Scholars Journal of Applied Medical Sciences (SJAMS) **ISSN 2320-6691 (Online)**
Sch. J. App. Med. Sci., 2016; 4(6E):2196-2199 **ISSN 2347-954X (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI: 10.36347/sjams.2016.v04i06.067

Original Research Article

Functional Independence Score in Hemophiliacs and Factors affecting it

Dr. S. Malathi [1],Dr. S. Kalaichelvi [2], Dr. Sivanesan [3], Dr. R. Vijay Usha Raj [4], Dr. V. Madhavan[5]
[1,2,4,5]Assistant Professor, Department of Medicine, Govt KMC Medical College, Chennai, Tamilnadu, India
[3]Junior Resident, Department of Medicine, Govt KMC Medical College, Chennai, Tamilnadu, India

***Corresponding author**
DR. S. Malathi
Email: malathidesigan@gmail.com

Abstract: Haemophilia is rare genetic disorder commonly affecting males with spontaneous bleeding. Recurrent joint bleed or hemarthrosis is very common in such patient. Chronic arthropathy sets in over a period of time leading to severe restriction of joint movements. In this study we have assessed the joint status of haemophilia patients using a tool called FISH – functional independence score in haemophilia patients, which is a performance based assessment tool. In this cross sectional study we have studied 58 subjects for 8 activities under 3 categories – self-care (eating and grooming, bathing and dressing), transfers (chair transfer and squatting) and locomotion (walking, stair climbing and running). Each activity was scored from 1 to 4 according to the level of assistance required. As a whole the mean age of patients were 28, mean FISH score was 26 lowest FISH score was for squatting, and stair climbing, Knee joint was the commonly involved joint. Presence of co-morbidities, ageing, and low I.Q and multi joint involvement significantly lowered FISH score, whereas bleeding episodes, severity, physiotherapy and distance travelled for treatment did not affect the score. This study gave insight into joint status of haemophilic patients, and the factors influencing their functionality.
Keywords: Haemophilia, functional independence score, FISH, haemophilic arthropathy.

INTRODUCTION:

Haemophilia is a group of related bleeding disorders that are inherited. INDIA has the second highest burden of haemophilia patients in the world. The pathophysiology of Haemophilia A and Haemophilia B is based on the insufficient generation of thrombin by the factor IXa/factor VIIIa complex through the intrinsic pathway of the coagulation cascade. Spontaneous hemarthroses are characteristic of severe disease. The most common sites of bleeding are into joints and muscles[1]. Approximately 80 per cent of haemorrhage occurs in the joints, Chronic and recurrent joint bleeds will lead to extensive destruction of articular cartilage, synovial hyperplasia and other reactive changes. This leads to joint deformity, muscle atrophy and soft tissue contractures, eventually leading to functional disability. Severe disability due to chronic hemarthroses is a tragic consequence of haemophilia patients in India [2]. The onus is on the treating physician to prevent this functional disability and hence more light has to be shed into factors leading to joint deformity unctional independence scoring in haemophiliais used to measure the disability in haemophilia patients. FISH is easily administered, reliable and inexpensive[3]. Aim of this study is to assess the functional joint status and the factors affecting it. Identifying the factors having positive or negative influence in functional status of haemophilia patients will help to improve the morbidity and mortality in haemophiliacs.

MATERIALS AND METHODS:

This study was done at Government Royapettah Hospital, Chennai for a period of eight months from December 2014 to September 2015. The study wasperformed after procuring informed written consent from all the participantsinvolved. Clearance was obtained from the Ethical Committee of theGovernment Kilpauk Medical College & Hospital Chennai.The study design is a cross sectional study.The study population included 58 patients who attended the HaemophiliaOP at Government Royapettah Hospital and in-patients in the same hospital.

Inclusion Criteria

1. Patients diagnosed as Haemophilia including
2. Factor VIII deficiency including inhibitors
3. Factor IX deficiency
4. Von Willebrand disease

Exclusion Criteria

1. New born babies with haemophilia
2. Patients with acute bleed
3. Bleeding disorders other than haemophilia

All patients, diagnosed and registered in the Haemophilia clinic were taken as the study population. The sample size was set to be 58. A detailed history regarding the onset and progression of the disease, family history, maternal carrier status, treatment history and the presence of complications were taken. After obtaining consent joint status assessment was done using Functional Independence Scoring in Haemophiliacs (FISH) scoring system. Single trained doctor assessed all patients to avoid inter observer bias. Each patient was evaluated for 7 activates, eating, grooming, bathing, dressing, chair transfer, walking, stair climbing and running. Each activity was graded from 1 to 4 according to the level of assistance required for each activity.The observations were statistically analysed using SPSS 20.0 software and results were interpreted.

RESULTS:

We studied 58 patients out of which 44(76%) were Hemophilia A, 10 (17%) were Hemophilia B and 4 (7%) were Von willebrand disease. All 4 Von willebrand patients werefemales (7%) and others were males (93%).The minimum age of the study population was 4years and maximum age was 85, with a mean age of 28.24.Mean FISH score was 26, lowest was 8 and maximum of 32. Lowest mean score was for squatting,running and then stair climbing, while running had the lowest mean score.

Table-1:Scores obtained in each category of FISH scoring in haemophiliacs (n=58)

FISH	MINIMUM	MAXIMUM	MEAN
Eating/grooming	1	4	3.88±0.46
Bathing	1	4	3.59±0.83
Dressing	1	4	3.59±0.83
Chair	1	4	3.47±1.01
Squatting	1	4	2.71±1.25
Walking	1	4	3.34±0.98
Stairs	1	4	2.97±1.1
Running	1	4	2.60±1.3
Total fish score	8	32	26.00±6.5

Table-2:Correlation between age and duration of disease to FISH score(n=58)

FISH	Age in years	Duration of disease(yrs)
Total fish score	-.678(**)	-.562(**)

There is a significant negative correlation between age and duration of disease and FISH score, i.e., as the age and duration of disease increases, the FISH score is reduced.

Table-3:Correlation between employment status and FISH score (n=58)

EMPLOYMENT STATUS	N	MEAN	P Value
Unemployed[a]	7	18.14±7.98	<0.01**
Student[c]	21	31.76±0.625	
Semi skilled[a]	19	22.11±5.065	
Unskilled[b]	6	26.50±5.505	
Professional[b]	5	27.00±2.12	

Mean FISH score was significantly lower in unemployed group and highest in students.

Table-4:Correlation between IQ and FISH score, normal IQ (n=52) sub normal IQ (n=6)

FISH	IQ	N	MEAN	P VALUE
Total fish score	Normal	52	26.58±5.886	0.047*
	Sub Normal	6	21±10.1	

From this table, FISH scoring was significantly low (level 5) in patients with sub normal IQ.Maximum bleed per month was 6 with a mean of 1.4 episodes. Mean bleed per year per patient was 16 episodes.

Table-5:Correlation of FISH with co morbidities, co morbid(n=7), no comorbities(n=51)

CO MORBID	N	MEAN	P VALUE
Present	7	19.00±7.61	0.002**
Absent	51	26.96±5.84	

FISH score was significantly lower in patients with co morbidities like HIV/HBSAG assessed using two tailed t test.

DISCUSSION

Analysis of FISH score shows that lowest scores are for squatting, followed by running and stair climbing. Mean FISH score was 26, the results are comparable to a where the mean FISH of Mexican haemophiliacs was 25.8. [4]. Since squatting was most difficult to perform, we should give early toilet training to haemophilia children and advice them to use western style lavatories to avoid stress on knee joint. Demographic and clinical characteristics of our study population are, Oldest patient was 85 years and the mean age of study population was 28 years, whereas a similar study conducted by A. Kar et al in Kolkata haemophilia society, INDIA had a mean age of to be 19.2 years [3]. Mean age of diagnosing haemophilia in our centre was 5.5 years, which was comparatively late than western population[5]. Age also seems to be an important factor in determining joint function, as we can see, as the age goes up, the FISH score becomes significantly lower.93% were males, this group majority were hemohilia A and 7 % were females[6]. All females had von willebrand disease. But the type of disease did not significantly influence joint function.70% of our patients are having severe disease i.e., factor levels <1%, but the severity of their disease did not affect the joint function, probably because arthropathy is a chronic problem due to repeated joint trauma, rather than the factor levels in the serum.7% were inhibitor positive but it did not significantly affect the FISH score.48% population were in the no income group , probably because most of our patients were students , hence they did not have stable income source, comparable to a similar study in Kolkata[7].Our study had 10% patients with sub normal I Q levels, and they were significantly having low FISH score, which shows that I Q is an important determinant of joint health. Most commonly involved joint in our study was knee joint followed by elbow and ankle joints were similar because these joints lack adequate muscle cover and are not able to withstand rotatory and angular stress[8]. We also found that 86% of our patients had multi joint disease i.e., arthropathy of more than one joint and it significantly lowered their FISH score.Mean bleeding episodes is 16 episodes per year[9]. We expected that as the bleeding episodes increase FISH score to come down but in our study the result was not significant. 7 patients had associated HIV or HBSAG infection and these patients had significantly lower Functional status compared to the rest of the population.20 % of the population are using orthotic support and their FISH score was significantly low compared to others [10]. Only 2 of patients were on prophylaxis treatment compared to the western population where majority of severe haemophiliacs take prophylaxisthese western studies have shown that prophylactic treatment prevents hemophilicarthropathy.48% of patients were on regular physiotherapy but that did no significantly improve their FISH score, this result was similar [11]which may be because the patients were doing exercise to reduce the effect of impairment rather than for strengthening their joints. More studies are required to probe into what type of exercise the patients are actually doing[12]. Our patients travelled upto maximum 300km for treatment and mean was 33km and most used their own vehicle for coming to hospital. Their travel was not significantly affecting the FISH score.. Opening of more treatment centres and starting home based self-administration of factors would help patients to avoid long travel for treatment[13]. More than 80% of the haemophilia patients are taking free treatment from the state, which is a good indicator how health care has improved in the state of Tamil Nadu. [14,15]

REFERENCES

1. Poonnoose PM, Thomas R, Bhattacharjee S, Shyamkumar NK, Manigandan C, Srivastava A; Functional Independence Score in Haemophilia (FISH): A new performance based instrument to measure disability. Haemophilia, 2005; 11:598-602.
2. Tlacuilo-Parra A, Villela-Rodriguez J, Garibaldi-Covarrubias R, Soto-Padilla J, Orozco-Alcala J. Functional independence score in hemophilia: A cross-sectional study assessment of Mexican children. Pediatric blood & cancer. 2010;54(3):394-7..
3. Kar A, Mirkazemi R, Singh P, Potnis-Lele M, Lohade S, Lalwani A, Saha AS;Disability in Indian patients with haemophilia. Haemophilia. 2007;13(4):398-404.
4. Buzzard BM; Physiotherapy for prevention and treatmentof chronic hemophilicsynovitis. Clin Orthop Relat Res, 1997; 343: 42–6.
5. Soucie JM, Cianfrini C, Janco RL, Kulkarni R, Hambleton J, Evatt B, Forsyth A, Geraghty S, Hoots K, Abshire T, Curtis R; Joint range-of-

motion limitations among young males with hemophilia: prevalence and risk factors. Blood. 2004;103(7):2467-73.
6. Dharmarajan S, Phadnis S, Gund P, Kar A; Quality of life in rare genetic conditions: a systematic review of the literature. Am J Med Genet A, 2010;152A : 1136-56.
7. Gilbert MS; Musculoskeletal complications of haemophilia: The joint. Haemophilia, 2000; 6:34.
8. Steven MM, Yogarajah S, Madhok SD, Forbes CD, Sturrock RD ; Hemophilic arthritis. Q J Med, 1986; 58:181
9. Bladen M, Main E, Hubert N, Koutoumanou E, Liesner R, Khair K; Factors affecting the Haemophilia Joint Health Score in children with severe haemophilia. Haemophilia. 2013;19(4):626-31.
10. Schoenmakers MA, Gulmans VA, Helders PJ, Van Den Berg HM; Motor performance and disability in Dutch children with haemophilia: a comparison with their healthy peers. Haemophilia. 2001;7(3):293-8.
11. Su Y, WONG WY, Lail A, Donfield SM, Konzal S, Gomperts E; Long-term major joint outcomes in young adults with haemophilia: interim data from the HGDS. Haemophilia. 2007;13(4):387-90.
12. Soucie JM, Cianfrini C, Janco RL, Kulkarni R, Hambleton J, Evatt B, Forsyth A, Geraghty S, Hoots K, Abshire T, Curtis R; Joint range-of-motion limitations among young males with hemophilia: prevalence and risk factors. Blood. 2004;103(7):2467-73.
13. Genderen FR, Fischer K, Heijnen L, Kleijn P, Berg HM, Helders PJ, Meeteren NL; Pain and functional limitations in patients with severe haemophilia. Haemophilia. 2006;12(2):147-53.
14. Monahan PE, Baker JR, Riske B, Soucie JM; Physical functioning in boys with hemophilia in the US. American journal of preventive medicine. 2011;41(6):S360-8.
15. Kar A, Phadnis S, Dharmarajan S, Nakade J; Epidemiology & social costs of haemophilia in India. The Indian journal of medical research. 2014; 140(1):19.