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Original Research Article

Impact of hemophilia on quality of life of affected children and their parents, a hospital based cross sectional study

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ABSTRACT

Background: Hemophilia can have a huge negative impact not only on the physical health but also on the psychological, economic and social well-being of the affected children and their family. Documentation of this negative impact on quality of life is vital in drawing the attention of relevant stakeholders to design and implement appropriate interventions.
Methods: This was a cross sectional study, Children with hemophilia aged 4 to 12 years, and their parents who were attending the Pediatric Hematology Clinic of the Advanced Pediatric Centre, PGIMER, Chandigarh, between July 2009 and June 2010 were included in the study. The Hemo-QoL questionnaire was used to assess the quality of life after translation into local language and validation.
Results: The study included 51 children with 10 children in 4-7 years age group and 41 children in 8-12 years age group and their parents. Perceived impact on family (77.3±14.7), poor physical health (62.9±29.8), sports and school (53.8±22.8) had the highest negative impact on QoL. Support from friends, family, and other persons seemed to have contributed positively towards the quality of life. Parents of older children had higher mean subscale scores in View of yourself subscale as compared to parents of younger children (40.8±14.2 vs 23.7±33.0, p value 0.018). Under Sports and school subscale also similar pattern was observed (48.2±20.7 Vs 34.1±13.8, P value 0.045).
Conclusions: The Quality of life was poor both among children affected with 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. Support from family, friends, and others is vital in maintaining QoL. The QOL worsens with increasing age of the child in both children and their parents.

Keywords: Haemophilia, Hemo-QoL, Quality of life

INTRODUCTION

Haemophilia A and B are congenital bleeding disorders caused by an X-chromosome linked deficiency in coagulation factors VIII or IX (FVIII, FIX. People with Hemophilia (PWH) are at a risk of spontaneous bleed for rest of their life. As per the recent population statistics by WFH (World Federation of Hemophilia) about 1,97,183 people are suffering from hemophilia globally. About 9.2% of the hemophilia burden is contributed by India.[1] Patients with hemophilia need to undergo frequent blood transfusions, which puts the person at risk of various blood-borne infections like HIV, hepatitis etc.[2] A study by Ghosh et al, have reported that around 50% of the patients with hemophilia are affected with hepatitis C virus and 10% with HIV.[3] Apart from the impact on Physical health, hemophilia will have a huge negative impact on the psychological, social and economic health

of the affected individual and their families. Children with hemophilia are reported to have a high burden of emotional difficulties and have been reported with conditions like lower self-perception and depression. Studies have reported that children with hemophilia are less joyful, less open and caring.[4,5] Another study examined and reported depression, anxiety and suicidal behavior in children and adolescents with hemophilia. It suggested that the rate of major depressive disorder was 6.0%.[6] Parents of the children with hemophilia undergo many challenges in day to day life and long-term impact as well. Evans et al., have studied the behavioral problems and family functioning in children with hemophilia and have reported higher incidence of these problems when compared to normal families.[7]

The need for repeated hospital visits may lead to school or work absenteeism of the PWH and their family members. Diseases like hemophilia have a serious impact on schooling and employment. A study by Kar A et al have reported 36.5% (n=148) of them have dropped out of school due to bleeding.[8] A prospective study has reported that children with hemophilia, on an average have lost 0 to 50 days of school days due to bleeding.[9]

Haemophiliacs, when compared to their classmates, tend to have more teacher-rated emotional, but not more behavioral disturbances, because of the impact of the chronic disorder on the child. The haemophilic child is greatly frustrated because of restrictions.[5]

A study on overall behavioral challenges reported that children with hemophilia were not found to be at increased risk of social or behavioral difficulties. However, both children with hemophilia and their parents identified more difficulties with emotional well-being.[4]

All these factors together may have tremendous negative impact on the persons and their family member's quality of life. Poor quality will also be at risk of increasing burden is seen on social and family aspects, not only by the patients themselves but also their parents. Due to challenges in studying rare disorders like hemophilia, it is difficult to study these diseases from public health point of view.

Very few studies have been carried out on hemophilia in India and there is very little data available. This study would help us understand the disease, its complications and the quality of life, from both the affected child and the parent's perspective and would benefit many patients, in terms of special care in educational institutions and employers, and providing some opportunity for improving the quality of life of patients. The objective of the study was to assess health-related quality of life of children with hemophilia and their parents attending a tertiary care teaching hospital. To assess the relationship between disease severity, type of bleeds and Demographic variables with Quality of life.

METHODS

Study design: This was a cross sectional study. Study Setting: This study was carried out in Pediatric Hematology Clinic of the Advanced Pediatric Centre, PGIMER, Chandigarh. Study population: Children who are diagnosed to have Hemophilia and their parents, in the Pediatric Hematology Clinic of the Advanced Pediatric Centre, PGIMER, Chandigarh, were the study subjects. Study duration: This study was carried out over a period of 12 months from July 2009-June 2010.

Inclusion criteria

- A child diagnosed to have Hemophilia A and B.
- Age 4-12 years

Study tools: The Hemo-QoL questionnaire (10) was used. This questionnaire was developed for children and adolescents with hemophilia by the European Hemophilia care centers and has sets of psychometrically tested questionnaires for different age groups of children as well as for their parents. Permission to use these questionnaires has been taken from the Centre of Psychosocial Medicine, Hamburg.

Children's questionnaire

Age group I (4-7 years)

This has 11 subscales and a total of 33 questions. Three response choices are given and the child has to decide one

Age group II (8-12 years)

This has 13 subscales and a total of 76 questions. Five response choices are given and the child has to decide one.

Parent's questionnaire, corresponding to the age groups

The age group I (4-7)

This has 11 subscales and a total of 33 questions. Five response choices are given and the parent has to decide one.

Age group II (8-12)

This has 13 subscales and a total of 76 questions. Five response choices are given and the parent has to decide one.

The dimension 'joint bleed' includes questions regarding no of episodes, severity and what they do whenever having joint bleeds. The 'injections' dimension includes questions about who had given their injections. The 'physical' dimension includes questions concerning pain and bleeding, etc., in the dimension 'feeling' it was asked

how children feel related to their hemophilia, questions about how children perceive themselves are included in the dimension 'view of yourself', the interaction in the family was questioned in the dimension 'family', the dimension 'friends' contains questions about the interaction with friends, questions about how children perceive the support they receive from others pertain to the dimension 'perceived support', in the 'sport and school' dimension children were asked about their school/kindergarten life, the dimension 'dealing' contains questions about how children deal with their hemophilia, questions in the dimension 'treatment' concern treatment issues, in the dimension 'others' the interaction with others is of interest. In the dimension 'global health' their overall health status is asked.

The original questionnaire was in UK English. Forward translation step was a translation of original questionnaire into Hindi which was done in Hindi section in our institute. Backward translation step was performed by translation of this version into English by one professional translator, who was native English speaker and fluent in Hindi. The source questionnaire and backward translation were compared. The Hindi questionnaire was tested in normal children before implementing in our study. This Hindi questionnaire was administered to the child and parent when he is admitted or attended the Pediatric Hematology Clinic.

Haemo-QoL questionnaire

Haemo-QoL questionnaires were translated into the regional language, Hindi. This was administered to children and parents in the 4-7 year and 8-12-year age group. In 4-7-year age group, children were unable to answer the questionnaire completely, they filled in barely 10% of the questionnaire. We are able to analyze only the parent's answers. In 8-12-year age group both the children and parents answered the questionnaire. Questionnaire for 4-7-year age group contains 11 subscales with a total of 33 questions and for 8-12-year age group, it contains 13 subscales with a total of 76 questions. Each subscale was assessed separately.

For scoring the questionnaire first, the numbers have to be assorted to the response scale, which is for age group II, 1= never, 2=seldom, 3=sometimes, 4=often, 5=all the time and for the age group I, scoring is 1=never, 2=sometimes, 3=very often.

Summing up the items belonging to a subscale yields the raw score (RS) per subscale. Its range lies between the lowest possible (number of items (n) x 1) and highest possible (number of items (n) x 5 in the older; or x 3 for the youngest) value of the respective scale.

Comparing scores across subscales is done. This raw score is divided by the number of items in the scale, the resulting standardized scale score (SSS) can have any (also decimal) value between 1 and 5 (or 3 for the youngest). A value of 1 represents the highest possible quality of life rating and a value of 5 (or three for the youngest) the lowest possible quality of life rating of the patient. Transferring a raw score to a transformed scale score (TSS) between 0 and 100 makes it possible to express the scale score in percent between the lowest (0) and the highest (100) possible value.

Statistical methods

Descriptive analysis was carried out by mean and standard deviation for quantitative variables, frequency, and proportion for categorical variables. The association between categorical explanatory variables and the quantitative outcome was assessed by comparing the mean values. The mean differences along with their 95% CI were presented. Independent sample t-test was used to assess statistical significance between the groups.

RESULTS

A total of 51 children with 10 children in 4-7-year age group and 41 children in 8-12 years age group were included in the study.

Table 1: Age and type of hemophilia.

Age group (years)	Haemophilia	
	A	B
4-7 (Group I) (N=10)	10(100%)	0 (0%)
8-12 (Group II) (N=41)	34(82.9%)	7(17.1%)
Total (N=51)	44(86.3%)	7(13.7%)

Out of 51, 44 (86.3%) children had Hemophilia A, 7 (13.7%) having a deficiency of factor IX. Among 4 to 7-year age group all the 10 children had Haemophilia A and among 8 to 12 years old 82.9% had hemophilia A and remaining 17.1% had hemophilia B (Table 1).

Table 2: Average of RS, SSS, and TSS of all attributes affecting quality of life.

Haemo-QoL scores	Children 8-12	Parents 8-12	Parents 4-7
Mean RS of all subscales	16.0±2.3	15.4±2.0	8.1±1.4
Mean SSS of all subscales	2.7±0.3	2.6±0.3	2.7±0.5
Mean TSS of all subscales	45.0±9.5	42.2±8.5	43.9±13.77

The mean raw score of all subscales was 16.0±2.3 in children 8 to 12-year-old and 15.4±2.0 in parents of the 8 to 10-year-old children. The mean RS score was lowest for parents of 4 to 12-year-old children. The mean SSS of all subscales was comparable in children and both the parent groups. The mean TSS of all subscale score was 45.0±9. in children 8 to 12-year-old. It was 42.2±8.5 in parents of the 8 to 12-year-old children and 43.9±13.77 in parents of 4 to 7-year-old children and total scores

indicate the quality of life is uniformly poor in both the affected children and their parents (Table 2).

Table 3: Comparison of Haemo-QoL subscale and global scores of children and their parents (8-12 year group).

Subscale	Kids (8-12yrs) (n=41)	Parents (8-12yrs) (n=40)	'p' value
Joint bleeds	38.7±16.1	36.9±14.9	0.732
Injections	30.6±9.1	31.5±8.7	0.736
Physical health	62.9±29.8	68.2±25.4	0.545
Feeling	53.2±25.9	48.9±21.8	0.311
View of yourself	38.2±19.6	40.8±14.2	0.586
Family	77.3±14.7	64.6±12.6	< 0.001
Friends	27.7±21.7	33.2±27.1	0.673
Perceived support	36.2±22.8	30.6±19.24	0.502
Other persons	28.0±21.2	21.3±19.0	0.172
Sports and school	53.8±22.8	48.2±20.7	0.277
Dealing with haemophilia	33.3±21.4	28.6±21.0	0.231
Treatment	40.0±16.5	31.6±13.4	0.054
Global health	64.6±19.3	63.7±19.5	0.671

Table 4: Comparison of Haemo-QoL subscale and global scores of parents in children with different age groups.

Subscale	Parents (4-7yrs) (n=10)	Parents (8-12yrs) (n=40)	'p' value
Joint bleeds	41.3±15.9	36.9±14.9	0.541
Injections	29.0±8.0	31.5±8.7	0.273
Physical health	68.1±25.5	68.2±25.4	0.942
Feeling	35.8±28.0	48.9±21.8	0.173
View of yourself	23.7±33.0	40.8±14.2	0.018
Family	63.7±20.7	64.6±12.6	0.557
Friends	50.0±28.8	33.2±27.1	0.115
Perceived support		30.6±19.24	
Other persons	26.2±24.6	21.3±19.0	0.635
Sports and school	34.1±13.8	48.2±20.7	0.045
Dealing with hemophilia		28.6±21.0	
Treatment	46.2±25.0	31.6±13.4	0.084
Global health	65.0±21.0	63.7±19.5	0.894

Among the children the aspects of life which had the highest impact on poor quality of life were perceived impact on family (77.3±14.7), followed by poor physical health (62.9±29.8), sports and school (53.8±22.8) and Feeling about the illness. (53.2±25.9). The occurrence of physical complications (Joint bleeds) and treatment as such did not have a high negative impact on global quality. Support from friends, family, and other persons seemed to have contributed in between coping up with the illness and contributed positively towards the quality of life. No significant differences were observed between the children and their parents in any of the subscales, except the family subscale. The children had a higher mean family subscale (77.3±14.7 vs 64.6±12.6, P value <0.001) as compared to their parents, indicating the stronger negative impact on Children (Table 3).

Parents of older children had higher mean subscale scores in View of yourself subscale as compared to parents of younger children (40.8 ± 14.2 vs 23.7 ± 33.0, p value 0.018). Under Sports and school subscale also similar pattern was observed (48.2 ± 20.7 Vs 34.1 ± 13.8, P value 0.045). Apart from these two domains, the mean subscale and global scores had no statistically significant differences between the two groups of parents (Table 4).

DISCUSSION

The current study has assessed the quality of life of children and parents of 51 children who were diagnosed as either Haemophilia A or B, using Haemo-QoL questionnaire. The mean TSS of all subscale score was 45.0±9. in children, 42.2±8.5 and 43.9±13.77 in parents of 8 to 12-year-old and 4 to 7-year-old children, indicating poor quality of life uniformly in both the affected children and their parents. Evans M et al in their comparative study reported more emotional, behavioral and family difficulties in hemophilia children and their families, compared with the healthy group.[7] The negative psychological impact can go beyond the poor quality of life as reported by Ghanizadeh A et al.[6] According to this study, about 6% of the children had signs of Approximately 36% of the subjects wished to die at least once during the last 6 months.

Among the children the aspects of life which had the highest impact on poor quality of life were perceived impact on family (77.3±14.7), followed by poor physical health (62.9±29.8), sports and school (53.8±22.8) and Feeling about the illness. (53.2±25.9). As per the current study, the QoL is worse in older children, as they develop more negative concern regarding self-perception and also face or difficulty in the participation of sports and other school activities. The occurrence of physical complications (Joint bleeds) and demands of treatment had no significant negative impact on global quality. In contrast to the current study Haghpanah S et al and Bullinger M et al have reported that quality of life was significantly associated with health status, being bothered by the disease and bleeding severity.[11,12] Goldstein G et al have reported that recurrent joint bleeds, change of physical appearance are associated with isolation, depression, anger, and aggressiveness among children with hemophilia.[13]

Support from friends, family, and other persons seemed to have vital contributing factors in coping up with the illness and contributed positively towards the quality of life. Parents of older children had higher mean subscale scores in View of yourself subscale as compared to parents of younger children (40.8±14.2 versus 23.7±33.0, p value 0.018). Under Sports and school subscale also similar pattern was observed (48.2±20.7 Vs 34.1±13.8, P value 0.045). This indicates worsening of the quality of life with increasing age of the child both in children and their parents. According to Goldstein G et al have reported that recurrent joint bleeds, change of physical appearance are associated with isolation, depression, anger, and aggressiveness among children with hemophilia.[13] Mothers are reported to bare the maximum burnt due to nature of transmission, resulting in accusations of boys and the family members and associated guilt and isolation within the family. Canclini M et al have reported a lower self-esteem, which tends to increase with age in hemophilic children, as compared to healthy subjects.[14]

There are other studies as in the study by Phadnis S et al have reported the high positive impact of a structured educational intervention of parents on the management of the illness and quality of life, which was lasting only up to a year.[15] The authors have emphasized the importance of continuous and repeated educational interventions to sustain the positive impact. This finding assumes even greater importance in the wake of current study findings, as the quality of life is found to worsen with advancing age of the child.

Limitation of the study was to be considering the relatively smaller sample size, the impact of various other sociodemographic factors on quality of life could not be assessed in the study. This would have helped us in an in-depth understanding of the issue and identification of highly vulnerable subgroups prone to the poor quality of life and more attention on them.

CONCLUSION

The Quality of life was poor both among children affected with hemophilia and their parents. The major aspects, which have contributed to poor quality of life were perceived impact of family, poor physical health and inability to participate in school/sports activities. The aspects which have contributed positively towards quality of life were supported by family, friends, and others. Parents of older children had a poor quality of life in how they view themselves and in school and sports activity domain as compared to parents of younger children.

Recommendations

There is need to devise more culturally appropriate tools to assess quality of life in hemophilia children and their parents and validate them in native populations, considering the heterogeneous nature of Indian population. All efforts should be made by clinicians and other stakeholders to address the poor quality of life in children with hemophilia and their parents. There is an urgent need to devise and test the effectiveness of different kinds of interventions to address poor quality of life in these children and their families.

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