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# Comprehensive Assessment of Joint Disability in Children with Hemophilia & Plan of Action for Effective Intervention

Authors
**Dr Devakumar V K MBBS DCH DNB[1], Dr P G Hariprasad MBBS DCH DNB[2]**
[1,2]Addl. Professor, Dept of Pediatrics, SAT Hospital, Govt. Medical College, Trivandrum.

**ABSTRACT**

**Background and objectives:** *Permanent disability follows chronic haemophilic arthropathy. So it is important to periodically assess joint function in haemophilic children and do early intervention. There are very few studies in Kerala regarding haemophilic children and their problems especially joint function. SAT hospital is a tertiary care centre in Trivandrum catering to patients from south Kerala. Haemophilia clinic is conducted in SAT every Saturday. Objectives of the present study are to find the extent of joint disability in haemophilia patients using Haemophilia joint health score, to find the outcome after standard care in the Hemophilia clinic.*

**Methods:** *Observational Hospital level prospective follow up study at SAT Hospital, Thiruvananthapuram. Joint health was assessed using Haemophilia Joint Health Score (HJHS 2.1). After intervention HJHS 2.1 repeated at 3 monthly interval twice.*

**Results & Discussion:** *Study was conducted among 60 haemophiliac children attending SAT haemophilia clinic. Majority were haemophilia A, 86.7%. Haemophilia B constituted only 13.3 % .76.7% were having severe disease. 20.0 % moderate disease, 3.3 % having mild disease. In the study group, 27 children (45%) had haemophilic arthropathy. In 48.1%, haemophilic arthropathy developed at more than 9 years of age. There was statistically significant improvement in HJHS Score in children receiving treatment from haemophilia clinic SAT, Hospital, Thiruvananthapuram*

**Conclusion:** *In the study group, 27 children (45%) had haemophilic arthropathy. There is significant improvement in HJHS2.1 score in haemophilia patients receiving treatment and care in hemophilia clinic.*

**Keywords:** *Haemophilia; HJHS2.1; haemophilic arthropathy,*

## Introduction

Hemophilia is an X linked congenital bleeding disorder caused by a deficiency of coagulation factor VIII (Hemophilia A) or coagulation factor IX (Hemophilia). They are divided in to three groups depending on the factor level. Mild hemophilia means factor level of 5-40%, moderate means 1-5%, severe means <1 %. .Haemophilia A occurs in 1 in 5,000 and haemophilia B occurs in 1 in 30,000 live male births[(7)]. Secondary data on haemophilia cases in the state of Maharashtra, India, when compiled and analyzed, it was found that ratio of haemophilia A to haemophilia B was 4.2:1[(8)]. The most common and troublesome problem for children with hemophilia are the joint bleeds. Initial bleeds recover completely. But when get repeated, as in severe hemophilics, recovery becomes more & more difficult and

ultimately chonic synovitis sets in, which eventually leads to chronic hemophilic arthropathy. But if you can intervene sufficiently early joint deterioration can be prevented to some extent. Moreover we expect Gene therapy for hemophilia to be successful in the coming years & if you can keep their joint healthy these young children can have a normal life in the future. Documenting & monitoring joint health in the person with haemophilia provides important information for assessing the efficacy of current treatment regimens. The international Haemophilia Joint Health Score (HJHS) has been developed to track an individual's joint health over time. It has been designed to assess the knee, elbow and ankle joints in which a person can manifest joint bleeding or Hemarthropathy. Each joint receives a numeric score, which can be compared to itself over time to determine whether a joint is showing degeneration. The validation of the Hemophilia Joint Health Score (HJHS) 1.0 led to the development of the HJHS 2.0and 2.1. This resulted in the deletion of redundant items and the improvement of current items. Reliability of HJHS was studied by Hilliard P et all and found to be very good.[5] SAT Hospital being a tertiary care centre caters to most of the bleeding disorders from south Kerala, and we are running a clinic exclusively for hemophilic children here.

**Methods:** Observational prospective follow up study

**Study Setting:** Hemophilia clinic, SAT hospital, Medical College, Trivandrum

**Sample Size:** Majority of the children with bleeding disorders in south Kerala attend SAT hospital the predominant tertiary care centre here. We included all the 60 haemophilic children among them in the study.

**Tools:** Data collected using interview utilizing semi structured questionnaire and clinical Assessment using HJHS 2.1.

**Data Collection:** Every child registered in hemophilia clinic was enrolled in the study, after getting informed consent. Using a predesigned schedule history was taken. Our physiotherapist who is specially trained in managing hemophilc joints will carry out physiotherapy. After physiotherapy & other interventions HJHS 2.1 repeated in the affected children in 3 monthly intervals twice

**Study Variables**

- Number of patients
- Type of hemophilia with severity
- Age of onset of joint problem
- initial joint assessment using HJHS 2.1
- Repeat HJHS 2.1 at 3 months & 6 months of intense physiotherapy and other interventions

Results Total no of children included were 60.As haemophilia is an x-linked disorder all are males. There were 7 children below 3 years,2 children between 3 & 6 years, 18 children between 6&9 years and 28 children above 9 years. Out of 60, 52 children were diagnosed to have Hemophilia A (86.7%) and 8 were Hemophilia B (13.3%). Among 60 children with Hemophilia 76,7% were of severe variety,20% were moderate & 3.3% mild.
In the study group 27 children (45%) had hemophilicarthropathy. In 48.1%, hemophilicarthropathy developed at more than 9 years of age. Among the 27 patients with hemophilicarthropathy, 23(85.1%) belonged to hemophilia A, and 4 belonged to hemophilia B

**Table 1:** Distribution according to age of onset of hemophilicarthropathy

| Age of onset of hemophilicarthropathy | Frequency | Percent |
|---|---|---|
| 3 – 5 | 4 | 14.8 |
| 6 – 8 | 10 | 37.0 |
| >9 | 13 | 48.1 |
| Total | 27 | 100.0 |

In 48.1%, hemophilicarthropathy developed at more than 9 years of age.Out of 14 children who developed arthropathy before 9 years 12 belong to severe haemophilia.

**Table 2:** Joint health assessment based on HJHS score

| HJHS score | Baseline | After 3 months | After 6 months |
|---|---|---|---|
| N | 27 | 27 | 27 |
| Mean | 17.6 | 11.2 | 8.5 |
| Sd | 11.9 | 10.5 | 9.9 |
| Minimum | 6 | 0 | 0 |
| Maximum | 49 | 31 | 31 |
| First quartile | 10.0 | 3.0 | 0.0s |
| Median | 12.0 | 9.0 | 4.0 |
| Third quartile | 24.0 | 22.0 | 17.0 |

Repeated measure Non parametric test
Friedman Test p <0.001

**Table 3:** HJHS Score

| HJHS score | N | Mean ± Sd | Median (Inter quartile range) |
|---|---|---|---|
| Baseline | 27 | 17.6±11.9 | 12 (10-24) |
| After 3 months | 27 | 11.2±10.5 | 9(3-22) |
| After 6 months | 27 | 8.5±9.9 | 4 (0-17) |

**Table 4:** Improvement in HJHS score

| | Wilcoxonsigned rank test | |
|---|---|---|
| | Z | P |
| Baseline and after 3 months score | -4.301 | <.001 |
| Baseline and 6 months score | -4.463 | <.001 |
| after 3 months and after 6 months score | -2.824 | 0.005 |

The data was not following the normal distribution. Non parametric test, was used. Improvement in baseline HJHS after three months is statistically significant. Base line to 6 months also, there is a significant improvement in score. Comparing score at 3months and 6 months also, the improvement is significant.

**Table 5:** Percentage distribution according to improvement in three months

| Improvement in 6 months | Frequency | Percent |
|---|---|---|
| No improvement | 1 | 3.7 |
| <50 % reduction | 8 | 29.6 |
| >50% reduction | 18 | 66.7 |
| Total | 27 | 100.0 |

After three months there was above 50 % reduction in HJHS score in 13/27 children. 11 had <50% reduction. No improvement in three children.

At 6 months, 18 children with hemophilicar-thropathy had >50% reduction in HJHS scores,< 50% reduction in 8, no improvement in one child.

**Discussion**

In our study it was found that majoriy of the 60 children belong to haemophilia A (86.7%) and the rest to haemophilia B. This is similar to a study done by kulkarny et all-[4]. Among 52 children with Hemophilia A 43 (82%) were of severe variety, 8 were moderate (15.4%) & 1 (3%) mild. Among 8 children with Hemophilia B, 3 (37.5%) belong to severe variety,4(50%) were moderate& 1 (12.5%) mild. In the US cohort also 65% were severe in type [(6)]

In the study group, 27 children (45%) had hemophilicarthropathy.

In 48.1%, hemophilicarthropathy developed at more than 9 years of age.

In Charles et al's clinical study of 40 hemophilia patients above 12 years of age, 36 out of 40 had joint disability. S Cross et al states haemophilic arthropathy can develop anytime from the second decade of life, and sometimes earlier, depending on the severity of bleeding and its treatment-[(1)]

Almost 80% of recurrent hemarthroses in persons with haemophilia occur in the six main joints of the elbows, knees and ankles[(2)] In his study Raoul et al, used Haemophilia Joint Health Score. The health status of the joints was assessed by using the Haemophilia Joint Health Score (HJHS), developed by the International Prophylaxis Study Group (Physical Therapy Working Group).

In the elbows, knees, and ankles, numeric scores for swelling, duration of swelling, muscle atrophy, axial alignment, crepitus on motion, flexion and extension loss, instability, joint pain, strength, and gait were scored. After adding all scores, raw scores and the percentage of joint impairment were scored (a score of 0%representing no joint impairment)[(3)]. Today the HJHS is increasingly used in paediatric and in adult studies. In the study by Sluiter et al[(2)] Hemophilia Joint Health Scoring was done in healthy adults playing sports. Evaluation of HJHS score in healthy adults

playing sports could improve the interpretation of this score in hemophilic patients.

Out of 27, hemophilicarthropathy there were a significant improvement in HJHS scores on subsequent follow up. After 3 months, there was >50% reduction in HJHS score in 13 out of 27 children with hemophilicarthropathy.11 had <50% reduction. No improvement in 3 children. At 6 months, 18 children with hemophilicarthropathy had >50% reduction in HJHS scores and <50% reduction in 8.No improvement was found in one child who was a 12 year old boy having severe hemophilia A. Probable causes for no improvement were found to be poor compliance to treatment and frequent hemarthroses. US Centers for Disease Control and Prevention (CDC) conducted a pilot 6-state surveillance study of hemophilia outcomes from 1993 to 1995, which concluded that there was reduced mortality rates (P =0.002) in persons receiving care from the federally funded network of specialized clinics which were dedicated to multidisciplinary comprehensive care of haemophilia[9]

## Conclusion

- 45% of haemophiliacs had chronic joint involvement
- Common age of onset of hemophilicarthropathy is above 9 years.
- Severe hemophiliacs can develop arthropathy in younger ages also.
- Hemophilicarthropathy is more common in hemophilia A.
- There is significant improvement in HJHS2.1 score in hemophilia patients receiving treatment and care in hemophilia clinic with a muitidisciplinary team including a physiotherapist trained in haemophilia care.

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## References

1. Cross S, Vaidya S, Fotiadis N. Hemophilicar-thropathy: a review of imaging and staging. Semin Ultrasound CT MR. 2013 Dec;34(6):516–24
2. Sluiter D, Foppen W, de Kleijn P, Fischer K. Haemophilia Joint Health Score in healthy adults playing sports. Haemophilia. 2013;20(2):282-286
3. Engelbert R Platinga M. Aerobic capacity in children with hemophilia. TheJournal of Pediatrics. 2008;152(6):833-838
4. Kulkarni R, Soucie JM, Lusher J, et al. Sites of initial bleeding episodes, mode ofdelivery and age of diagnosis in babies with haemophilia diagnosed before theage of 2 years: a report from The Centers for Disease Control and Prevention's(CDC) Universal Dsata Collection (UDC) project. Haemophilia 2009;15:1281–90.
5. Hilliard P, Funk S, Zourikian N, Berg-strom BM, Bradley CS, McLimont M, et al.Hemophilia joint health score reliability study. Haemophilia 2006;12:518-25.
6. Manco-Johnson MJ. Advances in the care and treatment of children withhemophilia. AdvPediatr. 2010;57(1):287–94.
7. Fomin ME, Togarrati PP, Muench MO. Progress and challenges in thedevelopment of a cell-based therapy for hemophilia A. J Thromb Haemost JTH.2014 Dec;12(12): 1954–65.
8. Kar A, Potnis-Lele M. Descriptive epidemiology of haemophilia in Maharashtra, India. Haemoph Off J World Fed Hemoph. 2001 Nov;7(6):561–7.
9. Manco-Johnson MJ. Advances in the care and treatment of children with hemophilia. Adv Pediatr. 2010;57(1):287–94.