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QUALITY OF LIFE OF PATIENTS WITH HEMOPHILIA IN BAGHDAD MEDICAL CITY

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ABSTRACT

Background: hemophilia is one of the most common inherited bleeding disorders. The chronic nature of the disease greatly affects the patients and their families and imposes a great burden upon their lives. And also, it exerts a psychological effect on the patients. And the expensive drugs of the disease make a burden on the healthcare system. **Objectives:** to assess the quality of life of patients with both hemophilia A and B, and to demonstrate the effect of some sociodemographic factors on the quality of life of these patients. **Methods:** this is a descriptive cross-sectional study with some analytical elements. It has done in children welfare hospital at Baghdad medical city, and data was collected using self-administered questionnaire, consisting of two parts, the first part demonstrates the sociodemographic and clinical characteristics of the patients, the second part was adapted from the world health organization WHOQOL-bref questionnaire. It included 320 participants. **Results:** the study showed that the mean age of the patients was 27.5 years and the most frequent age group was between 20- 29 years. 74% of the patients have fair to good socioeconomic status. 228 patients with hemophilia A and 92 with hemophilia B. 20 patients have recorded presence of inhibitors. 188 patients have severe disease. The lowest score was for the physical health domain (mean= 40.31, P value= 0.001) and the highest score was for the environmental health domain (mean= 52.60, P value= 0.001). **Conclusion:** the study concluded that the overall QOL was poor. There was a significant association between general QOL and age of diagnoses, marital status, number of children, the number of affected children, the presence of family history of hemophilia and the socio-economic status.

INTRODUCTION

Deficiencies of coagulation factors have been recognized for centuries, with genetic deficiencies causing lifelong recurrent bleeding episodes in joints, muscles, and closed spaces, either spontaneously or following injury. The most common inherited deficiencies are hemophilias, X-linked disorders caused by factor VIII (hemophilia A) or factor IX (hemophilia B). Rarer bleeding disorders due to deficiencies of factors such as FII, FV, FVII, FX, FXI, FXIII, and fibrinogen are usually autosomal recessive.[1] Advances in understanding the molecular bases of these deficiencies have improved disease characterization and hold promise for targeted therapies, including small molecules, recombinant proteins, and gene-based treatments.[2] Treatment typically involves replacing the deficient protein using recombinant or plasma-derived products or fresh-frozen plasma (FFP). Accurate diagnosis is crucial to optimize care and minimize the risks associated with inappropriate treatment[3] Hemophilia A, caused by factor VIII deficiency, affects approximately 1 in 10,000 individuals. Factor VIII, synthesized in the liver and endothelial cells, is stabilized by von Willebrand factor and has a half-life of 12 hours. Hemophilia A is classified into three groups based on factor VIII levels: severe (<0.01 units/ml), moderate (0.01–0.4 units/ml), and mild (>0.4 units/ml)[4] Severe hemophilia is associated with recurrent joint bleeding, leading to chronic hemophilic arthropathy if untreated, significantly impacting patients' daily lives and well-being. Management relies on prophylactic coagulation factor replacement or on-demand treatment, which is more common in resource-limited settings. Hemophilia B, or Christmas disease, caused by abnormalities in the factor IX gene, affects approximately 1 in 30,000 individuals. Although clinically similar to hemophilia A, it is less common. The frequency of bleeding depends on factor IX levels, and treatment involves recombinant factor IX concentrates.[5] Quality of life (QOL) assessment is vital in managing chronic diseases like hemophilia, as it reflects the patient-centered experience

and highlights the impact of the disease on health and daily life. The WHOQOL-BREF, developed by the World Health Organization and tested in 18 countries, is a widely used tool for QOL assessment. It contains 26 questions divided into four domains: physical health, psychological, social, and environmental. Evaluating QOL helps identify deficiencies in disease management and areas for improvement in patient care, ultimately enhancing patients' overall well-being.[6] Objectives: to assess the quality of life of patients with both hemophilia A and B, and to demonstrate the effect of some sociodemographic factors on the quality of life of these patients.

METHOD

The collected data were initially entered into Microsoft Excel 2016 and subsequently imported into the Statistical Package for Social Sciences (SPSS) software, version 25, for analysis. Descriptive statistics were utilized, with categorical variables presented as frequencies and percentages, while continuous variables were expressed as means ± standard deviation (SD). The study was conducted at the Child Welfare Hospital, Department of Bleeding Tendency, within Baghdad Medical City. A total of 721 hemophilia patients attended the bleeding tendency center, of whom 545 had hemophilia A and 176 had hemophilia B. The study sample comprised patients attending the center who were older than 15 years. A total of 320 patients meeting the inclusion criteria participated in the study. Inclusion criteria encompassed any patient with hemophilia A or B willing to participate. Exclusion criteria excluded any patient with other chronic illnesses affecting quality of life. The data were collected using self-administered questionnaire consisting of two parts, the first part consist of socio-demographic aspects which was designed by the researcher and approved by the supervisor and panel of experts in the community department and Hematology, Regarding the calculation of the socioeconomic status (SES), a simple equation that will calculate the SES as follows: SES= Education+ Occupation+ House ownership* 0.5 +Car ownership* 1+ [(AGE-20) /100]-retired/unemployed/deceased. The second part was based on WHOQOL-BREF questionnaire (English and Arabic version 1996), consisting of 26 items which was grouped into 4 domains.[7] The statistical analysis re-confirmed that categorical variables were summarized as frequencies and percentages, while continuous variables were reported as means ± SD, ensuring a comprehensive understanding of the demographic and clinical characteristics of the study population.

RESULTS

Figure 1 shows distribution of studied cases according to sociodemographic variables. Regarding the age of diagnosis; 62% were diagnosed before the age of 1 year, and 38% were diagnosed after the age of 1 years. Regarding marital status, 57.5% were single and 42.5% were married. 58.7% have no children, 13.7% have only one child and 27.5% have 2 and more children. 88.8% have no affected siblings, and 11.2% have their siblings affected with hemophilia. 45% had no family history of hemophilia, and 55% had a family history of hemophilia. 26.25% were poor socio-economically, 53.75% have fair socio-economic status and 20% have good socio-economic status.

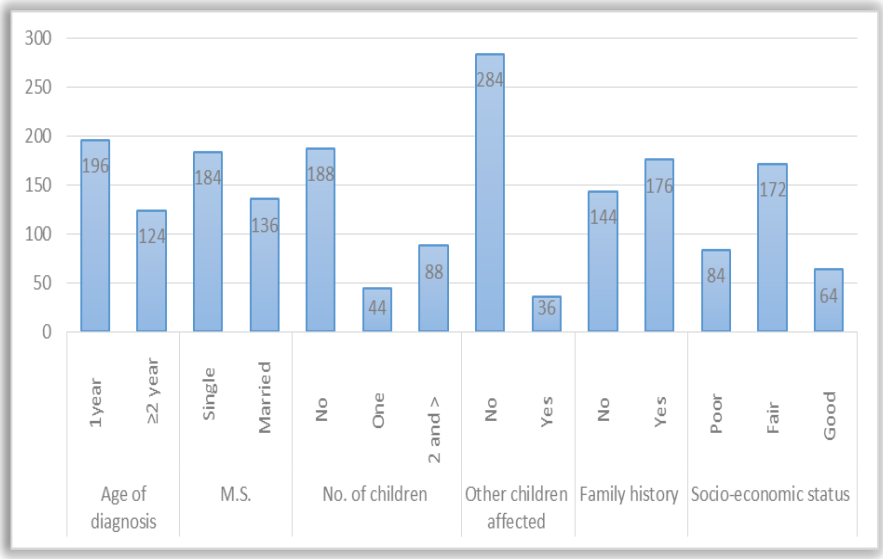

Figure 1: distribution of studied cases according to sociodemographic variables.

Table 1 shows that using the ANOVA test, the mean score of environmental domain = 52.60 ± 16.32 and it was the domain with the highest score, followed by social relationships (mean score of = 43.52 ± 9.88) and psychological wellbeing (40.42 ± 10.16). The lowest score was foe physical health (mean score 40.31 ± 11.15). (P. value <0.001).

Table 1: distribution of studied cases according to means of different QOL domains.

Domain	N	Mean	Std. Deviation	P value
physical health	320	40.31	11.15	0.001
psychological wellbeing	320	40.41	10.16	
social relationships	320	43.56	9.88	
environment	320	52.60	16.32	
Total QOL		44.21	18.06	

Figure 2 shows the distribution of studied cases according to general QOL and general health. Regarding general quality of life (QOL), 58% were having poor QOL, 27% were having fair QOL and 15% were having good QOL. Regarding general health, 55% had poor general health, 28% had fair general health and 17% had good general health.

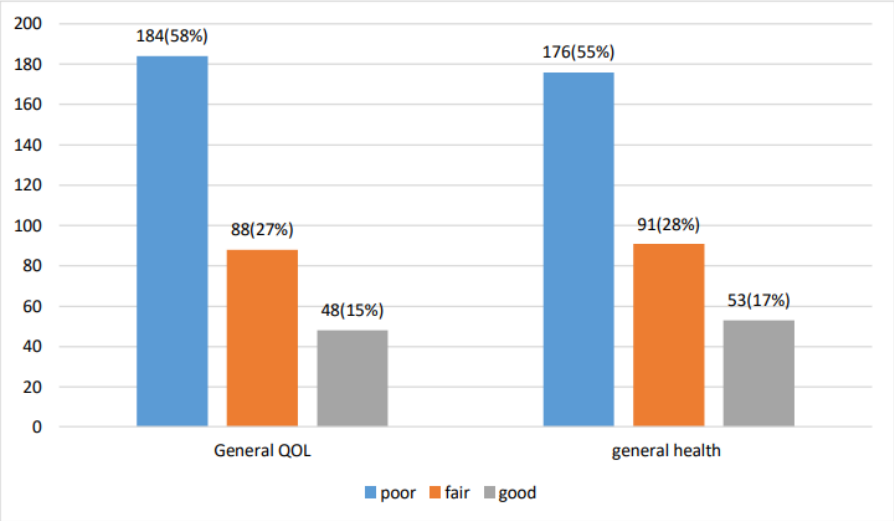

Figure 2: distribution of studied cases according to general QOL and general health.

Table 2: shows the association between socio-demographic variables and general QOL. 57.5% gave impression of poor general QOL, 27.5% gave impression of fair and 15% of good general QOL.

Table 2: association between socio-demographic variables and general QOL.

		Poor 184(57.5%)		Fair 88(27.5%)		Good 48(15%)		P.V.
		Count	N %	Count	N %	Count	N %	
Age of diagnosis	<1year 196	136	69.4%	32	16.3%	28	14.3%	<0.001
	≥1 year 124	48	38.7%	56	45.2%	20	16.1%	
Marital Status	Single 184	116	63.0%	24	13.0%	44	23.9%	<0.001
	Married 136	68	50.0%	64	47.1%	4	2.9%	
No. of children	No 188	124	66.0%	24	12.8%	40	21.3%	<0.001
	One 44	24	54.5%	16	36.4%	4	9.1%	
	2 > 88	36	40.9%	48	54.5%	4	4.5%	
children affected	No 284	172	60.6%	64	22.5%	48	16.9%	<0.001
	Yes 36	12	33.3%	24	66.7%	0	0.0%	
Family history	No 144	80	55.6%	64	44.4%	0	0.0%	<0.001
	Yes 176	104	59.1%	24	13.6%	48	27.3%	
Socio-economic status	Poor 84	44	52.4%	36	42.9%	4	4.8%	<0.001
	Fair 172	108	62.8%	36	20.9%	28	16.3%	
	Good 64	32	50.0%	16	25.0%	16	25.0%	

Table 3 shows the association between socio-demographic variables and Physical health. 38.7% gave the impression of poor physical health, 35% fair and 26.3% good. Of those who were diagnosed before the age of 1 year, 49% reported poor physical health, 38.8% reported fair and 12.2% reported good physical health. And consequently, those who were diagnosed after the age of 1 year reported their physical health as 22.6%

poor, 29% fair and 48, 4% as good. There was significant association between age of diagnosis and physical health (P. value= 0.001).

Table 3: association between socio-demographic variables and Physical health.								
		poor 124(38.7%)		fair 112(35%)		Good 84(26.3%)		PV
		N	%	N	%	N	%	
Age of diagnosis	<1year 196	96	49.0	76	38.8	24	12.2	0.001
	≥1 year 124	28	22.6	36	29.0	60	48.4	
M.S.	Single 184	68	37.0	60	32.6	56	30.4	0.138
	Married 136	56	41.2	52	38.2	28	20.6	
No. of children	No 188	72	38.3	64	34.0	52	27.7	0.311
	One 44	12	27.3	20	45.5	12	27.3	
	2 > 88	40	45.5	28	31.8	20	22.7	
children affected	No 284	104	36.6	104	36.6	76	26.8	0.078
	Yes 36	20	55.6	8	22.2	8	22.2	
Family history	No 144	44	30.6	64	44.4	36	25.0	0.003
	Yes 176	80	45.5	48	27.3	48	27.3	
Socio-economic status	Poor 84	28	33.3	44	52.4	12	14.3	0.001
	Fair 172	84	48.8	44	25.6	44	25.6	
	Good 64	12	18.8	24	37.5	28	43.8	

Table 4 demonstrates association between socio-demographic variables and Psychological wellbeing. 45% gave the impression of poor Psychological wellbeing, 44% was fair and 11% was good.

Table 4: association between socio-demographic variables and Psychological wellbeing.								
		Poor 144(45%)		Fair 140(44%)		Good 36(11%)		PV
		N	%	N	%	N	%	
Age of diagnosis	<1year 196	108	55.1	72	36.7	16	8.2	0.001
	≥1 year 124	36	29.0	68	54.8	20	16.1	
M.S.	Single 184	100	54.3	60	32.6	24	13.0	0.001
	Married 136	44	32.4	80	58.8	12	8.8	
No. of children	No 188	108	57.4	56	29.8	24	12.8	0.001
	One 44	12	27.3	20	45.5	12	27.3	
	2 > 88	24	27.3	64	72.7	0	0.0	
children affected	No 284	132	46.5	120	42.3	32	11.3	0.284
	Yes 36	12	33.3	20	55.6	4	11.1	
Family history	No 144	56	38.9	76	52.8	12	8.3	0.001
	Yes 176	88	50.0	64	36.4	24	13.6	
Socio-economic status	Poor 84	32	38.1	52	61.9	0	0.0	0.001
	Fair 172	96	55.8	60	34.9	16	9.3	
	Good 64	16	25.0	28	43.8	20	31.3	

Table 5 demonstrates association between socio-demographic variables and social relationships. 29% gave the impression of poor social relationships, 54% was fair and 17% was good.

Table 5: association between socio-demographic variables and social relationships.								
		Poor 92(29%)		Fair 172(54%)		Good 56(17%)		PV
		N	%	N	%	N	%	
Age of diagnosis	<1year 196	76	38.8	96	49.0	24	12.2	0.001
	≥1 year 124	16	12.9	76	61.3	32	25.8	
Marital status	Single 184	64	34.8	72	39.1	48	26.1	0.001
	Married 136	28	20.6	100	73.5	8	5.9	
No. of children	No 188	68	36.2	72	38.3	48	25.5	0.001
	One 44	4	9.1	32	72.7	8	18.2	
	2 > 88	20	22.7	68	77.3	0	0.0	
children affected	No 284	80	28.2	152	53.5	52	18.3	0.531
	Yes 36	12	33.3	20	55.6	4	11.1	
Family history	No 144	28	19.4	104	72.2	12	8.3	0.001

	Yes 176	64	36.4	68	38.6	44	25.0	
Socio-economic status	Poor 84	24	28.6	60	71.4	0	0.0	0.001
	Fair 172	68	39.5	72	41.9	32	18.6	
	Good 64	0	0.0	40	62.5	24	37.5	

Table 6 demonstrates association between socio-demographic variables and environment. 22.5% reported their environmental health as poor, 57.5% were fair and 20% were good.

Table 6: association between socio-demographic variables and environment.

		Poor 72(22.5%)		Fair 184(57.5%)		Good 64(20%)		PV
		N	%	N	%	N	%	
Age of diagnosis	<1year 196	60	30.6	108	55.1	28	14.3	0.001
	≥1 year 124	12	9.7	76	61.3	36	29.0	
Marital status	Single 184	48	26.1	92	50.0	44	23.9	0.007
	Married 136	24	17.6	92	67.6	20	14.7	
No. of children	No 188	48	25.5	96	51.1	44	23.4	0.006
	One 44	8	18.2	24	54.5	12	27.3	
	2 > 88	16	18.2	64	72.7	8	9.1	
children affected	No 284	68	23.9	152	53.5	64	22.5	0.001
	Yes 36	4	11.1	32	88.9	0	0.0	
Family history	No 144	24	16.7	96	66.7	24	16.7	0.010
	Yes 176	48	27.3	88	50.0	40	22.7	
Socio-economic status	Poor 84	16	19.0	64	76.2	4	4.8	0.001
	Fair 172	44	25.6	96	55.8	32	18.6	
	Good 64	12	18.8	24	37.5	28	43.8	

DISCUSSION

Hemophilia, a chronic disease with numerous complications, significantly impacts various aspects of life, making quality of life (QOL) assessment essential to refine treatment goals and improve care. The WHOQOL-BREF questionnaire, validated across cultures, proved reliable for estimating QOL in chronic diseases. However, patients with lower education levels faced difficulties understanding some questions[8] The study revealed that the mean patient age and the predominant age group (20–29 years) aligned with findings by Nunes A. in Brazil (2009).[9] Most patients were diagnosed before the age of 1 year, in contrast to Sarper N.'s study in Turkey (2010), where diagnosis occurred later. This earlier diagnosis in Iraq could be attributed to the prevalence of severe hemophilia symptoms manifesting during early circumcision practices.[10] Notably, patients diagnosed earlier had poorer QOL, possibly reflecting the severe disease's early onset and significant impact on physical health, psychological wellbeing, social relationships, and environmental health. Marital status significantly influenced QOL, with unmarried patients reporting poorer QOL compared to married individuals. Marriage provided stability, family support, and better care.[9] A significant association was also observed between the number of children and QOL, as a larger number of affected children increased caregiving burdens and frequent healthcare visits.[11] Similarly, family history of hemophilia correlated with poorer QOL across all domains due to the added challenges of managing multiple cases. The socioeconomic status (SES) of the study population, consistent with the Campinas study in Brazil (2001), revealed a significant association between SES and QOL in all domains.[7] Education level also played a role, with more than half of the patients having primary or intermediate schooling. This finding aligns with studies from Brazil, Spain, and Greece, indicating hemophilia's interference with schooling.[11-13] In this study, environmental health scored highest among QOL domains, as patients reported positive perceptions regarding security, income, leisure opportunities, and healthcare services. This contrasts with findings by other studies, where the psychological domain scored highest.[7,14,15] Conversely, physical health scored the lowest, consistent with Sarper N.'s (2010) and Ferreira A.'s (2013) findings, due to the significant impact of pain on daily activities, work, and overall wellbeing.[7,10] Psychological health also scored low, reflecting the effects of pain, disability, and life restrictions, contributing to depression and anxiety.[7] Social relationships scored second-highest, highlighting the supportive nature of Iraqi society. However, general QOL was poor, contrasting with Nunes A.'s findings in Brazil (2009), likely due to differences in treatment capabilities and rehabilitation programs.[9] Hemophilia B patients reported worse QOL scores across all domains than Hemophilia A patients, consistent with Varaklioti A.'s findings in Greece (2014), possibly due to limited access to recombinant factor IX treatment.[13] Patients with severe hemophilia experienced more impaired physical and environmental health, primarily due to increased bleeding episodes and financial and social burdens. This aligns with Sarper N.'s (2010) findings but diverges from the Hemo-QOL study in Europe.[10,17] Patients with inhibitors (20 documented cases) had worse outcomes across all domains due to resistance to

recombinant factor therapy, consistent with Ferreira A.'s (2013) study.[7] Serious bleeding episodes and target joints were common among patients, significantly affecting all QOL domains. The knee was the most frequently affected joint, while elbow joint issues led to the worst physical health scores due to their impact on mobility and upper-limb tasks.[18] Rehabilitation programs need improvement to address these challenges effectively.

CONCLUSION

The overall QOL was poor, mainly in the physical domain, and also for the social and psychological domains. With increasing age, the overall QOL was poor, this mainly due to limited treatment in the past and less access to hematologist. Most of the patients had target joints and this reflects limitations in rehabilitation programs and treatment. There was a significant proportion of patients who had negative feelings and despair among the sample.

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