# Quality of Life of People with Hemophilia

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## ABSTRACT:

**INTRODUCTION:** 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. Although life expectancy of hemophiliacs in developed countries is close to that of healthy persons this is not the case in developing countries.This study aims to identify the quality of life of people with Hemophilia, who are clinically diagnosed with either Haemophilia A or B.

**METHOD:** A descriptive study was conducted among 16 years and above male hemophiliac at hemophilia Care unit of NAMS, Bir Hospital. Sixty respondents were selected by using non-probability purposive sampling technique. The English version of Hemophilia Specific quality of life questionnaire. (Haemo-A-Qol) after getting approval from Dr. Sylvia Von Mackenson was translated in Nepali language and was administered to collect the data. Data processing was done using computer (SPSS version 19).The higher the score obtained in Haemo-A-Qol the poorer quality of life and the lower the score better the quality of life.

**RESULT:** This research study showed that 36.7% of respondents were between 21 to 25 years of age with the mean age of 24.9 +_6.25 years. Out of 60, 85% had Hemophilia "A" & 15% had Hemophilia "B" .Among them only 73% respondents know their factor activity level and of which 54% had severe, 29% had moderate form and 15% had mild form of hemophilia. The average Haemo-A- Qol total score was 43.19, ranging from 0.47 to 95.41. The dimension "view of yourself", "treatment ", "physical health" had the highest averages (62.91, 48.33 and 46.67) respectively indicating poorer quality of life where as the dimension "family planning and partnership" were the least impaired dimensions among the respondents with mean score of 13.54 & 19.86 respectively. By severity of the hemophilia, the severe form had the highest mean score 36.69 indicating poorer quality of life.

**CONCLUSION:** More than one- third of respondents had the highest averages indicating poorer quality of life.By dimension, "view of self" was the most impaired and "planning of the family" was the least impaired.

**KEY WORDS:** Hemophilia , Haemo-A-Qol, Quality of life.

## INTRODUCTION

The hemophilia is inherited, life-long, sex- linked disorders occurring predominantly in males.[1] Patients with hemophilia have little or no clotting factors IX or VIII.[2]Haemophilia 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.[3] Hemophilic patient bleeds either spontaneously or on trauma. About 75% of all bleeding in patients with hemophilia occurs into joints.

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Recurrent joint hemorrhages can result in damage so patient with severe factor deficiency can become crippled by the joint damage before they become adults. Spontaneous hemorrhage and GI bleeding can occur and the most dangerous hemorrhage is intracranial or extra cranial.[4]

Hemophilia is a condition that can be associated with significant morbidity and lifelong treatment. These individuals may suffer from joint pain, loss of range of motion, and crippling musculoskeletal deformity and disability, all of which impair their quality of life and have implications for employment, productivity, and other psychosocial aspects.[5]

In Italy, the project on the social burden and quality of life (QoL) of patients with haemophilia investigates

costs from a society perspective and provides an overview of their quality of life.On average, quality of life is worse in adult patients compared to child and caregivers: more than 75% of adult patients declare physical problems, 43% of adult patients and 54% of their parents have anxiety problems.[6]Similarly in another study from Italy among patients with severe haemophilia aged >65 years, the majority of patients with on-demand treatment, revealed more impairments in Health Related Quality of Life (HRQoL). Psychological and social well-being was perceived worse by patients compared to healthy controls, and a poor orthopedic status was negatively associated with HRQoL.[7] For individuals with bleeding disorders, and especially those with hemophilia, treatment can be a tremendously heavy financial burden. In fact, the treatment of hemophilia ranks among the most expensive chronic diseases in the United States.[8,9] In Nepal, the average cost of treatment is usually Rs.5, 20,000 per year. Complication can cause expenses to exceed this amount .[10]

The World Federation of Haemophilia (WFH) estimates that worldwide, approximately 70% of patients with hemophilia are underdiagnosed and untreated. Most live in developing countries. Although in developed countries the life expectancy of hemophiliacs is close to that of healthy persons, this is not the case in developing countries. Great disparity also exists in the treatment of hemophiliacs, especially when this relates to available factor concentrates. There are many reasons for the inadequate care of hemophilic patients: the perception of rarity of the disease; lack of laboratory facilities to diagnose the disorder; lack of understanding of the disorder by patients, their relatives, and even healthcare providers; poorly developed blood bank facilities; and lack of adequate factor supply are just some examples.[11] Similar is the situation of hemophiliac of Nepal. Although there has been many attempts to deliver minimum care by Nepal Hemophilia society still treatment of hemophiliacs is a challenge because of the exorbitant price of the factor concentrates, their unavailability and also the remoteness of the geography. In addition, lack of awareness regarding hemophilia, few diagnostic centers and all of which located in capital city, few hemophilia care centers etc. makes treatment much more difficult. Therefore the clinical presentation, health status and quality of life of people with hemophilia is different in our part of world.

Similarly, there are very few researches carried out in hemophilia as well as Research related to quality of life of people in hemophilia has not yet been studied or published in Nepal. So this study aimed to measure the quality of life of people with hemophilia in the context of developing countries like Nepal. Identifying the effects of hemophilia in those affected will be the first step toward a comprehensive patient Care as well as cost effective measure by preventing comorbidities, complications if programs to address such problems is carried out in earlier phase.

## METHOD

This study was conducted at Hemophilia care unit, Bir Hospital,amongst male patient aged 16 years and above who were clinically diagnosed with either Hemophilia A or B and willing to participate .

A descriptive research design was used.A total of 60 patients were included as per non-probability sampling technique. Semi-structured Interview Schedule and Likert Scale were used according to the objectives of the Study.Haemophilia Specific Quality of Life Questionnaire for Adults i.e. Haemo-A-Qol likert Scale was used after getting permission from Dr.Sylvia Von Mackensen.The English Version of Haemo-A-Qol was translated into Nepali language before administration. The tool consists of 46 items comprising 10 dimensions (physical health, feelings, view, sport & leisure time, work and school, dealing, treatment, future, family planning, & relationships/partners). Number for negative statements was assorted as: 0=Not applicable, 1= Never, 2=Rarely, 3=Sometimes, 4= Often, 5=All the time. The higher values represents poor quality of life. For positive statement, the number was reversed. Scoring was performed by transforming the scores achieved in each dimension, as well as the total score, on scales ranging from 0 to 100, with 0 representing the best and 100 the worst quality of life. To transform raw score into transformed Scale Score (TSS) following formula was used.

$$TSS = \frac{100 \times \text{Raw Score-minimal Possible raw Score}}{\text{Possible range of raw Score}}$$

Where possible range of raw score = maximal possible raw score – minimal possible raw score.

## RESULT

| Table 1: Socio-Demographic Information N=60 | | |
|---|---|---|
| Variables | Number | Percentage |
| Age groups | | |
| 16-20 | 13 | 21.7 |
| 21-25 | 22 | 36.7 |
| 26-30 | 16 | 26.7 |
| 31-35 | 4 | 6.7 |
| 36-40 | 3 | 5.0 |
| 41-45 | 2 | 3.3 |
| Marital Status | | |
| Married | 18 | 30 |
| Unmarried | 42 | 70 |
| Educational status | | |
| Primary | 1 | 1.7 |
| Lower Secondary | 4 | 6.7 |
| Higher Secondary | 35 | 58.3 |
| Bachelor & above | 20 | 33.3 |

The above table elucidates that majority of respondents (36.7%) were of 21-25 age group. The mean age of respondents was 24.9 +_6.29. Among 60 respondents only 30 % were married. All were literate and 58.3 % had education upto higher secondary level.

| TABLE 2: Information related to Disease N=60 | | |
|---|---|---|
| Variable | Number | Percentage |
| Types of Hemophilia | | |
| Hemophilia "A" | 51 | 85 |
| Hemophilia "B" | 9 | 15 |
| Factory activity level | | |
| <1 % | 24 | 40 |
| 1-5% | 13 | 21.7 |
| 5-40% | 7 | 11.7 |
| Unknown | 16 | 26.7 |
| Number of joint bleeding | | |
| Less than 3 bleeding episodes | 2 | 3.5 |
| Multiple episodes | 48 | 84.2 |
| None | 7 | 12.3 |
| Presence of joint impairment | 39 | 66.1 |

Out of 60 respondents 85% had hemophilia A and majority of respondents i.e. 40% had severe form of Hemophilia. In addition to that, 84.2% had multiple episodes of bleeding in joint & 66.1% had joint impairment.

| Table 3. Mean Scores of Haemo-A-Qol questionnaire in Total & by Dimension N=60 | | | |
|---|---|---|---|
| Dimension | n Valid | Mean | Standard deviation |
| Physical Health | 60 | 46.67 | 29.24 |
| Feelings | 60 | 45.31 | 29.78 |
| View of yourself | 60 | 62.91 | 29.93 |
| Sports and leisure | 57 | 43.33 | 40.75 |
| Work and School | 58 | 36.45 | 24.77 |
| Dealing | 60 | 40.83 | 32.29 |
| Treatment | 60 | 48.33 | 28.65 |
| Future | 60 | 44.71 | 35.65 |
| Family Planning | 57 | 13.54 | 18.10 |
| Partnership | 60 | 19.86 | 28.07 |
| Total | 60 | 43.19 | 29.72 |

The average Haemo-A- Qol total score was 43.19, ranging from 0.47 to 95.41.The dimension "view of yourself" , "treatment ", "physical health" has the highest averages (62.91 ,48.33 and 46.67) respectively indicating poorer quality of life where as the dimension "family planning and partnership" were the least impaired dimensions among the respondents.

| Table 4. Hemophilia-Specific Quality of Life Dimensions by Hemophilia Severity N=44 | | | |
|---|---|---|---|
| Dimension | Severe (n=24) | Moderate (n=13) | Mild (n=7) |
| Physical Health | 56.67±20.83 | 37.5±24.07 | 30.71±13.67 |
| Feeling | 43.26±22.14 | 41.67±15.15 | 39.28±18.65 |
| View of yourself | 52.29±12.06 | 44.16±11.24 | 50±7.07 |
| Sports and leisure | 26.92±28.81 | 40.20±30.24 | 36.97±30.9 |
| Work and School | 41.4±14.72 | 26.04±18.43 | 36.60±28.05 |
| Dealing | 34±15.3 | 42.36±28.3 | 21.42±15.85 |
| Treatment | 49.6±15.916 | 45.31±16.67 | 42.4±15.29 |
| Future | 48.74±18.012 | 35.41±17.51 | 41.2±19.5 |
| Family Planning | 14.06±15.01 | 9.61±10.7 | 17.96+_20.4 |
| Partnership | 20.83±23.69 | 6.94±15.42 | 17.83±31.33 |
| Total | 36.69±16.27 | 32.92±18.77 | 29.34±15.57 |

Quality of lifewas impaired with the increase in severity of hemophilia which is presented by the highest total averages obtained by people with severe hemophilia than people with moderate and mild hemophilia. i.e. 36.69 ,32.92&29.34 respectively.

## DISCUSSION

This study reveals that majority of respondents were diagnosed with Hemophilia "A" i.e 85% and 40% of the them had severe form of hemophilia which is supported by a study conducted in the National Centre for Congenital Bleeding Disorders in Laiko General Hospital of Athens, in which the predominant form of haemophilia was A (78%) and 63.6% of the individuals had severe haemophilia.[3]

According to this study, majority of respondents (84.2%) had multiple episodes of joint bleeding and 66.1% had functional impairment of joint of varying degrees which is supported by another 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.[12] A similar results were seen in a study conducted at five hemophiliac clinic across the country in India .The prevalence and risk factors for disability were studied in 148 patients with severe haemophilia A and the result showed only nine were free of disability out of 148.[13]

In this study, 76.67 % of the respondents reported it was painful for them to move & 5 % reported pain during movement all the time. Similarly in web-based cross-sectional survey in Italy, more than 75% of adult patients had problems in the physical sphere, specifically for mobility (75%) and pain/discomfort (76.7%).[6] 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 PWH reported that in the past 4 weeks, pain interfered with their daily life "extremely" or "quite a lot.[14]

The HRQol averages in this study were higher among severe hemophilia followed by moderate and mild form i.e 36.69 ,32.9 &29.3 respectively showing impaired quality of life in people with severe form of hemophilia than moderate and mild ones . In contradiction to a study conducted in Brazillian Blood center in which the HRQoL averages in some dimensions were higher among patients with moderate hemophilia than among those with severe hemophilia confirming the wide clinical variability among patients with moderate hemophilia.[15]

In this study , the highest averages on HRQol was in dimension " view of self" followed by "treatment "and "Physical health" with mean score of 62.9 , 48.33 & 46.67 respectively while in the naturalistic, multicenter, longitudinal COCHE Study among 233 adult haemophilia patients from 23 Italian Haemophilia Centers, patients showed mainly impairments in the dimensions "sport", "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 their haemophilia. [16]

By dimension ,"Famly planning "& "Partnership" had lowest averages with mean score of 13.54 & 19.86 showing the better quality of life in this study. Similar findings were seen in a study conducted in Brazilian Blood centre where the dimension 'Relationships and Partners' was the least impaired dimension among the participants with mean score of 21.41 & 17.52 respectively.[16]

## CONCLUSION

Health-related quality of life in hemophilia, measured by the Hemophilia-Specific Quality of Life questionnaire was influenced by severity of hemophilia and joint impairments. More than One-third of respondents had the highest averages indicating poorer quality of life. By dimension, "View of Self" was the most impaired and "Planning of family" and "partnership" was the least impaired.

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