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ROLE OF EARLY REHABILITATION IN HEMOPHILIC KNEE ARTHROPATHY

Physical Medicine & Rehabilitation

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ABSTRACT

Hemophilia A and B are genetic bleeding disorders that frequently result in joint hemorrhages, leading to hemophilic arthropathy, pain, and impaired mobility. Without timely rehabilitation, these joint complications can cause irreversible damage and long-term disability. Currently, no study from Bihar has evaluated the role of early rehabilitation in managing hemophilic arthropathy, highlighting a significant research gap. This study was conducted to address that gap and assess the efficacy of early intervention in improving outcomes. The results demonstrate that a structured rehabilitation program—comprising pharmacotherapy, physical therapy, and targeted exercises—significantly enhances joint health and functional independence in pediatric patients with hemophilia. Statistically significant improvements were observed in both Hemophilia Joint Health Score (HJHS) and Functional Independence Score in Hemophilia (FISH) over a three-month period ($p < 0.001$). The majority of patients presented with moderate hemophilia, predominantly affecting the right knee. Limitations included a short study duration, a pediatric-heavy sample, and limited follow-up. Future research should focus on a more diverse age range, larger sample size, and longer follow-up with multi-joint analysis for broader applicability.

KEYWORDS

INTRODUCTION

Hemophilia A and B are X-linked recessive bleeding disorders resulting from deficiencies of clotting factors VIII and IX, respectively. Hemophilia C, although rarer and not X-linked, arises due to a deficiency of factor XI. These coagulation abnormalities impair the body's ability to form effective blood clots, making affected individuals susceptible to prolonged bleeding. The hemostatic process involves a delicate interplay between procoagulant, anticoagulant, and fibrinolytic pathways. The extrinsic, intrinsic, and common coagulation cascades converge to activate factor X, which then catalyzes fibrin formation to stabilize the clot.

Hemophilia is a rare condition, with hemophilia A affecting approximately 1 in 5,000 male births, and hemophilia B around 1 in 30,000. A common clinical hallmark is spontaneous or trauma-induced joint bleeding (hemarthrosis), primarily affecting the knees, elbows, and ankles, though hips and shoulders may also be involved. Repeated bleeding episodes can lead to hemophilic arthropathy (HA)—a chronic, disabling condition characterized by joint pain, reduced range of motion (ROM), and functional limitation.

Inflammatory mediators such as iron, cytokines (IL-1β, IL-6, TNF-α), and vascular endothelial growth factor (VEGF) play key roles in the pathogenesis of HA. These factors contribute to synovial inflammation, pannus formation, cartilage erosion, and subchondral bone damage. Macrophages and monocytes amplify this process by promoting chondrocyte apoptosis and inhibiting matrix synthesis. Elevated VEGF and increased synovial vascularity are particularly prominent in severe HA, indicating a role for angiogenesis in disease progression. Over time, joint instability, muscle wasting, and recurrent hemarthroses compound the functional deterioration.

Methodology

This study was conducted in the Department of Physical Medicine and Rehabilitation (PMR) at PMCH, Patna. Pediatric patients diagnosed with Hemophilia A or B were enrolled after obtaining informed consent from their guardians. Detailed demographic and clinical histories—including family background, personal health, trauma history, and prior treatment—were recorded.

Comprehensive assessments were performed, including general physical examination, systemic review, and focused local joint evaluation. A total of 80 patients were initially admitted for evaluation. Key outcome measures used included the Hemophilia Joint Health Score (HJHS) to assess joint integrity, the Functional Independence Score in Hemophilia (FISH) to evaluate physical functionality, and Peterson's Radiological Score to determine structural joint damage.

Rehabilitation was delivered through a multi-modal approach comprising pharmacotherapy, physiotherapy, skin traction, orthotic support, and cryotherapy. Patients were also educated on hemophilia management and joint protection strategies. Follow-up assessments were conducted at one and three months using the same HJHS and FISH tools.

The FISH, a performance-based scale, tracked improvements in daily functional independence. Peterson's score provided radiological insights into joint degradation. The HJHS was primarily used to monitor joint health in major joints—specifically the knees, ankles, and elbows—in children aged 4 to 18 years, helping to determine the necessity for orthopedic or physiotherapy interventions.

Hemophilia joint health score 2.1(HJHS)

	Score
Swelling	0
No	1
Mild	2
Moderate	3
Severe	
Duration (swelling)	0
No or >6 months	1
>6 months	
Muscle atrophy	0
None	1
Mild	2
Severe	
Crepitus on motion	0
None	1
Mild	2
Severe	
Flexion loss	0
<5°	1
5–10°	2
11–20°	3
>20°	
Extension loss	0
<5°	1
5–10°	2
11–20°	3
>20°	
Joint pain	0
No pain through active ROM	1
No pain through active ROM, only pain on gentle overpressure or palpation	2
Pain through active ROM	
Strength	0
Holds test position against gravity with maximum resistance	1
Holds test position against gravity with moderate resistance	2
Holds test position against gravity with minimal resistance or against gravity	3
Able to partially complete ROM against or to move through ROM gravity eliminated	4
Trace or not muscle contraction	
All these items should be evaluated for both elbows, knees, and ankles, with a maximum score per joint of 20 and overall of 120	
Global gait (walking, stairs, running, hopping in 1 leg)	0
All skills are within normal limits	1
One skill is not within normal limits	2
Two skills are not within normal limits	3
Three skills are not within normal limits	4
No skills are within normal limits	
The maximum score for the assessment of the 6 joints plus gait is 124. For all items, if it cannot be scored, there is the option of non-evaluable (NE)	

Peterson's score

Radiographic Finding	Score
Osteoporosis	
Absent	0
Present	1
Enlarged epiphysis	
Absent	0
Present	1
Irregular subchondral surface	
Absent	0
Partially involved	1
Totally involved	2
Narrowing of joint space	
Absent	0
>1 mm	1
<1 mm	2
Subchondral cyst formation	
Absent	0
1 cyst	1
>1 cyst	2
Erosion of joint margins	
Absent	0
Present	1
Gross incongruence of articulating bone ends	
Absent	0
Slight	1
Pronounced	2
Joint deformity (angulation, displacement, or both between articulating bones)	
Absent	0
Slight	1
Pronounced	2

FUNCTIONAL INDEPENDENCE SCORE IN HEMOPHILIA (FISH)

	List of activities tested
Self-care	Eating Grooming Bathing Dressing
Transfers	Chair transferring Squatting
Locomotion	Walking Climbing stairs Running

Scores range from 1 to 4 for each activity depending on the degree of independence:

1, unable to perform;

2, requires the help of an assistant/aid;

3, able to perform the activity without an aid but not like a healthy subject;

4,able to perform the activity like other healthy subjects

Rehabilitation Protocol

The rehabilitation strategy for hemophilia patients at the PMR department involved an integrated and structured approach tailored to individual severity levels. The protocol consisted of pharmacological therapy, physical interventions, and progressive exercise regimens aimed at enhancing joint stability and function.

Pharmacological treatment was initiated based on the severity of hemophilia, followed by clotting factor infusions to manage acute bleeding episodes. Ice compression and joint protection techniques were employed in the early phase to minimize inflammation and prevent further damage.

Skin traction was used selectively to support joint rest and promote range of motion (ROM); however, its application required close monitoring to prevent the risk of re-bleeding. ROM exercises progressed in phases—beginning with passive movements in the supine position, followed by seated exercises, prone knee flexion, and eventually weight-bearing activities.

Strengthening exercises were gradually introduced, including prolonged isometric knee extensions, partial squats, and stair climbing to restore muscular strength and endurance. Proprioception and balance training began with basic standing tasks on stable surfaces, followed by more advanced challenges such as single-leg balancing with eyes open and closed on both even and uneven surfaces. These exercises aimed to improve neuromuscular coordination and joint stability.

RESULTS

This study analyzed clinical and rehabilitation outcomes in 60 pediatric patients with hemophilia. Various parameters such as age, gender, type and severity of disease, family history, joint involvement, and treatment response were evaluated.

The majority (65%) of the participants were between 4–10 years old, with a mean age of 9.31 years. Male predominance was observed (98%), and Hemophilia A was present in 95% of patients. Most cases (75%) had moderate severity, and 77% reported a positive family history. The right knee was the most commonly affected joint (80%).

Pre-rehabilitation, the Hemophilia Joint Health Score (HJHS) was primarily in the 12–15 range (mean = 13.85). After three months, a significant improvement was noted, with 35% of patients achieving a score of 6–7 and a mean score of 8.71 ($p < 0.001$).

HJHS Score Distribution:

Score Range	Pre-Rehab (%)	1st Month (%)	3rd Month (%)
6–7	0%	3%	35%
8–9	2%	25%	25%
10–11	13%	28%	33%
12–13	33%	30%	7%
14–15	28%	14%	0%
16–19	24%	0%	0%

Functional Independence Score in Hemophilia (FISH) also improved significantly, with the mean score increasing from 28.68 to 31.61 ($p < 0.001$). By the third month, 37% of patients reached the top score range (33–35).

FISH Score Distribution:

Score Range	Pre-Rehab (%)	1st Month (%)	3rd Month (%)
24–26	23%	0%	0%
27–29	32%	38%	8%
30–32	45%	58%	55%
33–35	0%	4%	37%

Pre-rehabilitation range of motion (ROM) loss was typically 20–30°. Following rehabilitation, ROM loss was reduced to less than 5° in mild cases, 6–10° in moderate cases, and 11–15° in severe cases. Swelling, which was initially tense with obscured bony landmarks, improved noticeably. In mild and moderate cases, bony landmarks became visible, and in severe cases, swelling was moderately reduced.

Summary & Limitations

This study presents the first regional effort in Bihar to evaluate the impact of early rehabilitation on hemophilic arthropathy. The findings demonstrate that timely and structured rehabilitation significantly improves joint health, range of motion, and functional independence in pediatric patients with Hemophilia A and B.

The average pre-rehabilitation HJHS and FISH scores were 13.85 and 28.68 respectively, both of which showed statistically significant improvements after three months of intervention. The highest benefit was seen in patients with moderate disease severity, while severe cases showed relatively lesser improvement. ROM loss was markedly reduced, and swelling significantly subsided in most patients.

Most participants were male children (mean age 9.3 years), consistent with the genetic pattern of hemophilia. Hemophilia A was predominant (95%), and a positive family history was noted in 77% of cases. The right knee was the most commonly affected joint, possibly due to frequent trauma in that limb.

Limitations:

- Short study duration limited long-term outcome assessment.
- Predominantly pediatric sample restricted age-related generalizability.
- Limited follow-ups may have missed late-onset changes.
- Single-joint (knee) focus limited broader applicability to other joints.

Suggestions for Future Research

- Include patients across all age groups for broader analysis.
- Enroll a larger number of participants to enhance statistical power.
- Conduct long-term follow-ups to assess sustained outcomes.
- Evaluate multiple joints beyond the knee for comprehensive insights.
- Encourage interdisciplinary management involving hematology, orthopedics, and physiotherapy.

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