Hemophilia Specific Quality of Life Index in Turkish Children and Young Adults. Cross-sectional Study from a Single Center

Türk Çocukları ve Genç Erişkinlerde Hemofiliye Özgü Yaşam Kalitesi İndeksi. Tek Merkezin Kesitsel Çalışması

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ÖZ

GİRİŞ ve AMAÇ: *Hemofiliye özgü hayat kalitesi ölçeği hemofili hastaları için düzenlenmiş bir ankettir. Hemofili tedavi standartlarını, hasta ve ebeveynlerinin duygularını ve yaşadıkları güçlükleri değerlendirmek için değerli bir araçtır. Bu çalışmanın amacı hemofili hastası çocuk ve genç erişkinlerin hayat kalitesini değerlendirmektir.*

YÖNTEM ve GEREÇLER: *Faktör düzeyi <1 olan 4-25 yaştaki hemofili hastaları poliklinik başvuruları sırasında çalışmaya alındı. Hemofiliye özgü yaşam kalitesi anketi çocuklar ve vasilerine (Haemo-QoL) ve erişkin hastalara (Haemo-A-QoL) uygulandı. (www.haemoqol.de). Hepatit ve insan immün yetersizlik virusu serolojisi için taramaları da yapıldı.*

BULGULAR: *Hemofili A ve hemofili B tanılı 43 hasta çalışmaya alındı. Çalışma grubu dahil etme kriterlerine sahip hastaların %74,1 ini oluşturuyordu. Tanıda ortanca yaş 4-16 yaştaki hastalarda 11,5 (5,5-25,5) ay, 17-25 yaştakilerde 12,0(6,0-27,0) aydı. Hastaların %60 ı (26/43) evde tedavi uygulayabiliyordu. Birincil ve ikincil koruyucu tedavi uygulayanlar sırasıyla grubun %18,6 ve 69,7 sini oluşturuyordu. On altı yaş üstündeki iki hasta hepatit B taşıyıcısıydı. Faktör enjeksiyonu 4-16 yaş grubunda en kötü puanı aldı. 4-7 yaş grubunda aile ilişkiler ve tedavi kötü etkilenmişti. 8-12 yaş grubunda algılanan destek, 13-16 yaş grubunda spor ve boş zamanlar kötü etkilenmişti. 4-16 yaş gruplarında toplam Haemo-QoL puanı 29,9-34,4 arasındaydı. Ağır eklem sorunu olan hasta yoktu.*

TARTIŞMA ve SONUÇ: *Bu çalışmada hemofili hastası çocuk ve genç erişkinlerin hayat kalitesi kabul edilebilir düzeydeydi. Profilaksiye uyumu arttırmak için hasta ve ailelerin eğitimi sürdürülmelidir.*

Anahtar Kelimeler: *hemofili A, hemofili B, yaşam kalitesi, çocuk*

ABSTRACT

INTRODUCTION: *Hemophilia-specific quality of life index (Haemo-QoL) is a questionnaire designed for patients with hemophilia. It is a valuable tool to evaluate standards of care and to understand patient's and parent's feelings and challenges. This study aims to evaluate the quality of life in hemophiliac patients.*

METHODS: *Hemophilia patients 4-25-year-old and having factor level ≤1 were enrolled during their outpatient visits. Turkish version of Haemo-QoL for children and proxy and adults was applied. (www.haemoqol.de) Patients were also screened for hepatitis virus and human immune deficiency virus serology.*

RESULTS: *Forty-three patients with hemophilia A and hemophilia B were enrolled. The study group was 74.1% of the patients carrying enrolment criteria. The median age of diagnosis was 11.5 (5.5-25.5) months for the 4-16 age group and 12.0(6.0-27.0) months for the 17-25-year-old group. Sixty percent of the patients (26/43) could perform the home treatment. Patients on primary prophylaxis and secondary prophylaxis were18.60% and 69.76% of the study group, respectively. Two patients in the>16 age group were hepatitis B carriers. Factor injection was the most impaired score in the 4-16-year-old group. In the 4-7-year-old group, relations with family and treatment dimensions were poorly impaired. In the 8-12 age group perceived support and in the 13-16 age group, sports and leisure scores were poor. In 4-16-year-old groups total haemo-QoL score range was 29.9-34.4. There was no patient with severe arthropathy*

DISCUSSION AND CONCLUSION: *Quality of life of children and young adults with hemophilia is acceptable in this study. Training of the patients and families should continue for more compliance to prophylaxis.*

Keywords: *hemophilia A, hemophilia B, quality of life, child*

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INTRODUCTION

Hemophilia-related quality of life (Haemo-QoL) questionnaire (Haemo-QoL) study was started in 1998, by several European Haemophilia Care Centers because there was no hemophilia specific instrument for evaluation of quality of life in pediatric haemophilia population. The project has ended in 2002 and 3 sets of psychometrically tested questionnaire versions were performed for three age groups of children and their parents. It is available in many languages (www.haemoqol.de) and used in many countries (1).

In Turkey, all the patients with hemophilia including Syrian refugees are in social coverage and have easy access to coagulation factors especially in the last 20 years. Activities of 13 hemophilia associations are important in the training of the patients and their families. Annual national congresses and hemophilia summer schools are organized every year for clinicians, nurses, patients and their parents. Patients and parents are encouraged for prophylaxis and home treatment.

Patient reported outcome is a valuable tool to assess standard of medical care, familial and social support in hemophilia. There are about 6000 registered hemophilia patients in Turkey. They are followed by hematologists in training and research hospitals and University hospitals of goverment. There are no hemophilia centers throughout our country and pediatricians and internal medicine specialists are also authorized to prescribe coagulation factors. Patients are not obliged to keep diaries to record bleeding episodes and factor administrations. Patients prefer to visit pediatricians or internal medicine specialists when they are near to their residence. This liberal organization is good as it provides easy access to factor prescription but some patients visit hematologists rarely which results with suboptimal joint health care.

Hemophilia specific, patient reported outcome is a valuable tool for clinical and epidemiological studies. This cross-sectional study from a University hospital located in the west of the country aims to evaluate treatment standards and quality of life of a group of children and young adults with hemophilia with factor level <1.

MATERIALS AND METHODS

Data regarding the age of diagnosis, presence of inhibitor, treatment and prophylaxis modality, bleeding frequency, sport activities, education and work status, any psychological disorder were recorded. Patients were also screened for hepatitis virus and human immune deficiency virus (HIV) serology. Turkish version of Haemo-QoL (for children and proxy) and Haemo-A-QoL questionnaire (for older than 16 years) were also applied. (www.haemoqol.de)

A declared brain-dead patient whose organs are being removed for donor purposes. Approval of the local ethics comittee (KOÜ KAEK 2015/227) for the study protocol in accordance with Helsinki Decleration 2008 agreements and written informed consents of the patients and legal guardians were obtained.

Haemo-QoL questionnaire for younger children (age group I: 4–7 years) has 21 items in 8 dimensions (physical health, feelings, view, family, friends, others, sport and school/kindergarten, treatment). For older children (age group II: 8–12 years) there is a self-administered questionnaire which has 2 more domains (received support and dealing) with a total of 64 items; for adolescents (age group III: 13–16 years), with 2 more domains (relationships, future) there are 77 items. Haem-QoL for adults (age group IV:16-25 years) contained 10 dimensions and 46 items (family

planning was also added). The "physical" dimension asks questions about pain and bleeding, restriction of movement, etc. The "feeling" dimension asks children's feelings about their disease. The dimension of "view" aims at how children perceive themselves. The "family" dimension aims evaluation of interaction within the family. The "friends" dimension evaluates the patient's interaction with friends. The dimension "perceived support" asks the patient's perception regarding the support received from others. The "sport and school" dimension is about restrictions from sports and social activities at school. The dimension "dealing" is about how children deal with their hemophilia. The dimension "treatment" is about replacement treatment etc. The dimension "others" is about interaction with others. Dimensions "future" and "partnership" aims evaluation of feelings about the future, having a girlfriend and family. Proxy versions of the Haemo-QoL questionnaire were also answered by the parents. Scores ranged between 0 and 100. High scores identified poor Haemo-QoL. The score calculation method can be found on the website www.haemoqol.de.

Statistics

Statistical Package of the Social Sciences version 22.0 was used for statistical evaluation. For evaluation of the distribution of numeric variables, the Shapiro-Wilk test was used. For the normal distribution, the mean and standard deviation was calculated. For comparison of categoric variables, the Ki-square test was used. For comparison of numeric variables with normal distribution Independent-Samples T-test and one-way ANOVA test; for abnormal distribution of numeric variables Mann-Whitney U test and Kruskal-Wallis test was used. Results were given in a 95% confidence interval with a p-value <0.5.

RESULTS

Forty-three of the patients with factor level ≤ 1 that visited the outpatient department during six month study period and had given consent for the study were enrolled. There were two patients (4.6%) with hemophilia A and high responding (>5BU) inhibitors. The highest inhibitor levels of these patients were 11.2 BU and 128 BU and they received treatment or prophylaxis with bypassing agents. The study group was 74.1% of the patients carrying enrolment criteria. Residences of only ten patients were in the city of the University hospital; whereas thirty-three patients were from neighboring cities where there were no hematologists.

Demographic data of the study group was shown in Table 1. Haemo-QoL scores of children in different age groups and young adults are shown in Table 2 and Figure 1. Haemo-QoL proxy scores of the study groups were shown in Table 3.

When study groups were compared, scores of family and treatment dimensions were statistically significant. In the family dimension, significance was between group I and III ($p<0.005$). In the treatment dimension, significance was between group I and II ($p<0.009$), between group I and III ($p<0.007$), between-group I and IV ($p<0.049$), between groups II and IV ($p<0.019$), and between groups III and IV ($p<0.027$) (Table 2).

When scores of children (group I-II-III) were compared with their proxy scores, there was no significance in any dimension.

In proxy reports, there was a significant difference between scores of different age groups in family and sport, and school dimensions. In family and sports and school dimensions, significance was between groups II and III ($p<0.013$ and $p<0.047$, respectively). (Table 3). There was no patient with severe arthropathy.

Table 1. Characteristics of the pediatric and adult hemophilia patients

Variables	Group I/III (4-16 years) n (%)	Group IV (17-25 years) n (%)	P
Hemofili A	19 (8.6)	17 (81.0)	N
Hemofili B	3 (13.6)	4 (19.0)	
Factor level <1	9 (40.9)	9 (42.9)	0.897[a]
Factor level= 1	13 (59.1)	12 (57.1)	0.897 [a]
Patients with inhibitor	1(4.5)	1(4.8)	
Patient with history of inhibitor	1(4.5)	0	1.000 [a]
Median age at diagnosis (month)	11.5 (5.5-25.5)	12.0 (6.0-27.0)	0.634 [a]
Age of first joint bleeding (year)	2.5 (1.2-4.5)	1.7 (1.0-6.5)	0.519 [a]
Patients with central nervous system bleeding	2 (9.1)	4 (19.0)	0.412 [a]
Patients with gastrointestinal bleeding	0	2(9.5)	0.233 [a]
Patients with hematuria	3(13.6)	2(9.5)	1.000 [a]
Patients with hepatitis B seropositivity	0	2(9.5)	0.233 [a]
Patients with hepatitis C seropositivity	0	0	
Patients with HIV seropositivity	0	0	
Patients with history of depression	1(4.5%)	2(9.5)	0.607 [a]
Patients visiting psychologist	0	3 (14.3)	0.108 [a]
Patients visiting psychiatrist	0	1(4.8)	0.488 [a]
Patients on primary prophylaxis	5(26.3)	3(15.8)	0.693 [a]
Patients on secondary prophylaxis	14(73.7)	16(84.2)	0.693 [a]
Median age for starting prophylaxis (years)	6.0(2.0-10.0)	13.0(11.0-16.0)	**<.001** [a]
Patients performing home treatment	9 (47.4)	17 (89.5)	**.005** [a]
Annual joint bleeding rate (median)*	4 (0-40)	7 (1-25)	
Patients underwent radiosynovectomy	5(22.7)	11(52.4)	**.044** [a]
Patients underwent artroscopic synovectomy	1(4.5)	4(19.0)	0.185 [a]
Patients performing sport activity	6(27.3)	4(19.0)	0.721 [a]

In group I-III 14 patients, in group IV only 7 patients answered, generally depends on memory a:Chi-square test

Table 2: Haemo-QoL scores of children in different age groups and in young adults.

	Grup-I (n=6) Mean±SD	Grup-II (n=5) Mean±SD	Grup-III (n:11) Mean±SD	Grup-IV (n=21) Mean±SD	p
Bleeding	14.10±25.0	39.03±26.38	29.86±29.12	-	.290 [a]
Factor injections	62.77±26.0	65.00±22.36	65.90±26.34	-	.912 [a]
Physical health	31.25±22.0	35.70±22.87	31.16±22.30	35.47±18.90	.983 [a]
Feelings	30.53±26.68	27.85±18.45	22.44±15.73	31.54±27.06	.863 [a]
View	16.66±25.81	18.84±25.83	23.86±17.47	35.0±19.87	.115 [a]
Family	58.33±12.90	40.0±19.36	33.62±16.30	-	**.022 *** [a]
Friends	41.66±37.63	33.75±24.04	43.18±16.64	-	.785 [a]
Others	20.83±18.81	14.98±13.37	17.42±19.43	-	.784 [a]
Sports and school/ kindergarden	33.32±21.08	36.87±16.74	42.14±16.38	-	.609 [a]
Treatment	70.83±33.22	17.13±14.59	25.81±11.0	38.98±16.38	**.003 **** [a]
Perceived support	-	62.50±25.38	44.88±20.88	-	.209 [b]
Dealing	-	21.42±10.71	24.34±13.34	16.11±21.90	.114 [a]
Relationship	-	-	17.04±33.66	-	-
Future	-	-	29.54±17.91	30.23±23.53	1.0[b]
Sports and leisure	-	-	-	62.14±25.57	-
Work/school	-	-	-	31.84±29.96	-
Family planning	-	-	-	15.17±19.82	-
Relationship/ partnership	-	-	-	3.17±8.92	-
General health	25.0±27.38	35.0±28.50	36.36±23.35	-	.717 [a]
Total score	36.8±16.4	34.4±12.5	32.5±8.1	29.9±14.3	.685 [a]

Significance was between group I and III.

****Significance was between group I and II (p<0.009), between group I and III (p<0.007), between group I and IV (p<0.049), between groups II and IV (p<0.019) and between groups III and IV (p<0.027).***

a:One Way ANOVA test

b:Independent Samples T test

Table 3. Haemo-QoL proxy scores of the study groups

	Group-I (n=6) Mean±SD	Group-II (n=5) Mean ±SD	Group-III (n:11) Mean ±SD	p
Bleeding	16.66±25.81	34.54±21.93	23.57±28.30	.473[a]
Factor injection	62.50±22.07	69.0±20.73	63.63±28.20	.857[a]
Physical health	33.33±31.04	47.11±15.62	42.52±25.22	.676[a]
Feeling	19.43±25.06	33.55±19.48	23.0±15.76	.301[a]
View	12.50±25.0	18.85±16.09	23.63±20.32	.146[a]
Family	46.87±15.68	58.0±15.24	35.22±15.44	**.029***[a]
Friends	25.0±15.81	27.5±26.73	56.81±29.77	.079[a]
Perceived support	-	47.5±9.47	51.70±31.63	1.0[b]
Others	6.25±15.30	16.64±4.15	8.70±9.40	.056[a]
Sports and School	-	53.12±11.26	35.06±17.57	**.047****[b]
Dealing	-	25.70±11.12	22.06±19.35	.357[b]
Kindergarten	45.78±29.69	-	-	-
Treatment	47.91±28.95	21.78±11.46	23.0±19.07	.125[a]
Future	-	-	17.04±15.58	-
Relationship	-	-	5.68±8.59	-
General Health	37.50±26.22	30.0±20.91	43.18±22.61	.561[a]
Total score	32.1±14.1	37.1±3.6	31.6±12.2	.669[a]

**Significance was between groups II and III (p<0.013)*
*** Significance was between groups II and III(p<0.047)*
[a]:One Way ANOVA test
[b]:İndependent Samples T test

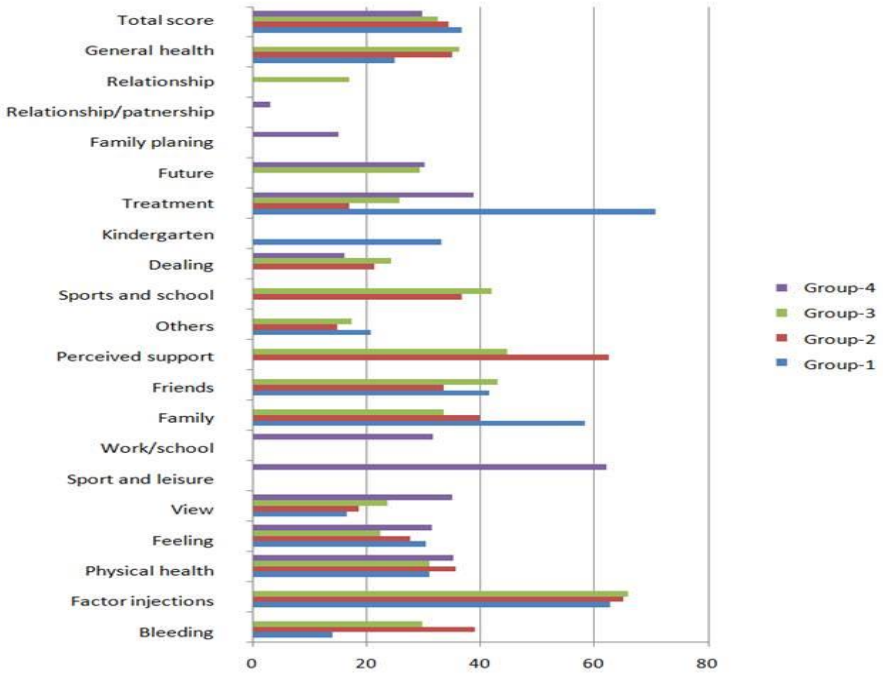

Figure 1. Comparison of Haemo-QoL scores of hemophiliac patients in different age groups.

DISCUSSION

In this single-center study comprising 4-25-year-old hemophilia patients with a factor level ≤1, demographic data showed that the age of diagnosis gets younger compared to the previous study of the center. In that study median age of diagnosis was 8.5±10.4 years in adult patients whereas it is about 12 months in the study patients. This is due to the availability of pediatric hematologists in the region since 2000 (2).

The incidence of intracranial hemorrhage (ICH) seems high (6/43) in the study. All except one ICH episode was before the diagnosis of hemophilia. A five-year-old hemophiliac boy who was not on prophylaxis suffered from ICH due to cranial trauma in the playground. In a term newborn, ICH was detected after an uneventful vaginal delivery. Neuromotor retardation of this boy was the only severe sequel among patients with ICH.

A multicenter international study of children with severe hemophilia showed that the overall incidence of intracranial hemorrhage was 0.7 /100 patient-years. Patients on full prophylaxis had 51 times lower incidence than patients on demand. The incidence for ICH was higher between the ages 1–3 years for patients treated on demand (3).

In our center due to the easy availability of factor concentrates, fresh frozen plasma is rarely used for the treatment of bleeding episodes in the last 20 years. Blood-borne virus contamination is not a problem for young hemophiliacs. Family screening showed that the hepatitis B carrier state of one patient is probably due to vertical transmission.

There were only two patients (4.6%) with high responding inhibitors in the study group. Immune tolerance induction (ITI) could not be started due to restrictions of social coverage. In our country, there were some experiences with low-dose ITI. In a case report, it was shown that 50 U/kg thrice-weekly eradicated low responding inhibitor in the 15th dose administration of factor concentrate (4). In a multicenter study from Turkey 21 hemophilia, patients with high titer inhibitors were enrolled. . A low dose regimen (50 U/kg thrice weekly) was used; immune tolerance could be achieved in 26.3% of the patients (5). In patients with inhibitors, prophylaxis with bypassing agents was administered but was not as effective as prophylaxis in patients without inhibitors. It was not possible to compare scores of patients with and without inhibitors due to the small number of patients with inhibitors. Patients with inhibitors were 4.5 and 14 years during the study. The latter patient with the highest inhibitor 11.2 BU, was dependent on a wheelchair for nine months due to knee arthropathy although radiosynovectomy was performed on the target joint. The first admission to the hematology department was when the boy was six years old although the diagnosis of the elder son was severe hemophilia. Inhibitors developed in the first 30 exposure to factor concentrate. Fortunately, inhibitors were lost at the end of the four years and prophylaxis was shifted to plasma-derived, von Willebrand factor rich factor concentrate from by-passing agents, and the joint health of the boy improved and he then lived an active life. The other boy was diagnosed when he was three-month-old and asymptomatic his sibling had already severe hemophilia. This boy also developed inhibitors in the first 30 exposure to factor concentrate. . The highest inhibitor level was 128 BU. Prophylaxis with by-passing agents was administered. There were periods when inhibitor level decreased to lower than 5 BU. But recovery was poor even in a period when the inhibitor was even <0.6 BU. He suffered rarely from spontaneous joint bleeds under

recombinant factor VII prophylaxis thrice weekly. The product was available with special permission of off-label prophylaxis.

Eighteen percent of patients (8/43) were on primary prophylaxis whereas 69% (30/43) were on secondary prophylaxis. The median age of starting prophylaxis was younger in children compared to young adults which can be explained by training and easily availability of factor concentrates. Sixty percent of patients (26/43) could perform the home treatment. The frequency of home treatment increased as the boys get older. Compared to historical data there is an increasing tendency for home treatment, especially in adult patients. In the previous study, only 52.9% of adults could perform the home treatment (2). Minority of the patients had bleeding diaries. Some patients on secondary prophylaxis either showed poor compliance or they had already target joints before initiation of prophylaxis. In a study of 39 adolescent boys with severe hemophilia from our country, compliance to prophylaxis was about 50%. They reported three reasons for non-compliance: time constraints, being tired of treatment, and problems with vascular access (6). Fortunately some young patients were reporting zero or only one annual joint bleeding. Prescribed prophylactic factor VIII dose was generally 25-40 U/kg twice or thrice weekly.

HaemoA-QoL scores of patients in the 4-7 age group were severely affected in family, treatment, and factor injections dimensions. The only statistically significant difference between patients in different age groups was in family and treatment dimensions. In the family dimension, the poorest score was in the 4-7 years-old age group and the best score was in the 13-16 years-old age group. Statistical difference was between 4-7 and 13-16 age groups. Families express that when the patients are older than 12 they are more familiar with the features of hemophilia, limitations from some activities, and show more compliance to treatment and relations with parents. Treatment scores also improve with age and in all groups including patients older than seven years, treatment scores are significantly improved compared to 4-7 years-old patients. The best treatment scores were in the 8-12 and 13-16 years-old group. Patients older than 16, try to gain autonomy and may refuse parents' control on their treatment and prophylaxis; treatment scores of these young adults were significantly worse compared to the 8-16 age groups. Family relations were severely impaired due to the limitations of the parents to save their children from trauma in young children. Some mothers stayed at the school garden to protect their children from trauma during brakes and they preferred factor injections to be performed by nurses instead of home treatment not to impair relations with their young children.

Novel agents, with their prolonged half-life, higher trough levels, and some with subcutaneous administration will be a solution to frequent injections and venous access problems. But they are not available for Turkish patients yet. Phase III study of 20 hemophilia B patients in 4-12 age group with rIX-FB weekly intravenous dosing in a median study period of 12 months showed improvement in HRQoL. There was also improvement in attending classes, physical activity, and treatment burden of caregivers related to their hemophilic child (7). Emicizumab is a recombinant antibody that mimics the cofactor function of FVIIIa and has a long half-life. It also has the advantage of subcutaneous administration. In hemophiliacs with inhibitors (>12 years), emicizumab prophylaxis was associated with meaningful improvements in health-related outcomes (8). Proxy scores showed that the poorest scores were in factor injections in

all age groups. Parents' poorest score was for the 4-7 age group. A European multicenter study including a Turkish center also showed that hemophilia care had a great psychosocial burden on parents; 66.2% reported that hemophilia affected their life; 26.8% reported an economic impact; 57.6% reported their child's inability to do certain things, they lost about 8.35 ± 14.5/year days due to hemophilia care, they spent ≥5 h/mo infusing (9).

In the 8-12-year-old group, the poorest scores were in factor injection (treatment) and perceived support dimensions. In the 13-16-year-old group factor injection score was again the poorest score. Also, friends and perceived support were seriously impaired. In young adults, sports and leisure was the poorest dimension. Family and treatment dimension scores were significantly different between the groups. (Table 2, Figure 1). Total haemo-QoL scores were similar among groups both in the patients and proxy; p=0.685 and p=0.669, respectively. In the 4-16-year-old group, the total score range was 32.5±8.10- 36.8±16.40. When compared with multicenter European study haemo-QoL scores were more impaired (1). In proxy groups, similar to patient scores total score range was 31.60±14.10-37.10±3.60. In proxy reports also factor injections were the poorest scores in all age groups similar to the patients' reports. The family dimension was severely impaired in 4-12-year-old patients whereas in 13-16 age-group friends dimension was more impaired. Perceived support was quite impaired in the 8-16-year-old group.

When parents' impression about their children's disease was evaluated with proxy scores of HaemoQoL, the significant difference between different age groups was in the only family and sports and school dimensions. Relations with family was better in the 13-16 age group. The poorest score was in the 8-12 age group and the difference with the 13-16 age group was significant.

Two young adult patients reported a history of depression and four patients visited a psychiatrist or psychologist. Studies,

Studies show that people with hemophilia have more anxiety and depression symptoms. It is suggested that these symptoms were mainly associated with their professional status, pain interference, physical activity, and the perception of consequences underlying hemophilia (10). In the Netherlands an organization for the psychosocial care of children with hemophilia and their parents is established. Social workers and psychologists are also the members of this multidisciplinary team of hemophilia comprehensive care centers. In addition to camps and other facilities, children and/or patients are invited at least four times during childhood to the outpatient clinic; before attending primary school, before attending high school, and at about 16-17 years. A multidisciplinary approach and support are performed for these patients and parents (11).

Training should be continued to increase patients' and parents' compliance to prophylaxis. A systematic analysis was performed to evaluate the long-term benefits of prophylaxis in patients with hemophilia. Nineteen studies published in English from Europe and North America between 2006-2016 (84% were children) with a median study follow of 5-19 years were analyzed. For patients on prophylaxis, joint health and bleeding rates were better and they had fewer hospitalizations or surgeries. They were not disabled and had good health-related Qo (12).

In a multi-center study of 43 patients with severe hemophilia from our country, 36-Item Short-Form Health Survey (SF36) and the State-Trait Anxiety Inventory (STAI) were used to evaluate the

quality of life and anxiety of patients or parents. Median QoL score was lower in patients>21 years old than parents of patients 2-13 years old and patients 14-21 years old ($p < 0.05$ for both). Median anxiety level was higher in parents of patients 2-13 years old and patients>21 years old ($p < 0.05$ for both).

The study showed that prophylaxis would improve QoL and reduce anxiety for young adults with severe hemophilia A. They also suggested that home treatment would reduce the anxiety of children about injections (13).

The limitation of the study is, joint evaluation is not performed and the annual bleeding rate is reported approximately by the patients.

CONCLUSION

Haemo-QoL scores of the patients in this study are acceptable. But improving quality of life in hemophilia may be possible a) If patients can be easily referred to pediatric hematologists and diagnosis is not delayed, b) Factor concentrates are easily available c) training of the patients and families for prophylaxis and home treatment is well-organized d) compliance to prophylaxis is good e) Products with extended half-life and/or subcutaneous administration are available.

Ethics Committee Approval: Ethical approval was obtained.

Kocaeli Univesity Clinical Research Ethics Committee (14/07/2015 date and 19/13 number)

Conflict of Interest: There is no conflict of interest.

Funding: No financial support was received.

Informed Consent: This a retrospective study.

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