

Functional Independence Score in Haemophilia: Ege University Adult Haemophilia Center Experience



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Introduction: Haemophilia A and B are X-linked hereditary disorders of factor 8 and 9 deficiency, respectively. Both are categorized according to the factor level in serum to mild (1%>), moderate (1-5%) and severe (>5%). Haemarthrosis is most common complication in haemophiliac patients, especially in elbows, knees and ankles. Synovial hypertrophy and the toxicity of ferrous in the joint due to recurrent haemarthrosis lead to complaints such as pain, limitation of motion, deformities, muscle weakness and atrophy, joint contracture and crepitus. Functional Independence Score in Hemophilia (FISH) has been developed as a measure of disability in patients with hemophilia. We aimed to evaluate the FISH and scores in adult haemophilia patients.

Material and methods: From June 2013 to August 2013, we evaluated 53 patients of whom 37 patients were with haemophilia A and 16 patients were with haemophilia B. All the patients are under supervision of Adult Hemophilia Clinic, Ege University Haematology Department. All of the patients are receiving support care, laboratory investigation and factor concentrates.

FISH evaluates three categories (eight activities): self care (eating, grooming, bathing and dressing), transfer (chair, squatting) and mobility (walking, going up stairs, running). Each activity is graded from 1 to 4 according to the level of independence and assistance required. The minimum and maximum scores are 8 and 32, respectively.

Results: Totally, 53 patients with the median age of 31 (20-45) were evaluated. 37 of patients were haemophilia A and 16 were haemophilia B. No inhibitor was detected.

23 of haemophilia A patients were severe, 12 were moderate and 2 were mild. 7 of haemophilia B patients were severe, 6 were moderate and 3 were mild. Patients were divided as group 1 and group 2 due to factor levels. Group 1 was mild and moderate patients and group 2 was severe patients for both haemophilia A and B. 70% of haemophilia A patients were in prophylaxis programme. (25 IU/kg per week) 40% of haemophilia B patients were in prophylaxis programme. (25 IU/kg per week)

Table 1. The mean score of eight activities of FISH in 53 haemophilic patients.

	Mean			
	Haemophilia A		Haemophilia B	
	Group 1	Group 2	Group 1	Group 2
Eating	3,78	3,86	3,65	4,0
Bathing	3,92	3,82	3,44	3,86
Dressing	3,78	3,13	3,44	3,71
Chair transfer	3,64	3,21	3,44	3,67
Squatting	2,71	1,69	2,0	2,43
Walking pattern	3,35	3,08	3,33	3,71
Stair climbing	3,42	2,95	3,0	3,71
Running	2,64	1,65	2,33	2,86
TOTAL	27,24	23,39	24,53	27,85

The mean FISH score for group 1 (mild+moderate) haemophilia A patients was 27,24 and 23,39 for group 2 (severe). The mean FISH score for group 1 haemophilia B patients was 24,53 and 27,85 for group 2. FISH score was lower in group 2 in haemophilia A, as expected. But surprisingly, group 2 FISH score was very high from group 1 in haemophilia B.

Discussion: FISH in haemophilia is a simple questionnaire assessing the disability in haemophilia A and B patients. We have a gap of education which results in lower level of function in home care. Adult patients would require some additional support and education to adopt to the adult life and survive with their disease and complications. More training projects are needed for adult haemophilia patients.

