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RESEARCH ARTICLE

JOINT PAIN AND QUALITY OF LIFE IN ADULT HAEMOPHILIA- A CASE SERIES

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ABSTRACT

Background: Haemophilia is a hereditary X-linked recessive disorder characterized by the deficiency of factor VIII or IX coagulant activity. Internal and prolonged bleeding into the joint space and muscles are the main symptoms of haemophilia which inadvertently results into haemophilic arthropathy and affects the individual's life. This study was conducted to gain an insight into the impact of haemophilia on the Quality of Life (QoL) of haemophiliac adults. **Methods:** This was a case series which included 30 patients, who were screened for the inclusion and exclusion criteria and enrolled in the study. Male patients with Haemophilia A or B who were undergoing treatment at the hospital and in the age group of 18-60 yrs. with complications of Haemophilia such as joint swelling, spontaneous bleeding etc. were included in the study. Patients with and musculoskeletal, neurological or cardiovascular conditions not associated with Haemophilia and other bleeding tendencies such as liver failure, anti- coagulant drugs, etc. were excluded from the study. The outcome measures used were the HAEM- A – QOL, which is an adult version of the quality of life in Haemophilic adults and the Numerical Pain rating scale (NPRS). **Results:** Total 30 patients were included in this study, the HAEM- A- QOL questionnaire was administered and NPRS was taken. The average age of the patients was 29 years (±3.97) and mean NPRS score was 4.2 (±1.31). 40% of the subjects had elbow as their target joint and rest 60% had knee as their target joint. The Mean and Standard Deviation for various subscales were: 1. Day to Day activities – 56(±6.99) 2. Mood and Feelings – 65.1(±7.32) 3. Work or school life, family life and Social Life – 62.8(±2.17) 4. Haemophilia Treatment – 30.5 (±7.56). Average QOL of Life was 53.6, with standard deviation ±10.46 in adult male haemophilia patients. **Conclusion:** The present study concludes that there was a moderate affection of the quality of life in these patients. According to the HAEM- A- QOL, the emotional and behavioural aspect was severely affected followed by the Social life and the Activities of Daily living, which were moderately affected. Worries/ concerns related to the treatment were mildly affected.

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INTRODUCTION

Haemophilia is a hereditary X-linked recessive disorder characterized by the deficiency of factor VIII or IX coagulant activity (Mishra, 2016). Haemophilia A (classical), caused due to deficiency of factor VIII is seen in 80% of the cases. Haemophilia B (Christmas disease), caused due to deficiency of factor IX is seen in 20% 0f the cases (Payal *et al*., 2016). Internal and prolonged bleeding into the joint space and muscles are the main symptoms of haemophilia, muscle bleeds can occur in any muscle due to direct blow or sudden stretch. The most commonly affected joints are knee, elbow and ankle (Strike *et al*., 2016).

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It is the most common congenital bleeding disorder encountered in clinical practice affecting men whereas women are generally carriers (Payal *et al*., 2016). The World Federation of Haemophilia estimates that there are 4,00,000 individuals worldwide with haemophilia, out of which 80% are in developing countries such as India (Payal, 2016). Haemophilia A occurs in 1 out of 10,000 male births, while haemophilia B, occurs in 1 out of 30,000 male births. The prevalence of haemophilia A varies with the reporting country, with a range of 5.4-14.5 cases per 100,000 male individuals (Kumar *et al*., 2019). The mainstay of haemophilia treatment includes clotting factor concentratedepending on the severity and type of factor deficiency (factor VIII or IX). Clotting factor concentrate is an orphan drug, meaning it is a pharmaceutical agent developed to treat medical conditions which are rare and hence would not be profitable to produce without government assistanc.

Owing to this its high cost and non- availability placesthe drug out of reach of the majority of Indian patients. The bleeding risk is related to the severity of the factor deficiency, and repeated joint bleeding can cause severe joint damage and pain, leading to disability (Limperg *et al*., 2017) which is called as Haemophilic Arthritis or Arthropathy. Haemophilic arthropathy is a disabling condition characterized by joint impairment, chronic pain, and reduced quality of life (Melchiorre, 2017). As mentioned by Melchiorre, it can even lead to subchondral bone damage and osteoporosis due to the haemarthrosis that occurs in the joints. The use of RICE and Ultrasound has been recommended to reduce muscle and joint haematomas (Melchiorre, 2017) Beyer suggests that it would be interesting to develop various physiotherapeutic regimes to treat the manifestations of haemophilia (Melchiorre, 2017). According to Ghosh & Ghosh in 2015, the following challenges are faced by haemophiliacs in the developing countries: 1. Rickety health care infrastructure 2. Lack of penetration of medical insurance services 3. Competing priorities for different diseases 4. Lack of awareness 5. Non availability of factor concentrates for PWH 6. Inadequate utilization of management modalities which can reduce factor usage 7. Cost of surgery and joint injections in Haemophilia 8. Challenge of inhibitor and chronic liver disease 9. Lack of Research in Haemophilia (Ghosh, 2015).

India is a developing nation with more than 50% of its population residing in rural and semi- urban areas. Patients suffering from haemophilia especially from these areas do not have easy access to the standard care and treatment as required for haemophilia. This is further compounded by the lack of awareness about the condition. Haemophilic arthropathy is a major cause offrequent absenteeism from school and work.This not only affects the patient at an individual level but also restricts his role as an active member of the family as well as in society. Self- reporting by respondents is important to capture the individual perception of health conditions and treatment regimes (Bullinger, 2002). Health Related Quality of Life (HRQoL) is a multidimensional concept that refers to the subjective impact of health and illness on an individual's daily functioning, which encompasses not only physical functioning but also social, cognitive and emotional functioning (Bullinger, 2012). In general, HRQoL questionnaires are either generic or disease-specific. Generic instruments allow for comparison of HRQoL across conditions, whereas diseasespecific questionnaires are more responsive to change in the area of interest and more sensitive to the impact of changes in clinical conditions or treatment (Bullinger, 2002). HRQoL instruments should include physical and mental domains and should possibly be based on patient report, by means of self-administered questionnaires or interviews that cover the different aspects of health (Gringeri, 2008). In India, there is a paucity of literature not only about haemophilia but also its affection on various aspects of life at an individual and social level. This study is thus being conducted to gain an insight into the impact of haemophilia on the Quality of Life (QoL) of haemophiliac adults.

Aim and Objective: To study the joint pain and quality of life in adult male haemophilia patients.

MATERIALS AND METHODS

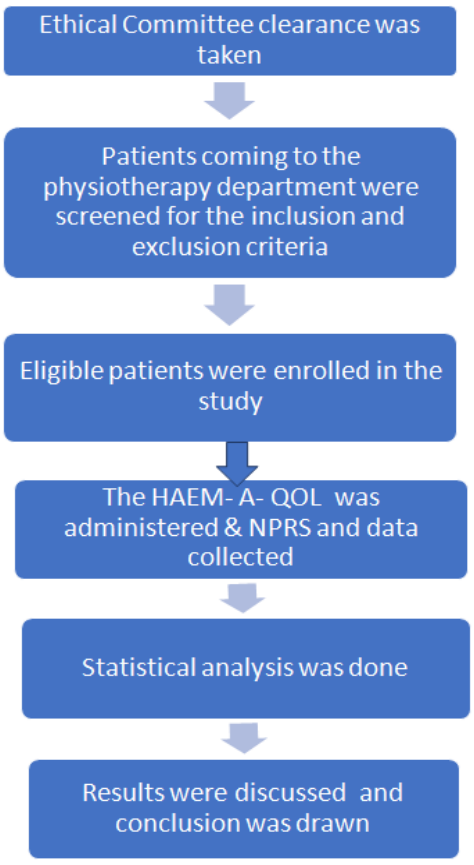
This was an Observational study conducted in K.J. Somaiya Medical College, K.J. Somaiya College of Physiotherapy, approved by the institutional ethical committee of K.J. Somaiya Medical College, Mumbai. Study participants were all adult male haemophilia diagnosed patients taking treatment at the Hospital. Total duration of the study was 6 months. Sampling method was convenient sampling. Kind permission of the author was taken for the usage of HAEM- A- QOL questionnaire for English, Hindi and Marathi languages (Mapi Institute ID-5691). Male patients with Haemophilia A or B who were undergoing treatment at the hospital and in the age group of 18-60 yrs. with complications of Haemophilia such as joint swelling, spontaneous bleeding etc. were included in the study. Patients with and musculoskeletal, neurological or cardiovascular conditions not associated with Haemophilia and other bleeding tendencies such as liver failure, anti- coagulant drugs, etc. were excluded from the study.

The following outcome measures were used

- Numerical pain rating scale (NPRS) for pain (Ferreira-Valente, 2011)
- HAEM- A- QOL questionnaire (Rentz, 2008)

In this study, the HAEM- A- QOL that is the adult version of the questionnaire has been used. The questionnaire consists of four domains namely day-to-day activities, moods and feelings, work or social life, family life and social life and haemophilia treatment which the patient has been subjected to in the last 4 weeks. Each domain has 11, 11, 14 and 5 questions respectively totalling to 41 questions. The participant has to mark the answer on a scale with 0 being None of the time and 5 being All of the time.There is no right or wrong answer. If the participant is unsure about how to answer a question, he has to choose the one response that best represents his opinion (Rentz, 2008)

Flowchart:



Statistical Analysis: We conducted descriptive analysis using Microsoft excel on 30 subjects. We calculated mean, standard deviation and range for the same.

RESULTS

Total 30 patients were included in this study, the HAEM- A-QOL questionnaire was administered and NPRS was taken. The average age of the patients was 29 years (±3.97) and mean NPRS score was 4.2 (±1.31). 40% of the subjects had elbow as their target joint and rest 60% had knee as their target joint.

The Mean and Standard Deviation for various subscales were:

- Day to Day activities – 56(±6.99)
- Mood and Feelings – 65.1(±7.32)
- Work or school life, family life and Social Life – 62.8(±2.17)
- Haemophilia Treatment – 30.5 (±7.56).

Average QOL of Life was 53.6, with standard deviation ±10.46 in adult male haemophilia patients.

DISCUSSION

This study was conducted to understand the quality of life and pain level of patients suffering from Haemophilia. Literature regarding the quality of life in haemophilic adults with respect to physical and mental well-being is scarce. Hence, such studies are required as they help us to understand our goals better and priorities during treatment of these patients. As mentioned earlier, the HAEM- A- QOL takes into account four domains which helps us in understanding the participant's whole perspective to the various aspects of their life. The average score of HAEM- A- QOL was 53.6 during this study. A study by Allhaidan concluded that the treatment concern is the most important domain in their study (Allhaidan, 2018). This particular study was performed in Riyadh, Saudi Arabia where treatment centres specializing in haemophilia are limited and hence, there is particular concern about the same.

The most affected domain in our study was mood and feelings of the patient with an average of 65.1. The worry about untimely bleeding is of constant concern to these individuals. This leads to a constant feeling of being less confident than others and ultimately frustration about not being able to take risks in life. The second most affected domain was work or school life, family life and social life with an average of 62.8. Many workplaces, in spite of having a disability reservation discriminate between their employees, or there is some sort of stigma associated with the quality of work these individuals will put in. The average age of patients in this study was around 29, who were mostly young adults, bread earners of their family, their role in planning for the future and supporting their family was crucial. Hence, this domain being the second most affected one was understandable. The treatment priority here would include patient education and lifestyle modifications for the same, along with the medical treatment for its compliance. India is a developing country and not fully disable- friendly and hence a few environmental modifications would be required for the betterment of these individuals. Future studies would be required to formulate exercise protocols for these individuals with adequate modifications due to their predisposition to haemarthrosis. Also, the effect of various exercises and modifications can be evaluated.

Conclusion

The present study concludes that there was a moderate affection of the quality of life in these patients. According to the HAEM- A- QOL, the emotional and behavioural aspect was severely affected followed by the Social life and the Activities of Daily living, which were moderately affected. Worries/ concerns related to the treatment were mildly affected.

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