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Original Research Article

To Study the Factors Affecting Quality of Life in Children Suffering from Haemophilia

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Abstract

Introduction: Congenital hemophilia predominantly affects males and is characterized by mutations in clotting factor genes located on the X chromosome. This study aims to evaluate the quality of life (QoL) of pediatric patients with hemophilia.

Materials and Methods: A cross-sectional comparative analysis was conducted involving 78 children with hemophilia treated at an Indian Hospital. A questionnaire encompassing demographic and socioeconomic information, along with a separate questionnaire focusing on quality of life, was utilized to gather data from the studied cases and their accompanying parents.

Results: The majority of patients experienced joint bleeding, yet most expressed satisfaction with their QoL. A significant portion of patients were able to complete homework, engage in daily activities, and move around freely. However, many patients reported difficulties and annoyances affecting their lifestyle, with some experiencing emotional challenges but not depression directly related to hemophilia. Despite these challenges, most children enjoyed school, had friends, made future plans, and were not apprehensive about their future. Although most patients could engage in muscle activities, some felt discomfort about visiting the hospital.

Conclusion: Hemophilia profoundly influences the health-related quality of life (HRQoL) in the studied cohort. Disease severity, bleeding frequency, joint bleeding occurrences, financial constraints, and therapeutic interventions collectively impact hemophilia-related HRQoL. Routine hemophilia management should include psychological assessments and caregiver support, integrating outcome measures that assess both HRQoL and caregiver burden.

Keywords: Children, Haemophilia, quality of life.

Introduction

Hemophilia is a congenital disorder predominantly affecting males, characterized by a mutation in the clotting factor gene on the X chromosome, leading to a deficiency of factor VIII or IX in hemophilia A or B, respectively. Common symptoms include spontaneous bruising, mucosal and joint bleeding, epistaxis, and severe bleeding events like intracranial hemorrhage. Repeated joint bleeding can cause severe joint damage and pain, resulting in disability [1-4]. The hallmark symptom of hemophilia is bleeding, typically occurring after trauma or surgery and correlating with the degree of clotting factor deficiency. Bleeding can manifest in muscles, joints, soft tissue, as well as in critical areas such as the neck, throat, chest, gastrointestinal system, or intracranially in life-threatening situations. Primary therapy focuses on preventing or treating bleeding episodes, often through on-demand administration of specific factor concentrates or prophylactic regimens, including non-replacement factors in recent advancements [5]. Numerous studies have assessed the impact of hemophilia on health-related quality of life (HRQOL), revealing associations with increased risks of hepatitis, AIDS, anxiety, depression, and social isolation as patients age [2]. HRQOL encompasses physical, psychological, and social aspects of functioning. Evaluating QoL has become crucial in guiding effective medical therapy and healthcare, with hemophiliacs generally experiencing lower QoL compared to healthy individuals [6]. Research by Soucie et al. indicates that hemophiliacs, particularly those with arthropathy, have lower QoL, emphasizing the need

to identify complications related to hemophilia and establish comprehensive therapy strategies [7]. This study aims to analyze the QoL of children with hemophilia and investigate potential factors such as the severity of hemophilia, age at diagnosis, and initiation of treatment that may influence QoL in this population.

Materials and Methods

A cross-sectional comparative study was conducted at an Indian hospital involving 78 children with hemophilia. The inclusion criteria comprised patients clinically and laboratory diagnosed with hemophilia, aged between 3-16 years, accompanied by parents or caregivers for younger children (4-8 years old) and children (8-16 years old) capable of reading, writing, and understanding all relevant aspects of the research, which were explained and discussed with them. Additionally, the patients were required to be outpatients, aware, possess good ability to fill out questionnaires, and be willing to follow up with the hospital. Exclusion criteria included patients with cognitive disabilities and mental health disorders, those with other chronic diseases or hematological disorders, recently diagnosed patients, and children (8-16 years old) who were unable to read or write or refused to participate in the study. Individuals unable to complete the questionnaire due to any reasons were also excluded. All cases included in the research underwent history taking, which involved gathering initial statements identifying the historian, their relationship to the studied case, dependability, age, gender, race, and age at diagnosis of the case. The evaluation of the quality of life of hemophilic children and their families was conducted using questionnaires. The study group received questionnaires to assess their quality of life, including demographic and socioeconomic information, as well as specific questions related to quality of life. The questionnaire was used to collect data from the studied cases and their accompanying parents. Statistical analysis was performed using Microsoft Excel software for data collection and encoding, followed by importing into Statistical Package for Social Sciences (SPSS version 20.0) for analysis. Qualitative data were represented as numbers and percentages, while quantitative data were represented by mean, standard deviation, median, and interquartile range. A p-value of <0.05 was considered significant for the results.

Results

Table 1 displays the socio-demographic variables of the study population. The majority of participants hailed from rural areas, and the disease was typically diagnosed within the first year of life. Table 2 presents the clinical variables observed among the study population. A significant proportion of children experienced bleeding episodes more than three times a month (39.74%), suffered from bleeding in joints (32.05%), had complete mobility (43.59%), and did not require blood transfusions (51.28%). Table 3 outlines the Health-Related Quality of Life (HRQoL) parameters among children with hemophilia. The majority of children expressed satisfaction with their quality of life (28%), had a neutral stance regarding their ability to perform duties (26.92%) and mobility (24.36%), and reported no experiences of depression (35.90%).

Table 1: Socio-demographic variables of study population

Variable	n	%
Rural Resident	64	82.05
Urban Resident	14	17.95
Age (years)	8.19 ± 3.5	
Date of Diagnosis		
1st Year of Life	65	83.33
2-6 years	11	14.10
7-12 years	2	2.56
>12 years	0	0.00

Table 2: Clinical variables among study population

Variable	n	%
Bleeding frequency		
Rarely	9	11.54
Once a month	6	7.69
Twice a month	20	25.64
Thrice a month	12	15.38
> 3 times/month	31	39.74
Rate of Haemarthrosis		
Absolutely	22	28.21
Rarely	3	3.85
Very often	3	3.85

Mostly	25	32.05
Always	25	32.05
Ability to move		
Absolutely	0	0.00
Little	8	10.26
Moderately	22	28.21
Especially	14	17.95
Completely	34	43.59
Rate of Blood Transfusion		
None	40	51.28
Little	19	24.36
Moderately	9	11.54
Much	2	2.56
Always	8	10.26
Rate of Arthoplasty		
Absolutely	61	78.21
Little	12	15.38
Much	5	6.41

Table 3: HRQoL parameters among children with Haemophilia

Variable	n	%
Satisfaction of quality of life		
Very upset	2	2.56
Dissatisfied	8	10.26
Neutral	18	23.08
Satisfied	21	26.92
Very Satisfied	1	1.28
Satisfaction of ability perform duties		
Very upset	11	14.10
Dissatisfied	11	14.10
Neutral	21	26.92
Satisfied	7	8.97
Very Satisfied	0	0.00
Effect of Mobility problem on lifestyle		
Absolutely	9	11.54
Little	19	24.36
Moderately	9	11.54
Very Much	11	14.10
Extremely	2	2.56
Satisfaction with ability to move		
Very upset	8	10.26
Dissatisfied	13	16.67
Neutral	19	24.36
Satisfied	10	12.82
Very Satisfied	0	0.00
Depression Frequency		
Never	28	35.90
Sometimes	9	11.54
Most of the times	9	11.54
Always	4	5.13

Discussion

Hemophilia is a bleeding disorder predominantly affecting boys due to an inherited deficiency of either factor VIII or factor IX. This condition is characterized by symptoms such as spontaneous bruising, mucosal and joint bleeding, epistaxis, and severe, potentially fatal bleeding events like intracranial hemorrhages [8]. In this cross-sectional comparative study, we enrolled 78 hemophilia patients. Our study is supported by Rodriguez-Santana et al. [9], who quantified differences in direct medical and societal costs and assessed the humanistic burden in severe pediatric hemophilia

cases. Their study included 794 participants, with an average age of 10.5 years across the sample. Additionally, Zhang et al. [10] conducted a Chinese study focusing on evaluating the long-term health-related quality of life (HRQoL) in children with hemophilia. They included 42 children and their parents, with a mean age of 5.48 years, and noted demographic distribution between urban and rural residences. Our findings align with those of Wardhani et al. [4], who reported that most patients were diagnosed with hemophilia at a young age, with a majority experiencing abnormal bleeding degrees and joint bleeding. However, they did not evaluate the severity of joint bleeding. Similarly, Kodra et al. [11] assessed the quality of life in hemophilia children, noting low frequencies of pain and mobility problems in the majority of cases.

Regarding joint health, Trzepacz et al. [12] highlighted the emotional challenges faced by children with hemophilia, including symptoms of depression, anxiety, and low self-esteem. Our results are consistent with Khair & Von Mackensen [13], who investigated the burden on caregivers' HRQoL, particularly in terms of negative impacts on family dynamics. Shahly et al. [14] emphasized the increasing role of family caregivers in managing chronic illnesses like hemophilia, highlighting the financial strain and societal contributions of uncompensated caregiving. Moreover, Baek et al. [15] evaluated HRQoL in hemophilia cases, noting associations between bleeding experiences, joint health, and impaired HRQoL. Similarly, Ferreira et al. [16] assessed joint status in hemophilia patients, linking arthropathy and target joint occurrences with overall HRQoL impacts.

Conclusion

In conclusion, this research successfully gathered crucial and reliable data concerning demographic characteristics, socioeconomic determinants of health, disease severity, healthcare services, and QoL in children with hemophilia and their caregivers for the first time. The study demonstrated that hemophilia significantly affects the HRQoL of the participants. Factors such as disease severity, frequency of bleeding episodes, occurrence of joint bleeding, financial burden, and the effectiveness of therapy all contribute to the impact on HRQoL among individuals with hemophilia. These findings underscore the importance of comprehensive care and support for individuals with hemophilia to improve their overall quality of life.

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