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Effect of Physiotherapy on Joint Health Status and Functional Independence of a 37-years Old Person with Hemophilia Suffering from Chronic Kidney Disease

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ABSTRACT

BACKGROUND- The common finding in hemophilia and the characteristic of a severe form is hemarthrosis, the recurrent intra-articular bleeding which further leads to synovitis and causes hemophilic arthropathy. The most common joints for bleeding are the knee, elbow, ankle, and wrist. Young adults with hemophilia are also more likely to develop age-related comorbidities, which adds to the clinical health issues already present.
CASE DESCRIPTION- A 37 years old, male subject with severe hemophilia A (factor assay<1%), high titre inhibitor and a medical diagnosis of bilateral hemophilic arthropathy in elbows, knees and ankles was included in the study. He was also diagnosed with stage-5 chronic kidney disease and had been on hemodialysis 2 times a week from the last two years prior to the study.
The joint health status using HJHS, functional independence using FISH and BI Scoring of Activities of Daily Living and pain using VAS was assessed before and after the treatment. The physiotherapy interventions were administered for 4 months, with 4 sessions a week. The Pre and post treatment scores of HJHS, FISH, BI and VAS were 49 vs 43, 9 vs 16, 5 vs 14 and 8 vs 4 respectively.
CONCLUSION- The findings of this particular case of a severe hemophiliac person who was inhibitor positive with hemophilic arthropathy of multiple joints and CKD showed that physiotherapy played a significant role in improving joint health status, functional independence and reducing pain.

Keywords: Chronic kidney disease, Hemarthrosis, Hemophilia, Hemophilic arthropathy, Physiotherapy.

INTRODUCTION

Hemophilia is described as a congenital disease which is caused by the deficiency of blood coagulation factor and characterized by recurrence of hemarthrosis, especially in joints [1]. Classical hemophilia is an X-linked bleeding disorder leading to a deficiency or complete absence of coagulation factor VIII (FVIII, hemophilia A) or factor IX (FIX, hemophilia B). The overall prevalence of classical hemophilia is reported as 1 in 5,000 male births across all ethnic groups, with a frequency of hemophilia A estimated at 11 cases per 100,000 men, four times more common than hemophilia B [2].

Clinical expression of hemophilia usually correlates with the coagulation factor activity in plasma and hemophilia is described as severe when the factor level <

0.01 IU/mL [2]. The classification of Hemophilia A (HA) provides a guidance to possible types of bleeding and the rate of occurrence of hemorrhagic episodes. The people with hemophilia (PWH) with severe form, experience spontaneous bleeding and hemorrhage after minor trauma about 1-6 times in a month, including hemarthrosis and intramuscular hemorrhage. Hemarthrosis is the recurrent intra-articular bleeding which further leads to synovitis and causes impaired cycle of circulation. The most common joints for bleeding are the knee, elbow, ankle, and wrist [3]. Joint and muscle bleeding in hemophiliacs causes arthropathy, joint damage, discomfort, reduced range of motion (ROM), and muscle weakening, which in turn affects functional mobility, balance, posture, and strength.

The main pharmaceutical treatment for hemophilia is clotting factor replacement therapy—the intravenous infusion of factor VIII or IX concentrate, to replace the missing or reduced coagulation factor [4]. While clotting factor are generally very safe and effective, a proportion of PWH will develop neutralizing anti-drug antibodies termed inhibitors, which, as their name implies, inhibit the ability of clotting factors to exert their corrective effect on the deficiencies of FVIII or FIX [5]. The development of neutralizing antibodies (inhibitors) to factor VIII (fVIII) or factor IX (fIX) is the most significant complication of hemophilia treatment, occurring in up to 33% of PWH with severe hemophilia A, in 13% of those with non-severe hemophilia A, and in 3% of PWH with severe hemophilia B. In the presence of an inhibitor, the risks of major morbidity and the cost of care increase substantially [6].

Clinical physicians, however, now face additional difficulties, mostly diseases linked to ageing. Adult hemophiliacs are susceptible to ageing comorbidities [7]. Chronic kidney disease (CKD) is a long-term condition, which is associated, in many PWH, with physical symptoms such as fatigue, muscle weakness and reduced ability to perform activities of daily living [8]. As soon as CKD occurs, it is associated with increased cardiovascular comorbid conditions [9]. With the kidney disease progressing, some coexisting conditions become more common and increase in severity. Hyperphosphatemia is an example of this, occurring because of insufficient filtering of phosphate from the blood by poorly functioning kidneys. High serum phosphate levels can directly and indirectly increase parathyroid hormone secretion, leading to the development of secondary hyperparathyroidism. Left untreated, secondary hyperparathyroidism increases morbidity and mortality and may lead to renal bone disease, with people experiencing bone and muscular pain, fracture, bone and joint abnormalities, and vascular and soft tissue calcification [10].

Patients who are ineligible for transplantation are typically appropriate for dialysis treatment if they choose to get it. The Peritoneal dialysis, Conventional hemodialysis, Daily and overnight dialysis, Hemodiafiltration or Self-care and home dialysis can be the options for management of CKD. The range of hemodialysis treatment times is typically between three and five hours, and some patients report hypotension symptoms during dialysis and increased exhaustion afterward, which restricts the acceptability of the procedure. Patients on dialysis who experience a variety of losses also experience higher depression and a decline in quality of life (QoL) [11].

CASE DESCRIPTION

The subject is 37 years old, male, severe hemophiliac with factor assay<1% and high titre inhibitor with BMI of 28.5 kg/m^2. He was diagnosed with hemophilia-A at the age of 03-months and was bed-ridden and totally dependent for activities of daily living from the last 02 years, prior to study because of the presence of co-morbidities along with severe hemophilia and numerous bleeds in the multiple joints in the past years. He had a medical diagnosis of bilateral hemophilic arthropathy in elbows,

knees and ankles 05 years back. He had repeated bleeds in the right elbow (07 hemarthrosis) and bilateral knees (03 in left and 02 in right knee) making them target joints. He gave an account of having been treated for episodes of excessive bleeding with On-demand FEIBA (Factor Eight Inhibitor Bypass Agent) intermittently, occasional cryotherapy, and rest. Additionally, he reported to have had daily painkillers due to moderate to severe pain in right elbow and bilateral knees for more than the last 2.5 years. He did not receive any physiotherapy management before this study period.

Along with this, he was also diagnosed with stage-5 chronic kidney disease [12] and had been on hemodialysis 02 times a week from the last two years prior to the study. The need of hemodialysis twice a week led to generalised progressive muscular weakness of upper and lower limbs (BMRC MMT scoring of most of the muscles of upper limb was 3/5 and that of muscles of lower limb was 2/5) followed by his inability to walk (FISH score 1 in locomotion domain).

One year prior to the study, he had an intracranial bleed for which he underwent burr hole surgery in left parietal bone. Within a month of surgery, he recovered from manifestations of intracranial bleed.

He was also diagnosed with bipolar disorder ten years ago at the age of 27 years, for which he was on medication.

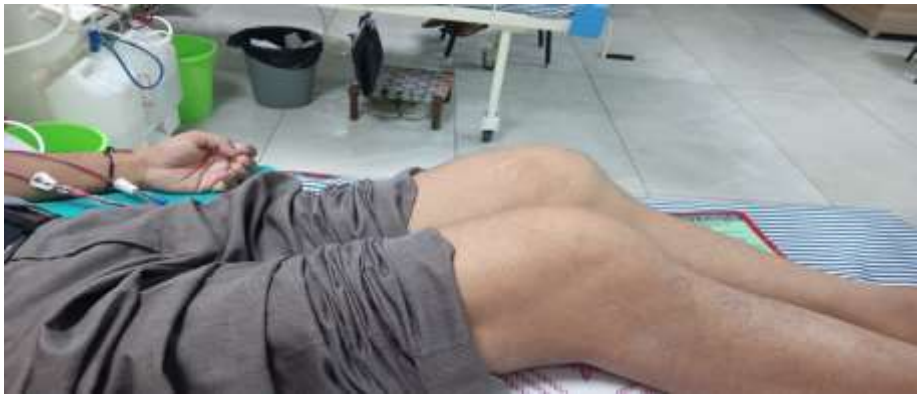

FIG-01: PWH with Bilateral knee arthropathy

After considering the demographic characteristics (including age, professional and personal history) and the medical history, the subject was further assessed for the joint health status using Hemophilia Joint Health Score (HJHS) [13], functional independence using Functional Independence Score in Hemophilia (FISH) [14] and Barthel Index (BI) Scoring of Activities of Daily Living whose total **scores** range from 0 (complete dependence) to 20 (complete independence) [15], pain using Visual Analogue Scale (VAS) with total scores range from 0 (no pain) to 10 (extreme pain) [16].

The HJHS is a score system for evaluating physical joint deterioration in PWH and is used for regular follow-up joint health exams. The six most often affected joints (elbows, knees, and ankles) are assessed in PWH using the HJHS, and overall scores fall between 0 and 124. High scores denote damage or impairment [13]. FISH is a performance-oriented evaluation tool for evaluating functional capacity in PWH. The tool consists of eight functional activities divided into three domains namely self-care, transfers and locomotion. The overall scores fall between 0-32. High scores indicate higher functional independence [14].

After the assessment, he received physiotherapy treatment which included exercise and electro-therapy for 4 months ie, 16 weeks, with an average of 4 sessions per week. The physiotherapy was in accordance with the guidelines of World Federation of Hemophilia (WFH). The sets, repetition and resistance of each exercise was dependent upon his ability to perform the exercise without pain and fatigue. The cold-packs

were applied after the physiotherapy session for 10-15 minutes at bilateral elbows, knees and ankles.

The exercises included passive range of motion exercises of bilateral upper and lower limbs, gradually progressing to active assisted range of motion exercises of the same along with breathing exercises (diaphragmatic, pursed lip and intercostal breathing) Glute squeezes and Glute bridge exercise for the first 8 weeks. All the exercises were done in supine lying. During this phase, the pain was managed by TENS application at both knees (frequency-50 to 100 Hz, duration-10 minutes each side) and Ultrasound at right elbow and both knees (mode- pulse with 1:4 ratio, intensity- 0.8 watt/cm^2, duration- 3 to 5 minutes at each site).

The exercises were progressed to strengthening of bilateral upper and lower limb including, isometric exercises of shoulder muscles, elbows and wrists in sitting position, isometric exercises of quadriceps, hamstrings, hip adductors and abductors, straight leg raise with a hold of 3-5 seconds, glute squeezes and glute bridge exercise in supine lying, along with backward and sideward straight leg raising with a hold of 3-5 seconds in side-lying position for next 4 weeks. Deep breathing exercises were incorporated in between the exercises and during periods of rest. The pain management continued the same as in the first 4 weeks. During this phase, for muscle stimulation, EMS was applied bilaterally at quadriceps (frequency- 45 Hz with a pulse of 300 µs and a contraction and relaxing cycle of 12 second ON and 8 second OFF, for a duration of 10 minutes).

Along with the continuation of the strengthening exercises and EMS, the mobility training including sit to stand with support, partial weight bearing on lower limbs with the use of a walker and supported standing was done in the last 4 weeks of intervention. All the parameters were re-evaluated after 4 months of physiotherapeutic intervention

RESULT

The Pre and post treatment scores of HJHS, FISH, BI and VAS were 49 vs 43, 9 vs 16, 5 vs 14 and 8 vs 4 respectively (TABLE 01,02,03).

TABLE 01 - The scores of HJHS and FISH before and after the physiotherapy administration. The results showed 4.84% decrease in HJHS from the baseline mean score and 21.87% increase in FISH score from the baseline mean score.

Outcome Measure	Pre	Post
HJHS	49	43
FISH	9	16

TABLE 02- Barthel Index Scoring of Activities of Daily Living Pre and Post treatment.

Barthel Index (BI) Scoring		
Activity	**Pre**	**Post**
Feeding	1	2
Bathing	1	1
Grooming	1	2
Dressing	1	2
Bowel Control	2	2
Bladder Control	2	2
Toilet Use	1	1
Transfers (e.g., from bed to chair and back)	0	1
Mobility (on level surfaces)	0	1
Stairs	0	0
Total	9	14

TABLE 03- Pain characteristics Pre and Post treatment.

Pain Sites	
Pre	**Post**
Right Elbow Both Knees	Right Elbow
Generic Vas Score	
Pre	Post
8	4

DISCUSSION

This case report represented a severe hemophilic person who had shown improvement in joint health and functional independence in daily activities after the administration of physiotherapy management despite of having complicated medical history. Due to the presence of other comorbidities along with the musculoskeletal conditions and higher BMI, physiotherapy was a challenge for him. He had to under-go for dialysis every 3rd day ie, twice a week which would cause fatigue and muscle weakness. On the day of dialysis, he could not perform exercises because dialysis itself was a very cumbersome process.

The first signs of recovery were registered after the first 3 months of physiotherapy. With the regular exercises, he gradually started to show functional recovery and was able to perform activities of self-care with

mild to moderate assistance (Barthel Index Scoring Pre and Post Treatment, 5 vs 14 respectively). But making him stand on his feet was still a challenge. Right elbow being the target joint, he could not bear weight on it because by putting weight on right elbow, it started to bleed instantly. So, the main focus was the strength training of glutes, and the muscles of the back to compensate the weaker part (ie, right elbow). After 4 months of regular physiotherapy, he started to walk at home with the help of a walker. He resumed back to his 6 hours job and also started driving a car. He needed minimal assistance while transferring from wheel-chair to driving seat and then back to wheel-chair.

A similar case study [17] of a 23 years old male hemophiliac, with hemophilic arthritis and major complaint of pain in different body sites, especially in ankles and right knee reported improvement in pain and quality of life of the patient after eight physiotherapy sessions, once a week, which aimed at muscle strengthening, mobility, balance, proprioception and gait training by means of weight bearing. The result of this study is consistent with the present study because our patient also received physiotherapy sessions which were based on lower limb strengthening and weight bearing along with pain management which resulted in reduction in pain as well as improved quality of life in terms of improvement in joint health status and functional independence.

Another case report [18] of a person with severe hemophilia A, showed improvement in quadriceps muscle strength and reduced joint pain after the 2 months of physiotherapy interventions including faradic muscle stimulation, ultrasonic therapy and progressive resistance muscle training. The findings of our study are in line with this study because of the improved joint health status and reduced pain with the use of muscle stimulation, progressive strength training and ultrasonic therapy.

A Systematic Review [19] regarding the efficacy of physiotherapeutic modalities in patients with hemophilia have also reported the strong evidence for using the therapeutic exercise modalities to improve joint health status, in combination with educational sessions to improve QoL of patients with hemophilic arthropathy.

CONCLUSION

The findings of this particular case of a severe hemophiliac person who was inhibitor positive with hemophilic arthropathy of multiple joints and CKD showed that physiotherapy played a significant role in improving joint health status, functional independence and reducing pain. Physiotherapy including exercise and electrotherapy improved muscle strength and reduction in pain which resulted in better joint health and functional independence status of the patient. It was also concluded that if one particular joint is bleeding, physiotherapeutic exercises can be focused on other alternate joint/s for the completion of the functional movement. Further research exemplifying the most effective physical therapy treatment for severe hemophilic persons with inhibitor and other medical complications are still needed.

List Of Abbreviations

BI- Barthel Index
CKD- chronic kidney disease
EMS- Electrical Muscle Stimulation
FISH- Functional Independence Score in Hemophilia
HA- Hemophilia A
HJHS- Hemophilia Joint Health Score
PWH- People with Hemophilia
VAS- Visual Analogue Scale

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