

## Original Research Article

## Effectiveness of Physiotherapy and Physical Rehabilitation in the Treatment of Chronic Hemophilic Arthropathy: A Prospective Study From a Comprehensive Hemophilia Care Center in Eastern India

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### Abstract

Hemophilia is a blood-clotting disorder caused by a deficiency in factor VIII (FVIII) or factor IX (FIX), which manifests itself through bleeding in the muscles and joints. Haemophilic arthropathy causes pain and deterioration in movement, thereby causing disability and permanent joint damage. Aim of the study was to determine the effect of physiotherapy on range of motion on target joints in people with hemophilia. This was a prospective study done on 50 patients of all age groups with hemophilia in the study period from March 2022 to February 2023

**Keywords:** Hemophilia, Arthropathy, Physiotherapy, Range of motion

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### Introduction

Hemophilia is a rare X-linked recessive coagulation disorder in which deficiencies of certain clotting factors are the cause of haemorrhage with minor trauma or surgery. Deficiency of Factor VIII or Factor IX, respectively, called hemophilia A or B, has a prevalence of one in 5,000 and one in 30,000 male births, respectively [1]. Depending on the amount of clotting factor, they are divided into three categories: severe (<1%), moderate (1–5%), and mild hemophilia (5–40%) [2]. About 80% of the episodes in person with haemophilia (PwH) involve bleeding in the joints (hemarthrosis), of which the most common is the knee joint, followed by the elbow and ankle [3]. The World Federation of Hemophilia (WFH) recommends regular factor replacement therapy to reduce bleed frequency in combination with physiotherapy aimed at preserving muscle strength and functional ability in order to prevent and treat chronic hemophilic arthropathy [4]. The ability to move is an essential element of health and well-being and is central to what it means to be healthy. Physical rehabilitation is directed towards the movement needs and potential of individuals, providing rehabilitation and services to develop, maintain, and restore maximum movement and functional ability and prevent musculoskeletal impairment throughout life [5].

### Materials and Methods

This was a prospective analysis of 50 patients with congenital haemophilia attending outpatient and inpatient departments over a period of one year, from March 2022 to February 2023, at Nil Ratan Sircar Medical College and Hospital, Kolkata, India.

All PwH with chronic hemophilic arthropathy who required physiotherapy were included in the study except those patients with

acute bleeds (<72 hours), symptoms and signs of infection in the joint, pseudotumors, and prior inhibitor positivity. The baseline range of motion (ROM) of the target joint was noted using a goniometer [6]. Physical therapy interventions were applied three days a week, for half an hour daily. Muscle strengthening, proprioception, flexibility, isotonic, and isometric exercises were initiated along with hot packs [7][8]. Exercises were taught, and home exercise courses were provided to all the patients with proper written instructions for recall. Both pre- and post-treatment joint ROM degrees were measured. Change in ROM was noted after one month, six months, and 12 months.

The data were entered into a Microsoft Excel sheet. Statistical analyses were done by Wilcoxon signed ranks test, t' test, and Kruskal-Wallis analysis for parametric and non-parametric data, respectively, which included frequency and percent distributions, the calculation of means, and standard deviations. Analyses were conducted using SPSS version 21, and the alpha value was set at 0.05 for statistical significance. Written consent was obtained from all the participants and/or their legal guardians, and all procedures were in accordance with the Helsinki Declarations of 1975.

### Results

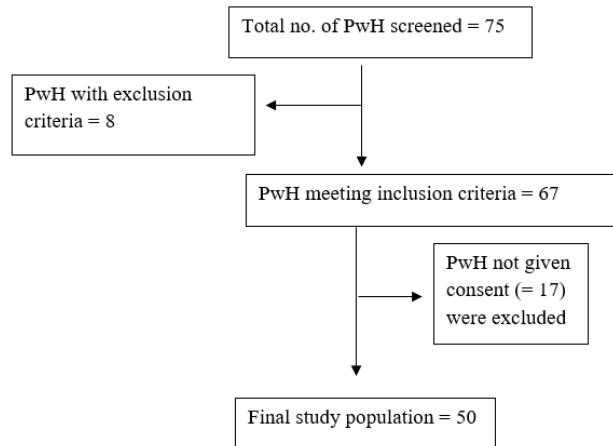
Seventy five PwH were screened out of which eight were excluded and 17 did not give consent for the study. Hence, total 50 patients could be included in the study based on the criteria.

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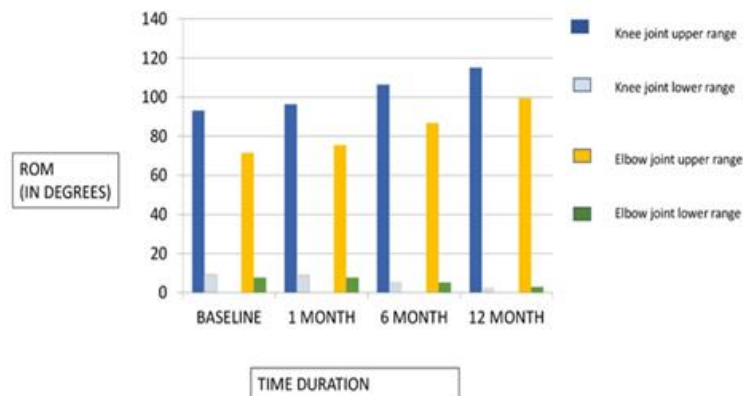
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**Figure 1: Console diagram showing number of patients screened, excluded, included and final study population**

Out of the total 50 PwH with hemophilia A included in the study, 38 (76%) had severe hemophilia, 2 (4%) had moderate and 10 (20%) had mild hemophilia. The mean age was 22.4 years (range 0–46 years). A total of 82 joints were studied, out of which 69 (84%) were knee joints and 13 (16%) were elbow joints. The mean baseline ROM in the knee joint was 9.27–94.170 (range 0-1200). After initiation of physiotherapy, the mean range of motion at 1 month, 6 months, and 12 months was 8.95- 96.340 (range 0 - 1220),

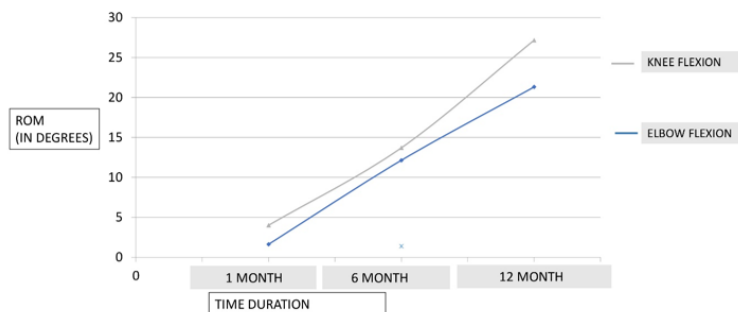
5.60- 106.450 (range 0- 1270) and 2.69 – 115.070 respectively (range 0- 1300). The mean baseline ROM in the elbow joint was 7.69- 71.540 (range 0-900). After initiation of physiotherapy, the mean range of motion at 1 month, 6 months, and 12 months was 7.69-75.540 (range 0 - 920), 5.23 - 86.770 (range 0- 1000) and 2.92- 99.460 (range 0-1050) respectively.



**Figure 2: Mean baseline ROM Knee joint and Ankle joint**

Mean improvements in ROM in flexion and extension on the knee joint at 1 month, 6 months, and 12 months were 1.620 (range 0-100), 12.130 (range 0-300), 21.130 (range 10-400), and 0.310 (range 0-50), 1.780 (range 0-200), 5.630 (range 0-25) respectively. (p value <0.001)

Improvements in ROM in flexion and extension on the elbow joint at 1 month, 6 months, and 12 months were 40 (range 0-100), 13.690 (range 0-200), 27.150 (range 5-250), and 0.20 (range 0-100), 1.380 (range 0-100), and 3.610 (range 0-150), respectively. (p value <0.001)



**Figure 3A: Mean improvement in ROM in flexion at Knee and Elbow joint (1 month, 6 month and 12 month)**

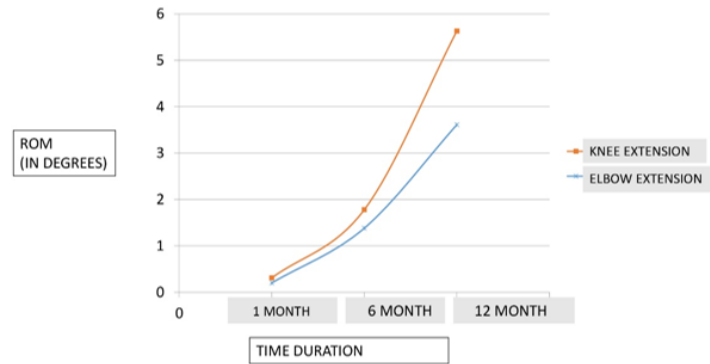


Figure 3B: Mean improvement in ROM in extension at Knee joint and Ankle joint (1 month, 6 month and 12 month)

**Discussion**

Hemophilia management needs a holistic approach to ensure better patient outcome. Educating patients and family members about the nature of the disease, replacement of the missing clotting factor and comprehensive care is of utmost importance. Evidence based therapeutic exercise and proper nutrition can help in controlling bleeding and maintain general health. Focus should be primary prevention of bleeding as well as treatment of acute bleeding episodes. Primary aims are- prevention of bleeding and treatment of acute bleeding. Multidisciplinary team approach can be instituted and is the key to efficient result. Home therapy, attention for psychosocial health and rehabilitation programs is an aid to restore joint and ensure

mobility [9].

Aim of physiotherapy is to facilitate resolution of the bleed, provide mobilization as well as to improve bone mineralization [10][11]. In our series, we showed that rehabilitation improves clinical evaluation scores, functional recovery and prevents disability. Rehabilitation in the form of physiotherapy should be initiated early in the course of disease as it increases joint ROM and muscle strength along with beneficial effects on proprioception and coordination [12]. It could also decrease the frequency of replacement therapy [13].

As shown in Table 1, the present study has shown comparable results to several other similar studies published in literature from different countries of the globe.

**Table 1: Summary of important findings of the present study and other similar published studies**

Parameters	The present study	Other similar studies
Number of patients	50	Singh et al [14]: 18, Timmer et al [15]: 10, Gurcay et al [16]: 25
Mean age of patients (on enrolment to study)	22.4 years	Singh et al [14]: 25 years, Timmer et al [15]: 57 years
Severity of hemophilia	- <ul style="list-style-type: none"> <li>Severe-75%,</li> <li>Moderate-4%</li> <li>Mild -20%,</li> </ul>	Singh et al [14] <ul style="list-style-type: none"> <li>Severe: 72.2%,</li> <li>Moderate: 5.6%</li> <li>Mild: 22.2%,</li> </ul>
Joints involved (studied)	- <ul style="list-style-type: none"> <li>Knee joint (84%)</li> <li>Elbow joint (16%)</li> </ul>	Gurcay et al [16] – <ul style="list-style-type: none"> <li>Knee joint (60%),</li> <li>Elbow joint (24.5%),</li> <li>Ankle joint (15.4%)</li> </ul>
Improvement in flexion of Knee joint	21.13 <sup>0</sup>	Barriuso et al [17]: 17 <sup>0</sup> , Gurcay et al [16]: 20 <sup>0</sup>
Improvement in extension of Knee joint	5.63 <sup>0</sup>	Barriuso et al [17]: 1 <sup>0</sup>
Improvement in flexion of Elbow joint	27.15 <sup>0</sup>	Gurcay et al [16]: 20 <sup>0</sup>
Improvement in extension of Elbow joint	3.61 <sup>0</sup>	Gurcay et al [16]: 0 <sup>0</sup>

Singh et al [14] evaluated the responsiveness to Hemophilia Joint Health Score (HJHS) and Functional Independence Score in Hemophilia (FISH) in 18 PwH with intermittent factor support and physiotherapy. Similarly, Timmer et al [15] evaluated the concept and feasibility of blended physiotherapy in 10 PwH. In the prospective series of musculoskeletal system rehabilitation of arthropathic joints by Gurcay et al [16], 25 young male PwH were studied. In the present study, we could include 50 PwH and follow-up over a period of 12 months for each of them.

Mean age in the present study was 22.4 years which was comparable to the study done by Singh et al [14] in which they included the PwH with a mean age of 25 years for joint evaluation. But, the study by Timmer et al [19] included the PwH with a mean age of 57 years. Majority of the PwH in the present study were severe (75%) followed by mild (20%) and moderate (4%). The study by Singh et al [14] got a

similar group in their study (table 1). Knee joint was the most affected joint (84%) followed by elbow joint (16%) as revealed in the present study. In the study done by Gurcay et al [16] knee joint was involved in 60% cases followed by elbow (24.5%) and ankle (15.4%).

At the end of 12 months, mean improvement in flexion and extension in the knee joint was 21.63<sup>0</sup> and 5.63<sup>0</sup> respectively. It was comparable to a case report done by Barriuso et al [17] in which the flexion and extension improvement in knee joint was 17<sup>0</sup> and 1<sup>0</sup> respectively. Mean improvement in flexion and extension in elbow joint was 27.15<sup>0</sup> and 3.61<sup>0</sup> respectively. Similar findings was reported in the study done by Gurcay et al [16] in which the mean improvement in ROM in flexion and extension on elbow joint was 20<sup>0</sup> and 0<sup>0</sup> respectively.

The present study showed significant improvement in ROM on both knee joints from 9.27<sup>0</sup> - 93.17<sup>0</sup> at baseline to 2.69<sup>0</sup> - 115.07<sup>0</sup> at 12

months and on elbow joint from 7.69° - 71.54° at baseline to 2.92° - 99.46° at 12 months after initiation of physiotherapy (p value <0.001). In our study, appropriate individualized physiotherapy treatment protocols were initiated and follow-up for a period of 12 months was done. There was dramatic improvement in overall ROM as well as flexion and extension improvement on both Knee joint and Elbow joint. Rehabilitation in the form of physiotherapy by increasing ROM improves muscle strength, proprioception, coordination, day to day activities and also reduced pain which is a major cause of morbidity among PwH [18]. It could also decrease the frequency of replacement therapy. It must be initiated before beginning of contracture development [19][20]. Rehabilitation programmes must be comfortable and suitable for PwH adapting exercise programs early in the course of disease [21][22].

The results of this study were very encouraging and important to show that physiotherapy and rehabilitation improves the range of motion of an affected joint including the target joint and thereby uplifting clinical status in people with hemophilia with chronic arthropathy. It also highlights the importance of multidisciplinary approaches including rehabilitation programs in the management of chronic hemophilic arthropathy. These programs protect the target joints from developing contractures, limit the impairment and improve quality of life in people with hemophilia.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

#### Conflicts of interest

There are no conflicts of interest.

#### References

- Iorio A, Stonebraker JS, Chambost H, Makris M, Coffin D, Herr C et al. Data and Demographics Committee of the World Federation of Hemophilia. Establishing the prevalence and prevalence at birth of hemophilia in males: a meta-analytic approach using national registries. *Annals of internal medicine*. 2019;171(8):540-6.
- White GC, Rosendaal F, Aledort LM, Lusher JM, Rothschild C, Ingerslev J. Definitions in hemophilia. *Thrombosis and haemostasis*. 2001;85(03).
- Atilla B, Güney-Deniz H. Musculoskeletal treatment in haemophilia. *EFORT Open Rev*. 2019;4:230-239.
- Srivastava A, Santagostino E, Dougall A, Kitchen S, Sutherland M, Pipe SW, Carcao M et al. WFH Guidelines for the Management of Hemophilia, 3rd edition. *Haemophilia*. 2020;26:1-158.
- Wells AJ, Stephensen D. The role of the physiotherapist in the management of people with haemophilia: defining the new normal. *Br J Hosp Med (Lond)*. 2020;81:1-8
- Jones P. Comprehensive care in hemophilia. In: Hilgartner MW, Pochedly C (eds) *Hemophilia in child and adult*. 3rd edn., Raven, New York. 1989;12:141-142.
- Gilbert M. Prophylaxis. musculoskeletal evaluation. *Semin Hematol*. 1993;30:3-6
- Högh J, Ludlam CA, Macnicol MF. Hemophilic arthropathy of the upper limb. *Clin Orthop*. 1987;218:225-23
- Cuesta-Barriuso R, Gómez-Conesa A, López-Pina JA. Physiotherapy treatment in patients with hemophilia and chronic ankle arthropathy: a systematic review. *Rehabil Res Pract*. 2013;2013:305249.
- Dalyan M, Tuncer S, Kemahlı S. Hemophilic arthropathy: evaluation of clinical and radiological characteristics and disability. *Turk J Pediatr*. 2000;42:205-20
- Heijnen L, De Kleijn P. Physiotherapy for the treatment of articular contractures in hemophilia. *Haemophilia*. 1999;5:16-19
- Santavirta N, Solovieva S, Helkama O, Lehto S, Kontinen YT, Santavirta S. Musculoskeletal pain and functional ability in hemophilia A and B. *Physiotherapy and rehabilitation in hemophilia patients*. *Rheumatol Int*. 2001;21:15-19
- Atkins RM, Henderson NJ, Duthie RB. Joint contractures in the hemophilias. *Clinical Orthopaedics and Related Research*. 1987;1(219):97-106.
- Singh R, Dinakaran M, Vandhiyadevan GD, Mathangi S, Pandey RA, John MJ. Responsiveness to hemophilia joint health score and functional independence score in patients with hemophilia with intermittent factor support and physiotherapy. *CHRISMED Journal of Health and Research*. 2021;8:187.
- Timmer MA, Kuijlaars IAR, Kloek C, de Kleijn P, Schutgens REG, Veenhof C et al. Proof of concept and feasibility of a blended physiotherapy intervention for persons with haemophilic arthropathy. *Haemophilia*. 2023;29:290-307.
- Gurcay E, Eksioğlu E, Ezer U, Cakir B, Cakci A. A prospective series of musculoskeletal system rehabilitation of arthropathic joints in young male hemophilic patients. *Rheumatol Int*. 2008;28(6):541-5.
- Cuesta-Barriuso R, Trelles-Martínez RO. Manual therapy in the treatment of patients with hemophilia B and inhibitor. *BMC Musculoskelet Disord*. 2018;19:26.
- Buzzard BM. Physiotherapy for the prevention of articular contraction in hemophilia. *Haemophilia*. 1999;5:10-15
- Heijnen L, De Kleijn P. Physiotherapy for the treatment of articular contractures in haemophilia. *Haemophilia*. 1999;5:16-9.
- Pelletier JR, Findley TW, Gemma SA. Isometric exercise for an individual with hemophilic arthropathy. *Phys Ther*. 1987;9:1359-1364
- Gualtierotti R, Solimeno LP, Peyvandi F. Hemophilic arthropathy: Current knowledge and future perspectives. *J Thromb Haemost*. 2021;19(9):2112-2121.
- Doly EA. Role of physiotherapy in hemophilia patient: a case study on new dimension of physiotherapy application. *Med Crave Moj Yoga Physical Ther*. 2017;2(3):79-80.

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