ORIGINAL ARTICLE

The Quality of Life of Children with Hemophilia in Shiraz, Iran

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Received: 21 December 2012 **Revised:** 10 March 2013 **Accepted:** 13 March 2013

Abstract

Background

Hemophilia might impact the quality of life (QOL) in children and adolescent. This study aimed to assess the quality of life in children with hemophilia and identify the factors that predict their QOL.

Methods

It was a cross-sectional study. A consensus sample of twenty-seven male children aged 8-16 years old with hemophilia participated in this study during 2011. The Haemo-QoL questionnaire version age group II (8–12 years old) or age group III (13–16 years) was used to measure the quality of life in children with hemophilia. Data were analyzed in SPSS using Spearman's rho, Pearson correlation coefficients, and multiple regression analysis.

Results

The total mean score on the Haemo-QoL was 54.1±7.3. Haemo-QoL correlated with the number of bleeding event and school absence, family income, mother s' education level, severity of hemophilia, person who performs his/her infusions, insight to health status and insight to suffering from hemophilia ($P<0.05$). QOL was predicated by family income ($\beta=-0.69$; explained 73% of variance) and infusion of the clotting factor replacement by parents ($\beta=-0.33$; explained 81% of variance).

Conclusion

The QOL of a sample of male children with hemophilia was impaired. Moreover, number of bleeding event, number of school absences, mother s' education level, severity of hemophilia, insight to health status and insight to suffering from hemophilia were associated with Haemo-QOL. In addition, family income and parent- administration of the product were the factors which predict the QOL in children with hemophilia.

Keywords: Adolescents; Children; Hemophilia; Quality of life; Predication

Please cite this article as: Bagheri Sh, Beheshtipoor N, Rambod M, Karimi M, Zare N, Hashemi F. The Quality of Life of Children with Hemophilia in Shiraz, Iran. IJCBNM. 2013;1(2): 110-120.

Introduction

Hemophilia is a rare, chronic, inherited disease primarily affecting males.[1] It is an X-linked bleeding disorder caused by a defective Factor VIII or Factor IX allele and leading to impaired clotting characterized by spontaneous bleeding and excessive bleeding after surgery or trauma.[2]

There are 6496 patients with congenital bleeding disorders who were registered by the Ministry of Health of Iran by December 2006. Among them, there are 3957 (61%) Factor VIII deficiency hemophilia A (frequency of severe, moderate and mild hemophilia A was 47, 33 and 20%, respectively).[3]

Children with hemophilia may lose opportunities to achieve their potentialities during school and later in life.[4] Individuals with severe hemophilia A (Factor VIII level ≤1%) experience frequent bleedings, often into soft tissue and joints, leading to joint damage. Repeated bleeding can lead to development of target joints and arthropathy,[2] as well as pain, permanent deformities, loss of mobility and disability.[5] It has been indicated that hemophilia patients who develop target joints and suffer from severe arthropathy have decreased Quality of Life (QOL).[6] Moreover, joint bleeding results in emotional and behavioral problems, family problems,[7] and impairment of health-related quality of life (HRQOL).[8,9] Hemophilia in young patients indicates lower physical functioning, bodily pain, general health and social functioning QOL in comparison to the general population.[10]

According to Dekoven et al. individuals with hemophilia experience the impairment in QOL and among children aged 8-16 years old the highest impairment was in the "physical health" subscale.[11] It was also shown that the dimension of "physical health" was lowest in the subscale of QOL in children and adolescents with hemophilia A.[12]

One of the factors that might affect the QOL could be the context. In an international multicenter West European study of children, it was indicated that there were differences between the QOL in various areas of study and the QOL in the Turkish patients was more impaired in the subscales of physical health, feeling, view, school and sport, and treatment.[13]

Beside the probable effect of different contexts on QOL of hemophilia, it could be possible that different ages might influence the QOL. Result of the studies indicated that impairment in QOL was different in various age groups.[11] Moreover, it was shown that with increase in age of hemophilia clients, the levels of QOL decreased.[11]

The QOL of children with hemophilia might be differed among countries; these differences can be explained by characteristics such as severity of hemophilia, frequency of bleeding and type of treatment (prophylaxis or on-demand therapy),[9,14,15] age,[16,17] and psychosocial factors.[14] It has been indicated that adults who have always been on prophylaxis reported the highest HRQOL scores in all domains when compared to other adults.[18] Another study reported that pediatric QOL scores among children generally decreased with increasing hemophilic severity as well as severity of joint pain.[19] De Wee et al. reported that a more severe bleeding phenotype was associated with lower scores on the domains of physical functioning, role limitations due to physical functioning, bodily pain, general health, social functioning and physical component summary in adult patients with moderate and severe von Willebrand disease.[20] In a study on adult patients with hemophilia from Sweden, regression analysis showed that 30.8% of the variance in QOL for the "Physical Component Score" could be explained by the age of start of prophylaxis, bleeding frequency and orthopaedic joint score independently.[21]

Although the QOL of young patients with hemophilia has been determine in some studies, there are few studies in our country in this field and also few studies have been conducted to assess the factor related to QOL in hemophilia patients. Therefore, this study was designed to assess QOL in children

with hemophilia and identify the factors that predict their QOL. Identifying these factors might help to know a high risk patient and predict QOL.

Materials and Methods

The present study was a cross-sectional research. It was conducted on children with hemophilia during 2011 in order to determine the most effective factors in predicting QOL. The target population consisted of hemophilia patients aged 8-16 years old. All of the hemophilia patients at this age group were recruited from the list of the Hemophilia Clinic affiliated to Shiraz University of Medical Sciences (SUMS), Iran.

Based on the power of 80%, α=0.05, and r=-0.10, a sample size of 30 subjects was selected for this study. In the list of the Hemophilia Clinic, there were 34 male patients aged between 8-16 years old. During the study, seven subjects did not participate in the study due to lack of interest (n=3), lack of referral to the Hemophilia Clinic (n=2) and no reason (n=2). Therefore, twenty seven male children participated in the study.

A consensus sample of 27 male children with hemophilia was asked to respond to the questionnaires. The inclusion criteria were male gender, mild, moderate or severe hemophilia A or B; alert and oriented, and ability to read and speak Persian. On the other hand, the subjects with congenital, chromosomal, or neurological anomalies, physical limitation, any systemic diseases (such as diabetes, etc), and psychiatric disorders, as well as those with emotional crisis during the previous month were excluded from the study.

The study was approved by the Ethics Committee of Shiraz University of Medical Sciences. The written informed consents were obtained from all the children's parents. The purpose of the study and data collection procedures was explained for them. The parents were given an opportunity to withdraw from the study at any time. The subjects' parents were assured of anonymity and confidentiality. They were informed that their participation doesn't pose any threat to treatment and care.

Data Collection

All of the subjects and their parents were invited to the Hemophilia Clinic affiliated to SUMS by calling them and after the written informed consents were signed by the children and their parents, they were asked to fill out two questionnaires. A specially-designed questionnaire was used to gather information about demographic/clinical and socioeconomic characteristics and Haemo-QoL questionnaire was used to measure the QOL of hemophilia children. Some of the information about medical history was collected from the patients' medical records held at the hemophilia center.

Instruments

A specially-designed questionnaire assessed the type of hemophilia, severity of hemophilia (clotting factor VIII or IX≤1%, 1-5% and ≥5% indicated mild, moderate or severe, respectively), previous history of inhibitor development, type of treatment (prophylactic or on- demand), type of product (plasma-derived or recombinant), age, number of bleeding event (in the 4 weeks before the study), number of school absences (4 weeks ago), number of referral to the hemophilia clinic (4 weeks ago), person who gave his/her infusions (nurses, parents and self-administration), insight to health status, insight to suffering from hemophilia, positive history of AIDS and hepatitis, parents' income and mother s' education level.

Quality of life was assessed with Haemo-QoL questionnaire (long version). The Haemo-QoL is a disease-specific self-report questionnaire for children and adolescents with hemophilia developed by Von Mackensen et al.[22] in Europe. There are three versions for three age groups: version I for children aged 4–7 years (21 items in eight dimensions), version II for children aged 8–12 years (64 items in ten dimensions), and version III for

adolescents aged 13–16 years (77 items in 12 dimensions). Because the subjects in the present study were children and adolescents between 8 and 16 years old, version II or III was used as appropriate. For children aged 8–12 years (age group II), the questionnaire comprised ten dimensions (physical health, feelings, view, family, friends, others, sports and school, treatment, perceived support and coping). For adolescents aged 13–16 years (age group III), the questionnaire was expanded with two additional domains (relationships and future).[9,22] Subjects scored each item on a 5-point scale according to how often they encountered problems during the previous 4 weeks (1=all the time, 2=often, 3=sometimes, 4=seldom, 5=never).[22]

Because the number of items in each dimension differs in the two versions of the questionnaire for different age groups, raw scores were transformed on a scale from 0 to 100 to make it possible to compare the age groups and the levels of impairment in different dimensions. Higher scores on the Haemo-QoL reflect higher levels of impairment and thus lower levels of QOL.[9] After the subjects were instructed about how to complete the questionnaire, the time it took them to complete the instrument was 20 minutes.

The original Haemo-QoL questionnaire in English,[22] was translated into Persian and given to ten experts for evaluation. Their corrective feedback was incorporated into the Persian translation, and then the questionnaire was back-translated into English by a bilingual translator to identify inconsistencies and loss of or changes in meaning. The content of the translated questionnaire did not differ from the original version.

The Haemo-QoL questionnaire was originally developed and tested in six countries (France, Germany, Italy, the Netherlands, Spain and the United Kingdom) for psychometric properties in 339 children with hemophilia and their parents. The three age group versions of the Haemo-QoL demonstrated acceptable internal consistency (Cronbach's α=0.70) and retest reliability values, and were shown to have sufficient discriminatory and convergent validity.[22] Von Mackensen et al. reported a Cronbach's for the Haemo-QoL questionnaire of 0.91, and a test–retest correlation for the total score of 0.92.[22] Mercan et al. confirmed that Cronbach's alpha coefficient for the total score was >0.70 (range 0.77 to 0.96) in the Turkish version of the Haemo-QoL questionnaire.[13]

In this study, reliability coefficients for internal consistency (Cronbach's α) for each of the subscales ranged from 0.61 to 0.81 in the age group II (8–12 years) and from 0.49 to 0.84 in the age group III (13–16 years). Cronbach's α for the total score was 0.89 for the age group II and 0.78 for the age group III. Ten faculty members of Shiraz University of Medical Sciences verified the face and content validity of the translated versions of the instrument.

Data Analysis

Statistical analyses were done with SPSS v. 16.0. Descriptive statistics (frequency distribution in percentage or mean and standard deviation) were used to report the demographic/clinical and socioeconomic characteristics. Spearman's rho (r_s) and Pearson's correlation coefficients were calculated to identify associations among variables. To determine the factors predicting QOL, multiple regression analysis was used. P values less than .05 were considered significant.

Results

Demographic and Clinical Characteristics

Twenty seven male children with hemophilia participated in the study. None of patients was on prophylaxis. All the patients were on-demand treatment and were treated with plasma-derived products; none of them was treated with recombinant products. In addition, none of the patients was HIV-positive or had hepatitis or a history of inhibitors. Mean age of the entire sample was

11.5±3.9 years (range 8.0 to 16.0). Most of the subjects were 8 to 12 years old (n=16, 59.26%), had hemophilia A (n=23, 85.1%) and severe hemophilia (n=11, 40.7%). The majority of subjects were infused clotting factors at the hemophilia clinic by nurses (n=14, 51.9%). The education levels in most of the mothers of the subjects were elementary (n=15, 55.6%). Sixteen (59.3%) parents of patients perceived their income as sufficient. Twelve subjects (44.4%) experienced fair health status and eleven (40.7%) suffered from hemophilia severely. Table 1 displays the demographic/clinical characteristics of hemophilic subjects.

Tables 1: The demographic/clinical characteristics of hemophilic subjects

Variable	n	%
Age (yr)		
8-12	16	59.26
13-16	11	40.74
Type of hemophilia		
A	23	85.2
B	4	14.8
Severity of hemophilia		
Mild	11	40.7
Moderate	5	18.5
Severe	11	40.7
Infusion products by		
Nurse	14	51.9
Parents	11	40.7
Patient	1	7.4
Mothers educational levels		
Illiterate	3	11.1
Elementary	15	55.6
Diploma	7	25.9
University	2	7.4
Patient income		
Poor	10	38.5
Sufficient	16	61.5
Health status		
Excellent	1	3.7
Very good	2	7.4
Good	8	29.7
Fair	12	44.4
Poor	4	14.8
Suffering from hemophilia		
Somewhat	3	11.1
Moderate	5	18.6
Consider	8	29.6
Very much	11	40.7

Mean age at the first bleeding episode was 9.7 (SD=12.7) months. For all subjects, the mean bleeding event's rate was 4.9 (SD=4.5) per month. The mean number of subjects referring to the hemophilia clinic was 2.2 (SD=1.9) per month. The mean number of school absences was 2.7 (SD=1.9) days per month.

Quality Of Life of Children with Hemophilia

The mean overall score in the Haemo-QoL was 54.1±7.3 for the entire sample. The greatest impairment in total subjects was in the friends (68.0±17.2), perceived support (66.0±16.1) and family dimensions (63.6±11.0). The dimensions with the lowest levels of impairment were relationship (44.3±12.9), coping (37.3±13.5) and others (36.9±12.6).

The mean overall scores on the Haemo-QoL for age group II and III were 53.0±8.7 and 55.6±4.6, respectively and the degree of impairment in overall QOL was high in both age groups. Table 2 shows the mean scores of the overall Haemo-QoL and each of the subscale in the age group II and III. The results for overall Haemo-QoL scores and scores for each dimension in the age groups II and III are summarized in figure 1.

Correlation between Quality of Life and Other Variables

The total score of the QOL of subjects significantly correlated with age (r=0.43, P=0.023), number of bleeding event (r=.51, P=0.006), number of school absences (r=0.47, P=0.012), and family income (r=-0.87, P<0.0001). In addition, there was an association between the mother s' education levels (r_s=-0.71, P<0.0001), severity of hemophilia (r_s=0.58, P<0.05), person who gave his/her infusions (r_s=0.67, P<0.0001), insight to health status (r_s=0.86, P<0.05) and insight to suffering from hemophilia (r_s=0.79, P<0.0001) with Haemo-QoL. However, there was no association between total Haemo-QoL score and the number of referral to the hemophilia clinic (r=0.35, P=0.07).

Table 2: Total Haemo-QoL score and scores on individual dimensions for age groups II and III

Haemo-QoL	Mean±SD	Age group	
		II (n=16)	III (n=11)
		Mean±SD	Mean±SD
Physical health	61.6±11.5	59.81±12.47	64.28±9.97
Feeling	60.9±16.4	56.91±18.13	66.75±11.96
View	47.5±16.7	40.96±13.81	57.04±16.53
Family	63.6±11.0	62.18±12.24	65.62±9.05
Friend	68.0±17.2	72.26±20.15	61.93±9.45
Perceived support	66.0±16.1	61.73±16.14	72.15±14.62
Others	36.9±12.6	32.28±10.91	43.55±12.27
Sport	57.3±13.0	53.32±13.57	63.12±10.09
Coping	37.3±13.5	40.17±16.47	33.11±5.99
Treatment	49.3±10.7	50.64±11.69	47.44±9.14
future	48.3±10.9	0.00	48.29±10.85
Relationship	44.3±12.9	0.00	44.31±12.94
Total Haemo-QoL	54.1±7.3	53.03±8.72	55.63±4.62

Age group II=8-12 years old; age group III=13-16 years old

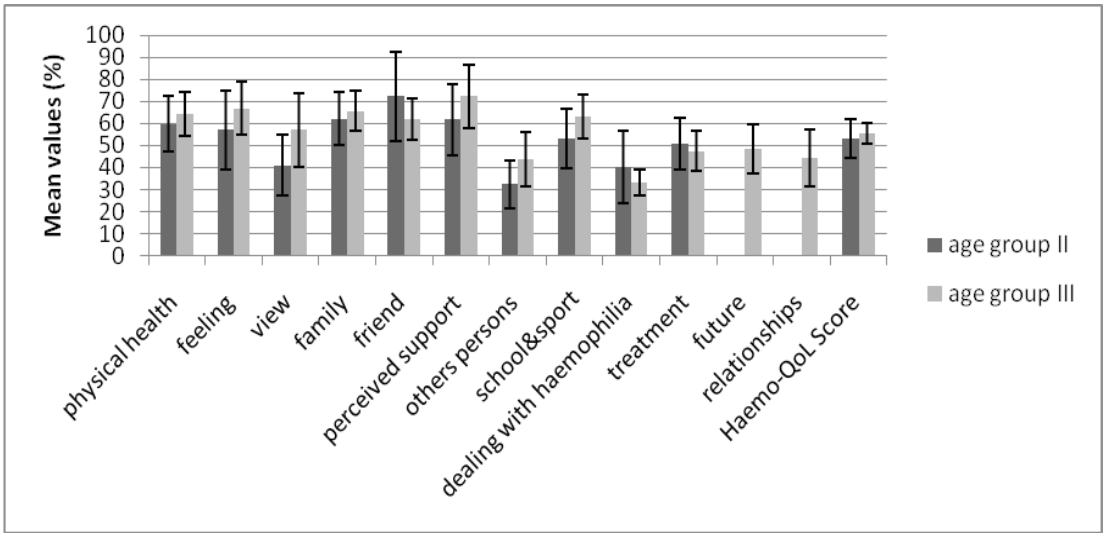

Figure 1: Mean scores for different Haemo-QoL dimensions in age group II and age group III.

Predictors of Quality of Life

A multiple stepwise regression was applied to distinguish the contribution of each selected number of variables on children and adolescent's QOL. These predictor variables were family income, age, mothers' education, number of referral to the hemophilia clinic, number of bleeding event, number of school absences, severity of hemophilia, person who gave his/her infusions, insight to health status and insight to suffering from hemophilia. It was indicated that 90% of variance of the QOL was explained by these variables. Moreover, QOL was predicted by family income (β=-0.69; explained 73% of variance) and infusion by parents (β=-0.33; explained 81% of variance). Hemophilic subjects who had a higher income and his/her infusions was done by parents reported better QOL. Other variables did not contribute to the explained variance (table 3).

Discussion

This study determined the QOL of children and adolescents with hemophilia with the Haemo-QoL questionnaire (long version). It was found that adolescents were more impaired in total Haemo-QoL scores compared to children aged 8–12 years. The highest impairment

Table 3: Predictors of quality of life in children with hemophilia (n=27; results from a multiple regression analysis).

Independent variables	β	T	P value
Family income	-0.76	-6.23	<0.0001
Parents infusion of products	-0.33	-3.11	0.005
Number of bleeding event	0.16	1.39	0.17
Number of school absences	0.13	1.16	0.25
Severity of hemophilia			
Sever	0.21	1.99	0.05
Moderate	-0.02	-0.21	0.82
Mild			
Mother s' educational levels			
Illiterate			
Elementary	-0.77	-0.62	0.54
Diploma	0.02	0.17	0.86
University	0.002	0.01	0.98
Suffering from hemophilia			
Somewhat	0.01	0.11	0.90
Moderate	-0.08	-0.72	0.47
Consider	-0.06	-0.56	0.57
Very much	0.20	1.46	0.15
Not at all			
Self-infusion of products	0.04	0.40	0.69
Health status			
Excellent	-0.07	-0.67	0.50
Very good	0.08	0.64	0.52
Good	-0.19	-1.97	0.06
Fair	0.07	0.66	0.51
Poor			
Age	0.20	1.98	0.06

r^2=0.90; Linear multiple regression, stepwise

was in the friends, perceived support and family dimensions of QOL. Total Haemo-QoL scores were associated with the number of bleeding event and school absences, family income, mother s' education level, severity of hemophilia, person who gave his/her infusions, insight to health status and insight to suffering from hemophilia. Haemo-QoL was predicted by income and infusion by parents.

The result of this study revealed that mean overall scores on the Haemo-QoL for the age group II and III were 53.0±8.7 and 55.6±4.6, respectively. Dekoven et al. reported that the mean total score of HAEMO-QOL in children aged 8-16 years and children aged 4-7 years was 33.8 (SD=15.5), and 35.0 (SD=16.1).[11] It was also shown that Haemo-QoL scores in children aged 4 to 16 years were 39.6±15.0 for the children.[13] These findings indicated that the impairments in QOL in our population was higher than that in other studies in developed and developing countries. Comparison of the QOL between the present study and others indicated that the QOL of children and adolescents with hemophilia in our study is unsatisfactory. However, Gringeri and the Haemo-QoL Group, Van der Net et al. and Broderick et al. found that QOL in children with hemophilia in developed countries (Western Europe and Australia) is satisfactory.[9,23,24] The QOL of children in these countries has improved greatly because of the widespread access of patients to home treatment and prophylaxis and participation in sports. In some of these countries, patients with hemophilia participate in sports almost as much as their healthy peers.[23,25] About two-thirds of the children with hemophilia in these developed countries were on prophylaxis,[9,26] whereas none of our study subjects was

receiving prophylactic treatment and only four of 27 children exercised regularly.

The present study suggests that the most impaired dimensions of QOL, as in developed countries,[9,23,26] were friends and perceived support; however, the impairment in our study was much greater. The absence of prophylaxis and participation in sports may account for the low QOL in children with hemophilia in our study. In comparison, younger children (age group II) had greater impairments in the friends, coping and treatment dimensions. Parental supervision and control are greater in younger children than in adolescents, so the former may not learn how to deal with their disease and are kept away from interaction with their friends. Adolescents had a higher number of bleeding episodes, referrals to the hemophilia clinic and school absences; they also showed greater impairment in other dimensions. Their problems with how they perceive themselves (reflected in the view dimension) and their interactions with others (in the others dimension) were significantly greater than children in the age group II. In the study by Gringeri et al.[9] as in our study, older children had a higher number of bleeding episodes and had lower total scores on the Haemo-QoL, although the differences were not significant in the former study.

Household income was associated with the Haemo-QoL and family income was the most important variable for prediction of QOL. It was demonstrated that income highly correlated with QOL in pediatric cancer patients.[27] Risk of adverse health status is greatest among childhood cancer patients with low household income.[28-30] Devinsky et al. denoted that lower socioeconomic condition was associated with poorer HRQOL in adolescents with epilepsy as a chronic disease.[30] In addition, this study indicated the relationship between Haemo-QoL with mothers' education levels. Family socioeconomic status such as parents' education, family income and family structure was associated with limited resources and enhanced stressors.[28-30] Family stressor influences the normal adolescents' development task,[31,32] and at least, this might negatively impact the QOL.

This study indicated a relationship between Haemo-QoL and the person who gave his/her infusions and QOL was predicted by parental administered products. It was shown that pediatric patients who were infused by family members demonstrated better adherence than pediatric self-infused ones.[18] It seems that injection impacts the relationship of parents and children. Quality of parent-child relationship is potentially a modifiable resistance factor in the teenagers,[24] and improving QOL outcome.[33]

Health status had a high impact on Haemo-QoL. This result was consistent with the study of Von Mackensen et al. They revealed patients highly impaired in orthopedic status had a worse health related QOL as compared to those with a less impaired orthopedic status.[34]

Age was the factor with a great influence on QOL in our population. As in our study, Miners *et al.* and Trippoli et al. found that age was the main predictor of QOL.[16,17] In this study with increase in age, the number of bleeding events, school absences and severity of hemophilia enhanced. These issues might negatively impact the Haemo-QoL.

As shown in the present study, a higher number of bleeding episodes and school absences correlated with impairment in QOL. Brown et al. demonstrated that the number of bleeding episodes correlated negatively with physical QOL and was associated with the number of missed work or school days.[15] Other research also found that more frequent or more severe bleeding episodes were associated with lower health-related QOL.[20] In a study by Shapiro et al. the number of bleeding events was positively associated with school absenteeism. This means that inadequate treatment and more severe disease can lead to more bleeding and may eventually cause further damage and impairment.[4]

The association between the severity of hemophilia and QOL in our study was consistent with other studies.[19,20] Poon et al. was demonstrated that individuals with more

severe hemophilia and higher self reported joint pain and motion limitation had poorer HRQOL scores, particularly in the physical aspects of HRQOL.[19] It was also revealed that the patients with various severities of hemophilia showed different impairments in their HRQoL. Severely affected patients were more impaired in their 'physical functioning' than moderate or mild patients.[21] The severity of the disease may influence joint movement, physical activity, treatment and relationships with others, and these factors might negatively impact the QOL.

None of the children or adolescents in our sample had AIDS or hepatitis. Currently in Iran, blood screening tests and the use of coagulation factors free from the viruses have led to a reduction in the incidence of infectious blood diseases in hemophilia patients. Mahdaviani et al. reported that almost all patients infected with blood-borne agents were infected before 1995.[35]

The mean age at the first bleeding episode in our study population was 9.7±12.7 months; Gringeri et al. reported that the mean age at the first bleeding episode in European children with severe hemophilia was 11 months.[9] The lower mean age at the first bleeding event in children with hemophilia in Iran is probably due to circumcision in the first months of life among boys.

Children had on average 4.9 bleeding events per month, but according to Gringeri et al., the incidence of bleeding episodes per month was 0.4 ± 0.4 in the prophylaxis group and 1.1±1.6 in the on-demand group.[9] Their findings suggested that patients who were treated on-demand had more bleeding. In a study by Woolf et al., absence from school was problematic for hemophilia patients, with an average of 18 school days missed in an academic year.[36]

The interpretation of the present study is constricted by some limitations. The main limitation was the small sample size. Another limitation of this study was that only males were studied. Both genders should be considered in future studies, as comparisons of QOL between genders may be informative. A further limitation of this study was that the age range was between 8 and 16 years. Future studies should determine the QOL for all persons with hemophilia and compare as many age groups as possible.

A family member with a chronic illness such as hemophilia may affect the parents' QOL. Therefore, additional research is needed to examine the QOL of the parents of children with hemophilia. Factors such as personal and familial experience, parents' awareness of hemophilia and the patient's functional capacity, joint functioning and social support might also be related to QOL and well-being in young persons with hemophilia. Further studies are needed to determine the relationships among these factors and how they impact the QOL.

Conclusion

The result of this study indicated that the QOL of a sample of male children with hemophilia was impaired, and that impairment according to the total Haemo-QoL score was greater among adolescents than children aged 8-12 years. Moreover, number of bleeding event, number of school absences, mother s' education level, severity of hemophilia, insight to health status and insight to suffering from hemophilia were associated with Haemo-QoL. In addition, family income and parent- administration of the product were the factors which predict the QOL in children with hemophilia. Therefore, in order to improve Haemo-QoL of adolescents, patients who were given drug infusion by third party care givers excluding parents and also patients with lower familial income are suggested to receive more effective interventions by authorities, managers and health care providers. For evidence based practice, further studies on Haemo-QoL in children are recommended.

Acknowledgement

We thank all the children, adolescents and their families who participated in this study, and the staff of Dastgheib Hospital and Hemophilia

Clinic for their support. We would particularly like to thank Professor Von Mackensen for sending the original Haemo-QoL questionnaire to us. We thank K. Shashok and S. Parand for improving the use of English in the manuscript. Financial support was from Shiraz University of Medical Sciences. The research reported in this article was done in partial fulfillment of the requirements for the Master of Science degree in pediatric nursing awarded to Shahpar Bagheri and supported by project No. 5525.

Conflict of interest: None declared

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