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"OUTCOME ANALYSIS OF TIMELY CONSERVATIVE MANAGEMENT OF HAEMOPHILIC KNEE ARTHROPATHY WITH FACTOR REPLACEMENT AND PHYSIOTHERAPY."

Orthopaedics

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ABSTRACT

INTRODUCTION: Haemophilia is a disorder of the initiation of coagulation, and is due to reductions in the concentrations of, or the presence of a less active version of, one of two coagulation factors, factor VIII and factor IX. There are several orthopaedic problems linked to haemophilia including recurrent hemarthroses, chronic synovitis, flexion deformities, hypertrophy of the growth epiphyses, damage to the articular cartilage and hemophilic arthropathy

MATERIALS AND METHODS : 290 patients of all age group of haemophilia A and B coming to orthopaedic OPD with Knee Haemarthosis between October 2018 to May 2020 were included in our study. These patients were prospectively analyzed for complications and severity. HJHS Scoring system were used to assess the severity of joint disease.

RESULTS : . out of 290 patients it was found that haemophilia was more prevalent in rural population (53.10%) as compared to urban population (46.90%).

In our study it was found that majority of the patients with haemophilia had single joint involved (57.94%) as compared to 42.06% who had multiple joint involved.

It was revealed that in majority right knee was involved (52.41%) followed by left joint in 47.58% patients with haemophilia. It was found that most common complication in patients of knee haemarthrosis was recurrent haemarthrosis (57.24%) followed by synovitis (27.93%). arthropathy (9.31%) . It was found that most common complication in patients of knee haemarthrosis was recurrent haemarthrosis (57.24%) followed by synovitis (27.93%). arthropathy (9.31%) . It was found that out of 290 patients with haemophilia, majority were performing regular physiotherapy (57.59%) followed by 42.41% of the patients who were occasional in physiotherapy. It was found that mean HJHS score was increasing with increasing the severity of the disease. Mean HJHS score for mild cases was 10.53±4.495 as compared to 13.06±7.575 of moderate cases and 17.82±7.991 of severe cases.

CONCLUSION

In present study haemophilia was more prevalent in rural population. Majority of the patients with haemophilia had single joint and in majority right knee was involved. Most common complication in patients of haemophilia was recurrent haemarthrosis followed by synovitis and arthropathy. Majority of our patients were having severe form of the disease. Adequate factor replacement along with good active physiotherapy early detection and prompt treatment with active life style makes muscles and joints healthier and stronger reduces tendancy of bleeding and further damage of joint. reduce the frequency and longterm complications of knee haemarthrosis like recurrent haemarthosis , synovitis ,arthropathy, fracture and deformity.

Prophylactic factor replacement is having definitely positive role as per available literature over western countries, unfortunately it needs more specific clinical trial to know about longterm good results in India.

KEYWORDS

haemarthrosis, haemophilia, arthopathy, haemophilic factor, knee joint, synovitis

INTRODUCTION

Haemophilia is a disorder of the initiation of coagulation, and is due to reductions in the concentrations of, or the presence of a less active version of, one of two coagulation factors, factor VIII and factor IX. Orthopaedics is a large and complex field of medicine that caters not only to patients with fractures and soft tissue injuries but also musculoskeletal conditions.(Gani A 2016).The major morbidity experienced by patients with haemophilia today is joint disease, a result of repeated bleeding episodes into joint spaces.There are several orthopaedic problems linked to haemophilia including recurrent hemarthroses, chronic synovitis, flexion deformities, hypertrophy of the growth epiphyses, damage to the articular cartilage and hemophilic arthropathy. Haemophilia is an X-linked recessive condition ad it affects one in 5000 men.

The deficiency of these factors results from a defect in one of two genes on the X chromosome, F8 (which codes for factor VIII) and F9 (which codes for factor IX). Hemophilia A is caused by deficiacy of factor VIII and haemophilia B is caused by deficiacy of factor IX .haemophilia A is the most common form of haemophilia .haemophilia has X –linked mode of inheritance.most of the females are carrior and are asymptomatic. Based on clinical manifestations, three stages can be distinguished. Acute hemarthrosis mostly resolves with well-conducted replacement therapy and rehabilitation, usually without clinically detectable sequelae. Subacute hemarthrosis occurs after repeated hemarthrosis episodes in the same joint – at this stage, the joint and surrounding soft tissues do not fully recover, while clinical signs of joint damage persist and are detectable between bleeding episodes. Chronic arthropathy develops following numerous repeated bleeding episodes in joints, resulting in a significant loss of muscle function and muscle–tendon contractures. The management of acute bleeding episodes is based currently on immediate replacement therapy with clotting factor concentrates (prophylaxis) is effective in preventing recurrent bleeding episodes into joints and muscles and the RICE (Rest, Ice, Compression, and Elevation) regime. In the acute phase, the first therapeutic measure consists in injecting clotting factor within 2 hours of bleeding onset. The classically recommended dose is 25 to 40 IU/kg of FVIII, or 50 IU/kg in cases of severe haemarthrosis.

Several injections, spaced out over time, are generally required. In developed countries (20-30% of the world population), the orthopedic problems of haemophilia have reduced since the introduction of prophylaxis using concentrates of the deficient coagulation factor. The use of prophylaxis has the effect of turning serious haemophilia into a milder form by maintaining the factor level above 1% permanently; doses are titrated to the concentration of factors and the incidence of bleeds. While improvements in medical prophylaxis and treatment for haemophilia have improved outcomes and reduced the need for orthopaedic interventions in developed countries, musculoskeletal problems remain common in haemophiliacs.

There is limited data available highlighting the complications associate with the routine replacement of clotting factors in patients of

haemophilia with knee haemarthrosis. Hence present study is an attempts to assess the long-term outcome of prophylaxis antihaemophylic factor to prevent pattern and response of knee haemarthrosis in haemophilia patients. Knee haemarthrosis occurs more who belongs to lower socioechonomic group and where lack of avialibilty at local places .Patients belongs to remote area are usually takes 3-5 days or even more to reach hamidia hostipital and sometimes even after the development of complication of haemarthrosis.

So there is need of improvement of awareness about haemophilia and to trained doctors and paramedical staff at district level and community level hospitals For the improvement of rehabilitation ,physiotherapy facities should be present at community level.

MATERIALAND METHODS

This is a prospective observational study conducted in department of orthopaedics with the collabotation of haemophilia day care centre Gandhi medical college and Hamidia hospital ,Bhopal ,Madhya Pradesh,India .

Total 290 haemophilic patients with knee haemarthrosis are taken for the study duration of October 2018 to may 2020

All the patient presented to department of Orthopaedics, detailed history was taken then general examination and local examination done. Based on history and physical examination target joint were assessed using Hemophilia Joint Health Score (HJHS) 2.1 for functional assessment of knee joint on every follow-up at 3week, 6week, 3month, 6 month and one year. Over the knee haemarthrosis creppe bandage were applied. In case of acute bleeding ice fomentation was done. X-rays and ultrasonography for the suspicion of synovitis and arthropathy was done. All patients received on demand factor replacement therapy as per protocol.

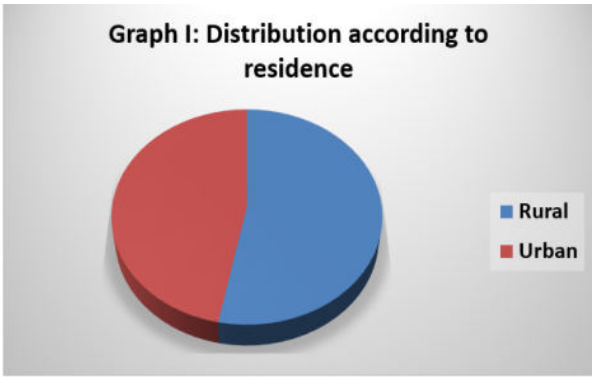
INCLUSION CRITERIA: All the patients of haemophilia A and B coming to orthopaedic OPD of Hamidia hospital with knee haemarthrosis of all age group

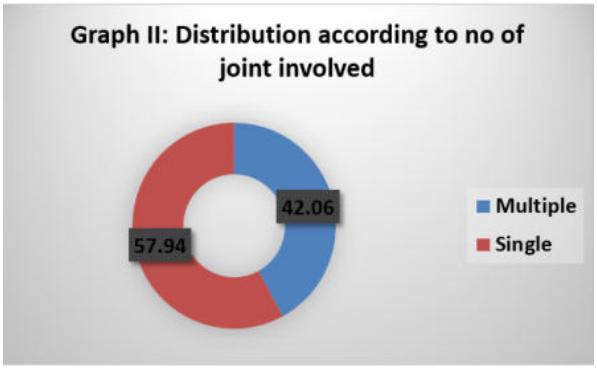
EXCLUSION CRITERIA: Patients not giving consent for study**.** Patients who lost to follow up. Non haemophilic patient.

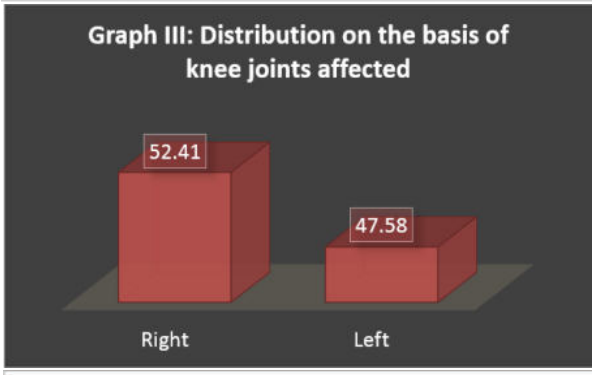
RESULTS:

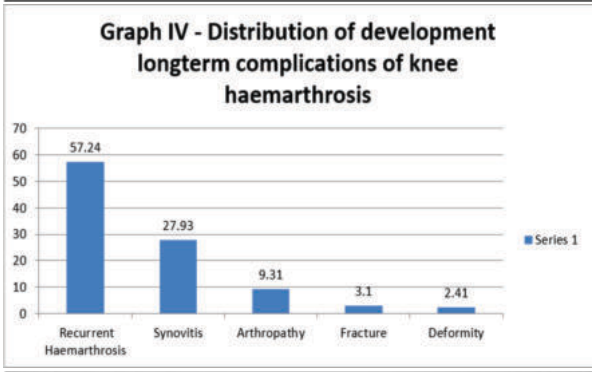
Out of 290 patients it was found that haemophilia was more prevalent in rural population (53.10%) as compared to urban population (46.90%). In our study it was found that majority of the patients with haemophilia had single joint involved (57.94%) as compared to 42.06% who had multiple joint involved. It was revealed that in majority right knee was involved (52.41%) followed by left joint in 47.58% patients with haemophilia. It was found that most common complication in patients of knee haemarthrosis was recurrent haemarthrosis (57.24%) followed by synovitis (27.93%). arthropathy (9.31%) . It was found that most common complication in patients of knee haemarthrosis was recurrent haemarthrosis (57.24%) followed by synovitis (27.93%). arthropathy (9.31%) . It was found that out of 290 patients with haemophilia, majority were performing regular physiotherapy (57.59%) followed by 42.41% of the patients who were occasional in physiotherapy. It was found that mean locomotion score was similar for each severity of the disease as revealed by the insignificant p value of 0.070.

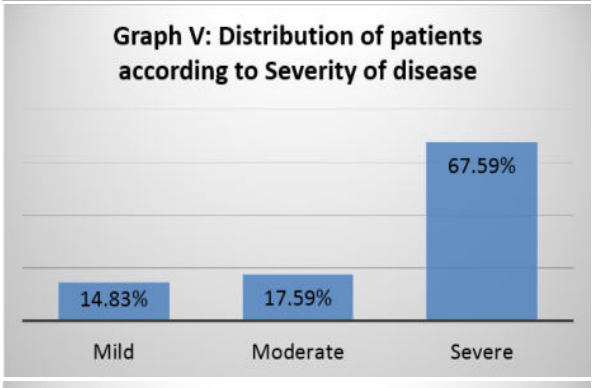
It was found that mean HJHS score was increasing with increasing the severity of the disease. Mean HJHS score for mild cases was 10.53±4.495 as compared to 13.06±7.575 of moderate cases and 17.82±7.991 of severe cases. The post hoc analysis between groups was also highly significant with p value of <0.001 for each severity.

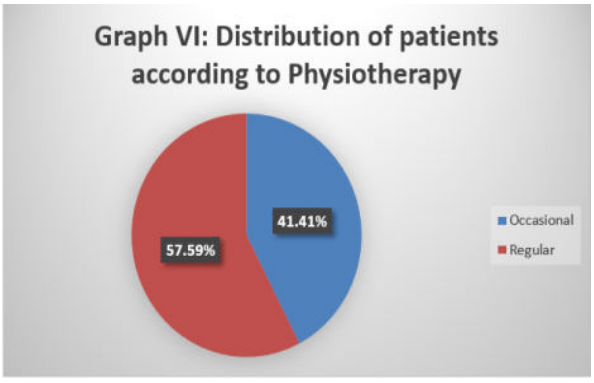


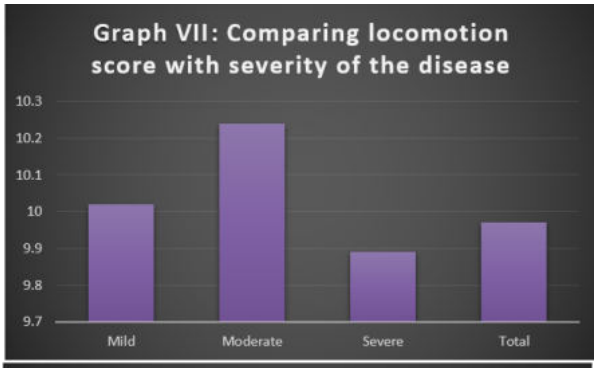


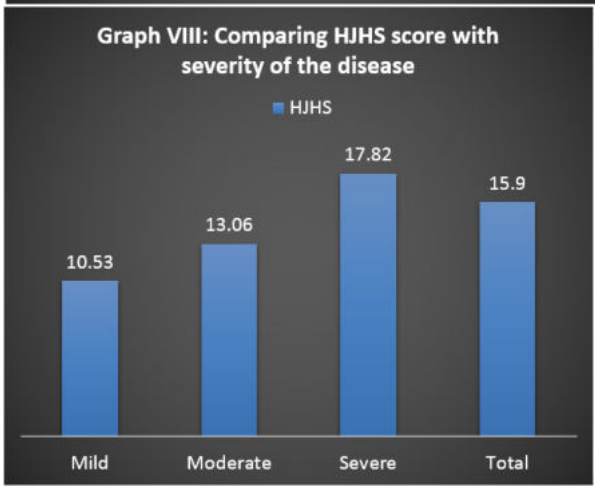












DISCUSSION

The early damage to joints in patients with haemarthrosis that often escapes diagnosis because of the insufficient investigations for biomechanical changes. Arthropathy in haemophilia needs complex assessment with several tools. Different reports have shown high rates of complications for bleeding, deep infections and prosthetic failures. There was limited data available to demonstrate good results in a larger collective with a long follow-up period. Present study was carried out to evaluate hemophilic joint clinically and functionally using Hemophilic joint health score (HJHS) and locomotion score and global gait score.

In present study haemophilia was more prevalent in rural population than urban population. In a similar study by Ayyed Mohammad Namoos AL – Zubaidyin a descriptive study of haemophilia involving 60 patients with haemophilia. Residency distribution revealed 65% were rural and 35% were urban.[1]

In present study majority of the patients with haemophilia had single joint involved as compared to multiple joint involved.

In present study, in majority right knee was involved. In a similar study by Jansen et al[2] and Abdel Ghany et al[3] found similar results.

The incidence and severity of haemorrhagic complications of haemophilia are directly related to the severity of the underlying coagulation defect. Although the intrinsic pathway of coagulation is severely impaired in haemophilia, the extrinsic tissue factor dependent pathway remains intact and is probably the major haemostatic regulatory system. It has been found that normal synovial tissue and cultures of synovial fibroblast were deficient in tissue factor, which suggests that in synovium – lined joints, haemophiliacs have the functional inactivity of intrinsic and extrinsic coagulation pathways.[4] This may explain the marked propensity towards haemorrhage in joints compared with the other tissue sites in these patients.

In present study, most common complication in patients of haemophilia was recurrent haemarthrosis followed by synovitis and arthopathy. In our study joint bleeding (recurrent haemarthrosis) was most common complication with haemarthrosis of knee joint being most common, similar to study of M. Varadrajan et al who has got haemarthrosis in 52.9% of the cases along with knee being the most common joint involved followed by elbow and ankle as in our study.[5] As per Rawand Polus Shamoon et al[6] Balkan C. et al[7] and Rodiguez-Merchan E.C[8] the overall incidence of haemarthrosis was more than 75%.

Greene et al (1994) showed that hemophilia can result in musculoskeletal bleeding, including hemarthroses, leading to musculoskeletal complications. The articular problems of the hemophiliac patients starts in infancy which includes recurrent haemarthroses, chronic synovitis, flexion deformities, hypertrophy of the growth epiphyses, damage to the articular cartilage, and haemophilic arthropathy. The most commonly affected joints are the ankle, the knee, and the elbow.[9]

In present study out of 290 patients with haemophilia had severe disease (67.59%) followed by moderate disease (17.59%) and mild disease (14.83%). A study from Bhopal Madhya Pradesh by Choudhary et al reported that majority of patients (52%) were severe haemophiliac (42.32%-61.54%) at 95% confidence interval.[10]

Out of 290 patients with haemophilia, majority were performing regular physiotherapy (57.59%) followed by 42.41% of the patients who were occasional in physiotherapy.

In present study mean locomotion score was similar for each severity of the disease as revealed by the insignificant p value of 0.070. In a study of orthopaedic assessment results, including knee motion analysis, from 273 children (median age 9.8 years) with haemophilia A, B or von Willebrand disease, compared with 200 age-matched and sex-matched controls, reported similar findings.[11]

However, on follow up there was significant improvement in locomotion score from 6.97±0.45 at baseline to 9.97±0.97 at 3 weeks, 12.97±0.66 at 6 weeks and 13.92±0.97 at the end of 3 months, 14.68±0.58 at the end of 6 months and 16.97±0/23 at the end of 1 year
To date, HJHS is the internationally recognized tool for assessing joint structure and function. In present study mean HJHS score was increasing with increasing the severity of the disease. Mean HJHS score for mild cases was 10.53±4.495 as compared to 13.06±7.575 of moderate cases and 17.82±7.991 of severe cases. The post hoc analysis between groups was also highly significant with p value of <0.001 for each severity. Recent study by Oymak et al demonstrated agreement between the HJHS and MRI scores, indicating that the HJHS may be used safely as a first-line tool to evaluate structure[12], and a study in China by Sun et al has proven the reliability, internal consistency and global transferability of HJHS version 2.1, even for administration by physiotherapists and physicians with limited haemophilic experience.[13] Present study is in agreement to both above mentioned studies which highlight the HJHS score in evaluating the severity of the disease. We also performed the follow of the patients for the HJHS score to get further idea on its usefulness. It was found that on follow up there was significant improvement in HJSH score from 12.90±3.45 at baseline to 15.90±6.78 at 3 weeks, 18.90±3.12 at 6 weeks and 19.88±2.34 at the end of 3 months, 20.92±1.46 at 6 months and 22.90±1.14 at the end of 1 year.

World Federation of Haemophilia recommends the HJHS, which picks up the early signs of joint damage. It monitors joint changes over time for assessing efficacy of treatment.

The last subtitle of the HJHS was the global gait score and its maximum score is 4 points. In present study It was found that on follow up there was significant improvement in global gait score from 1.48±0.46 at baseline to 1.59±0.76 at 3 weeks, 3.46±0.56 at 6 weeks, 3.58±0.34 at 3 months, 3.88±0.29 at 6 months and 4.00±0.00 at the end of 1 year.

In present study age at haemarthrosis also had a significant negative correlation with locomotion score ($r=-0.161$, $p=0.006$) and HJHS score ($r=-0.240$, $p<0.001$). Which means lower the age at haemarthrosis higher will be the average bleeding event per event, locomotion score and HJHS score.

CONCLUSION:

In our study most of the patients belongs to rural background and they present late to the hospital. In present study haemophilia was more prevalent in rural population. Majority of the patients with haemophilia had single joint and in majority right knee was involved. Most common complication in patients of haemophilia was recurrent haemarthrosis followed by synovitis and arthropathy. Majority of our patients were having severe form of the disease. Adequate factor replacement along with good active physiotherapy early detection and prompt treatment with active life style makes muscles and joints

healthier and stronger reduces tendency of bleeding and further damage of joint. reduce the frequency and longterm complications of knee haemarthrosis like recurrent haemarthosis , synovitis ,arthropathy, fracture and deformity.

Prophylactic factor replacement is having definitely positive role as per available literature over western countries, unfortunately it needs more specific clinical trial to know about longterm good results in India.

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