment of quality of life in children with hemophilia.
2. Assessment of self esteem in children with hemophilia.

Subjects:

One hundred hemophilic children aged (4- 16) years were included in the study in addition to 50 age matched healthy controls. Patients were selected on the basis of the following criteria:

1. Inclusion Criteria:
 - a. Age (6- 18) years.
 - b. IQ \geq 70.
2. Exclusion Criteria: Associated Chronic Medical Condition.

Methods:

The study participants were submitted to the following:

1. History Taking.
2. Physical Examination:
 - a. General examination: Head and neck, limbs, skin, back and spine, genitalia.
 - b. Systemic examination: Neurological, cardiovascular, chest, abdominal.
3. Investigations:
 - a. IQ Test: IQ was performed to assure the children's ability to participate in the QOL and self esteem assessment. By using Wechsler Intelligence Scale for Childhood (Wechsler, 1991). The Wechsler Intelligence Scale for Children, often abbreviated as WISC, is an individually administered measure of intelligence intended for children aged (6- 18) years. The WISC is designed to measure human intelligence as reflected in both verbal and nonverbal (performance)

abilities.

- b. Quality of life assessment: using Arabic version of the Hemophilia- QOL questionnaire (Tantawy et al., 2011): The Haemo- QOL self- reported disease- specific questionnaire for the quality of life in hemophilia was translated into the Arabic language, including the questionnaire for different age groups (child and parent versions). The translation was undertaken by a team of 2 professional pediatric hematologists, a psychologist, and a professional English language expert. Backward and forward translations were carried out to ensure efficient translation. An interview version for smaller children age group I: (4- 7) years was available with 21 items pertaining to 8 dimensions (Physical Health, Feelings, View, Family, Friends, Others, Sport and school/ Kindergarten, Treatment); for the schoolchildren aged (8- 12) years (age group II), the self- administered questionnaire consisted of 2 additional domains (Perceived Support, Dealing) with overall 64 items; and for adolescents (age group III): (13- 16) years, it was expanded with another further additional domain Future and consisted of 75 items as well as a questionnaire for parent rating containing 9 to 11 subscales (depending on age- group versions).

Statistical Analysis:

Data obtained from the present study were computed using SPSS versions 17 under the platform of Microsoft Windows 7. Continuous data were expressed in the form of mean \pm SD while categorical data were expressed in the form of count and percent. Comparison of continuous data was performed utilizing student t test, while categorical data were done using Chi- square test. Relation between variables were investigation by Pearson's correlation coefficient. P value less than 0.05 was considered statistically significant

Results And Discussion:

Table (1) Demographic characteristics of the studied patients (n= 100)

Age (Years)	Range	4- 16
	Mean \pm SD	10.09 \pm 4.44
	4- 7	40 (40.0%)
	8- 12	30 (30.0%)
BMI	13- 16	30 (30.0%)
	Normal	52 (52.0%)
	Obese	48 (48.0%)

This table shows the demographic characteristics of the studied patients. They had a mean age of 10.09 \pm 4.44 years distributed as 40 in the age from (4- 7) years, 30 in the age from (8- 12) years and 30 in the age from (13- 16) years. BMI distribution showed that 52 children were within normal ranges while 48 children were obese.

Table (2) Socioeconomic status (SES) in the studied patients (n= 100)

	No	%
Low	55	55.0
Intermediate	41	41.0
High	4	4.0

This table shows SES in the studied patients. It was low in 55 patients, intermediate in 41 patients and high in 4 patients.

Table (3) Quality of life domains in the studied patients (n= 100)

	Range	Mean± SD
Physical_Health	15- 80	47.74±17.89
Feeling	32- 93	62.3±17.82
View	22- 95	56.34± 20.38
Family	25- 79	48.33±13.74
Friends	16- 81	43.82±15.13
Support	25- 70	45.36± 13.15
Other_People	26- 94	56.78±17.66
Sports_School	00- 98	46.95± 27.5
Dealing	32- 54	42.65± 6.4
Treatment	20- 98	53.02± 21.15
Total	31.25- 63.8	50.9± 6.1

This table shows the quality of life domains in the studied patients. It is clear that the studied children had generally poor quality of life domains. This is manifested by the mean total QOL score which is 50.9.

Table (4) Relation of the total quality of life score to the clinical data

		Total QOL Score	Student	
			t	p
Circumcision Bleeding	+	50.9± 6.3	0.211	0.833
	-	51.8± 5.7		
Epistaxis	+	50.92± 6.16	0.126	0.9
	-	51.07± 6.22		
Gum Bleeding	+	51.35± 5.83	0.938	0.35
	-	50.06± 6.95		
Bleeding Dental	+	50.9± 5.63	0.127	0.899
	-	51.06± 6.6		
Previous Surgery	+	51.8± 4.35	0.293	0.771
	-	50.9± 6.5		
Hemarthrosis	+	52.9± 6.05	3.4	0.0009*
	-	49.0± 5.6		
Muscle Hematoma	+	51.57± 6.15	0.925	0.357
	-	50.43± 6.16		
Target Joint	+	52.5± 5.6	4.9	0.0001*
	-	46.2± 5.2		
Limited Joint Movement	+	55.2± 5.7	5.45	0.0001*
	-	48.9± 5.2		

This table shows the relation between QOL score and the clinical data. Patients with joint problems including hemarthrosis, target joint and limited joint movement had significantly worse QOL score when compared with patients without.

Table (5) Comparison between Coppersmith self- esteem inventory score in patient& controls

	Patients (n= 100)	Controls (n= 50)	Student T Test	
			t	P
Range	20- 70	50- 80	7.6	0.0001*
Mean± SD	45.94± 16.77	65.2± 8.82		

This table shows a significantly lower Coppersmith self- esteem inventory in patients when compared with controls.

Table (6) Relation of self esteem to the demographic data

			Self Esteem	
			r	p
Age			- 0.05	0.6
Age Groups	4- 7	44.6± 18.0	ANOVA	
	8- 12	51.1± 15.1	F	P
	13- 16	42.5± 15.8	2.22	0.11
			Student T Test	
BMI	Normal	50.07± 15.4	t	p
	Obese	41.4± 17.1	2.64	0.01*

This table shows no significant relations between self esteem and patients'

age. Obese children had significantly lower self esteem scores when compared with children with normal weight.

Table (7) Relation of self esteem to SES

	Self Esteem	One Way Anova	
		F	P
Low	44.3± 16.6	0.62	0.53
Intermediate	47.5± 16.8		
High	51.0± 19.3		

This table shows general improvement of the self esteem with better SES. However, the differences between different SES categories was not statistically significant.

In the present study, it was shown that 48% of the studied children are obese. This is in accordance with the conclusions the systematic review of Wong et al. (2011) who noted that overweight and obesity are now more prevalent in the hemophilia population than previous generations, with rates similar to and, in certain subsets even higher, than that of the general population. Increased BMI leads to limitations in joint range of motion in the general population and even more so in persons with hemophilia.

In the current study, the most commonly encountered manifestation is target joint was target joint 75.0% followed by gum bleeding 72%, circumcission bleeding 69.0%, epistaxis 54.0%, hemarthrosis 50.0% muscle hematoma 49.0%, dental bleeding 45%, and limited joint movement 33.0%.

These data are in agreement with many studies. In the study of Kern et al. (2004), 15 out 16 children with hemophilia had target joint. Also, in the study of Ragni et al. (2012), 54.9% of children who underwent circumcission had bleeding.

In an Egyptian study, hemarthrosis, muscle hematoma and dental bleeding were also frequently reported in children with hemophilia (Mokhtar et al., 2012).

In our study, 73 children were having on demand therapy while only 23 were on secondary prophylaxis. This is in accordance with the study of Tantawy et al. (2011) who studied who noted that 68% of the studied children are on demand therapy.

Regarding the QOL, the present study found that it is clear that the studied children had generally poor quality of life domain. This is manifested by the mean total QOL score which is only 50.9. In our study, comparison between individual and total QOL scores in the studied age groups had revealed that children of the middle age had significantly better QOL scores when compared with the other two groups. The relatively poor QOL scores in the present study is explained by the higher frequency of joint problems which had detrimental effects on the studied children physical health and other QOL domains.

Regarding the relation of quality of life to the demographic data, the present study found that no significant correlation between patients age and total QOL score. However, comparison between the studied age groups had shown that the age group of (8- 12) years had better score when compared to the other two groups. Also, it was revealed that obese children had significantly worse QOL score when compared with normal weight children. This is supported by the study of Khair et al. (2012) who noted that hemophilic children with sedentary life and overweight /obesity had significantly worse QOL when compared with normal weight children.

In our study, children with high SES had better total QOL score when compared with children with intermediate and low SES. Also, patients with intermediate SES had better QOL than patients with low SES. This is

explained by the economic burden of supplying the appropriate treatment (Tezanos Pinto and Ortiz, 2004).

In the present study, patients with joint problems including hemarthrosis, target joint and limited joint movement had significantly worse QOL score when compared with patients without. This is evidenced by the study of van der Net et al. (2006) who investigated the level of physical fitness, functional ability and quality of life in children with hemophilia. They noted that patients with haemophilia with good joint health and no limitations of activities have comparable physical fitness and physical active lifestyle with healthy peers and good HRQOL.

Also, the study of Gringeri et al. (2014) concluded that more than two to three bleeding into the same joint may cause irreversible and progressive structural damage that compromise health-related quality of life (HRQOL).

Regarding the relation between treatment regimen and QOL, the present study found that patients on secondary prophylaxis had significantly better QOL score when compared with patients on demand.

This is supported by the study of Valentino et al. (2012) who compared the efficacy of two prophylaxis regimens (primary outcome) and of on-demand and prophylaxis treatments (secondary outcome). In their study, prophylaxis significantly reduces bleeding compared with on-demand treatment with subsequent improvement in quality of life scores.

This was directly confirmed by the recent study of Santagostino et al. (2014) who investigated the HRQOL of patients with haemophilia A treated prophylactically with a new recombinant factor VIII. They found that switching to prophylaxis was identified as a potential driver of improvement of HRQOL in patients with haemophilia A.

In the present study, assessing patients self-esteem revealed significantly lower Coppersmith self-esteem inventory in patients when compared with controls. This is in agreement with the study of Canclini et al. (2003) who carried out a case-control study in which some psychological dimensions (social expectations, tendency to depression, state of anxiety and self-esteem) were evaluated in a group of 60 haemophiliacs. A control group was formed of 78 healthy subjects matched for age, socio-economic class and level of education. The results showed that the haemophiliacs have a good psychological adaptation to their disease with the exception of their greater tendency to have less self-esteem than do the healthy subjects.

Also, in the study of Hegeman et al. (2011), the authors aimed to explore: (i) perceived competence, (ii) perceived impact of illness, and (iii) analyze associations between perceived competence and demographic factors, disease-related factors and joint status in young haemophiliacs in the Netherlands. Fifty-four children age (8-12) years and 72 adolescents (12-18) years with haemophilia participated in this cross-sectional, multi-centre, explorative study. Measurements included perceived competence (Self Perception Profile for Children/Adolescents), impact of disease (Revised Perception Illness Experience; range 1-5), demographic factors, disease-related factors, joint status and functional status. In this study, children with haemophilia had a lower global self-worth score.

However, in the study of Alioglu et al. (2013) who aimed to research how the disease which creates limitations in different areas of life affects emotions, feelings and attitudes of a child about himself. The research was conducted on 23 patients with severe haemophilia A (factor VIII level <1%) aged between (9-15) years and 23 controls at the same age interval without any known

chronic disease or mental deficiency. Self-esteem was assessed using Piers-Harris Children's Self-Concept Scale which is comprised of six subscales that aim to assess emotions, feelings and attitudes of children between 7 and 18 years of age about themselves. In this study, self-esteem levels of haemophiliac and healthy children were compared. There was no statistically significant difference between the total self-esteem scores of haemophiliac and healthy subjects. A comparison of the subscale scores of Piers-Harris Children's Self-Concept Scale used in study revealed a statistically significant difference between haemophiliac patients and controls in terms of "Behavior and adaptation" ($P=0.03$). A statistically significant difference was found between the haemophiliac patients and the controls when anxiety subscale points were compared ($P=0.044$).

This discrepancy is explained by the variable sample size, selection criteria and importantly, the different utilized self-assessment tools.

In the present study, obese children had significantly lower self-esteem scores when compared with children with normal weight. This is in accordance with the study of Danielsen et al. (2012) who aimed to identify factors related to low domain-specific self-esteem in children with overweight/obesity. In this study, children with overweight scored significantly lower than normal-weight children on all self-esteem domains.

We also found that patients with limited joint movement had significantly lower self-esteem when compared with patients with normal joint movement. In spite of the fact that patients with target joint had worse self-esteem than patients without, the differences is not statistically significant. These data finds support in other studied evaluating children's self-esteem in diseases affecting joint movement including arthritis (Yadav and Yadav, 2013).

Finally, we showed a statistically significant inverse correlation between self-esteem and QOL i.e. better (low) quality of life scores are associated with better self-esteem scores. The relation of QOL to self-esteem was previously reported in many diseases including obesity (Kim et al., 2013), and inflammatory bowel disease (Ross et al., 2011).

Conclusions:

1. Hemophilic children have poor quality of life and lower self-esteem when compared with healthy peers.
2. SES, obesity, joint affection, on demand treatment are the main determinants of QOL and self-esteem in these patients.
3. QOL was significantly correlated with self-esteem.

Recommendations:

1. A larger nationwide study is recommended to delineate the precise impact of hemophilia on QOL, self-esteem and other psychological variables.
2. Psychiatric consultation should be an essential element in management of hemophilic children.
3. Centers for treatment of hemophilia should have multi-specialty team (pediatrician, hemtologist, psychatrist, physiotherapist and social worker).
4. Health education to families and teachers of children with hemophilia to know how to make the environment more save and supported to them.
5. More official efforts must be exerted to assure adequate supplementation of drugs instead of frequent blood transfusion.
6. More non official efforts must be done to support these children in form of social committee.

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