

Functional Independence Score in Haemophilia: A Hospital Based Observational Study in a Tertiary Care Center in Central India

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ABSTRACT

BACKGROUND

Functional Independence Score in Haemophilia (FISH) is a performance-based assessment tool, to objectively measure the patient's functional disability. The present study was done to assess the functional abilities of haemophiliac patients presenting to a tertiary care centre.

METHODS

This hospital based observational study was performed on all patients (100) diagnosed as haemophilia referred to Hamidia Hospital from 1st March 2017 to 30th June 2018. Patients with other bleeding disorders and patients on medication which can alter coagulation profile were excluded.

RESULTS

Mean FISH for mild, moderate and severe haemophilia was 25.26 ± 2.62 , 23.65 ± 3.67 and 21.81 ± 2.65 and overall mean was 23.51 ± 3.23 .

CONCLUSIONS

The functional components like transfer and locomotion were mainly determined by joint bleeds and total joint score respectively which were observed by the positive correlation between the respective components. Hence more the bleed and joint score, higher the dependence for transfer and locomotion.

KEYWORDS

Haemophilia, Coagulation Profile, Haemarthrosis, FISH

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BACKGROUND

The X-linked recessive disorders associated with deficiency of coagulation factors, Haemophilia A (factor VIII deficiency) and Haemophilia B (factor IX deficiency) are the commonest forms of Haemophilia. The overall incidence of Haemophilia A is 1:5,000 male birth and that of Haemophilia B is 1:25,000 male birth.¹ In this disease, the process of Haemostasis is hampered and predisposing haemophiliacs to either spontaneous or post traumatic bleeding.² Coagulation factor deficient individuals have severe, moderate and mild forms of disease, defined by plasma factor levels of <1%, 2-5%, and 6-40% respectively. Haemarthrosis (Intra-articular bleed) is most common, most painful as well as most physically, economically and psychologically debilitating manifestation of the haemophilia.³ Functional Independence score in Haemophilia (FISH) is a performance-based assessment scale, to objectively measure the patient's functional disability consists of 8 basic routine activities. FISH is reliable, inexpensive and easily applied scale to evaluate joints status. The present study was done to assess functional abilities of haemophiliac patients presenting to a tertiary care centre.⁴

METHODS

This hospital based observational study was performed on all patients (100) diagnosed as haemophilia referred to Hamidia Hospital from 1st march 2017 to 30th June 2018. Patients with other bleeding disorders and on medication which can alter coagulation profile were excluded. After taking informed consent, a thorough clinical review was done consisting of history including family history, physical examination & relative laboratory tests. All patients were clinically evaluated by using functional independence score in haemophilia (FISH) scale. Function independence score - Developed by poonoose et al,⁵⁻⁷ this scoring system consists of 8 basic activities and aims to assess activities of daily living (ADL), under three categories (self-care, transfer and locomotion). (Figure 1) This score can also be applied to evaluate change in functional independence over a period, or after a therapeutic intervention. The recent version of FISH includes the assessment of eight activities of daily life which include- Eating and grooming, bathing, dressing chair transfer (getting up from a chair), squatting (this assesses the ability to squat on the floor and rise to an erect posture), 10-meter walking pattern, climbing stairs, running.

Scored on a 4-point Likert score according to the level of assistance required. 1. Unable to perform the activity 2. Need assistance or adaptation is 3. Activity is performed, but the individual has discomfort 4. Activity is performed normally.

Overall FISH score ranged from 8 to 32.

Score 8: unable to perform any activity

Score 32: perform all activities normally.⁸⁻¹⁰

Statistical average was done by mean value and dispersion measured by standard deviation.

FUNCTIONAL INDEPENDENCE SCORE IN HEMOPHILIA (FISH)				
Performance based instrument				
Patient Name:		Patient Code:		
Today (dd/mm/yyyy) : / /				
A. Self Care				
1. Eating and grooming	<input type="radio"/> 1	<input type="radio"/> 2	<input type="radio"/> 3	<input type="radio"/> 4
2. Bathing	<input type="radio"/> 1	<input type="radio"/> 2	<input type="radio"/> 3	<input type="radio"/> 4
3. Dressing	<input type="radio"/> 1	<input type="radio"/> 2	<input type="radio"/> 3	<input type="radio"/> 4
B. Transfers				
4. Chair	<input type="radio"/> 1	<input type="radio"/> 2	<input type="radio"/> 3	<input type="radio"/> 4
5. Squatting	<input type="radio"/> 1	<input type="radio"/> 2	<input type="radio"/> 3	<input type="radio"/> 4
C. Locomotion				
6. Walking	<input type="radio"/> 1	<input type="radio"/> 2	<input type="radio"/> 3	<input type="radio"/> 4
7. Stairs (12 - 14 steps)	<input type="radio"/> 1	<input type="radio"/> 2	<input type="radio"/> 3	<input type="radio"/> 4
8. Running	<input type="radio"/> 1	<input type="radio"/> 2	<input type="radio"/> 3	<input type="radio"/> 4
Total Score				

RESULTS

Mean age of patients was 19.02 ± 12.58 years with a range of age is 1-65 years. All cases were male and no female case was found in our study. Regarding age of onset of first clinical manifestation, majority of patients were below 1 year (41.35%-60.58% at 95% confidence interval). Mean age of onset was 2.44 ± 2.77 years. Haemophilia A was the most common type of haemophilia i.e. 89% (82.21%-93.91% at 95% confidence interval). There was no significant difference seen between BPL and APL population. In this study 57% patients were having positive family history supporting its mode of genetic inheritance. Most common clinical presentation was Haemarthrosis (62.48%-79.90% at 95% confidence interval) and knee joint was the involved in majority of the cases (56.78%-76.61% at 95% confidence interval). Musculoskeletal complications were commonest & associated with disabilities. Activated partial thromboplastin time (APTT) was raised in all patients.

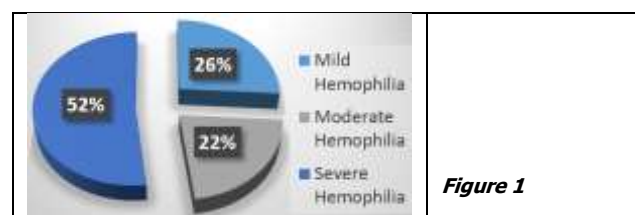


Figure 1

Majority of patients (52%) were severe haemophiliac (42.32%-61.54%) at 95% confidence interval. Mean Functional Independence Score in Haemophilia (FISH) for mild, moderate and severe haemophilia was 25.26 ± 2.62 , 23.65 ± 3.67 and 21.81 ± 2.65 respectively and overall mean FISH was 23.51 ± 3.23 . FISH score was higher in those with mild haemophilia then in patients who were having moderate or severe type of haemophilia.

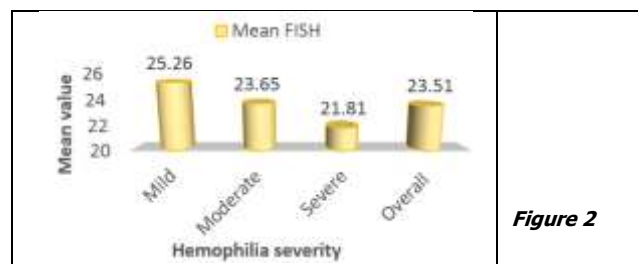


Figure 2

DISCUSSION

Mean FISH for mild, moderate and severe haemophilia in study done by Shamoon et al¹¹ was 29.5, 28.3 and 26 and overall mean was 28. Overall Mean FISH in study done by Thangamani¹² et al was 25.48. They did not mention FISH in mild, moderate and severe haemophiliac patients separately. The functional components like transfer and locomotion were mainly determined by joint bleeds and total joint score respectively which were observed by the positive correlation between the respective components. Hence more the bleed and joint score, higher the dependence for transfer and locomotion.¹³⁻¹⁵

Despite long term improvement in life expectancy of Haemophilia patients over the years, activities of daily life (ADL) measures are a subject of intense discussion and every attempt in research is a part to constantly improve issues related to it. As mentioned previously, since ADL is a multifaceted topic which varies with culture, beliefs, etc., we aimed to study this function comprehensively in small sample of patients with Haemophilia in Indian population. Based on the results from the baseline indicators, the sample population that we assessed were predominantly of Haemophilia A (Factor level <1, 89% with a range of 82.21% - 93.91% at 95% confidence interval), the most common type in general population. And at the time of study, most of the patients in the sample population studied were of severe type of Haemophilia, which was 52% (42.32%-61.54% at 95% confidence interval).¹⁶⁻¹⁹

Functional independence score is indicating that Psychometric values of emotions are clearly related to pain and inability to move among haemophiliac patients and issues related to pain dominate the Psychological health of a patient compared to locomotion. As observed, pain in major joints (Knee, Ankle and Elbow) seemed to dominate the mobility and feeling of self-satisfaction among the sample population. Compared to other factors which are commonly affecting Haemophilic patients, transfer ability dominated the Social relationships domain and it could be due to improper usage of appropriate walking aids for mobility and it is expected that a proper evaluation of walking aid is warranted.²⁰⁻²³

CONCLUSIONS

The assessment of joint health is one of the most important goals in haemophilia care. FISH assessment is a useful tool

to clinically assess functional defects in hemophiliacs due to feasibility and it may be able to detect haemophilic arthropathy as well. Advancement in medical services and positive public awareness may lead to better quality of life for hemophiliac patients. Study to access reliability & responsiveness of FISH in all variables also remains to be established.

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