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Evaluation of the Quality of Life in Children with Haemophilia

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ABSTRACT

Background: Congenital haemophilia primarily affects males. Haemophilia A or B is caused by mutation of clotting factor genes on X chromosome.

Objective: The aim of the current study is to assess quality of life of children with haemophilia.

Patients and methods: A cross sectional comparative study was conducted at Hematology Clinic at El-Helal Hospital and Sohag University Hospital on 50 children with haemophilia.

Results: Most patients had joint bleeding and most of them were satisfied about their quality of life (QoL). Most of patients were able to do homework completely, work well and get around. Most of patients had difficulties annoyance and difficulties effect on lifestyle. Most patients never had a depression due to the haemophilia. Most children like school and had friends which made with future plans and never feared from the future. Most patients could perform muscle activities but some of them felt upset going to hospital.

Conclusion: Haemophilia has effect on studied cases health-related quality of life (HRQOL). The severity of the disease, bleeding frequency, rate of joint bleeding, financial burden, & therapy all have impact on haemophilia HRQoL. Routine haemophilia care should contain psychological assessment & caregiver support, which should be measured using result measures that account for both HRQoL and caregiver burden.

Keywords: Haemophilia, Quality of life, Children, Caregivers, Cross sectional study, Al-Azhar University.

INTRODUCTION

Hemophilia is congenital disorder that impacts mostly men. It is caused by clotting factor gene mutation on X chromosome, which results in lack of factor VIII or -IX in haemophilia A or B [(1)]. Spontaneous bruising, mucosal and joint bleeding, epistaxis, & severe bleeding events like intracranial haemorrhage are all common symptoms [(2)]. Repeated joint bleeding results in severe joint damage and pain, resulting in disability [(3)].

The most common symptom of haemophilia is bleeding, which occurs after trauma or surgery, with severity correlated with degree of clotting factor deficiency. Bleeding can happen in muscles, joints, or soft tissue, as well as in neck, throat, chest, gastrointestinal system, or intracranially in life-threatening cases [(1)].

Primary therapy aim is to prevent or treat bleeding; bleeding is usually treated by on-demand administration of specific factor concentrate to compensate for deficient clotting factor, and prevention has contained prophylaxis regimens of these factor-replacement treatments, with non-replacement factors becoming more widely available in recent years [(4)].

Many studies have looked at the impact of hemophilia on health-related quality of life (HRQOL). The disease is also linked to an increased risk of hepatitis, AIDS, anxiety, depression, & isolation, all of which reduce patients' quality of life as they age. HRQoL is a multidimensional concept that describes an individual's ability to function in terms of physical, psychological, and social health [(2)].

Quality of life (QoL) Evaluation is becoming increasingly popular as guide to effective medical therapy & healthcare. According to studies, hemophiliacs have a lower quality of life than healthy people [(5)].

Soucie et al. discovered that hemophilia studied cases of all years old have a lower quality of life because of arthropathy [(6)].

The goal of the QoL assessment is to identify children who are experiencing difficulties as a result of their hemophilia disease, estimate complications related to hemophilia, & define comprehensive & satisfactory therapy.

As a result, it is essential to investigate & analyze whether degree of bleeding, severity of hemophilia, years old of diagnosis, & years old at which treatment is initiated affect QoL in children with hemophilia

The aim of the current study was to analyze QoL of children with hemophilia.

PATIENTS AND METHODS

A cross sectional comparative study was conducted at Hematology Clinic at El-Helal Hospital and Sohag University Hospital on 50 children with haemophilia.

Inclusion Criteria:

Patients diagnosed clinically and laboratory with hemophilia A, aged between 3-16 years, parents or caregivers (in case of children 4-8 years old) and children (8-16 years old) who can read, write and understand all relevant aspects of research which will be explained and discussed with them, outpatients, aware, and have a good ability to fill out questionnaires and following up with Hematology Clinic at El-Helal Hospital and Sohag University Hospital.

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Exclusion Criteria:

Patients with cognitive disabilities and mental health disorders, patients with other chronic diseases, recently diagnosed patients, patients with other hematological disorder and patients 8-16 years old who can't read and write, and refuse to take part in study, and people who can't complete the questionnaire due to any problems will be excluded from research.

Study Tools:

All cases contained in research were subjected to following:

History taking:

Initial statement identifying historian, that person's relationship to studied case and their dependability, studied case's years old, gender, race, and other important identifying information, and age at diagnosis.

Evaluation of quality of life of hemophilic children and their families:

The studied group was given questionnaire to assess their quality of life. Questionnaire on demographic and socioeconomic information, and questionnaire on quality of life, was used to collect data from studied cases and their accompanying parents. Questionnaire was used to assess haemophiliacs' quality of life. Despite fact that study team presented necessary explanations and assisted illiterates in completing questionnaires, studied cases were free to choose appropriate answers in presence of their parents.

Ethical Approval:

Al-Azhar University Ethics Board approved this research, and each participant signed informed written consent form. This work has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for studies involving humans.

Statistical Analysis

Data collected and encoded using Microsoft Excel software. Data were then imported into Statistical Package for Social Sciences (SPSS version 23.0) software for analysis. According to the type of data, qualitative represented as numbers and percentages, while quantitative data represented by mean, standard deviation, median, and interquartile range. P-value was set at ≤ 0.05 for significant results.

RESULTS

Table 1 showed demographic features among research population.

Table (1): Demographic features among research population

Variable	Research population (n = 50)	
	N	**%**
Residence - Rural N (%) - Urban n (%)	**41** **9**	**82%** **18%**
Age (years) Mean ± SD. Median (IQR) Range (Min-Max)	**8.28 ± 3.8** **7.5 (5.25 - 11)** **13 (3 - 16)**	

SD: standard deviation **IQR:** interquartile range

Figure 1 showed date of disease detection among the study population. Number of participants that answered with first year of age was 42 (84 %). Number of participants that answered with 2 to 6 years was 7 (14 %). Number of participants that answered with 7 to 12 years was 1 (2 %). No participants answered with more than 12 years.

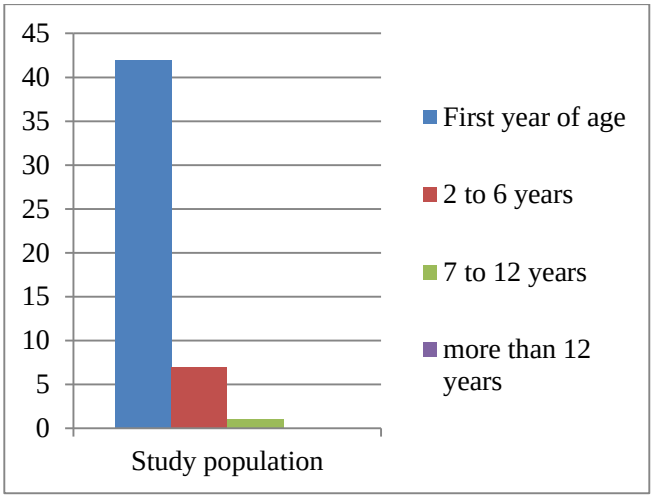

Figure (1): Bar chart showing study population data regarding date of disease detection.

Table 2 showed bleeding frequencies among the study population.

Table (2): Bleeding frequency among the study population.

Variable	Study population (n = 50)	
	n	%
Bleeding frequency		
Rarely n (%)	**6**	**12%**
Once a month n (%)	**4**	**8%**
Twice a month n (%)	**13**	**26%**
Three times a month n (%)	**8**	**16%**
More than three times n (%)	**19**	**38%**

Table 3 showed rate of joint bleeding among the study population.

Table (3): Rate of joint bleeding among the study population.

Variable	Study population (n = 50)	
	N	%
Rate of joint bleeding		
Absolutely n (%)	**14**	**28%**
Rarely n (%)	**2**	**4%**
Very often n (%)	**2**	**4%**
Mostly n (%)	**16**	**32%**
Always n (%)	**16**	**32%**

Table 4 showed ability to move among the study population.

Table (4): Ability to move among the study population.

Variable	Study population (n = 50)	
	N	%
Ability to move		
Absolutely		
n (%)	**0**	**0%**
Little		
n (%)	**5**	**10%**
Moderately		
n (%)	**14**	**28%**
Especially		
n (%)	**9**	**18%**
Completely		
n (%)	**22**	**44%**

Figure 2 showed rate of receiving blood transfusion among the study population.

Number of participants that answered with I did not receive was 26 (52%). Number of participants that answered with a little was 12 (24%).

Number of participants that answered with moderately was 6 (12%). Number of participants that answered with Much was 1 (2%). Number of participants that answered with always was 5 (10%).

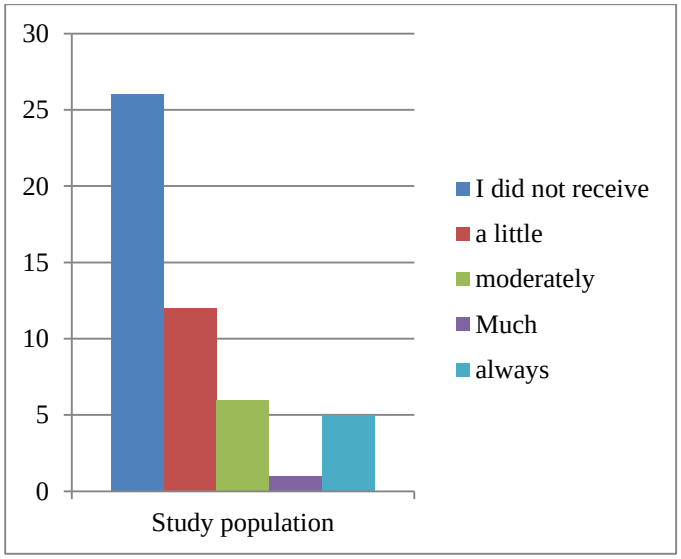

Figure (2): Bar chart showing study population data regarding Rate of receiving blood transfusion.

Figure 3 showed Incidence of arthroplasty among the study population. Number of participants that answered with absolutely was 39 (78%). Number of participants that answered with little was 8 (16%). Number of participants that answered with Yes was 3 (6%).

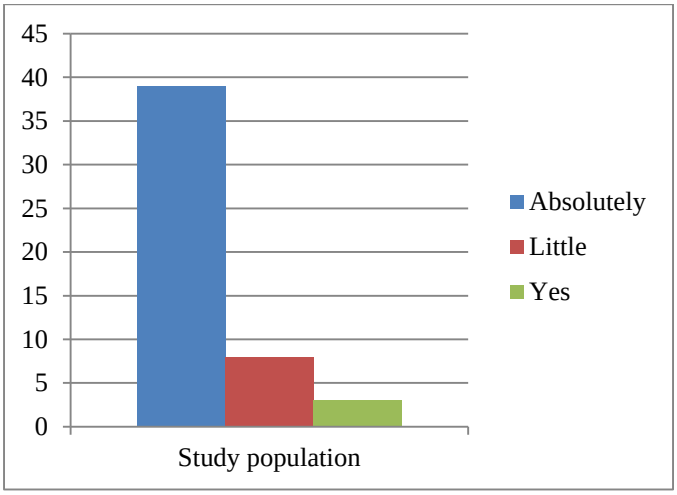

Figure (3): Bar chart showing study population data regarding incidence of arthroplasty.

Table 5 showed Satisfaction of quality of life among the study population.

Table (5): Satisfaction of quality of life among the study population.

Variable	Study population (n = 50)	
	n	%
Satisfaction of quality of life		
Very upset n (%)	**2**	**4%**
Dissatisfied n (%)	**8**	**16%**
Neither satisfied nor upset n (%)	**18**	**36%**
Satisfied n (%)	**21**	**42%**
Very satisfied n (%)	**1**	**2%**

Figure 4 showed Ability to do homework among the study population. No participants answered with Absolutely. Number of participants that answered with Little was 8 (16%).

Number of participants that answered with Moderately was 12 (24%). Number of participants that answered with Especially was 12 (24%). Number of participants that answered with completely was 18 (36%).

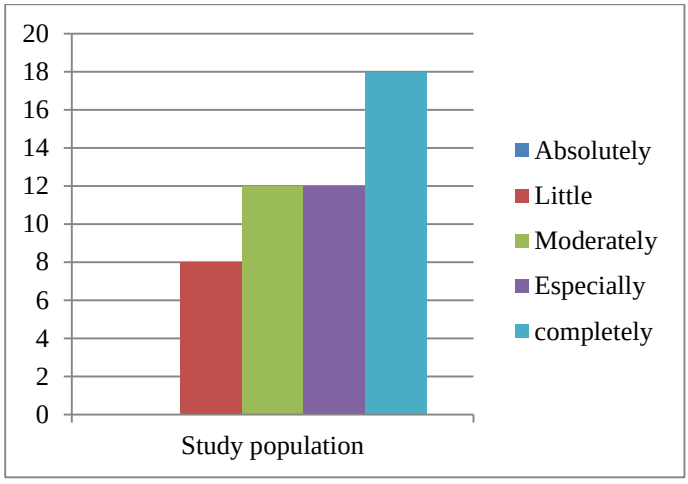

Figure (4): Bar chart showing study population data regarding Ability to do homework.

Figure 5 showed Satisfaction of ability to carry out duties among the study population. No participants that answered with Very upset. Number of participants that answered with dissatisfied was 11 (22%).

Number of participants that answered with not satisfied or upset was 11 (22%). Number of participants that answered with Satisfied was 21 (42%). Number of participants that answered with Very Satisfied was 7 (14%).

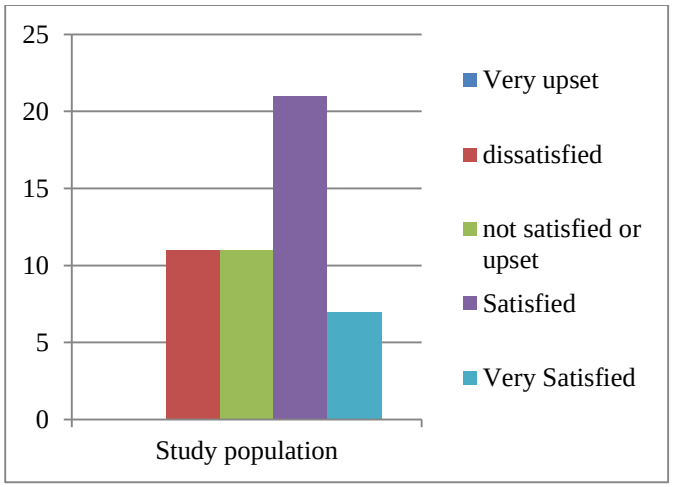

Figure (5): Bar chart showing study population data regarding Satisfaction of ability to carry out duties.

Figure 6 showed Mobility diff iculties effect on lifestyle among the study population. Number of participants that answered with Absolute was 9 (18%). Number of participants that answered with little was 19 (38 %). Number of participants that answered with Moderate amount was 9 (18 %). Number of participants that answered with Very much was 11 (22%). Number of part icipants that answered with extreme amount was 2 (4%).

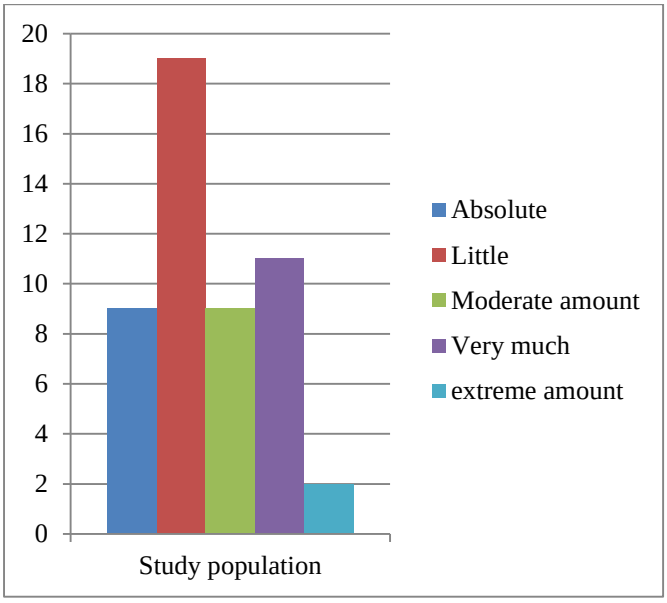

Figure (6): Bar chart showing study population data regarding Mobility difficulties effect on lifestyle.

Figure 7 showed Satisfaction of ability to move among the study population. No participants answered with Very upset .Number of participants that answered with dissatisfied was 8 (16%). Number of participants that answered with neither satisfied nor upset was 13 (26%). Number of participants that answered with Satisfied was 19 (38%). Number of participants that answered with Very Satisfied was 10 (20%).

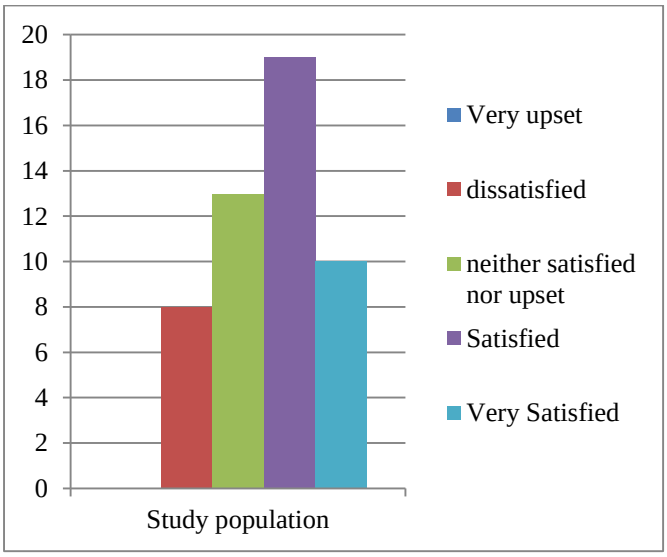

Figure (7): Bar chart showing study population data regarding Satisfaction of ability to move.

Figure 8 showed depression frequency among the study population. Number of participants that answered with Never was 28 (56%).

Number of participants that answered with Sometimes was 9 (18%). Number of participants that answered with Most of the time was 9 (18%). Number of participants that answered with Always was 4 (8%).

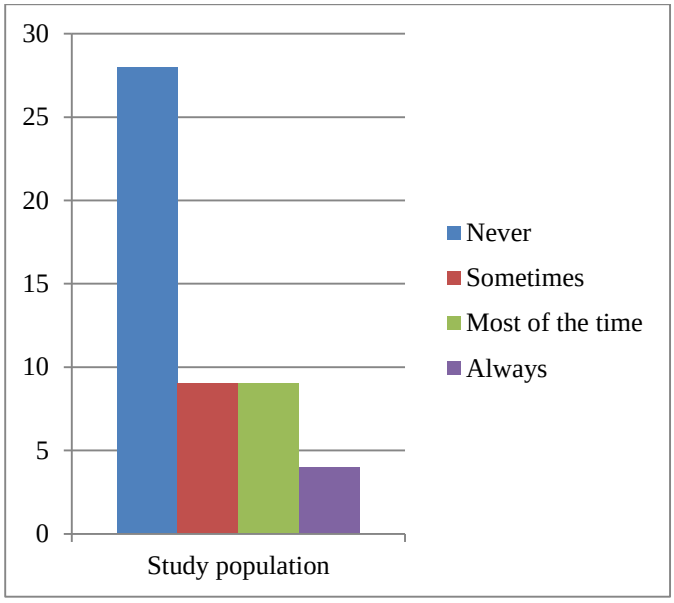

Figure (8): Bar chart showing study population data regarding Depression frequency.

Table 6 showed Analysis of factors affecting QoL.

Table (6): Analysis of factors affecting Quality of life

Variable	Quality of life			
	OR	95% Confidence Interval		P-value
		Lower	Upper	
Bleeding frequency	**3.856**	**1.417**	**10.493**	**0.008**
Rate of joint bleeding	**1.336**	**0.830**	**2.151**	**0.233**

DISCUSSION

Haemophilia is bleeding disorder that primarily impacts boys & is affected by inherited deficiency of factor VIII or factor IX. It is characterised by symptoms like spontaneous bruising, mucosal bleeding, joint bleeding, epistaxis, and severe or even fatal bleeding events like intracranial haemorrhages [(7)].

This cross-sectional comparative study was conducted at Out-Patient Clinic of hematology and Pediatrics Department, El-Helal Hospital and Sohag University Hospital, Egypt. This study was conducted on 50 Hemophilia patients. The studied group was given a questionnaire to assess their QoL.

As regard demographic data of the studied patients, we found that 82% of the patients were from Rur al Residence and 12% were from Urban Residence. Age in the study population ranged from 3 to 16 with mean 8.28 (SD 3.8) years.

The present study can be supported by **Rodriguez-Santana *et al.*** [(8)] who aimed to quantify differences in direct medical & societal costs & studied case humanistic burden in severe paediatric haemophilia studied cases. The study enrolled 794 people. Average years old of entire sample were 10.5 years (years old one–seventeen).

As well, **Mousavi *et al.*** [2] who aimed to provide updated account on health-related quality of life in children with hemophilia. The study included 65 randomly selected hemophiliac children. Studied cases' years old averaged 12.9 (SD 3.9) years.

Furthermore, another Chinese study by **Zhang *et al.*** [9] aimed to evaluate long-term HRQoL of haemophilia children. 42 children with haemophilia and their parents were included in research. Mean (SD) years old of the studied cases was 5.48 (4.63) years. However, they stated that 59.5% of the patients were Urban and 40.5% were from Rural Residence.

Regarding date of disease detection among our study population, we found that 42 (84%) of the patients were detected at the First year of age, 7 (14 %) were detected at 2 to 6 years, 1 (2 %) was detected at 7 to 12 years and no participants were detected at more than 12 years.

Outcomes were in agreement with **Wardhani *et al.*** [3] as Age at diagnosis (years old) ranged from 0-1 years in 61.9% of patients, 3-6 years in 14.3% of the patients and 6-12 years in 23.8%. Also, no participants were detected at more than 12 years.

Regarding Bleeding frequency among our study population, we found that 38% of the participants bleeding more than three times and Number of participants that answered with twice a month was 13 (26%). Number of participants that answered with Three times a month was 8 (16%).

However, **Wardhani *et al.*** [3] reported that 14.3% of the study population had normal degree of bleeding and 85.7% had abnormal degree of bleeding. Also they reported that 28.6% of the study population had joint bleeding but they didn't evaluate severity of joint bleeding.

Outcomes were in agreement with **Zhang *et al.*** [9] who aimed to assess HRQoL & its related factors between patients with haemophilia. They reported low frequency of Bleeding

Regarding frequency of pain feeling in our study, Number of participants that answered with Absolutely was 3 (6%). Number of participants that answered with rarely was 11 (22%). Number of participants that answered with very often was 9 (18%). Number of participants that answered with Mostly was 5 (10%). Number of participants that answered with Always was 22 (44%).

Kodra *et al.* [10] showed research to evaluate QoL of studied cases with haemophilia. In their research 84.6% of the haemophilia children rarely had pain feeling but 15.4% had pain problems.

In the current study, most of our patients had no difficulty regarding Ability to move as number of participants that answered with completely was 22 (44%) and Number of participants that answered with Moderately was 14 (28%). Also, 96 % of our study population had no complications. Furthermore, only 6% of our study population had arthroplasty and 16% had little incidence of arthroplasty.

Also**, Kodra *et al.*** [10] found that 76.9% of the haemophilia children rarely had pain feeling but 23.1% had Mobility problems.

However, **Zhang *et al.*** [9] reported a significant association between arthroplasty and haemophilia (P <0.001).

Regarding Satisfaction of quality of life among the study population we found that 42% were satisfied, 36% were neither satisfied nor upset, 16% dissatisfied and only 4% were Very upset.

Regarding ability to work among the study population, we found that 54% had good ability to work moreover, 38% were satisfied of ability to move.

Kodra *et al.* [10] reported that most of the haemophilia children were satisfied with their ability to carry out duties and 92.3% of them had no problems to carry out duties.

Regarding Depression frequency among our study population, most of the patients never had depression as number of participants that answered with never was 28 (56%). Number of participants that answered with sometimes was 9 (18%). Number of participants that answered with most of the time was 9 (18%). Number of participants that answered with always was 4 (8%).

However, **Trzepacz *et al.*** [11] Children with haemophilia have difficulty managing their emotional well-being, such as symptoms of depression, anxiety, and low self-esteem, according to research.

Moreover, we found that 36% of parent had an overly anxious feeling about the affected child. Also, 46% reported that having an injured child gives me a pessimistic view of the future.

Our results were in agreement with **Khair & Von Mackensen,** [12] who aimed to investigate effect of burden on hemophilia caregivers' HRQoL. Caregivers found highest burden associated with disease of their sons in dimension 'negative impact' of Impact on Family Scale (Mean 60.08, SD 20.7).

As well, **Shahly *et al.*** [13] caregiving for those with chronic illnesses is progressively falling on family caregivers, who are vulnerable to financial strain, with uncompensated family care-giving being important societal asset that offsets rising formal healthcare costs.

In the current research, we found that there is a significant correlation between bleeding frequency and rate of joint bleeding with quality of life of children with hemophilia. Odds ratio of bleeding frequency was 3.856, and the confidence interval was ranged from 1.417 to 10.493. Odds ratio of rate of joint bleeding was 1.336, and the confidence interval was ranged from 0.830 to 2.151.

Outcomes were in agreement with **Rodriguez-Santana *et al.*** [8] who described that the disease severity and frequency of bleeding was important predictor of rising direct medical & societal costs & were associated with worse HRQoL scores.

Also, our results were in agreement with **Baek *et al.*** [14] who aimed to evaluate HRQoL of moderate and

severe haemophilia studied cases. The univariate analysis of study population revealed that bleeding experience and joint bleeding, and haemophilic arthropathy were related to impaired HRQoL (P <0.001).

As well, **Ferreira *et al.*** [15] aimed to evaluate joint status of hemophiliac studied cases at Brazilian blood center. They conclude in general that HRQoL in people with hemophilia is affected by attendance of arthropathy; meanwhile occurrence of target joints was related to Haem-A-QoL total score.

However, **Wardhani *et al.*** [3] There is no high relationship among bleeding frequency, haemophilia severity, years old at diagnosis, and years old at treatment initiation with QoL of children with haemophilia A ($p = 0.330$, $p = 0.608$, $p = 0.516$, $p = 0.864$; $r = -0.223$, $r = -0.119$, $r = 0.150$, $r = 0.040$, respectively).

In conclusion, this research was successful in gathering important and reliable data on demographic characteristics, socioeconomic determinants of health, disease severity, healthcare services, and QoL in children haemophilia and their caregivers for 1st time. Haemophilia has effect on studied cases' HRQoL. The severity of the disease, bleeding frequency, rate of joint bleeding, financial burden, and therapy all have an impact on haemophilia HRQoL.

DECLARATIONS

- **Consent for Publication:** I verify that all authors have agreed to submit manuscript.
- **Availability of data & material:** Available
- **Competing interests:** None
- **Funding:** No fund
- **Conflicts of Interest:** authors confirm that they have no conflicts of interest with regard to publication of this paper.

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