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Health – Related Quality of Life Assessment in Filipino Children with Hemophilia Aged 4 To 16 Years in a Tertiary Hospital

Anna Marie D Espaldon[1]* and Flerida G Hernandez[2]

[1]Fellow, Pediatric Hematology/Oncology Department of Pediatrics, University of Santo Tomas Hospital, Philippines
[2]Consultant, Pediatric Hematology/Oncology, Department of Pediatrics, University of Santo Tomas Hospital, Philippines

Abstract

Background: Determining how hemophilia and its treatment may affect the quality of life of children with this bleeding disorder is feasible by using hemophilia-specific Quality Of Life (QoL) questionnaire.

Objectives: To determine the health-related QoL of Filipino children with hemophilia treated in a tertiary hospital, specifically to identify the factors that significantly influence the quality of life of these children, and correlate sociodemographic data with patient and parent reported health-related quality of life.

Methods: A cross-sectional study was conducted on all children with hemophilia aged 4 to 16 years and their parents who are seen at the University of Santo Tomas Hospital Hemophilia Out-Patient Clinic for follow up care from July to December 2012 using the Haemo-QoL questionnaire for Filipinos.

Results: Fifty one patients were included in the study. Group I showed most impairment in the subscale of Family (43.75 ± 36.8), while Groups II and III showed most impairment in Sports and School subscale (Group II: 58.2 ± 18.77; Group III: 59.27 ± 17.46). Group I showed least impairment in Attitude (6.25 ± 7.60) while Groups II and III had least impairment in Treatment subscale (Group II: 12.5 ± 15.26; Group III: 23.99 ± 11.02). Parents' scores reflect the same areas of concern with that of the child's. The total mean TSS in our study is 28.39 ± 4.76, reflecting the good QoL of our patients.

Conclusion: Identifying the areas of impairment among children with hemophilia and their parents can help clinicians address these concerns; improve their understanding of the disease and their quality of life.

Keywords: Hemophilia; Quality of Life; Sociodemographic Data

Introduction

Throughout the years, medical science has garnered vast progress in terms of diagnosis and management of chronic diseases and parallel to this achievement is the emergence of disease-related long term sequelae and complications. Hemophilia is a life-long bleeding disorder due to deficiency of clotting factors VIII and IX for Hemophilia A and Hemophilia B, respectively. These coagulopathies are characterized by bleeding that may occur with or without trauma, and management necessitates replacement of the deficient clotting factor, therapeutically or prophylactically [1]. In the absence of or insufficient medical support, bleeding into joints, muscles and other organ systems such as the genito-urinary and the central nervous system, leads to severe joint damage, pain and multi-organ bleeding, causing permanent disability and significant impairment in the Quality Of Life (QoL) of patients [2].

The World Health Organization has defined QoL as "an individual's perception of their position in life in the context of the culture and value systems in which they live, and in relation to their goals, expectations, standards and concerns" [3]. Health – Related Quality of Life (HRQoL), a type of QoL concept, is defined by Kleinman as "the individual experience of illness with the psychosocial response to disease- related and treatment-related symptoms" [4]. Measuring the HRQoL in a quantifiable basis can be of benefit in identifying the subsets of patients who are at risk for hemophilia-related problems, give an estimate of the disease burden and determine treatment satisfaction. Defining how these factors may affect the quality of life of children with this bleeding disorder is feasible by evaluating the physical, emotional, mental, social, and behavioral elements of well-being and function as determined by the patients and observers, leading to the development of the hemophilia-specific QoL questionnaires [2,5].

The European Haemo-QoL Project was spearheaded by Dr. Sylvia von Mackensen of Germany, aims to develop and test a quality of life assessment tool for children and adolescents with hemophilia. The project included 339 children with hemophilia from 20 centers, as well as their parents or guardians from from six West European countries (Germany, Italy, France, Spain, Netherlands, and the United Kingdom) active in developing a pilot test version of the Haemo-QoL questionnaire. In 2002, the project produced 3 sets of the Haemo-QoL questionnaire for three age groups of respondents based from expert discussions and literature reviews. It has been tested in a pilot study within three age groups: children aged 4-7 years, 8-12 and adolescents aged 13-16 years. Pilot testing of the Haemo-QoL questionnaire was carried out and was then linguistically validated in 33 language versions [6,7].

Several domains were addressed by the Haemo-QOL questionnaires. These domains were defined by the Haemo-QoL group as follows: The Physical scale includes questions regarding pain, mobility and anxiety; Feeling is concerned with the children's mood, emotional consequence and actions in relation to hemophilia; Attitude scale deals with relationships to others and to own person; the Family scale deals with restrictions, the child's position in the family, problems at home, activities of the parents and own feelings in the family; Friends scale deals with the child's relationship, feelings of anxiety and activities with his friends; Perceived Support scale has questions about how the

***Corresponding author:** Anna Marie D Espaldon, University of Santo Tomas Hospital, Manila, Philippines, Tel: +639228537986; E-mail: mgy_md@yahoo.com

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children recognize social support or estrangement and isolation they receive from others; Sports and School asked about sports participation and school activities and the Coping asked questions about the children's control and emotional acceptance of their hemophilia. Hemophilia care and treatment side effects were asked in Treatment scale; how they interact with others was asked in the Other Persons; and Future and Relationships asked about their views of the future and the possibility of having relationships with the opposite sex [5].

In 2011, translation to the Filipino language and pilot testing of the Haemo-Qol questionnaire was done on 34 children with hemophilia and their parents by Dr. Puno-Mendoza and colleague in collaboration with Dr. Sylvia von Mackensen of the Haemo-QoL group. The linguistic validation process was done by professional bilingual translators from the University of the Philippines, Sentrong Wikang Filipino. Revisions were made based on child's responses, giving rise to a 16-item Haemo- QoL questionnaire from 21 items for Group 1, 60-item questionnaire from 63 items for Group 2 and 73-item questionnaire from 76 items for Group 3. Likewise, modifications were made based on parents' responses. A 12-item questionnaire for parents was made from 21 items for Group 1, 61-item questionnaire from 64 items for Group 2 and a 69-item set from 74 items for Group 3. The internal consistency of the Peds-QL, which is the generic measure used in the validation process, and the Haemo-QOL was measured by Cronbach's Alpha Coefficient. Values of ≥ 0.70 were considered acceptable for comparison. The revised Haemo-QoL for Filipinos showed sufficient reliability and validity to measure HRQoL of children with hemophilia [8].

Objectives

General objective

To determine the health-related quality of life of Filipino children with Hemophilia treated in a tertiary hospital.

Specific objectives:

1. Identify the factors and domains that significantly influence the quality of life of children with hemophilia.
2. To compare the HRQoL as reported by children (Child Reports) versus the HRQoL reported by parents or guardians (Parent Reports).
3. To correlate demographic data with child and parent reported HRQoL.

Definition of Terms

1. Moderate Hemophilia- persons with 1-5% clotting factor activity. Bleeding may occur with moderate injury.
2. Severe Hemophilia-persons with <1% clotting factor activity; bleeding may occur spontaneously.
3. Inhibitors-are antibodies that either inhibit the activity or increase the clearance of a clotting factor.
4. Prophylaxis- a routine scheduled replacement of clotting factor concentrate in people with hemophilia.

Methodology

After approval from the Department of Pediatrics' Ethics Committee was secured, the Filipino version of the Haemo-QoL questionnaire was administered to all children diagnosed with Hemophilia, aged 4 to 16 years and their parents or guardians who were seen at the University of Santo Tomas Hospital Hemophilia Out-Patient Clinic for follow up care from July to December 2012. The diagnosis of hemophilia was made by determining the serum factor VIII or factor IX level for hemophilia A and B respectively. Factor VIII and IX levels were measured by coagulation assay.

Informed assent and consent were signed by both child and parent, respectively. Eligible in the study were children with moderate and severe hemophilia without inhibitor, not on factor prophylaxis, can read and comprehend the questionnaires and has no co-morbid conditions. Data such as age, type of Hemophilia, severity of the disease, relationship of respondent to the patient and Socioeconomic Status (SES) were obtained apart from the clinical parameters included in the Haemo-QoL questionnaire. SES was classified as either bottom class or upper class, based on assigned hospital class. Families with >P20, 000.00 monthly income were classified as upper class while service patients or those with a monthly income <P20, 000.00 were classified as lower class.

The subjects were divided into 3 age groups namely Group I: 4 to 7 years, Group II: 8 to 12 years and Group III: 13 to 16 years, based from the age grouping done by the Haemo-QoL group. The 4 to 7 year age group had the researcher-administered questionnaire while the 8 to 12 - year and 13 to 16-year age groups had self-administered questionnaires. Correlation between self and parent reports were determined by repeated measurements. Comparison between age group, severity of disease and social economic status were measured using ANOVA F-test. Analyses were performed with two-sided tests with statistical significance set at $P<0.05$. The SPSS for Windows (version 16) was used for data analysis.

An interview version for Group I child respondents included items pertaining to seven subscales (Physical Health, Feelings, Attitude, Family, Friends, Sports and School and Treatment. Group II questionnaire included two additional subscales (Perceived Support and Coping) while two more subscales were added (Future and Relationships) to Group III. Parents' questionnaires included 12, 60 and 73 items for Groups I, II and III respectively. The subscales of Attitude, Perceived Support and Other Persons were deleted from the Group I parents' questionnaires since the Cronbach's Alpha is <0.7 [8].

The questionnaires have five Likert responses scaled from 1 to 5 for groups II and III and 1 to 3 for group I. Scores were calculated by converting the raw score to a Transformed Scale Score (TSS) which would allow comparison between answers of different age groups. Essentially, a high score means a low quality of life. A scoring manual is provided in the website http://www.haemoqol.de.

Results

A total of 51 children with hemophilia and their parents or guardians were recruited for this study with 8, 19 and 24 respondents, with mean age of 6, 10 and 15 years respectively. All subjects were able to complete the questionnaires. Ninety percent (46/51) have hemophilia A, 84% of whom have moderate disease (43/51) and 78% (40/51) of respondents had <2 episodes of bleeding/month. Mothers comprised 74% (38/51) of parent respondents. Eighty six percent (44/51) belonged to the lower socioeconomic status (Table 1).

Means and standard deviations of the respondents' score in the Haemo-QoL domains were presented in Table 2. Group I child respondents showed most impairment in the scale of Family (mean TSS 43.75 ± 36.8), which was also reflective of the parents' score of 64.06 ± 38.11. Group I children showed least impairment in the scale

Group	I	II	III	Total
Demographic characteristics	n = 8 (%)	n = 19 (%)	n = 24 (%)	51 (%)
Mean Age (years)	6	10	15	
Type of Hemophilia				
Hemophilia A	8 (100)	17 (89.47)	21 (87.5)	46 (90)
Hemophilia B	0	2 (10.52)	3 (12.5)	5 (10)
Severity of Disease				
Moderate	6 (87.5)	16 (84.21)	21 (87.5)	43 (84)
Severe	2 (12.5)	3 (15.79)	3 (12.5)	8 (16)
Number of Bleed/Month				
<2	5 (62.5)	15 (78.94)	20 (84)	40 (78)
>2	3 (37.5)	4 (21.05)	4 (16)	11 (22)
Parent Report respondent				
Mother	7 (87.5)	15 (78.95)	16 (66.66)	38 (74)
Father	1 (12.5)	3 (15.79)	5 (20.83)	9 (23)
Others	0	1 (5.26)	3 (12.5)	4 (3)
Socio-economic Status				
Lower	6 (75)	17 (89)	21 (88)	44 (86)
Upper	2 (25)	2 (11)	3 (12)	7 (14)

Table 1: Demographic and medical characteristics of patients with Hemophilia

	GROUP I			**GROUP II**			**GROUP III**		
HRQoL Scales	**Child Reports Mean TSS (SD)**	**Parent Report Mean TSS (SD)**	***p***	**Child Reports Mean TSS (SD)**	**Parent Report Mean TSS (SD)**	***p***	**Child Reports Mean TSS (SD)**	**Parent Report Mean TSS (SD)**	***p***
Physical Health	35.42 (35.78)	33.09 (24.15)	0.321	31.23 (18.66)	36.68 (18.92)	0.956	36.11 (16.43)	50.54 (22.38)	0.145
Feelings	16.67 (20.41)	22.32 (11.81)	0.172	30.91 (20.42)	39.28 (22.42)	0.713	28.68 (23.94)	40.92 (15.6)	**0.040**
Attitude	**6.25**§(7.60)	NA		32.52 (12.01)	45.34 (13.63)	0.619	31.36 (14.71)	36.46 (18.03)	0.335
Family	**43.75***(36.80)	**64.06***(38.11)	0.951	29.26 (17.21)	46.35 (20.24)	0.523	31.55 (18.78)	31.77 (11.74)	0.280
Friends	12.50 (16.67)	39.16 (19.03)	0.850	32.35 (22.41)	36.12 (25.39)	0.623	30.97 (22.2)	34.03 (17.16)	0.225
Perceived Support	NI	NI		35.29 (15.15)	46.69 (12.88)	0.524	35.68 (14.93)	36.79 (15.93)	0.759
Other Persons	NI	NI		33.08 (12.68)	40.59 (19.32)	0.102	25.17 (12)	30 (15.91)	0.183
Sports and School	10.42 (13.46)	**19.84**§(17.12)	0.644	**58.20*** (18.77)	**56.76*** (11.25)	0.480	**59.27*** (17.46)	**52.82*** (12.92)	0.156
Coping	NI	NI		30.38 (13.32)	37.01 (11.24)	0.505	29.68 (12.4)	27.43 (16.84)	0.150
Treatment	27.08 (20.69)	NA		**12.50**§ (15.26)	**23.99**§ (11.02)	0.204	**16.15**§ (14.94)	**23.09**§ (18.98)	0.285
Future	NI	NI		NI	NI		28.6 (20.72)	36.46 (18.1)	0.522
Relationship	NI	NI		NI	NI		17.24 (21.25)	NI	

TSS – Transmuted Scale Score; NA – Not Applicable; NI – Not Included in the Group Questionnaire; * - Most Impaired; § - Least Impaired

Table 2: Comparison between HRQoL means TSS between child reports and parent reports.

of Attitude. Both Groups II and III child respondents showed most impairment in the scale of Sports and School (Group II: 58.2 ± 18.77; Group III: 59.27 ± 17.46) and least impairment in the Treatment scale (Group II: 12.5 ± 15.26; Group III: 16.15 ± 14.94).

Parents' scores reflect similar areas of most impairment as the child's. Group1 parents had most impairments in the scale of Family (64.06 ± 38.11,) while Groups II and III parents showed most impairments in the scale of Sports and School, with TSS of 56.76 ± 11.25 and 52.82 ± 17.46 respectively. Least impairments were seen in the scales of Sports and School (19.84 ± 17.12) for Group I, and Treatment for Groups II (23.99 ± 11.02) and III (16.15 ± 14.94). Significantly different child - parent responses was only noted in Group III respondents, in the scale of Feelings (28.68 ± 23.94 vs. 40.92 ± 15.6; p = 0.040).

Tables 3, 4 and 5 show the comparison between HRQoL scales and demographic characteristics.

Statistically significant correlations between HRQoL and demographics were seen in Group III respondents only. The scale Attitude was significantly affected across all demographic characteristics with people with Hemophilia B (TSS 47.22 versus Hemophilia A TSS 29.10; p=0.013), severe disease (TSS 45.36 versus moderate disease TSS 29.36; p=0.040) and >2 bleeding episodes/month (TSS 36.80 versus <2 bleeding episodes/month TSS 30.27; p=0.017) having more impediments in their dealings with themselves and their relationships with others. Concerns about the future was significantly impaired among those with severe disease (TSS 54.16 versus moderate disease TSS 24.95; p=0.022) and those with>2 bleeding episodes/month (TSS

	Physical	Feelings	Attitude	Family	Friends	School	Treatment
Severity of Disease							
Moderate	25	13.89	2.78	33.33	12.5	5.56	27.78
Severe	66.66	25	16.66	75	12.5	24.99	25
P	0.422	0.258	NA	0.852	0.873	0.851	0.362
Number of Bleeds/Month							
<2	26.67	23.33	10.00	60.00	5.00	10.00	26.67
>2	50.00	5.55	0.00	16.67	25.00	11.11	27.78
P	0.658	0.261	NA	0.695	0.163	0.595	0.880

* No statistics were computed as to the type of Hemophilia because there is no subject with Hemophilia B.

Table 3a: Correlation between HRQoL and demographic characteristics – Child Report Group I.

	Physical	Feelings	Attitude	Family	Friends	School
Socioeconomic Status						
Lower	38.72	25.59	22.40	68.75	35.54	16.39
Upper	5.77	25.00	0.00	87.50	25.00	11.90
P	0.438	NA	NA	0.732	NA	0.541

Table 3b: Correlation between HRQoL and demographic characteristics – Parent Report Group I.

	Physical	Feelings	Attitude	Family	Friends	Perceived Support	Other Persons	Sports School	Coping	Treatment
Type of Hemophilia										
A	27.06	30.28	31.85	30.30	35.00	34.58	32.77	55.96	32.77	13.33
B	62.49	35.71	37.49	21.42	12.5	40.62	35.41	75	12.48	6.25
P	0.242	0.993	0.912	0.841	0.905	0.852	0.259	NA	0.717	0.801
Severity of Disease										
Moderate	25.93	28.87	32.94	31.45	34.38	35.71	32.73	55.36	31.54	11.90
Severe	55.95	40.47	30.55	19.04	22.92	33.33	34.72	71.43	24.99	15.28
P	0.764	0.607	0.652	0.631	0.888	0.826	0.658	0.179	0.099	0.726
Number of Bleeds/Month										
<2	*23.90*	*27.52*	30.56	*29.19*	*33.65*	*34.62*	*32.69*	56.32	33.65	14.42
>2	55.04	41.96	38.88	29.46	28.13	37.50	34.37	64.28	19.78	6.25
P	0.427	0.293	0.215	0.921	0.431	0.554	0.913	0.528	0.372	0.176

Table 4a: Correlation between HRQoL and demographic characteristics – Child Report Group II.

	Physical	Feelings	View	Family	Friends	Perceived Support	Other People	School	Dealings	Treatment
Socioeconomic Status										
Lower	39.96	36.46	45.18	44.77	36.19	45.83	41.18	53.77	34.88	24.14
Upper	0.00	53.57	31.25	46.88	31.25	37.50	25.00	59.46	42.14	16.67
P	NA	0.784	0.987	0.826	NA	0.990	0.351	0.282	0.096	0.835

Table 4b: Correlation between HRQoL and demographic characteristics – Parent Report Group II.

51.56 versus <2 bleeding episodes/month TSS 24.01; p=0.019). Sports and School was significantly impaired among those with severe disease (TSS 66.99 versus moderate disease TSS 58.16; p=0.032).

Correlations between HRQoL and the 2 socioeconomic brackets of parent responders did not show significant difference; however, parents in the lower socioeconomic bracket had higher TSS across all scales.

The child respondents' answers to the open-ended questions revealed most concerns with the following: limitation in play and fear of injections for Group I; missed school days, concerns regarding how other people view their illness for Groups II and III. Parents expressed fear of possible life-threatening bleeds and costs of factor concentrate and rehabilitation when the need arises.

Discussion

Determination of the quality of life of children and adolescents with hemophilia is an important part in the holistic management of this chronic disease, and this is attainable through the use of disease - related QoL questionnaires. Our study showed that majority of the scales of the Filipino version of the Haemo-QoL had a mean TSS below 50 for both child and parent respondents with moderate and severe hemophilia, showing a good quality of life in our patients and their families.

Compared to the Turkish, Egyptian and Iranian over-all mean TSS of 39.6 ± 15.0, ≥ 50 and 54.1 ± 7.3 respectively, the total mean TSS in our study is 28.39 ± 4.76, reflecting a good QoL of Filipino children with hemophilia seen in our center [9-11]. This, however, should be interpreted with caution since majority of the respondents had moderate disease as compared with the Turkish and Egyptian studies where majority of the respondents had severe hemophilia.

Younger children showed most impairment in the scale of Family which was attributed to parental supervision, with parents exerting more control over their children, thus keeping the patient away from interacting with friends and participating in physical activities. Older children (Groups II and III) were most impaired in the scale of Sports and School. Like the younger children, over protectiveness may be the reason, this time from the teachers and school administrators that may deter them from participating in school activities. Parents,

	Physical	Feelings	Attitude	Family	Friends	Perceived Support	Other Persons	Sports School	Coping	Treatment	Future	Partner
Type of Hemophilia												
A	34.12	26.14	29.10	29.59	32.42	38.10	25.79	60.08	27.57	16.67	25.85	15.54
B	49.99	46.43	47.22	45.24	20.83	18.75	20.83	53.57	44.44	12.50	47.90	29.17
P	0.397	0.099	**0.013**	0.815	0.964	0.346	0.950	0.160	0.293	0.909	0.075	0.239
Severity of Disease												
Moderate	32.99	24.61	29.36	29.58	30.95	35.11	24	58.16	29.35	15.47	24.95	14.34
Severe	58.33	57.14	45.36	45.26	31.11	39.58	33.33	66.99	31.94	20.88	54.16	37.5
P	0.812	0.901	**0.040**	0.759	0.884	0.539	0.712	**0.032**	0.683	0.879	**0.022**	0.143
Number of Bleeds/Month												
<2	30.62	26.07	30.27	30.35	30.29	35.00	24.16	57.02	27.70	16.87	24.01	16.31
>2	63.54	41.71	36.80	37.52	34.38	39.06	30.21	70.53	39.58	12.50	51.56	21.88
P	0.910	0.348	**0.017**	0.870	0.783	0.621	0.807	0.062	0.898	NA	**0.019**	0.284

Table 5a: Correlation between HRQoL and demographic characteristics – Child Report Group III.

	Physical	Feelings	Attitude	Family	Friends	Perceived Support	Other Persons	Sports School	Coping	Treatment	Future	Partner
Socioeconomic Status												
Lower	53.24	40.79	36.52	31.92	35.55	38.92	32.90	51.82	27.91	22.98	38.34	NA
Upper	20.83	33.33	30.21	23.89	16.67	13.89	11.11	47.61	20.83	22.22	16.67	NA
P	0.882	0.115	0.765	0.099	0.077	0.950	0.564	0.485	0.314	0.120	0.305	

Table 5b: Correlation between HRQoL and demographic characteristics – Parent Report Group III.

particularly mothers, and caregivers tend to overprotect because they perceive their child to be more susceptible and at risk because of his illness. Holmbeck and colleagues, in their study of 68 families with a chronically ill child, have illustrated that over protectiveness has detrimental effects on the growing child such as symptoms of depression and oppositional behavior. Overprotective parents are also less expected to grant independence to their child in the future. Parents may be torn between allowing self-sufficiency to their child and the need to protect the child from injuries and further medical harm [12]. A review done by Giordano and colleagues reiterated that children with hemophilia should be allowed self-sufficiency and be encouraged to participate in physical activities such as swimming, walking and running to strengthen muscles and joints. Hemophiliacs should also be informed of their illness so that they can manage their future and be able to adapt to their environment accordingly [13].

In contrast to Groups II and III, Group I children with hemophilia showed least impairment in the subscale of Attitude. This may be due to the reason that preschoolers are naturally curious about their surroundings and are just beginning to hone socialization skills as they discover and experience day to day activities. Preschoolers in general, may not be able to understand the concept of cause and effect, which is learned during the school – age years [14,15]. Groups II and III showed least impairment in the subscale of Treatment. This may be due to better understanding of their illness as well as the necessity for factor infusions and supportive measures when bleeding occurs. A study of 108 hemophiliacs done by Lindvall et al. showed that patients started to become responsible for his illness in terms of treatment at a mean age of 14.1 years, showing their deep of understanding of the disease process [16].

Group III child – parent responses revealed significantly different TSS in the scale Feelings showing that the child has better acceptance, less emotional concerns and can adapt better with his illness than his parent. Beeton et al. stated that parents are affected by how their child communicates the way they feel about their illness and how well these children adapt to their disorder [17]. This is in contrast with a later study done by Weidebusch comparing parents of children with hemophilia with parents of children with other chronic illness (juvenile idiopathic arthritis, diabetes mellitus type 1). The study showed that parents of hemophiliacs had better QoL and lower psychosocial strains due to strengthened adaptive coping mechanisms [18].

Only Group III respondents showed statistically significant correlations between HRQoL and demographic characteristics. Severe hemophilia affects how children see themselves, how they participate in school and how they see the future. Because these older children incurred more number of bleeds in their lifetime, they are therefore more at risk for the development of joint disabilities. Stehl showed that adolescents with severe hemophilia were found to be more compliant and cautious than those with mild or moderate disease. Also, disease severity affects the personality traits and behavior patterns especially in this age group where self-sufficiency is becoming the norm [19]. The severity of hemophilia and how it affects the QOL are consistent with other studies done by Arzu et al., Tantawy et al. and Bagheri et al. [9-11].

Barriers to hemophilia care may significantly affect the QOL. Studies by Zhou and colleagues, Saxena and Remor, showed that educational barriers (e.g.: lack of awareness regarding signs of bleed, importance of early treatment), financial difficulties (families belonging to the lower income bracket, insurance coverage and costs of treatment), treatment adherence (if on factor prophylaxis), distance from hemophilia treatment center and psychological barriers (denial, fear of pain and injections, risky behavior, parental burden over providing care and discipline) are factors that lowers QoL [20-22]. Mehramiri et al. presented that socioeconomic factors such as unemployment, low income, occupational problems caused by physical disability, low educational level and Hepatitis C infection were hindrance to the improvement of the QoL of persons with hemophilia [23].

Factor prophylaxis was known to improve QoL in terms of significantly reduced the frequency of hemarthrosis, chronic hemophilic arthropathy, number of hospital consults and admissions, less school absenteeism, less pain and handicap and less need for intervention to impaired joints [9,24,25]. None of the children in the present study are on factor prophylaxis, mainly for economic reasons. Factor VIII infusion is given on demand when bleed arises. Despite this, the TSS

among all groups and demographic characteristics were not statistically different between the groups compared. Best evidence for the use of factor prophylaxis was showed in a study done by Buchbinder and Ragni [26]. A review of 21 QOL studies combining adult and pediatric patients showed mixed impact of factor prophylaxis, hence, their recommendation against the use factor concentrate prophylaxis versus on-demand therapy to improve the HRQOL of hemophiliacs.

Our study showed a satisfactory QoL despite the absence of factor prophylaxis and low economic status of our patients. This may be due to the presence of support groups (Hemophilia Association of the Philippines for Love and Service (HAPLOS) Foundation Inc., Philippine Hemophilia Foundation Inc.) and charitable institutions that extend assistance through factor donations and financial aid as reflected by the relatively low TSS in the subscale of Perceived Support, which means that they recognize the help extended to them. These organizations link the patient to community groups, providing education, promoting self-reliance and support to families in terms of seminars, workshops and factor concentrate availability through humanitarian means.

Subscale impairments, severity of hemophilia, barriers to hemophilia care, presence of support groups are factors that significantly affect and predict the QOL of Filipino children with hemophilia. Addressing these concerns may amplify their understanding of the disease and further improve their quality of life.

Recommendations

The HaemoQoL assessed the quality of life of a group of patients seen at a Hematology clinic in a tertiary hospital. The same questionnaire may be used for individual patients seen in the private setting. A multi-center approach may also be done get a wider perspective of the QoL of Filipino hemophiliac to help clinicians further improve patient care. Strategies to improve QoL medically, socially, emotionally and psychologically may be crafted and scaled nationally.

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