

ORIGINAL ARTICLE

Correlations between hemarthrosis, physical assessments and functionality of upper extremity in hemophilic arthropathy of the elbow

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Purpose: The aim of this study was to examine the correlations between hemarthrosis, physical assessments, and functionality of upper extremity in hemophilic arthropathy (HA) of the elbow.

Methods: This cross-sectional study includes 17 adolescents and young adults with HA of elbow aged between 12 and 30 years. In this scope 23 elbow joints were evaluated. Number of hemarthrosis in elbow was asked to the patients. Pain level was assessed with the Numerical Pain Scale (NPS), range of motion with universal goniometer, muscle strength with digital dynamometer. Hemophilia Joint Health Score-Elbow Point (HJHS-EP), Hospital for Special Surgery Elbow Scoring System and Quick-Disability of Arm, Shoulder and Hand (Q-DASH) were also used for assessing physical status and functionality of elbow and upper extremity.

Results: There were correlations between HJHS-EP and loss of extension ($r=0.54$, $p=0.008$) and between activity NPS and pronation/supination angles ($r=-0.55$, $p=0.007$ and $r=-0.60$, $p=0.003$; respectively). Loss of extension was showed moderate negative correlation with triceps muscle strength ($r=-0.53$, $p=0.01$). The Q-DASH was strongly negative correlated with triceps muscle strength and pronation angle ($r=-0.58$, $p=0.004$ and $r=-0.74$, $p<0.001$; respectively). There was strong positive correlation between number of hemarthrosis and NPS ($r=0.56$, $p=0.005$).

Conclusion: We thought that extension loss and pronation angle were important variables in the physical examination of the elbow in hemophilic arthropathy and functionality may be increased with improvement of pronation angle and triceps muscle strength also the number of hemarthrosis can be decreased by reducing activity pain.

Keywords: Arthropathy, Elbow joint, Hemophilia, Range of motion.

Dirsek hemofilik artropatisinde hemartroz, fiziksel değerlendirmeler ve üst ekstremitte fonksiyonelliği arasındaki ilişkiler

Amaç: Bu çalışmanın amacı, dirsek hemofilik artropatisinde, hemartroz ve üst ekstremitenin kas-iskelet sistemi ve işlevselliği arasındaki ilişkiyi incelemektir.

Yöntem: Bu kesitsel çalışma yaşları 12-30 arasında 17 adolesan ve genç erişkin içermektedir. Bu kapsamda 23 dirsek eklemi değerlendirildi. Hastalara dirsek eklemindeki kanama sayıları soruldu. Ağrı seviyesi Nümerik Ağrı Skalası (NAS) ile eklem hareket açıklığı evrensel gonyometre ile kas kuvveti dijital dinamometre ile değerlendirildi. Hemofili Eklem Sağlığı Skoru-Dirsek Puanı (HESS-DP), Hospital for Special Surgery Dirsek Skorlama Sistemi ve Kol, Omuz ve ve El Sorunları Hızlı Anketi (Q-DASH: Quick-Disability of Arm, Shoulder and Hand) dirsek ve üst ekstremitenin fiziksel durumunu ve fonksiyonelliğini değerlendirmek için kullanıldı.

Bulgular: HESS-DP ile ekstansiyon kaybı arasındaki ($r=0,54$, $p=0,008$) ve aktivite NAS ve pronasyon/supinasyon açıları arasındaki güçlü ilişkiler (sırasıyla, $r=-0,55$, $p=0,007$; $r=-0,60$; $p=0,003$) vardı. Ekstansiyon kaybı, triceps kas kuvveti ~~güçü~~ ile ters yönlü güçlü ilişki ($r=-0,53$, $p=0,01$) gösterdi. Q-DASH, triceps kas kuvveti ve pronasyon açısı ile ters yönlü güçlü ilişkiydi (sırasıyla, $r=-0,58$, $p=0,004$; $r=-0,74$, $p<0,001$). Kanama sayısı ile aktivite NAS arasında pozitif yönlü güçlü ilişki vardı ($r=0,56$, $p=0,005$).

Sonuç: Ekstansiyon kaybı ve pronasyon açısının dirsek hemofilik artropatisinde fiziksel değerlendirmesinde önemli değişkenler olduğunu ve fonksiyonelliğin, pronasyon açısı ve triceps kas kuvvetinin iyileşmesiyle gelişebileceğini ayrıca aktivite ağrısı azaltılarak hemartroz sayısı düşürülebileceğini düşündük.

Anahtar Kelimeler: Artropati, Dirsek eklemi, Hemofili, Eklem hareket açıklığı.

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Recurrent hemarthrosis is the most prominent finding of severe hemophilia and lead to disability with aging and frequently occur in hinge joints such as the knee, elbow and ankle.^{1,2} The common features of these joints are that they have a high synovial fluid content and prone to trauma.³ The elbow joint is the most commonly affected upper extremity joint in patients with hemophilia. Recurrent hemarthrosis makes the joint more vulnerable to new a hemarthrosis.⁴ The inflammatory process that begins with recurrence of bleeding results in synovitis, osteochondral degeneration and advanced HA. Hemophilic arthropathy (HA) is inevitable if recurrent hemarthrosis cannot be controlled.⁵ Symptoms in the early stages of HA include swelling, muscle atrophy or weakness, and limited range of motion (ROM). In the end stage, there are increased pain, joint destruction and loss of ROM. Treatment depends on the stage of the HA and the condition and location of the affected joint.⁶

The elbow is a complex joint in upper extremity and recurrent hemarthrosis causes structural alterations. Radial head hypertrophy is first sign in HA of the elbow. This pathology lead to compression of the proximal ulnar facet then restricting rotational movements.⁷ Over time osteochondral defects in the olecranon fossa and contractures in the soft tissues cause limitation of flexion and extension.⁸ Secondary problems such as valgus instability and cubital tunnel syndrome may be seen due to progression of HA.⁹

In the literature there are scanty data according the detailed physical evaluations and the functionality of the elbow joint and upper extremity in the patients with hemophilia. Poonnoose et al. used only The Hemophilia Joint Health Score 2.1 (HJHS) for physical evaluation of the elbow in their study.¹⁰ Cuesta-Barriuso et al. performed the manual therapy in HA of elbow.¹¹ Ernstbrunner et al. surgical treatment applied in advanced HA of elbow.¹² But The HJHS was not used in both studies. Musculoskeletal system detailed evaluation of the affected joints should be performed in order to perform the treatment correctly. In addition to The HJHS International Prophylaxis Study Group-Physical Health and Joint Function Expert Working Group recommends assessment of functionality in addition to joint health.^{13,14}

Due to the lack of detailed evaluation studies in the literature, we planned this study. The hypothesis of our study is that hemarthrosis adversely affects the health of elbow joint and functionality of the upper extremity. The aim of this study was to evaluate the elbow joint and upper extremity and examine correlations between hemarthrosis, physical assessments and functionality of upper extremity in HA of the elbow.

METHODS

This cross-sectional study was to evaluate the elbow joint and upper extremity in the patients with hemophilia. Ethics committee approval of the study was obtained from the Cukurova University on 6 July 2018 with decision no: 79/48. In order to calculate the sample size of the study we used to pain variable in study performed by Cuesta et al. (Effect size: 0.77, 80% power, $\alpha=0.05$) and found that at least 16 cases.¹⁵ The patients were informed by the system of Association of Cukurova Hemophilia and invited to the study for evaluation. The inclusion criteria were to be treated with prophylactic factor replacement. The exclusion criteria were affected of the shoulder and/or wrist joints in addition to the elbow. Participants were given verbal and written consent about the study from themselves (aged over 18 years) and their families (aged 12-18).

Physical examination

In the study, 17 adolescents/young adults aged 12-30 were included and 23 elbow joints were evaluated. In socio-demographic evaluation, age, height, weight, type and severity of hemophilia, affected elbow joint and number of hemarthrosis in the this joint over the last 5 weeks were questioned. In the physical evaluation of the musculoskeletal system, Numerical Pain Scale (NPS) was used for pain, universal goniometer for ROM and digital dynamometer (Lafayette Hand-Held Dynamometer®) for muscle strength. In the scale-based physical evaluation of the elbow joint, the elbow point of the HJHS and the Hospital for Special Surgery Elbow Scoring System (HSS-ESS) were performed by the physiotherapist. The Quick-Disability of Arm, Shoulder and Hand (Q-DASH) questionnaire

was used to assess functionality. Thoracic spine was evaluated for scoliosis and kyphosis.

In Numerical Pain Scale (NPS) which was used for pain assessment patients with hemophilia were asked to mark the appropriate 0 to 10 pain levels on a 10 cm bar separately for rest and activity pain.¹⁶ Elbow flexion/extension and forearm pronation/supination angles were measured with a universal goniometer.¹⁷ Muscle strength assessment was performed on flexors/extensors of elbow, flexors/extensors/abductors of shoulder flexors/extensors of wrist. Elbow flexors and extensors were evaluated with resistance for flexors from the anterior forearm and extensor muscle posterior to the forearm while the patient was in the sitting position the arm next to the trunk and the elbow was flexed approximately 45°. Force was applied proximal to the elbow while the shoulder flexors were in the 90° flexion of the shoulder and the elbow in the flat position. The shoulder extensors were evaluated in the shoulder 90° flexed and the elbow flexed. The individual was asked to perform the appropriate muscle movement with maximal isometric contraction.¹⁸ In the strength measurement of the wrist flexor/extensors the dynamometer was placed on the palm of the hand while the arm was facing upwards while the arm was hanging from the table to the half of the forearm while the arm was sitting in the sitting position next to the trunk. For extensors the dynamometer was placed on the back of the hand with the palm facing down and the same procedure was applied.¹⁹

The HJHS is a physical examination tool that evaluates knee, elbow and ankle joints bilaterally and total score is obtained by adding a global gait score. In our study we only used the elbow point of HJHS. In the evaluation of HJHS, 8 parameters such as swelling, swelling duration, crepitus in motion, atrophy, loss of flexion, loss of extension, joint pain and muscle strength are scored according to HJHS's own assessment system. The validity and reliability study of HJHS in adolescent and young adult hemophilia patients was conducted by Fischer and De Kleijn in 2013.²⁰ Assessments were performed by physiotherapist who was non-blind to the study.

The HSS-ESS is a scoring in which the physical assessment of the elbow joint is performed by the clinicians. Turchin et al.

conducted the validity study of HSS-ESS.²¹ The assessment consists of 8 sections: pain, function, sagittal arch, muscle strength, flexion and extension contracture, pronation and supination angles. If the total score is 90-100 it is perfect, 80-89 is good, 70-79 is moderate, 60-69 is weak and a score of 60 or less is considered to be a poor outcome.²²

The Q-DASH is a quick form of DASH, which evaluates the functionality of the upper extremity and the findings related to the disease. In Q-DASH 11 questions in the questionnaire and the questions are answered as 1-5 between 1=No difficulty, 5=Unable. The total score is calculated from 0-100, and as the score approaches 100, it indicates that functionality and symptoms worsen and the Turkish validity and reliability study was conducted by Duger et al.²³

Statistical analysis

The suitability of the variables to normal distribution was analyzed by analytical methods (Kolmogorov-Smirnov/Shapiro-Wilk tests). Descriptive statistics are given as mean±standard deviation (X±SD) for normally distributed variables. For non-normally distributed variables, median values and minimum-maximum values are given. In order to determine whether there is a relationship between the variables, direction and severity of the relationship, Pearson correlation is preferred if the data has normal distribution and Spearman's correlation is preferred if the data is not normally distributed. Statistical significance level was accepted as $p < 0.05$. SPSS 22.0 package was used.

RESULTS

The mean values of age, height and weight of the 17 patients with hemophilia were 22.94±5.65 years, 172.53±6.74 cm, and 69.71±16.72 kg, respectively. Sixteen patients had severe hemophilia, 1 had moderate hemophilia, 3 had hemophilia B, 14 had hemophilia A and 1 had inhibitory positive. Six patients had right joint, 5 had left joint, 6 had both elbows. Eight of the evaluated elbow joints belonged to individuals between 12-18 years and 15 belonged to individuals between 18-30 years. One adolescent had right thoracic scoliosis. In the statistical analysis of the variables used in

the study, the mean and standard deviation values of those with normal distribution and the median and minimum-maximum values of those without normal distribution are given in Table 1. The *r* and *p* values obtained from the correlation analysis between number of hemarthrosis, activity/resting NPS, HJHS-EP, Q-DASH and other variables are given in Table 2. In tables, ** and * indicates respectively strong and moderate correlation, - shows negative correlation.

DISCUSSION

In our study, physical and functional evaluations of the elbow joint and upper extremity were evaluated in individuals with HA of the elbow and the relationships between the variables were also determined. As a result, loss of extension and pronation angle in elbow joint showed strong correlation with some other variables. This finding considered the importance of these two variables. Correlations between Q-DASH and triceps/biceps muscle may indicate that function improve if muscle strength increases.

In our study, 17.6% of the patients with HA of the elbow had hemophilia B and the rest was hemophilia A. It has been reported in the literature that approximately 85% of hemophilia patients are hemophilia-A patients.¹ In one patient, the inhibitor was positive and proportionally corresponded to 5.8%. According to the results of the screening programs performed in our country in the literature, inhibitor development rate in hemophilia A was reported as 10% and in hemophilia B as 3%.¹ These data of the individuals are consistent with the literature. A 13-year-old adolescent with positive inhibitor had advanced HA and loss of 72° elbow extension in his left elbow. Thoracic spine examination of this individual revealed a right thoracic scoliosis, the opening of which was left-sided. This may be due to advanced HA of the elbow. The case of patients with inhibitor having scoliosis considered that inhibitor increases joint bleeding frequency and eventually, causes advanced arthropathy at an early age and other musculoskeletal problems such as scoliosis. Our findings are consistent with the literature knowledge including that joint conditions are worse in patients with

inhibitor and the incidence of advanced stage arthropathy is higher.^{24,25}

In the study of Atalar et al. the mean Q-DASH score of the patients with HA of the elbow aged between 15-48 years who underwent radial head excision surgery was reported as 41.4.²⁶ The median value of Q-DASH in our study was found to be 21.5 (0-61.3). The low Q-DASH scores in our study can be attributed to the relatively small number of patient required surgical treatment. The median values of HSS-ESS were 83 and the minimum-maximum values were 15-96. In the internal evaluation of the questionnaire, 80-89 were classified as "good". This result may be due to different stages of arthropathy among patients. The median value of the pronation angle of the individuals in our study was 70°, and the angle of supination was 90°. In the surgical treatment studies in the patients with HA of the elbow, the supination and pronation angles are consistent with our study.^{27,28} It was seen in our study that the median value of the supination angle was closer to normative values than the pronation angle. We may say that the pronation angle is affected more than the supination but more clinical studies larger sample size is needed.

A strong correlation between HJHS-EP and loss of elbow extension may indicate importance of this angle. A moderately negative correlation between loss of elbow extension and triceps muscle strength is consistent with the information in the literature knowledge about that as muscle strength decreases, the joint is more exposed to bleeding and arthropathy and joint ROM limitation progresses.²⁹ The negative correlation between Q-DASH and HSS-ESS may considered that improving of clinical symptoms can restore functionality. The strong correlation of Q-DASH with the pronation angle may indicate the clinical significance of the pronation angle. Strong correlation of Q-DASH with triceps muscle strength and moderate correlation of Q-DASH with biceps muscle strength may indicate that muscle strength is related to function. In the literature, there are studies showing that muscle strength and functions are related to each other.^{30,31}

Activity NPS strong correlated with pronation and supination angles considered that rotational movements in daily activities cause compression in the proximal ulnar facet and provoking pain.⁷ The elbow joint is bleeding in

Table 1. Descriptive statistical values of the all variables.

	Median (Min-Max)	Mean±SD
Number of Hemarthrosis *	1 (0-5)	
Resting NPS* (0-10)	0 (0-5)	
Activity NPS (0-10)		3.48±2.25
HJHS-EP (0-20)		7.52±3.65
Q-DASH* (0-100)	21.50 (0-61.3)	
HSS-ESS* (0-100)	83 (15-96)	
Elbow Flexion (°)		122.96±8.22
Loss of Elbow Extension* (°)	13 (3-72)	
Supination* (°)	90 (32-95)	
Pronation* (°)	70 (40-80)	
Biceps MS (lbs)		25.33±5.71
Triceps MS (lbs)		15.29±4.77
Shoulder flexors MS (lbs)		24.94±6.62
Shoulder abductors MS (lbs)		25.22±6.68
Shoulder extensors MS (lbs)		18.31±5.61
Scapular adductors MS (lbs)		16.56±5.49
Wrist extensors MS (lbs)		16.42±4.0
Wrist flexors MS (lbs)		15.17±3.93

NPS: Numerical Pain Scale, HJHS-EP: Hemophilia Joint Health Score-Elbow Point, Q-DASH: Quick-Disabilities of the Arm, Shoulder and Hand Questionnaire, HSS-ESS: Hospital for Special Surgery-Elbow Score System, MS: Muscle Strength lbs: Pound. Kolmogrov-Smirnov and Shapiro-Wilk tests were used in statistical analysis. * As the variables do not show a normal distribution, median (minimum-maximum) values are given.

Table 2. Correlation between hemarthrosis, pain, Hemophilia Joint Health Score, Q-DASH, and other variables.

	N. of hemarthrosis		Activity-NPS		Resting-NPS		HJHS-EP		Q-DASH	
	r	p	r	p	r	p	r	p	r	p
N. of hemarthrosis	-	-								
Activity NPS	0.56	0.005*	-	-						
Resting NPS	0.14	0.532	0.30	0.168	-	-				
HJHS-EP	0.24	0.26	0.22	0.321	0.36	0.088	-	-		
Q-DASH	0.32	0.133	0.52	0.023*	0.34	0.113	-0.43	0.040*	-	-
HSS-ESS	-0.13	0.548	-0.36	0.092	-0.70	0.001*	-0.46	0.025*	-0.48	0.020*
Flex. angle of elbow	-0.12	0.570	-0.40	0.059	-0.51	0.012*	0.54	0.008*	-0.27	0.219
Loss of extension	0.19	0.376	0.07	0.752	-0.07	0.750	-0.14	0.518	0.29	0.174
Supination	-0.12	0.585	-0.55	0.007*	-0.18	0.401	-0.22	0.309	-0.29	0.182
Pronation	-0.34	0.111	-0.59	0.003*	-0.21	0.335	-0.23	0.280	-0.74	0.001*
Biceps MS	0.01	0.981	-0.08	0.706	-0.31	0.145	-0.24	0.270	-0.42	0.047*
Triceps MS	-0.08	0.716	-0.09	0.670	-0.11	0.619	-0.29	0.183	-0.58	0.004*
Shoulder flexion-MS	0.24	0.275	0.11	0.629	-0.06	0.784	-0.07	0.751	-0.23	0.297
Shoulder abd.-MS	0.10	0.651	0.21	0.346	0.05	0.827	0.05	0.817	-0.07	0.762
Shoulder ext.-MS	-0.29	0.184	-0.33	0.127	-0.10	0.637	-0.02	0.921	-0.36	0.094
Wrist extension-MS	-0.25	0.245	-0.05	0.823	0.04	0.843	-0.16	0.462	-0.37	0.086
Wrist flexion-MS	-0.11	0.611	-0.05	0.834	-0.03	0.900	-0.43	0.040*	-0.07	0.734

* p<0.05. r: Correlation coefficient. N.: Number. NPS: Numeric Pain Scale. HJHS-EP: Hemophilia Joint Health Score-Elbow Point. Q-DASH: Quick-Disabilities of the Arm, Shoulder and Hand Questionnaire. HSS-ESS: Hospital for Special Surgery-Elbow Score System, MS: Muscle Strength. Flex.: Flexion. Abd.: Abduction.

cases of trauma and excessive exposure to force. In cases where muscle strength is insufficient, the inability of the muscle to compensate the overloads of the joints may cause joint bleeding.²⁹ Activity NPS strongly correlated with number of hemarthrosis may mean that muscle strength is insufficient to cover loads on joint during movement. Shoulder and wrist muscle strengths did not show a significant relationship with other variables. As a result of this finding, it may be considered that the HA of the elbow did not adversely affect the muscles of other joints of the upper extremity. However, studies comparing muscle strength of healthy individuals are needed.

Limitations

The limitations of this study, radiological imaging of the joint could not be performed in patient with hemophilia. If these measurements could be made, the elbow joint with HA would have been comprehensively evaluated.

Conclusion

We thought that extension loss and pronation angle in HA of the elbow may be important variables in examination due to its correlation with other variables and elbow flexor and extensor strengthening may be necessary because of its relationship with functionality. We have also suggested that number of hemarthrosis can be decreased by reducing activity pain.

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Ethical Approval: The protocol of the present study was approved by the Cukurova University Faculty of Medicine Non-interventional Clinical Research Ethics Committee (issue: 79/48 date: 06.07.2018).

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