

ABSTRACT

BACKGROUND

Haemophilia is a commonest hereditary bleeding disorder . It includes Haemophilia A ,Haemophilia B. It occurs due to deficiency of clotting factors Factor VIII and Factor IX respectively. People with Haemophilia bleed continuously as blood is unable to clot due to deficiency of clotting factors. They present with haemarthrosis, soft tissue hematomas or prolonged bleeding after trauma. Functional Independence Score in Haemophilia (FISH) is used to assess the degree of disability. Basic laboratory investigations like CBC and APTT can be used to assess the severity of the disease.

METHODOLOGY

This study was done as a prospective study at Haemophilia day care centre Tirunelveli Medical college hospital from March 2017 to September 2018(18 months). The study population included 35 cases. Detailed history regarding onset and progression of the disease, family history,age of diagnosis of the disease ,treatment history were asked. Patients were examined for physical disability, Disability assessment is done using Functional Independence Score in Haemophilia (FISH) in which every patient is examined for 7 activities under 3 categories for a total score of 28. Basic laboratory investigations like CBC and APTT are done using semi automated analyser. APTT values are correlated with the FISH scores and the severity of the disease is assessed.

RESULTS

In our study out of 35 cases 33 cases were severe haemophilia , 2 cases were mild haemophilia. FISH score was low in patients with severe haemophilia <28 compared to mild haemophilia cases whose FISH score was equal to 28. APTT values in these patients were compared with the FISH score, out of 35 cases, 34 cases had elevated activated partial thromboplastin time . Among them 28 patients had APTT between 38-58 sec with a mean FISH score of 22, 2 patients had APTT between 59-78 sec and their mean FISH score was 20 , another 2 patients had APTT between 79-98 sec with a mean FISH score of 19 , APTT value of >99 was noted in 2 patients and they had a mean FISH score of 16. 1 patient had APTT within normal range (i.e.,) below 38 sec and his FISH score was 25. It was found that for progressive increase in APTT values the mean FISH score was decreasing indicating that increase in APTT is associated with the increasing severity of the disease.

CONCLUSION

Though FISH score has been a vital tool in assessing the severity of the disease, haemoglobin and APTT could help in effective management of haemophilic patients on regular therapy.

KEY WORDS

Haemophilia, Disability, Haemoglobin, APTT, Functional Independence Score in Haemophilia (FISH) .