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Self-Care Practice among Iraqi Patients with Inherited Bleeding Disorders: A Cross-Sectional Study

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Abstract: This study aimed to evaluate the performance of Iraqi patients with inherited bleeding disorders (InBDs) on self-care practice in terms of general preventive measures, physical activity, and healthy dietary patterns. A total of 75 male patients with InBDs were included. Self-care practice performance was assessed using a self-report questionnaire containing 27 items related to three domains: general preventive measures, physical activities, and dietary pattern practices. Total and component scores for self-care practice performance were evaluated regarding patient and disease characteristics. Overall, mean total scores for self-care practice indicated good performance in 22 (29.3%) patients. The mean scores for general preventive measures (27.2, ranging from 19 to 33), physical activity (19.9, ranging from 13 to 28), and dietary pattern (10.8, ranging from 8 to 13) components were in the range of good, moderate, and moderate-to-good performance, respectively. Good performance scores in overall self-care were more likely in students (p=0.031), in normal-weight patients (p=0.047), in hemophilia A patients (p<0.001). And good performance scores in physical activity were more likely in patients with higher educational attainment (p=0.026), in students (p=0.002), and patients with injection administration made by others (p=0.041). Our findings in Iraqi patients with InBDs revealed an overall favorable performance on self-care practice, while a need for improved self-practice regarding physical activity, particularly after a decrease in parental supervision with the start of self-infusion. Overweight/obesity was evident in a considerable percentage of patients. A need for strategies to promote self-motivation towards better self-care was revealed in obese patients and those with constant joint pain.

Keywords: inherited bleeding disorders, self-care practice, preventive measures, physical activity, obesity.

伊拉克遗传性出血性疾病患者的自我保健实践：一项横断面研究

摘要：本研究旨在评估伊拉克遗传性出血性疾病(InBDs)患者在一般预防措施、身体活动和健康饮食模式方面的自我保健实践表现。共纳入75名男性InBDs患者。使用自我报告问卷评估自我保健实践表现，该问卷包含与三个领域相关的27个项目：一般预防措施、身体活动和饮食模式实践。根据患者和疾病特征评估自我护理实践表现的总分和组成分。总体而言，自我护理实践的平均总分表明22名(29.3%)患者表现良好。一般预防措施（27.2，范围从19到33）、身体活动（19.9，范围从13到28）和饮食模式（10.8，范围从8到13）组成部分的平均得分在良好、中等、和中到好的表现，分别。学生（p=0.031）、正常体重患者（p=0.047）和血友病A患者（p<0.001）更有可能在整体自我保健方面取得良好的成绩。具有较高教育程度的患者（p=0.026）、学生（p=0.002）和他人注射给药的患者（p=0.041）更有可能在体育活动中获得良好的成绩。我们在伊拉克InBDs患者中的研究结果显示，在自我保健实践方面总体表现良好，同时需要改进身体活动方面的自我实践，特别是在开始自我输液后父

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母监督减少之后。相当大比例的患者明显超重/肥胖。在肥胖患者和持续关节疼痛的患者中揭示了促进自我激励以实现更好的自我保健的策略的需要。

关键词: 遗传性出血性疾病、自我保健实践、预防措施、体力活动、肥胖.

1. Introduction

Inherited bleeding disorders (InBDs) refer to a heterogeneous group of diseases almost always inherited as an autosomal recessive, dominant, or X-linked pattern. They are characterized by a bleeding diathesis attributed primarily to the quantitative or qualitative abnormalities of coagulation factors, platelets, or both [1-4].

According to a recent annual global survey of the World Federation of Hemophilia, there are more than 287.000 people affected by a coagulation disorder across 106 countries worldwide, including hemophilia A or B in 62.2% of cases, Von Willebrand disease in 24.3%, and other bleeding disorders in 13.5% [5].

The choice of therapy in managing a patient with InBD depends on the type of clotting factor deficiency, disease severity, type of bleeding episode and minimal residual activity, and the patient's responses to previous systemic treatment modalities and factor replacement therapies [6]. Implementing personalized management strategies in compliance with the patient's disease and lifestyle characteristics is critical to ensure optimal outcomes [7]. Moreover, patient education to enable sufficient knowledge and proper understanding of the disease and his/her commitment to essential health-related, physical and nutritional practices are considered vital to prevent disease-related complications [7].

However, repeated soft tissues and joint hematomas (hemarthrosis) in patients with bleeding tendency progressively lead to chronic arthropathy, chronic pain, and disability in the long-term, as a result of the reduction in joint range of movement, joint contracture, or pseudo-tumors, which in turn adversely affect the quality of life [8].

The recognition and prevention of avoidable and undesirable complications of InBD, motivational factors, and autonomy are considered central to improving adherence and self-care behaviors in InBDs, as in many other more prevalent chronic conditions such as diabetes [7], [9]. Fewer studies are available on self-care behaviors in patients with InBDs than those on adherence to medical treatment [9].

Hence, due to the scarcity of studies that address self-care practice among patients with InBDs in Iraq, this research was designed to activate the initiation of the forthcoming interventional programs related to this topic at a national level and to evaluate the performance of Iraqi patients with InBDs on self-care practice in preventing disease-related complications in terms of general preventive measures, physical activity, and healthy dietary patterns.

2. Materials and Methods

2.1. Study Population

A total of 75 male patients with InBDs were included in this prospective case series study conducted from January to June 2019. All adolescent and adult patients with InBDs in the National Center of Hematology were included by consecutive sampling technique. All children below ten years with InBDs were excluded since they could not perfectly describe their performance regarding the investigated issues.

This research was approved by the Ethics Committee of the National Center of Hematology. Written informed consent was obtained from each participant following a detailed explanation of the research objectives and protocol, which was conducted following the ethical principles stated in the Declaration of Helsinki.

2.2. Study Parameters

Socio-demographic data (age, educational status, occupational status) and clinical details (body mass index [BMI, kg/m^2], diagnosis, severity of disease, joint motion, joint pain, injection administration, bleeding control, physiotherapy and analgesic usage) were recorded for each patient. Total and component (general preventive measures, physical activity, and healthy dietary patterns) scores for self-care practice performance were calculated and evaluated regarding patient and disease characteristics.

2.3. Assessment of Performance on Self-Care Practice

The performance of self-care practice was assessed using a self-report questionnaire consisting of 27 individual items related to three domains, including general preventive measures (12 items), physical activities (10 items), and dietary pattern practices (5 items) (Table 1). Each item is scored from 1 (rarely performed: poor performance) to 3 (always performed: good performance) that produce domain scores ranging from 12 to 36 for general preventive measures, 10 to 30 for physical activities, and 5 to 15 for dietary pattern

practices. The overall score ranges from 27 to 81, with higher scores indicating better performance.

For general preventive measures, final scores were categorized as poor (scores <18), moderate (scores 18-27), or good (≥27). For physical activity, final scores were categorized as poor (scores <15), moderate (scores 15-22), or good (≥23). For dietary patterns, final scores were categorized as poor (scores <7.5), moderate (scores 7.5-11), or good (≥11).

Table 1 Items included in the assessment of self-care practice performance scores

	GENERAL PREVENTIVE MEASURES	(1) Rarely "Poor performance"	(2) Sometimes "Moderate performance"	(3) Always "Good performance"
1	I inspect the injured limb periodically after bleeding has stopped			
2	I rest the joint			
3	I use ice bags frequently			
4	I cover with ice (5-10 minute)			
5	I look for numbness, coldness, color change after bandaging the limb			
6	I put injured limb above the heart level			
7	I report fever over 38°C during self-infusion of coagulation factor			
8	I inspect the motion of injured limb frequently			
9	I ware compress for swelling and pain			
10	I consult my doctor before using drug			
11	I adhere to vaccination recommendations (i.e. hepatitis vaccination)			
12	I refer to medical center if I have worries about successful completion of my treatment process			
	PHYSICAL ACTIVITY	**(1) Rarely "Poor performance"**	**(2) Sometimes "Moderate performance"**	**(3) Always "Good performance"**
1	I consult doctor or physiotherapist about type of sport			
2	I exercise at health center			
3	I inform the coach about my condition			
4	I use protection for teeth, heart and pads			
5	I have an ID about my disease			
6	I examine after factor injection peak effect			
7	I avoid fatigue, hunger and too much effort			
8	I inform the coach or manager of school about essential intervention at the time of an incident at school			
9	I have regular school attendance			
10	I participate in regular sports activity (swimming, body building, cycling, football, walking)			
	DIETARY PATTERN	**(1) Rarely "Poor performance"**	**(2) Sometimes "Moderate performance"**	**(3) Always "Good performance"**
1	I adhere to dietary advices (i.e. eating non-stimulating and less spicy foods)			
2	I use laxative at the time of constipation			
3	I get appropriate diet that involve essential nutrients and minerals (like iron)			
4	I apply dietary restriction to avoid gaining excessive weight			
5	I avoid eating solid and hot foods after tongue and mouth bleeding			

Table 2 Socio-demographic and clinical characteristics of patients (n = 75)

Socio-demographic characteristics		
Age (year), n(%)	10-19	29(38.7)
	20-29	28 (37.3)
	30-39	12(16.0)
	40-49	6(8.0)
Educational level, n(%)	Primary education	19(25.3)
	Intermediate education	29(38.7)
	Secondary education	16(21.3)
	Higher education	11(14.7)
Occupational status, n(%)	Student	35(46.7)
	Unemployed	10(13.3)
	Employed	10(13.3)
	Self-employed	20(26.7)
Clinical characteristics		
Body mass index category, n(%)	Underweight	13(17.3)
	Normal	40(53.3)
	Overweight	14(18.7)
	Obese	8(10.7)
Diagnosis, n(%)	Hemophilia A	56(74.7)
	Hemophilia B	13(17.3)
	Factor VII deficiency	6(8.0)
Severity of disease, n(%)	Moderate	16(21.3)
	Severe	59(78.7)
Joint motion, n(%)	Yes	30(40.0)
	No	45(60.0)
Joint pain, n(%)	Always	20(26.7)
	Sometimes	55(73.3)
Injection administration, n(%)	Self-administration	37(49.3)
	By others	38(50.7)
Bleeding control, n(%)	Within 1 h	55(73.3)
	Within 2 h	10(13.3)
	Within 3 h	3(4.0)
	After 6 h	7(9.4)
Physiotherapy, n(%)	Yes	34(45.3)
	No	41(54.7)
Type of analgesic, n(%)	Paracetamol	53(70.7)
	Opioid analgesic	15(20.0)
	None	7(9.4)

2.4. Statistical Analysis

Statistical analysis was performed using IBM SPSS Statistics for Windows, version 23.0 (IBM Corp., Armonk, NY, USA). Chi-square (χ^2) test, Yates Continuity Correction, and Fisher Exact tests were used to investigate the association between categorical data. Data were expressed as the mean (standard deviation, SD), minimum-maximum, and percent (%) where appropriate; $p \leq 0.05$ was considered statistically significant.

3. Results

3.1. Socio-Demographic and Clinical Characteristics

All of the patients were males, and the majority of them (76%) were either adolescents or young adults up to 29 years of age. Overall, 46.7% of patients were students, 26.7% were self-employed, and 38.7% had intermediate education (Table 2).

BMI values were normal in 53.3% of patients, while overweight and obesity were evident in 18.7% and 10.7% of patients, respectively (Table 2).

Hemophilia A was the most common diagnosis (74.7%), followed by hemophilia B (17.3%) and factor VII deficiency (8.0%). The underlying disease was severe in 78.7% of patients with limited joint mobility and constant joint pain in 60.0% and 26.7% of patients, respectively, along with the use of paracetamol type of analgesics by 70.7% of patients (Table 2).

Most of the patients (73.3%) received factors within one hour from the onset of bleeding, while 54.7% of patients were not receiving physiotherapy. More than half of patients were dependent on other persons at the health center or at home to receive the therapeutic injections (Table 2).

3.2 Performance Scores for Overall Self-Care Practice and Components

Overall, the mean total score for self-care practice was 57.9 (SD 5.3, ranged 46 to 70), indicating moderate-to-good performance, while moderate performance was noted in 53 (70.7%) patients and good performance in 22 (29.3%) patients. Considering component scores, the mean scores were 27.2 (SD 33.0, ranged 19 to 33) for general preventive measures, 19.9(SD 3.6, ranged 13 to 28) for physical activity, and 10.8 (SD1.2, ranged 8 to13) for dietary pattern; indicating good, moderate and moderate-to-good performance, respectively.

Table 3 Overall self-care practice performance according to patient and disease characteristics

	Overall self-care practice, n(%		p value
	Moderate performance	Good performance	
Age (years)			
10-19	14(26.4)	15(68.2)	0.453
20-29	25(47.2)	3(13.6)	
30-39	10(18.9)	2(9.1)	
40-49	4(7.5)	2(9.1)	
Educational status			
Primary education	16(30.2)	3(13.6)	0.443
Intermediate education	18(34.0)	11(50.0)	
Secondary education	11(20.8)	5(22.7)	
Higher education	8(15.1)	3(13.6)	
Occupational status			
Student	19(35.8)	16(72.7)	**0.031**
Unemployed	8(15.1)	2(9.1)	
Employed	8(15.1)	2(9.1)	
Self-employed	18(34.0)	2(9.1)	
BMI (kg/m²) category			
Underweight (<18.5)	7(13.2)	6(27.3)	**0.047**
Normal (18.5-24.9)	29(54.7)	11(50.0)	
Overweight (25-29.9)	9(17.0)	5(22.7)	
Obese (≥30)	8(15.1)	0(0.0)	
Diagnosis, n(%)			
Hemophilia A	43(81.1)	13(59.1)	**<0.001**
Hemophilia B	5(9.4)	8(36.4)	
Factor VII deficiency	5(9.4)	1(4.5)	
Severity of disease, n(%)			
Moderate	13(24.5)	3(13.6)	0.234
Severe	40(75.5)	19(86.4)	
Time to control bleeding			
Within 1 h	36(67.9)	19(86.4)	**0.013**
Within 2 h	7(13.2)	3(13.6)	
Within 3 h	3(5.7)	0(0.0)	
After 6 h	7(13.2)	0(0.0)	
Joint motion, n(%)			
Yes	22(41.5)	8(36.4)	0.679
No	31(58.5)	14(63.6)	
Joint pain, n(%)			
Always	19(35.8)	1(4.5)	0.05
Sometimes	34(64.2)	21(95.5)	
Injection administration, n(%)			
Self-administration	27(50.9)	10(45.5)	0.516
By others	26(49.0)	12(54.5)	
Physiotherapy, n(%)			
Yes	24(45.3)	10(45.5)	0.989
No	29(54.7)	12(54.5)	
Type of analgesic, n(%)			
Paracetamol	36(67.9)	17(77.3)	**<0.001**
Opioid analgesic	12(22.6)	3(13.6)	
None	5(9.4)	2(9.1)	

3.3 Self-Care Practice Performance According to Socio-Demographic and Clinical Characteristics

For general self-care practice, including all the three measures, good performance scores were more likely in students (p=0.031), in normal-weight patients (p=0.047), in hemophilia A patients (p<0.001), in patients with faster (within the first hour) control of bleeding (p=0.013), and in patients on paracetamol type of analgesics (Table 3).

There was a non-significant tendency for lower rates of good self-care performance in patients with constant joint pain compared to those experiencing joint pain sometimes (4.5 vs. 95.5%, p=0.05). No significant difference was noted between patients with moderate and good performance on overall self-care practice in terms of age, educational status, disease severity, joint status, injection administration, and physiotherapy (Table 3).

According to patient and disease characteristics, no significant difference was noted in self-care performance on general preventive measures and dietary patterns. For physical activity, good performance scores were more likely in patients with higher educational attainment (p=0.026), in students (p=0.002), in patients with injection administration made by others (p=0.041), and those without the analgesic need (p=0.002) (Table 4).

Table 4 Physical activity according to patient and disease characteristics

	Physical activity, n(%)		p value
	Moderate performance	Good performance	
Age (years)			
10-19	20(33.9)	9(56.3)	0.175[1]
20-29	22(37.3)	6(37.5)	
30-39	12(20.3)	0(0.0)	
40-49	5(8.5)	1(6.3)	
Educational status			
Primary education	19(32.2)	0(0.0)	**0.026[1]**
Intermediate education	22(37.3)	7(43.8)	
Secondary education	12(20.3)	4(25.0)	
Higher education	6(10.2)	5(31.3)	
Occupational status			
Student	23(39.0)	12(80.0)	**0.002[1]**
Unemployed	22(37.3)	0(0.0)	
Employed	10(16.9)	0(0.0)	
Self-employed	4(6.8)	3(20.0)	
BMI (kg/m²) category			
Underweight (<18.5)	9(15.3)	4(25)	0.451[1]
Normal (18.5-24.9)	30(50.6)	8(50)	
Overweight (25-29.9)	13(22)	4(25)	
Obese (≥30)	7(11.9)	0(0.0)	
Diagnosis, n(%)			
Hemophilia A	47(79.7)	9(56.3)	0.056[1]
Hemophilia B	7(11.9)	6(37.5)	
Factor VII deficiency	5(8.5)	1(6.3)	
Severity of disease, n(%)			
Moderate	12(20.3)	4(25)	0.735[3]
Severe	47(79.7)	12(75)	
Time to control bleeding			
Within 1 h	41(70.7)	14(93.3)	0.243[1]
Within 2 h	10(17.2)	0(0.0)	
Within 3 h	3(5.2)	0(0.0)	
After 6 h	4(6.9)	1(6.7)	
Joint motion, n(%)			
Yes	24(40.7)	6(37.5)	1.00[2]
No	35(59.3)	10(62.5)	
Joint pain, n(%)			
Always	19(32.2)	1(6.3)	0.054[3]
Sometimes	40(67.8)	15(93.8)	
Injection administration, n(%)			
Self-administration	44(74.5)	7(43.8)	**0.041[2]**
By others	15(25.4)	9(56.3)	
Physiotherapy, n(%)			
Yes	28(47.5)	6(37.5)	0.670[2]
No	31(52.5)	10(62.5)	
Type of analgesic, n(%)			
Paracetamol	43(72.9)	10(62.5)	**0.002[1]**
Opioid analgesic	14(23.7)	1(6.3)	
None	2(3.4)	5(31.3)	

[1] *Chi Square test*, [2] *Yates Continuity Correction*, [3] *Fisher Exact test*

4. Discussion

Our findings in Iraqi patients with InBDs revealed moderate-to-good performance regarding self-care practice overall and slightly better performance for adherence to general preventive measures and healthy dietary patterns rather than physical activity recommendations.

The association of hemophilia A with better self-care practice than other InBDs in our study seems notable given that an appropriate level of protection is considered crucial, particularly in the context of hemophilia to avoid potentially dangerous behaviors without triggering psychological distress related to overprotection or overindulgence [9].

Although a reduction in infection-related mortality/morbidity rates and advances in factor replacement therapy has improved the overall life expectancy and quality of life among patients with hemophilia in recent years, the high prevalence of overweight and obesity in patients with target joints remains an important problem [10]. The inability to maintain a healthy weight has been linked to barriers towards engagement in physical activity, such as reduced activity to prevent bleeding and protect the joints and the association of target joints with chronic joint pain and restriction of movement due to progressive arthropathy [10]-[11]. Muscle atrophy due to reduced mobility and loss of muscle function is also considered to increase the risk of weight gain [10], [12].

Accordingly, nearly one-third of patients in the current study were overweight or obese, while the presence of overweight or obesity was associated with poorer performance scores in overall self-care practice related to general preventive measures, dietary patterns,

and physical activity. This seems notable given that obesity, particularly in young patients with hemophilia, is considered a health issue and an aggravating factor for poor joint health and poor cardiometabolic health [11].

Notably, patients with InBDs are considered to have a two-fold higher likelihood of obesity than the same-age persons in the general population [13], while high rates of overweight and obesity were reported in a relatively younger hemophilia cohort [14]. This observation is important given that the overweight/obesity itself was associated with an increased number of joint bleeds, and reduced function in the lower limbs was reported in a Dutch hemophilia cohort [11].

In fact, the lower performance scores for physical activity measures in our study population comprising young patients seems important given the association of appropriate physical activity with improved fitness, neuromuscular development, and healthy body weight to prevent joint damage in hemophilia patients [15]. Notably, a study among 459 young hemophilia patients across the US reported that 60% of patients managed their hemophilia by simply avoiding entire physical activity in contrast to guideline recommendations encouraging non-contact sports such as swimming [16]. Better physical activity scores per se were associated specifically with certain factors such as lack of analgesic need and being a student, and higher educational attainment in our cohort. This finding seems notable given the data from a past study with 74 young patients with hemophilia in Germany, in which authors reported that "having fun" and "social aspects" rather than "health benefits" were indicated by patients as their primary motivations for participating in sports activities [17].

Although our findings revealed no significant impact of age or self-infusion on overall self-care practice, being a student was associated with a higher likelihood of dependency on others for injection administration. Furthermore, both factors (being a student and dependency on others for infection administration) increased adherence to physical activity recommendations. This seems to support the consideration that children and adolescents are more likely to avoid general recommendations with a decrease in parental supervision after they start self-infusion [18]. Nonetheless, overall self-care practice scores indicated good performance only in one-third of our study population. Most patients were in the 10-19-year or 20–29-year age groups, and half performed self-infusions.

Hence, our findings emphasize the need for improved self-care practice, particularly in terms of physical activity recommendations after the start of self-infusion practice. They also highlight the likelihood of overall poor performance in overweight/obese patients and a tendency for poorer self-care performance in patients with constant joint pain. In this regard, healthy dietary patterns and engaging in appropriate physical activities should be encouraged in Iraqi patients with InBDs to adequately address illness-related issues properly and timely by developing and maintaining motivation for their treatment and health behaviors (9).

The major strength of the current study is that it represents the first approach to identify and possibly enhance the management of InBDs in Iraq. Certain limitations to this study should be considered. The first small sample size due to the single-center design of the present study is an important limitation for generalizing our findings to the overall InBD patient population. It emphasizes the need for nationwide studies involving all hemophilia treatment centers in Iraq to provide more comprehensive data on the performance of Iraqi patients to initiate the appropriate future interventional programs in the country. Lack of cooperation experienced by some participants in answering the questionnaire items due to loss of concentration, misunderstanding, or disease-related conditions is another limitation of the current study.

5. Conclusion

In conclusion, our findings in Iraqi patients with InBDs revealed an overall favorable performance on self-care practice. At the same time, a need for an improved self-practice was noted considering the physical activity, particularly after the decrease in parental supervision with the start of self-infusion, rather than general preventive measures or dietary habits. Our findings also indicate the presence of overweight/obesity in a considerable percentage of patients and a need for strategies to promote self-motivation towards better self-care, particularly in obese patients and those with constant joint pain for joint health and cardio-metabolic health.

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