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Health Related Quality of Life (HRQoL) in patients with Hemophilia

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Abstract: ***Background:****Besides striving to achieve greater strides in treating Hemophilia, the importance of assessing patient's perspective with Hemophilia is the need of the hour.*
Aim:*A study to assess the Health Related Quality of Lifeamong patients with Hemophilia in the Hematology Outpatient Department of Christian Medical College, Vellore*
Methodology: *A Non experimental descriptive design was undertaken. A total of 120 subjects with Hemophilia aged between 18-59 years were selected based on total enumeration sampling technique. The HRQoL was assessed using 'A36 Hemophilia-QoL'questionnaire. Descriptive and inferential non parametric statistics such as frequency distributions, mean, standard deviation, ANOVA, Independent't' test were used in this study*
Results: *83.3% of the population had Hemophilia 'A'. 87.5% had severe Hemophilia. 87.5% of the population had joint bleed. 74.2% had knee as a target joint. Range of motion was predominantly impaired in knee(75.8%) and elbow(60.8%) joints. Administration of factor concentrates was the primary treatment option for 72.5%. 94.2% reported that the treatment was available. The overall score of HRQoLin Hemophilia is moderate(67.98). HRQoL is significantly better in Hemophilia patients not having knee(p-0.000**)** and elbow(p-0.001) as target joints, absence of joint swelling (p-0.000)and joint pain (p-0.000) and when range of motion is unimpairedin ankle (p-0.018), knee (p-0.000) and elbow (p-0.000) joints*
Conclusion:*The study reveals that there is moderate HRQoLin patients with Hemophilia. This warrants for further explorationand development of strategies focusing on specific QOL domainsinorderto improve the HRQoL in patients with hemophilia*
Key variables: *HRQoL, Patients with Hemophilia*

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I. Introduction

Hemophilia is an X-linked congenital bleeding disorder caused by a deficiency of coagulation factor VIII (Hemophilia A) or factor IX (Hemophilia B) which is the result of mutations of the respective clotting factor genes. Hemophilia has an estimated frequency of approximately one in 10,000 births. India harbours the second highest number of global patients with HemophiliaA(Kar et al., 2014). Hemophilia is characterized by spontaneous and post-traumatic bleeding complications in joints and muscles leading almost inevitably to pain, severe joint damage, disability and a dramatic impairment of quality of life. It is obvious that the availability of safer products and early prophylaxis have greatly improved the management of haemophilic patients with a consequent dramatic impact not only on symptoms and survival of these patients, but also on their quality of life. Besides evolution of advanced treatment strategies in treating Hemophilia, the importance of assessing patient's perspective on his quality of life with Hemophilia is crucial(Knobe&Berntorp, 2011). Assessment of HRQoLas a patient-reported outcomebrings out the individual experience and perception of illness/health including psychosocial response todisease & treatment related symptoms. It also helps to evaluate the quality of care rendered and the need for modifications in interventions while caring for patients with Hemophilia(Gringeri A, Von Mackensen S., 2008). QoL is a recent focus of research in Hemophilia. In the guidelines for the management of Hemophilia by the World Federation of Hemophilia, QoL is considered as one of the outcomes of treatment which should be monitored every 6 to12 months (Srivastava et al., 2013)

II. Objectives Of The Study

1.To assess the overallHRQoL in patients with Hemophilia
2. To assess the HRQoL in various domains in patients withHemophilia
3.Tocompare and identify the difference inHRQoLwith selecteddemographicand clinical variables

III. Hypotheses

1. The HRQoL is significantly affected in patients with Hemophilia
2. There is a significant relationship between HRQoL with demographic and clinical variables in patients with Hemophilia

IV. Methodology

Approach and design: Quantitative non experimental approach and descriptive design was used.
Setting: The study was conducted in the Hemophilia clinic of Hematology Department, Christian Medical College,Vellore.
Sampling: A total of 120 subjects with Hemophilia who fulfilled the study criteria, were selected using total enumeration sampling technique

V. Data Collection Instrument

The data collection instrument was a structured questionnaire with 3 parts:

Part A: Demographic variables
Assesses thebaseline demographic characteristics of the subjects.It includes age, gender, religion, location, marital status, education, employment status, insurance, personal income and family income

Part B: Clinical variables
Assesses age at diagnosis, type, severity, joint involvement, site of bleed, range of motion limitation, pain, treatment options, availability of factor concentrates, co-morbidities, body mass index and viral status

Part C:A36 Hemophilia-QoLquestionnaire
'A36 Hemophilia-QoL' is a 36 item standardised questionnaire to assess the HRQoLin patients with Hemophilia. It includes 9 domains: physical health, daily activities, joint damage, pain, treatment satisfaction, treatment difficulties, emotional functioning, mental health, relationships and social activity. The response options are in the form of 5 point Likert scale ranging from 0 to 4. Minimum score possible is 0 and the maximum score is 144. Higher the score, better the quality of life

Reliability & validity:
It has an internal consistency reliability score of 0.95 (Cronbach's alpha coefficient) and test retest reliability score of 0.92($p<0.001$). Numerous studies using A36 Hemophilia-QoL internationally supported high validity and reliability (Remor E, Et al, 2005)

VI. Results

a**. Demographic variables:**

52.5% of the population was between 18-29 years. 97.5% of them were males. 83.3% were Hindus. 55% were from the rural areas. 67.5% were single. 40.8% were graduates. 53.33% were unemployed. Majority of them (80.33%) had no insurance coverage for treatment. 42.5% of them had no personal income.45.9% had a family income between Rs.5000-Rs.15,000

b. **Clinical variables:**

83.3% of the population had Hemophilia'A'. The median age at diagnosis of Hemophilia is 2 years (0.6,6.5).Majority of them (87.5%) had severe Hemophilia. 56.7% had a family history of Hemophilia.74.2% of them had knee, 33.6% had elbow, 16.7% had ankle and 9.2 % had shoulder as a target joint. 89.2% had joint involvement. 80% had joint swelling. 45.8% had impaired range of motion of the ankle. Range of motion was predominantly impaired in knee(75.8%) and elbow(60.8%) joints. 41.7% had soft tissue bleed. 87.5% of the population had joint bleed. Administration of factor concentrates was the primary treatment option (72.5%) followed by combination of factor concentrates and blood products administration (20.8%), use of inhibitors alone (1.7%), factor concentrates and inhibitors (1.7%), factor concentrates and surgery (0.8%). 94.2% reported that the treatment was available.91.7% of the population had no associated co-morbidities. 57.5% of them reported pain mainly over the knee(39.4%), knee & elbow together (16.7%) and elbow (12.1%). The mean height, weight and BMI was 167.5± 8.6, 62.9±13.2 and 22.5±8.59 respectively. 56.7% of them had normal BMI. 0.8% of the population was found to have HIV and Hepatitis B positive status. 2.5% of the population had Hepatitis C positive status

c. **Health Related Quality of Life**

Table 1:*HRQoL in patients with Hemophilia (N=120)*

Sl. No.	HRQoL Domains	Mean HRQoL score	Confidence Interval (95%)	Minimum to maximum possible score
1.	Physical health	15.82	14.56- 17.09	0-32
2.	Daily activities	6.83	6.15- 7.50	0-16
3.	Joint damage	5.09	4.57-5.61	0-12
4.	Pain	3.57	3.17-3.96	0-8
5.	Treatment satisfaction	5.15	4.86-5.44	0-8
6.	Treatment difficulties	9.2	8.79-9.74	0-16
7.	Emotional functioning	5.61	5.32-5.92	0-20
8.	Mental health	6.19	8.79-9.74	0-12
9.	Relationships & social activity	10.44	9.45-11.43	0-20
	Overall HRQoL	67.98	63.80-72.15	0-144

Table 1 reveals that the mean totalHRQoLscore in patients with Hemophilia is moderate(67.98). There is moderate HRQoLin domains such as Physical health(15.82), Daily activities (6.83), Joint damage (5.09), Pain (3.57%), Treatment satisfaction (5.15), Treatment difficulties (9.2), Mental health (6.19), Relationships & social activity (10.44) and poor HRQoLscore is noted in the Emotional functioning domain (5.61)

d. Comparison of HRQoL with selected demographic and clinical variables

Table 2:*Comparison of HRQoL means with selected demographic variables*

Sl. No.	**Demographic variables**	**Overall HRQoL Mean ± Standard Deviation**	**t/F value**	**p value**
1.	Age		0.470*	0.639
	18-29 years	68.92±24.82		
	30-59 years	66.93±21.19		
2.	Marital status		0.979*	0.329
	Married	65.00±22.76		
	Single	69.41±23.25		
3.	Employment status		0.730*	0.466
	Employed	69.63±22.18		
	Unemployed	66.53±23.94		
4.	Location		0.692*	0.489
	Urban	69.59±21.77		
	Rural	66.65±24.20		
5.	Insurance		0.636*	0.525
	Yes	70.74±26.94		
	No	67.32±22.19		
6.	Education		0.800**	0.598
	Primary	57.60±26.33		
	Secondary	69.18±23.36		
	Higher secondary	64.16±21.93		
	Graduate	69.94±24.66		
	Post graduate	70.65±18.06		
7.	Personal income		1.040**	0.461
	Nil	64.47±25.06		
	<5000	67.59±19.17		
	5000-15000	69.59±21.18		
	15000-30000	83.60±16.15		
	>30000	72.77±26.62		
8.	Family income		0.540**	0.841
	<5000	64.58±25.43		
	5000-15000	66.80±22.87		
	15000-30000	71.41±21.00		
	>30000	71.68±23.81		

* t test, **ANOVA

Table 2 reveals that there is no statistical significance but the mean HRQoL is higher in age group between 18-29 years (68.92±24.82) compared to 30-59 years. Mean HRQoL is higher in unmarried male (69.41±23.25) than who were married, employed Hemophilia patients have better HRQoL (69.63±22.18) than unemployed patients.Mean HRQoL was better among the urban patients (69.59±21.77) than the rural counterparts. Hemophilia patients having medical insurance coverage have better HRQoLscore(70.74±26.94)

than those who do not have. The mean HRQoL score increased with higher educational status (70.65±18.06). Haemophilia patients with higher personal income (72.77±26.62) and family income (71.68±23.81) have better HRQoL than those who have lesser income

Table 3:*Comparison of HRQoL means with selected clinical variables*

Sl. No.	Variables	Overall HRQoL Mean ± Standard Deviation	t value	p value
1.	Type Hemophilia A Hemophilia B	 66.62±22.70 74.75±24.41	1.443	0.151
2.	Family history Present Absent	 71.35±24.12 63.56±21.08	1.851	0.066
3.	Target joint-ankle Yes No	 59.00±19.90 69.77±23.35	1.925	0.056
4.	Target joint-knee Yes No	 62.20±19.43 84.55±24.99	5.105	**0.000****
5.	Target joint –elbow Yes No	 58.75±17.45 72.59±24.25	3.213	**0.001****
6.	Joint swelling Yes No	 63.83±19.95 84.54±27.49	4.194	**0.000****
7.	Impaired ROM –Ankle Yes No	 62.62±19.35 72.51±25.10	2.383	**0.018***
8.	Impaired ROM-Knee Yes No	 63.29±20.08 82.69±25.95	4.208	**0.000****
9.	Impaired ROM- Elbow Yes No	 61.62±19.43 77.85±24.99	3.987	**0.000****
10.	Pain Present Absent	 61.12±18.74 77.25±25.27	4.018	**0.000****

*p<0.05 **p<0.005

Table 3 depicts that HRQoL is significantly better in Hemophilia patients not having knee (p-0.000**)** and elbow(p-0.001) as target joints, absence of joint swelling (p-0.000) and joint pain (p-0.000) and when the range of motion is unimpaired in ankle (p-0.018), knee (p-0.000) and elbow (p-0.000) joints. Patients withHemophilia 'B' (74.75± 24.41) had better HRQoLthan in Hemophilia 'A', patients with a family history of Hemophilia had higher HRQoL (71.35± 24.12). Mean HRQoL was better in those not having ankle as a target joint (69.77± 23.35)

VII. Discussion

The study reveals that majority of the population had Hemophilia 'A' (83.3%) and severe Hemophilia (87.5%). 56.7% had a family history of Hemophilia. These findings were congruent with studies done in Brazil and India, which revealed higher prevalence of Hemophilia 'A'(84.6% and 88.3% respectively) in its severe form(80%) and with a positivefamily history (58.4%) (Adriana Aparecida Ferreira et al.,2013, Denise Rodrigues Holsbach, et al, 2016&Saurabh Mishra, et al, 2016)

The present studyalso shows remarkable physical complaints, where majority of the population had joint involvement (89.2%), joint swelling (80%), joint bleed (87.5%) and knee as a target joint (74.2%). Nearly half of them had impaired range of motion of the ankle (45.8%) and soft tissue bleed (41.7%).0.8% of the population was found to have HIV and Hepatitis B positive status. 2.5% of the population had Hepatitis C positive status.This is in line with an Indian study done by Saurabh Mishra (2016) in Lucknow among 71 Hemophilia patients whichrevealed that joint involvement (77.9%), joint swelling (76.6%) and joint bleed (15.6%) resulted in compromised joint movement. Knee joint was observed to be as the target joint among 57.1% of the patients. Soft tissue bleed occurred in 62.3%. Hepatitis B and C infection were observed in 6.5% and 9.1% of the patients, respectively. Similarly in a Brazilian study, viral infections were more prevalent in those who had Hemophilia where Hepatitis C was the most frequent infection (Denise Rodrigues Holsbach, et al, 2016)

The study reports a moderate HRQoL score of 67.98.This finding is congruent with a study done in Iran assessing the QoL of 100 adults with Hemophilia, where the total QoL score was 71.88 (Dolatkhah et al.,2014). The reason for moderate HRQoL score could be attributed to the unmet needs ofHemophilia patients specific to emotional and psychosocial aspects, chronic morbidity presenting with constant pain, joint damage & disability and frequent visits to the hospital for factor replacements and other treatment. Contradicting to the HRQoL scores in the present study, poor HRQoL scores were seen in studies done in India among 51 adult Hemophilia patients(47.4 ± 14.), Turkey assessing 31 adult patients (47.4 ± 14) and in Brazil studying 39 Hemophilia patients (35.55) (ArzuMercan, 2010, Adriana Aparecida Ferreira et al.,2013 & Saurabh Mishra, et al, 2016). In these studies the decline in HRQoL score was influenced by the presence of arthropathy and infectious diseases transmitted by blood products

There is moderate HRQoL in domains such as Physical health (15.82), Daily activities (6.83), Joint damage (5.09), Pain (3.57%), Treatment satisfaction (5.15), Treatment difficulties (9.2), Mental health (6.19), Relationships& social activity (10.44).Poor HRQoL score is noted in the Emotional functioning domain (5.61).This emphasizes on optimizing the care in meeting the emotional needs of the patients, which requires a specially trained professional in the field who could be a part of the Hemophilia management team. In a Brazilian study, 'Sports and leisure' and 'Physical health' were the most impaired dimensions and the dimension 'Relationship and partners' was the least impaired (Adriana Aparecida Ferreira et al.,2013).In an Indian study, the highest impairments were found in dimensions such as 'physical activity & leisure', 'physical health' and 'view'(Saurabh Mishra, et al, 2016)

Mean HRQoL is higher in single (69.41±23.25), urban patients (69.59± 21.77) and in those with higher educational status (70.65± 18.06).This could be due to better access to treatment centers and support services among the educated urban population. Contrarily, in a similar study in Iran, the QoL was very poor in Hemophilia patients who lived in urban area (p-0.013) but as in the present study, here too single patients had a better QoL than married patients (p-0.155). Low education and lack of awareness of the diseases among patients with Hemophilia led to reduced QoL and more disease complications (Dolaketh et al., 2014)

The present study revealed that HRQoL was significantly better in unimpaired ROM of joints [ankle (p-0.018), knee (p-0.000) and elbow (p-0.000)], absence of target joint (p-0.001), joint swelling (p-0.000) and joint pain (p-0.000). This is in line with a study done in the US, where patients with more severe Hemophilia and higher self-reported joint pain and motion limitation had poorer scores, particularly in the physical aspects of HRQoL. In adults, significant correlations ($p < 0.01$) were found between ROM measures and self-reported measures (Poon et al,2012). This stresses the need for appropriate musculoskeletal assessment and prompt treatment in managing arthropathyand pain

VIII. Nursing Implications of the Study

The following nursing and collaborative interventions can be implemented to improve the QoL in patients with Hemophilia:

- Focus on interventions to reduce joint swelling,arthropathy and reducing range of motion limitation of ankle, knee and elbow joints
- Pain reduction strategies, assistance in daily activities, improvement of physical and mental health
- Paying keen attention to emotional functioningamong patients with Hemophilia by means of a 'Hemophilia-Nurse Counsellor' is essential

IX. Recommendations

- A long term study with large samples can be done to assess the HRQoL in Hemophilia patients
- A qualitative study can help to further study the perception of HRQoL in Hemophilia patients
- A study can be done on the perception of Health care providers on Hemophilia
- A similar comparative study can be conducted to assess HRQoL in Hemophilia as well as healthy adults

X. Conclusion

The study has thrown light into the unexplored Indian patient's perspectives with Hemophilia. Thestudy reveals moderate HRQoL among patients with Hemophilia. Specific treatment options aimed to manage physical symptoms as well as psychosocial issues is of prime importance. This poses a great challenge for the health care team to address the issues hindering QoL in order to provide a patient-centered holistic care inHemophilia

CONFLICTS OF INTEREST

The authors have declared no conflicts of interest.

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