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Rehabilitation Science

HEALTH-RELATED QUALITY OF LIFE IN PATIENTS OF HAEMOPHILIA FROM RAJASTHAN

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ABSTRACT

Introduction: Haemophilia is an X-Linked recessive bleeding disorder affecting men. Haemophilia A & B are defined as severe with the factor level below(<1%), moderate(1-5%), and mild(5-40%). Haemophilia have an impact on the patients quality of life and can affect the everyday life of their families. Quality of life is an important patient based outcome criterion which can be used to describe the impact of the health condition on the patients as well as the effects of the treatment strategies.

Objective: To analyse health related quality of life (HRQoL) in haemophilia patients

Method: North Indian patients with haemophilia A & B, aged >13yrs were compared with normative individual matched for age, sex, geography and social status. The SF-36 questionnaire used to measure HRQoL, allows to calculate 8 score (on a 0-100 scale)the Physical component (PCS) and Mental component(MCS) summary. Mean Mental (MCS) and physical (PCS) component summary scores were assessed and compared with Indian normative data.

Results: In order to assess HR-QoL in patients with haemophilia A and B , 88 individuals with severe, moderate and mild haemophilia were enrolled out of which 67 patients received MOS short Form 36(SF-36) questionnaire Patients mean age was 21years (SD=9.9), 91% were type A , and 9% were type B, the results from these questionnaire showed that the Mean PCS and MCS was significantly lower in the haemophilia patients compared to the normative data.

Conclusion: patients with haemophilia have poor health related quality of life as compared to normative individual. That's why early intervention in haemophilia is must to reduce the functional disability. These observations should help to optimise health care delivery in this increasing and neglected population of people with haemophilia.

KEYWORDS : Quality Of Life, Physical And Mental Component, Haemophilia

INTRODUCTION

Haemophilia is an X-linked inherited recessive bleeding disorder that is characterized by a deficiency of clotting factor VIII (classic haemophilia, or haemophilia A) or IX (haemophilia B). Haemophilia has a frequency of 1 in 5000 male births, whereas haemophilia B has a frequency of 1 in 30000 male births [1].

India has approximately one lakh people affected with Haemophilia with an estimated prevalence rate of 1:10000. Around 14000 are registered patients. It has been estimated that in India 1300 children with haemophilia are born each year and there are nearly 50,000 patients with severe haemophilia [2].

The level of clotting factor grades the severity of the disease. Patients with severe haemophilia have <1% clotting factor activity, moderate affected patients 1-5% and mild patients 6-40% [3].

Lower levels of clotting factor, especially in the severe patients result in spontaneous haemorrhages in muscles and joints, but may affect other organs as well. Especially repetitive haemorrhages in joints ultimately result in crippling haemophilic arthropathy[4]. One reason for this is that a large majority (approximately 80%) of the patients [5] live in developing countries where financial constraints limit the use of factor concentrates [5].

Another reason is the formation of inhibitory antibodies (inhibitors) to factor concentrates in about 30% of young patients with haemophilia A and in 1-6% of patients with haemophilia B [6]. When inhibitors are present, the treatment is less effective and patients may be more affected by haemophilia in terms of joint status and quality of life [7]. Thus, despite improved treatment strategies, still many patients with haemophilia experience the serious consequences of this disease.

Health-related quality of life is increasingly recognized as a important outcome in the care of patients with haemophilia. During the course of disease, haemophilia patients health related quality of life may be compromised because of joint bleeds, chronic pain, and other clinical complications. Variables which have been shown to be associated with reduced HRQL among haemophilia patients include: the severity of the clotting defect, bleeding frequency and orthopaedic problems. As well as, treatment may improve HRQL (2009) [8]. Until recently, the most widely used instruments to assess Quality of life in Haemophilia research have been the generic measures, such as SF-36(Medical Outcome Study) or EQ-5D (EuroQoL) [9].

There are a very few data about haemophilia in India. Many patients in India end up with severe disability due to late recognition, unavailability of factor or inappropriate treatment.

This study is designed to measure Health Related Quality of Life among hemophilia patients, the study explores clinical and treatment factors and their impact on Health Related Quality of Life (HRQL). Many previous studies [12,13] ,have correlated Health Related Quality of life (HRQL) in hemophilia, we have also tried to explore Health related Quality of Life (HRQL) in Indian scenario.

Material and methods

The present study was conducted in the department of Physical Medicine and Rehabilitation, Sawai Mansingh Hospital Jaipur during the period of April 2012 to November 2013.

Source of Data:

All diagnosed cases of haemophilia from 14yr to 45yr attending outpatient department and those admitted to the hospital.

Inclusion Criteria:

- Patients age > 13 yrs
- Willingness to participate in the study
- Mild , Moderate, Severe Hemophilia patients

The Regional Committee for Medical Research Ethics approved the study. Our research was approved by the committee on research ethics at the institution in which the study was conducted and in accordance with the declaration of the World medical Association.

Methodology:

All patients admitted to Department of Physical Medicine and Rehabilitation (PMR) with diagnosis of hemophilic arthropathy was clinically evaluated and screened and explained about the type and nature of study. Informed consent was taken and enrolled for study. A detailed history and thorough clinical examination was done and patient demographic data was filled and Evaluation of Health Related Quality of Life in Hemophilia patients was done by WHO SF-36 scale[10,11].

Outcome variables

SF-36 score was used to measure quality of life at admission and at 6 months from admission also the SF-36 score of normative individual were calculated

SF-36 questionnaire measures health related quality of life on eight dimensions: physical functioning, social functioning, physical and mental role limitations, mental health, energy/vitality, pain and general health perception. Results for each dimensions are scored and transformed on to a scale from 0(worst health) to 100(best score). Results from SF-36 can also be reported as a physical component scale(PCS) and mental component scale (MCS)

Statistical analysis

All the haemophilia patients were analyzed at admission and at 6months from the admission, on scale as mentioned previously. Comparison between mean values of each scale from admission to normative individual using unpaired t-test and from admission to 1st follow up was done using paired t-test.

Results

Eighty- eight patients with haemophilia A(n=80) and haemophilia B (n=8) were enrolled for the study. Out of 88 patients 66 patients were eligible for the study. Out of 88 patients 53 patients had severe haemophilia, 30 had moderate and 5 had mild haemophilia. Mean age of the patient is 21.

Table 1 : Distribution of haemophilia

Haemophilia- A	80
Haemophilia -B	8
Severity -Mild	5
-Moderate	30
-Severe	53
Age , mean(SD)	21(9.9)

Results from the mean SF -36 score and respective summary statistics are shown in table 2. Combining the SF-36 data from the haemophilia and the normative populations all the 8 domains of quality of life in haemophilic patients show p value(<.05) on unpaired t test and are highly significant as compared to normative individuals this shows that all the aspects of quality of life were poor in haemophilia patients as compared to normative individuals.

Physical component score(PCS) and mental component score(MCS) of haemophilia patient compaired with normative individuals using unpaired t-test there was a highly significant difference in the physical component score as well as mental component score(p value <.05). This signifies that haemophilia patients have a poor quality of life as compared to normal individuals in both domains i.e. physical and mental

Table 2 : Comparison of SF-36 different domains in haemophilia patients and normative individuals

SF -36 Components	Haemophilia Patients	Normative Individuals	P*value
PHYSICAL FUNCTIONING	53.61	91.11	0.0001
ROLE PHYSICAL	23.35	93.58	0.0001
BODILY PAIN	49.42	87.66	0.0001
GENERAL HEALTH PERCEPTION	37	79.15	0.0001
ENERGY/VITALITY	53.04	76.93	0.0001
SOCIAL FUNTIONING	57	85.27	0.0001
ROLE EMOTIONAL	57	78.82	0.0001
MENTAL HEALTH	74.51	93.47	0.0044
PCS	24	46.86	0.0001
MCS	27.77	42.49	0.0001

Table 3 : Age wise distribution of SF-36 PCS and MCS

AGE GROUP	GILBERT SCORE	HAEMOPHILIA SF 36 PCS	MCS
11-20	3.77	30.96	29
21-30	4.5	22.6	25.86
31-40	4.67	25.26	29.52
41-50	4.75	20.55	19.24

Discussion

In recent years, several studies addressing quality of life issues in the haemophilia population have been published. However, those which focus on adult patients have tended to use generic quality of life questionnaires, such as SF-36 or EQ-5D, as no disease-specific measures were available. The results of these studies point to impairment in pain, general health, physical role and vitality comparing haemophiliac patients to general population.

However studies into the quality of life of adult patients as a function of haemophilia treatment and integral health care are still rare. Also functional disability in haemophilia is also a very less discussed issue and no study in India yet correlated the functional disability impact on quality of life of patients.

In the present study mean SF-36 scores in haemophilia patients were well below the normative individuals . Similar findings were seen in study by A.H.Miner et al.[12] and C.A.Sabin et al.[13] except for Role Physical and General Health perception which is very low in our study as compared to them. Whereas T.M Brown et al.[8] study also suggests that the most impaired aspect of Health Related Quality of Life were physical domains: physical functioning (PF) and Role physical (RP). We also found that the mental component is more compromised in our patients as compared to other study showing that lack of infrastructure in haemophilia care, lack of awareness and family support to the patient.

In the present study the mean physical component scale (PCS) was 24.77 and mean mental component scale (MCS) was 27.77 , which was well below the general population i.e. PCS is 46.86 and MCS is 42.49. Similar results were found in the other study done by A.H.Miner et al. in the PCS i.e. 30.7 but the MCS is 51.5 which was comparable to the general population i.e. 51.4. In the other study done by C.A.Sabin et al. they found PCS 31.9 and MCS 52.5 which was comparable to the general population i.e. 51.4[12]. Study performed by T.M.Brown et al. suggest the mean patient PCS of 39.9, is well below the general US population of 49.6, whereas the mental component score MCS= 49.2 was comparable with general US norms (49.4). Study done by Gingeri et al.[14] among Italian Haemophilia patients with inhibitors, obtained a PCS of 36.9 and MCS of 50.2. So from the above we can say that haemophilic patients had generally poor Health Related Quality of Life when compared to general population. On comparing with other studies in which physical component score was low but mental component score was comparable to general population. Whereas in our study we found that both the physical component score and the mental component score was low this difference in the mental component score could be due to 1. In the other studies factor is also used prophylactically whereas we only used factor on demand basis and prophylactic use of factor might had a positive effect on the quality of life 2. In the Indian subpopulation that we studied there could be a psychological component such as depression which leads to decrease in the mental component score and quality of life of haemophilia patients.

With the use of SF-36 questionnaire we have shown that compared to the general population, patients with haemophilia A and B who were registered at our centre experienced lower levels of HR-QoL, both in mental component and physical component. However, because these data are cross-sectional they are currently limited in their use and further prospective analysis is needed. Nevertheless, although these results show that these patients currently experience significantly decreased levels of HRQoL, it is hoped that significant improvements in HR-QoL will be detected over time as the effect of free factor availability in our centre.

Conclusion

In our study, there was a significant difference in the health related quality of life between haemophilia patients and the normative individuals in both the physical and mental component of quality of life.

In haemophilia, factor replacement is not the only available treatment but a concomitant rehabilitation programme, community support and psychological support together can make a difference in the quality of life of haemophilia patients

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