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Quality of Life of People with Haemophilia of Selected Districts, Nepal

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Abstract

Background: Hemophilia is an inherited, lifelong, sex-linked bleeding disorders predominantly occurring in males. People with bleeding disorder face life-long physical, psychological, financial and employment challenges. This study aims to identify the quality of life of people with Hemophilia, who are clinically diagnosed with either Hemophilia A or B.

Methods and materials: A descriptive survey design was conducted among 16 years and above male hemophiliac of Bara and Parsa districts, Nepal. Altogether 61 respondents were selected by using convenient sampling technique. A valid tool (Haemo-A-QoL) after getting approval from Dr Sylvia von Mackenson was translated in Nepali and Bhojpuri language and was administered to collect the data. Data processing was done using a computer (SPSS version 20).

Results: This study showed that the maximum number of respondents was of age group 16-20 (32.8%) and mean age group and standard deviation were 21-25 and 1.382 respectively. Out of 61 respondents 86.1% of the respondents had hemophilia A and 13.9% had hemophilia B. Among them 75.4% respondent know their factor activity level and of which 24.6% had severe, 26.2% had moderate form and 24.6% had mild form of hemophilia. The average Haemo-A-QoL total mean score was 32.72.

Conclusion: The dimension “Physical Health”, Feelings”, “Sports and leisure”, has the highest averages (72.76, 62.09 and 68.5 respectively) indicating poorer quality of life whereas the dimension “future” and “partnership and sexuality” was least impaired dimension between the respondents.

Keywords: Quality of life; Hemophilia; Sex-linked bleeding disorders; Physical health

Introduction

Globally, it is estimated that there are 400,000 people suffering from hemophilia. In 2016, total people with hemophilia were 184,723 among which 1,49,764 were diagnosed with Hemophilia A and 29,712 were hemophilia B among 113 countries. There were 12,996 people with hemophilia A and 3,953 with hemophilia B in USA, 6,559 and 1,472 in UK, 12,533 and 3,953 in China, 15,218 and 2,379 in India and 500 and 73 in Nepal respectively [1].

Hemophilia is a rare genetic disease that results from mutations in the genes that code for proteins necessary for normal blood clotting, called coagulation factors. There is a wide variation in the reported prevalence of hemophilia across countries [2].

Hemophilia A (deficiency in factor [F] VIII) and hemophilia B (deficiency in FIX) are the most common serious congenital coagulation factor deficiencies. Hemophilia A is common, occurs in 1:5000 male births, whereas hemophilia B occurs in 1:30,000 male births. Hemophilia is found in all ethnic groups; there is no geographic or racial predilection [3].

Hemophilia is usually innate. Genes transmit messages regarding the way the body’s cells will build up as a child grows into an adult. Occasionally hemophilia can happen when there is no family history of it. This is called sporadic hemophilia. About 30% of hemophilic people did not get it through their parent’s genes. It was caused by a change in the person’s own genes [4].

Severity of the illness is linked with the quantity levels of factors VIII and IX in the patients’ plasma; hemophilia is described as severe, moderate, and mild, when the residual plasma activity of the clotting factor is <1%, between 1% and 5%, and between 5% and 40%, respectively [5].

Hemophilic patient either bleed spontaneously or on trauma. About 75% of all bleeding in patients with hemophilia occurs into joints. Recurrent joint hemorrhages can result in damage so patient with severe factor deficiency can become crippled by the joint damage before they become adults. Bleeding also occurs in muscle, subcutaneous tissues resulting in hematomas. Spontaneous hemorrhage and GI bleeding can occur, and the most dangerous hemorrhage is intracranial or extracranial [6].

Quality of Life is an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns. It is a broad-ranging concept affected in a complex way by the person's physical health, psychological state, personal beliefs, social relationships and their relationship to salient features of their environment. Quality of life is defined as an all-inclusive concept incorporating all factors that impact upon individual’s lives [7].

A study conducted in National Center for Congenital Bleeding Disorder in Laiko general hospital of Athens showed that the predominant form of hemophilia was A (78%) and 63.6% of the individuals have severe hemophilia. Most of the patients suffered from hemarthrosis (78%), muscle hematomas (89.8%) and other types of bleeding (74.6%). Regarding, viral infections, 84 patients (71.2%) were HCV positive and 33 patients (28.0%) were HIV positive [8].

There is currently no cure for hemophilia. However, treatment has advanced remarkably in the past 30 years. Children with hemophilia who receive comprehensive treatment can now look forward to a near-normal life expectancy. For individuals with mild hemophilia A, the Medical and Scientific Advisory Council (MASAC) of the National Hemophilia Foundation recommends that desmopressin (DDAVP) should be used whenever possible. DDAVP is available in both an injectable form (DDAVP Injection) and as a highly concentrated nasal spray [9].

A Study in India showed that the Quality of life was poor both among children affected by hemophilia and their parents. Perceived impact on family, poor physical health and inability to participate in school/sports activities are the major contributors to poor QoL [10].

A cross-sectional study in Nepal showed that the dimension "view of yourself", "treatment", "physical health" had the highest averages indicating poorer quality of life whereas the dimension "family planning and partnership were the least impaired dimensions among the respondents [11].

Similarly, the conditions of a patient with hemophilia are also poor in Nepal. Nepal Hemophilia Society attempts to deliver services but still, the treatment of the patient with hemophilia is a challenge in Nepal due to the exorbitant price of factor concentrates and its unavailability. Lack of awareness regarding Hemophilia, few diagnostic centers, unidentified cases, few hemophilia care centers made treatment of hemophilia more difficult in Nepal. There are very few researches carried out in hemophilia as well as research related to the quality of life of people with hemophilia. This study aimed to measure the quality of life of people with hemophilia of Bara and Parsa district which would be the first step towards comprehensive patient care as well as cost-effective measure by preventing co-morbidities and complications. So, researcher is interested to study the quality of life of people with hemophilia in Bara and Parsa districts of Nepal examining how patients perceive and experience these manifestations in their daily life. The findings of the study might be helpful to give baseline data regarding the quality of life of people with hemophilia. Also, this study will be helpful to health care planner, health care provider, health researcher, as well as policymakers to plan program in improving and maintaining the quality of life of the patient with hemophilia.

Materials and Methods

A descriptive survey design was used to find out the quality of life of people with Hemophilia in selected districts. All hemophilic patient of Bara and Parsa district of Province 2 of Nepal who was 16 years and above was selected for the study. There were total of 137 people with hemophilia in Bara and Parsa districts among them 61 respondents were selected. The convenient sampling method was used to select the samples.

The sample size was calculated at 95% confidence level and 5% confidence interval.

Level of significance (α) assumed 5%, $Z=Z_{5\%}=1.96$,

E stand for difference of sample proportion and population proportion of prevalence=error=10%=0.1

p=0.8(proportion of characteristics) [11]

q=1-0.8=0.2

The sample size was calculated by using formula,

$$n = PQ\left(\frac{Z\alpha}{E}\right)^2$$

$$= PQ\left(\frac{Z_{5\%}}{E}\right)^2$$

$$= 0.8\times 0.2\left(\frac{1.96}{0.1}\right)^2$$

$$= 61$$

All hemophiliac patients who are willing to participate and who gives consent were selected for the study.

Hemophilia-Specific Quality of Life questionnaire for adult i.e., Haemo-A-QoL, likert scale was used after getting permission from Haemo-QoL Group. The English version of Haemo-QoL Group was translated to Nepali language and Bhojpuri language before distribution.

Pretesting was done on 10% of the total sample i.e., 6 respondents and necessary modification of the instrument was done, and those respondents were excluded during data collection.

The study was conducted only after the approval of research committee of campus. Prior to data collection written permission was taken from concerned organization i.e., Nepal Hemophilia Society, Parsa, Nepal. Informed consent was taken from respondents to ensure the right of the respondents. Confidentiality of all the participants was maintained.

The researcher introduced herself to the respondents ascertain their cooperation for the study. The verbal informed consent was taken from each respondent prior to data collection. Data was collected by the researcher in the morning and evening time through a household survey by using a semi-structured interview schedule. Each respondent was interviewed for 15-20 minutes accordingly, 5-6 sample were collected every day and total sample collection was completed within 2 weeks.

All collected data were reviewed and checked for its completeness, consistency and accuracy. The collected data were coded and entered in SPSS version 20. Data were summarized using descriptive statistics like frequency, percentage, mean and standard deviation and inferential statistics.

Results

Characteristics	Frequency	Percentage
Age Group		
16-20	20	32.8
21-25	16	26.2
26-30	14	23
31-35	6	9.8

36-40	2	3.3
41-45	3	4.9
Marital Status		
Married	19	32.8
Unmarried	41	65.6
Widower	1	1.6
Educational Status		
Primary	8	13.1
Secondary	24	39.3
Higher Secondary	9	14.8
Bachelors	17	27.9
Master's and Above	3	4.9
Occupation		
Service	18	29.5
Business	18	29.5
Agriculture	12	19.7
Student	13	21.3
Religion		
Hindu	54	88.5
Buddhist	3	4.9
Islam	4	6.6
Ethnicity		
Dalit	1	1.6
Madhesi	32	52.5
Janajati	16	26.2
Muslim	4	6.6
Brahmin/Chettri	8	13.1
Economic Status		
Able to Fulfill basic needs for a year	21	34.4
Can save extra even after a year	39	63.9
Not enough even for a year	1	1.6

Table 1: Socio-Demographic Information of the respondents (n=61). Mean age group was 21-25 years and SD =1.382.

Table 1 shows that the maximum number of respondents were of age group 16-20 (32.8%) and minimum from 36-40 (3.3%). About 41(65.6%) of the respondents were unmarried and 1 (1.6%) respondent was a widower. All the respondents were literate among them, the maximum of the respondents 24 (39.3%) had completed secondary educational status. Services and Business were the occupation maximum respondents were engaged i.e., 18 (29.5%). 54 (88.5%) of the respondents followed Hindu religion and a maximum of the respondents belong to ethnic group Madhesi i.e., 32(52.5%). 39 (63.9%) of the respondents can save extra money even after a year.

Characteristics	Frequency	Percentage
Age at diagnosis		
Below 1 year	4	6.6
1-5 year	45	73.8
6-10	7	11.5
11-15	5	8.2
Types of Hemophilia		
Hemophilia A	53	86.1
Hemophilia B	8	13.1
Level of factor activity		
Below 1%	15	24.6
1-5%	16	26.2
6-40%	15	24.6
Unknown	15	24.6
Presence of Inhibitor		
Yes	1	1.6
No	15	24.6
Unknown	45	73.8
Age of first bleeding episode in joints		
1-5	42	68.9
6-10	12	19.7
11-15	7	11.5
Number of joint bleeding		
Less than 3 episodes	9	14.8
Multiple episodes	52	85.2
Joint Impairments		
Yes	34	55.7
No	27	44.3
Mode of Treatment		
Conservative Treatment	9	14.8
Prophylactic	12	19.7
On demand	40	65.6

Table 2: Information Related to Disease Condition and Treatment (n-61).

Table 2 depicts that the majority (73.8%) of the respondents, were diagnosed with hemophilia at the age of 1-5 years. Out of 61

respondent's 86.1% of the respondents had hemophilia A and majority of the respondent's i.e., 26.2% had a mild form of Hemophilia. The majority of the respondents (68.9%) had joint bleeding at the age of 1-5 years. In addition to that 85.2% of the respondent had multiple episodes of bleeding in joint and 55.7% had a joint impairment. Maximum (65.6%) of the respondents treat their hemophilia according to demand.

Haemo-A-QoL information

Physical Health characteristics	Never	Rarely	Sometimes	Often	All the time
Swelling in the joints or muscle	3 (4.9%)	3 (4.9%)	30 (49.2%)	25 (41%)	0
Pain in my joints	2 (3.3%)	3 (4.9%)	35 (57.4%)	21(34.4%)	0
Painful for me to move	2 (3.3%)	4 (6.6%)	34 (55.7%)	21(34.4%)	0
Difficulties in walking as far as wanted	1 (1.6%)	4 (6.6%)	31 (50.8%)	25 (41%)	0
Needed more time to prepare myself	0	4 (6.6%)	26 (42.6%)	31 (50.8%)	0
Sports and Leisure					
I had avoided sports that I like because of hemophilia	6 (9.8%)	2 (3.3%)	2 (3.3%)	23 (37.7%)	28 (45.9%)
I had to avoid sports like football	4 (6.6%)	1 (1.6%)	2 (3.3%)	25 (41%)	29 (47.5%)
I did as much as sports as others	21 (34.4%)	35 (57.4%)	1 (1.6%)	2 (3.3%)	2 (3.3%)
I didn't have freedom to travel where I wanted	2 (3.3%)	48 (78.7%)	9 (14.8%)	1 (1.6%)	1 (1.6%)
It was necessary for me plan everything in advance	0	0	7 (11.5%)	53 (86.9%)	1 (1.6%)

Table 3: Physical Quality of Life related information (n=61).

Table 3 shows the Physical quality of life of the respondents with Hemophilia. About 35 (57.4%) of the respondents complained about the pain in their joints sometimes during the past 4 weeks. Besides, 48 (78.7%) of the respondents stayed at their home because of hemophilia which restricted their freedom to travel where they wanted.

Feelings Characteristics	**Never**	**Rarely**	**Sometimes**	**Often**	**All the time**
Hemophilia was burden for me	2 (3.3%)	4 (6.6%)	25 (41%)	29 (47.5%)	1 (1.6%)
Hemophilia made me angry	0	5 (8.2%)	32 (52.5%)	24 (39.3%)	0
Was worried because of Hemophilia	0	4 (6.6%)	26 (42.6%)	31(50.8%)	0
Was felt excluded	0	0	12 (19.7%)	47 (77%)	2 (3.3%)
Opinion about yourself					
Wanted to be healthy like other people	0	2 (3.3%)	1 (1.6%)	19 (31.1%)	39 (63.9%)
Felt satisfied about my body	5 (8.2%)	43 (70.5%)	12 (19.7%)	0	1 (1.6%)
Hemophilia made my life more difficult	0	2 (3.3%)	40 (65.6%)	13 (21.3%)	6 (9.8%)
Felt different from others because of hemophilia	0	2 (3.3%)	34 (55.7%)	20 (32.8%)	5 (8.2%)
I managed not to think about my hemophilia	0	1 (1.6%)	34 (55.7%)	22 (36.1%)	4 (6.6%)
Future					
Difficult to lead a normal life	1 (1.6%)	2 (3.3%)	34 (55.7%)	22 (36.1%)	2 (3.3%)
Things will get better in the future	5 (8.2%)	36 (59%)	15 (24.6%)	5 (8.2%)	0
Worried about the worsening condition	0	5 (8.2%)	35 (57.4%)	19 (31.1%)	2 (3.3%)
Life plans are influenced by Hemophilia	0	1 (1.6%)	36 (59%)	19 (31.1%)	5 (8.2%)

Afraid that I will need a wheel chair	1 (1.6%)	2 (3.3%)	40 (65.6%)	15 (24.6%)	3 (4.9%)

Table 4: Psychological Quality of Life related information (n=61).

Table 4 reveals that 77% of the respondents felt excluded due to hemophilia. Approximately 65.6% of the respondent's life was difficult due to hemophilia and 65.6% of the respondents were afraid that they will need a wheelchair in future.

Characteristics	Never	Rarely	Sometimes	Often	All the time	Not applicable
Work and Studies						
Able to go to work/college regularly	1 (1.6%)	6 (9.8%)	17 (27.9%)	37 (60.7%)	0	
Able to work/study like healthy colleagues	1 (1.6%)	14 (23%)	12 (19.7%)	34 (55.7%)	0	
Everyday activities were endangered by Hemophilia	0	7 (11.5%)	30 (49.2%)	23 (37.7%)	1 (1.6%)	
Difficulties in paying attention at work/college because of pain	0	4 (6.6%)	32 (52.5%)	25 (41%)	0	
Planning of Family						
Problems having children	7 (11.5%)	12 (19.7%)	0	0	0	42 (68.9%)
Afraid of not having children	12 (19.7%)	6 (9.8%)	1 (1.6%)	0	0	42 (68.9%)
Afraid not to be able to take care of children	1 (1.6%)	0	4 (6.6%)	12 (19.7%)	2 (3.3%)	42 (68.9%)
Worry not to be able to raise a family	1 (1.6%)	0	4 (6.6%)	12 (19.7%)	2 (3.3%)	42 (68.9%)
Partnership and Sexuality						
Finding difficult to date because of Hemophilia	35 (57.4%)	22 (36.1%)	3 (4.9%)	1 (1.6%)	0	
Uncertain in relationship with women	36 (59%)	20 (32.8%)	0	4 (6.6%)	1 (1.6%)	
Can't have a normal relationship	26 (42.6%)	23 (37.7%)	11 (18%)	1 (1.6%)	0	

Table 5: Social Quality of Life related information (n=61).

Table 5 shows that 60.7% of the respondents were able to go to work/ school regularly. Majority of the respondents i.e., 19.7% worry sometimes not to be able to raise a family.

Dealing with Hemophilia	Never	Rarely	Sometimes	Often	All the time
Recognize timely when blood loss started to happen	1 (1.6%)	25 (41%)	28 (45.9%)	7 (11.5%)	0
Able to tell whether bleeding or not	1 (1.6%)	30 (49.2%)	22 (36.1%)	8 (13.1%)	0
Was able to control bleeding	2 (3.3%)	42 (68.9%)	11 (18%)	6 (9.8%)	0
Treatment					
Was dependent on factor concentrate	1 (1.6%)	1 (1.6%)	12 (19.7%)	42 (68.9%)	5 (8.2%)
Dependent on doctors for treatment	0	39 (63.9%)	16 (26.2%)	6 (9.8%)	0

Annoyed about the amount of time spent having the injection	0	30 (49.2%)	16 (26.2%)	14 (23%)	1 (1.6%)
Felt interrupted in daily activities by the injection	1 (1.6%)	23 (37.7%)	24 (39.3%)	13 (21.3%)	0
Afraid of complications	0	2 (3.3%)	10 (16.4%)	31 (50.8%)	18 (29.5%)
Problems with how treatment was administered	0	8 (13.1%)	20 (32.8%)	26 (42.6%)	7 (11.5%)
Afraid that in case of emergency other doctors don't know how to treat	0	0	9 (14.8%)	31 (50.8%)	21 (34.4%)
Was satisfied with Hemophilia center	0	0	0	0	61 (100%)

Table 6: Dealing with Hemophilia and Treatment (n=61).

Table 6 shows that the majority of people are able to recognize and tell and tell if there is any internal bleeding into joints i.e., 45.9% and 49.2% respectively. Maximum of the respondents depend on factor concentrate (68.9%).

Characteristics	Mean	Standard Deviation
Physical Health	72.7617	22.88699
Feeling	62.0902	24.99658
Opinion About Yourself	43.7158	27.73099
Sports and Leisure	68.5544	20.23102
Work and Studies	38.0328	23.15309
Dealing with Haemophilia	57.377	32.84709
Treatment	40.1639	23.33445
Future	34.0619	22.75915
Planning Your Family	36.4299	56.33275
Partnership and Sexuality	20.9472	21.95191

Table 7: Mean score of Haemo-A-QoL questionnaire in total and by dimension (n=61).

Table 7 reveals that the average Hemo-A-QoL total mean score was 32.72. The dimension "Physical Health", Feelings", "Sports and leisure", has the highest averages (72.76, 62.09 and 68.5 respectively) indicating poorer quality of life whereas the dimension "future" and "partnership and sexuality" were the least impaired dimension among the respondents.

Severity	Frequency	Percent
Unknown	15	24.6
Mild	15	24.6
Moderate	16	26.2
Severe	15	24.6
Total	61	100

Table 8: Severity of Hemophilia (n=61).

Table 8 indicates that 75.4% respondents had known their severity of hemophilia and among them, the maximum of the respondents had a moderate form of hemophilia (26.2%).

Characteristics	Severe	Moderate	Mild
Physical Health	54.57 ± 18.52	35.51 ± 22.01	28.52 ± 15.83
Feeling	40.52 ± 21.36	42.17 ± 13.01	37.22 ± 15.63
Opinion About Yourself	53.19 ± 11.08	43.14 ± 10.24	48 ± 9.07
Sports and Leisure	28.92 ± 12.53	35.12 ± 18.36	39.57 ± 21.91
Work and Studies	43.4 ± 16.72	23.03 ± 16.43	25.42 ± 13.29
Dealing with Haemophilia	38 ± 13.5	44.35 ± 20.51	23.42 ± 13.85
Treatment	48.6 ± 17.01	43.32 ± 16.53	40.31 ± 12.29
Future	47.69 ± 19.03	38.49 ± 15.52	39.56 ± 16.48
Planning Your Family	14.08 ± 9.18	10.52 ± 8.7	15.93 ± 8.8
Partnership and Sexuality	21.84 ± 14.32	7.7 ± 4.32	18.94 ± 30.12
Total	38.67 ± 14.21	33.16 ± 17.52	27.34 ± 13.56

Table 9: Hemophilia Specific Quality of Life Dimension by Hemophilia Severity (n=46).

Table 9 shows that the impaired quality of life with the increase in the severity of hemophilia which is presented by the highest total averages obtained by the people with severe hemophilia than people with moderate and mild hemophilia i.e., 38.67, 33.16 and 27.34 respectively. For people with severe hemophilia, the dimension "Physical Health" was the most impaired dimension (54.57) and the dimension "opinion about yourself" had the highest averages 53.19, 43.14 and 48 among all (severe, moderate and mild) form of hemophilia showing poorer quality of life.

Variables	Transcribed Mean Score
Physical health	75
Psychological Health	69
Social Health	60
Dealing with Hemophilia and treatment	53

Total	64.25

Table 10: Information related to Quality of life.

Table 10 shows that the total mean score is 64.25 and the dimension "Physical Health" has the highest average i.e., 75 showing poor quality of life.

Discussion

In this study the age of diagnosis of Hemophilia ranged from 0-15 years of age and in 73.8% of respondents, the diagnosis was at 1-5 years of age whereas in a study among 39 people with hemophilia at Juiz de Fora Regional Blood Center, Brazil, age at diagnosis ranged from 0 to 67 years and in 58.8% of the patients the diagnosis was made after two years of life [12].

This study reveals that the majority of the respondents were diagnosed with hemophilia A i.e., 86.1% which is supported by a study conducted in the National Centre for Congenital Bleeding Disorders in Laiko General Hospital of Athen, in which the predominant form of hemophilia was A (78%). In this study 39.3% had mild form of hemophilia whereas 63.6% of the individual had severe hemophilia in a study conducted in the National Centre for Congenital Bleeding Disorders in Laiko General Hospital of Athen which does not support this study as 24.6% of the respondents were unknown about their severity of hemophilia [8].

According to this study, majority of the respondents 85.2% had multiple episodes of joint bleeding and 55.7% had functional impairment of joint of varying degree which is supported by a study conducted in 75 people with hemophilia at the Hemophilia Care Unit, Bir Hospital, NAMS, in which total of 176 bleeding episodes into various sites were recorded. Bleeding into joints was the most common, occurring in 144 episodes (81.8%). Many of the joint bleeding was into multiple joints simultaneously [13].

In this study, 55.7% of the respondents reported it was painful for them to move. Similarly, in web-based cross-sectional survey in Italy, more than 75% of adult patients had problems in the physical sphere, specifically for mobility and pain and discomfort (76.7%). A similar findings are presented in a HERO study among 675 adult hemophiliac from ten countries where 89% of adult PWH reported that they experienced pain that interfered with activities in the previous 4 weeks, 26% of adult with PWH reported that in past 4 weeks, pain interfered with their daily life extremely or quite a lot [14].

In this study the highest averages on HRQoL was in dimension "Physical Health", Feelings", "Sports and leisure", has the highest averages (72.76, 62.09 and 68.5 respectively) while in the naturalistic, multicentre, longitudinal COCHE study among 233 adult hemophilia patients from 23 Italian Hemophilia centers. Patients showed mainly impairment in the dimension "sports"," future", and "Physical health". Most of them (75.6%) had to refrain "often or always" from sports like soccer, concerning their "future" 48.5% thought "often" or "always" that their life plans are influenced by hemophilia [15].

By dimension "Future" and "partnership" had the lowest averages with a mean score of 34.0 and 20.9 respectively showing the better quality of life in this study. Similar findings were seen in a study conducted in Brazil in Blood Center where the dimension "relationship and Partners" was the least impaired dimension among the participants with a mean score of 21.41 and 17.52 respectively.

Conclusion

On the basis of the findings, it can be concluded that health-related quality of life in hemophilia was influenced by joint impairments. More than one-third of the respondents had the highest averages indicating poorer quality of life. By dimension "Physical Health", was the most impaired and "Planning of family" and "Partnership" was the least impaired. This study only includes two districts of Nepal. Further study can be conducted in other districts to generalize the finding.

References

1. World Federation of Hemophilia (2017) Report of the Annual Global Survey 2016. World Federeation Hemoph, Montréal, Québec, Canada.
2. Iorio A, Stonebraker JS, Brooker M, Soucie JM, Rezende SM, et al. (2017) Measuring the quality of haemophilia care across different settings: a set of performance indicators derived from demographics data. Haemophilia 23: e1-e7.
3. National Hemophilia Foundation (2019) National Hemophilia Foundation for all bleeding disorders. National Hemophilia Foundation, New York, United States.
4. World Federation of Hemophilia (2016) How do you get hemophilia? World Federation of Hemophilia, Montréal, Québec, Canada.
5. Mansouritorghabeh H (2015) Clinical and laboratory approaches to Hemophilia A. Iran J Med Sci 40: 194-205.
6. Lucile Packard Children's Hospital Stanford (2019) Hemophilia. Stanford Children's Health, Stanford, California.
7. World Health Organization (2019) WHOQOL: Measuring Quality of Life. Health statistics and information systems, WHO, Geneva, Switzerland.
8. Varaklioti A, Kontodimopoulos N, Katsarou O, Niakas D (2014) Psychometric properties of the Greek Haem-A-QoL for measuring quality of life in Greek haemophilia patients. Biomed Res Int 2014: 968081.
9. Indiana Hemophilia and Thrombosis Centre (2019) Types of Hemophilia | Indiana Hemophilia andamp; Thrombosis Center. Indiana Hemophilia and Thrombosis Centre.
10. Manikandasamy V, Arumugasamy S, Mathevan G (2017) Impact of hemophilia on quality of life of affected children and their parents, a hospital based cross sectional study. Int J Contemp Pediatr 4: 1620.
11. Karki S, Niraula HK (2014) Quality of Life of People with Hemophilia. J Law, Healthc Ethics. 6: 235-243.
12. Ferreira AA, Leite ICG, Bustamante-Teixeira MT, Corrêa CSL, Cruz DT, et al. (2013) Health-related quality of life in hemophilia: results of theHemophilia-Specific Quality of Life Index (Haem-a-Qol) at a Brazilian bloodcenter. Rev Bras Hematol Hemoter 35: 314.
13. Rewati MR, Malla R (2007) Pattern of clinical presentations in hemophiliacs in Nepal. Post-Graduate Medical Journal of NAMS.
14. Varaklioti A, Kontodimopoulos N, Katsarou O, Niakas D (2014) Psychometric Properties of the Greek Haem-A-QoL for Measuring Quality of Life in Greek Haemophilia Patients. Biomed Res Int 2014: 1-12.
15. Von Mackensen S, Scalone L, Ravera S, Mantovani L, Gringeri A (2005) PHM5 assessment of health-related quality of life in patients with haemophilia with the newly developed haemophilia-specific instrument HAEM-A-QOL. Value Heal 8: A127.