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1. Background:

1.1. Haemophilia

Haemophilia A and B are single gene disorders that occur due to mutations in the coagulation factor VIII gene (haemophilia A) or the coagulation factor IX gene (haemophilia B). The gene mutation results in deficient synthesis of coagulation factor VIII or IX, presenting as haemorrhagic tendencies in patients\(^1, 2\). The frequency of bleeding depends on the residual coagulation factor level\(^3\). Patients are classified as having severe, moderately severe or mild haemophilia based on <1%, 1-5% and 5-40% of normal clotting factor levels\(^4, 5\). Replacement therapy with clotting factor concentrate (CFC) is the main treatment for haemophilia. The high cost of this product places it out of reach of a majority of patients from developing nations including India\(^6, 7\). The estimated annual cost of treating a patient with severe haemophilia may be as much as Rs.320 000 ($5000)\(^8\). Sub-optimal treatment results in chronic pain, places the patient at risk of life threatening haemorrhagic episodes, progressive disability due to haemarthrosis\(^7\), and premature mortality. Families incur extensive out of pocket (OOP) expenditures\(^8\). The chronic nature of the disorder affects the quality of life (QOL) and wellbeing of patients and their family members\(^9\).

1.2. The public health challenge of haemophilia in India

Other than haemoglobinopathies, genetic disorders are not considered to be a public health problem in India, as the debilitating nature of these conditions limit the number of patients\(^10\). There is no government funded haemophilia service in the country. This service gap has been filled by non-governmental organization (NGO), the Hemophilia
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Federation of India (HFI), which imports and distributes CFC through 76 haemophilia treatment centres (HTCs) across the country. Recently, this NGO has used judicial recourse to demand subsidized treatment through government health services. In several states of the country, the judiciary has instructed state public health services to provide free treatment, heralding the emergence of government supported haemophilia services in India. India reports 11,586 patients, second only to the United States (13,276)\(^{11}\). It is estimated that only one fourth of Indian patients are reported\(^{12}\). But, India may have an estimated 54,454 patients with haemophilia A\(^{12}\). These evidences suggest the urgent need for a comprehensive haemophilia programme with components of prevention, management of patients, surveillance, education and psychosocial support services for patients and parents.

1.3. Psychosocial support (PSS)

In developed countries, management of haemophilia involves comprehensive care with the aim of achieving the best possible QOL for patients and their family members. Psychosocial support services for parents and caregivers are an integral component of comprehensive care services. The adverse impact of the chronic bleeding affects the QOL of patients\(^{13-16}\). However, fewer studies have documented the impact of haemophilia on the QOL of parents/caregivers\(^{17-19}\). The available studies reveal that parental QOL is affected. Various studies have shown that psychosocial support to caregivers/parents of patients with haemophilia improves their QOL, resulting in a reduction in depression and an improvement in their coping skills\(^{20,21}\).
2. Research gap:

In contrast to developed countries, there is a lack of data on the QOL and psychosocial needs of parents of children affected with haemophilia in India and other developing countries. In these countries, parental distress is likely to be exacerbated by lack of economic ability to purchase CFC for treatment. Psychosocial support interventions are likely to improve the QOL of parents of children with haemophilia. Although regional language brochures providing information on haemophilia are available in India, validated psychosocial support interventions are unavailable. Available interventions from developed nations are inappropriate in the healthcare and socio-cultural context of India.

3. Objectives:

The objectives of the study are:

1. To use peer experiences to identify the psychosocial needs of parents of children affected with haemophilia,

2. To develop a psychosocial intervention tool,

3. To measure the health related quality of life (HRQOL) of parents of children with haemophilia using a validated tool, and

4. To test the impact of the developed psychosocial support intervention on the HRQOL of parents.
4. Materials and Methods:

4.1 Psychosocial needs assessment of parents of children with haemophilia

Needs assessment of parents of children with haemophilia was undertaken through a qualitative study carried out in Pune city, Maharashtra, India. Participants were mothers of children with haemophilia. After obtaining written informed consent, mothers were enrolled sequentially till saturation in responses was noted (n=6). In-depth interviews were conducted in the regional language (Marathi) using an interview guide. Directed content analysis was carried out from the transcribed and translated interviews to assess the psychosocial needs of parents.

4.2 Development of an audio-visual PSS module

A PSS module was developed based on the data derived from the in-depth interviews of mothers, and from available literature. The PSS module was initially developed as a series of PowerPoint slides using Microsoft Office PowerPoint 2007 and reviewed through three separate meetings of stakeholders from the HTCs from Maharashtra, including doctors, physiotherapists, psychiatrists, parents, and patients. Changes were incorporated at all stages and the intervention material was converted into an audio-visual presentation using the Microsoft Windows Movie-maker 2007. This version was reviewed for a final time by invited members from HTCs across Maharashtra and subsequently used for the study.

4.3 Study design, setting and sample

The validated PSS module was used for intervention. Participants were parents of children with haemophilia. The “before and after intervention” HRQOL, knowledge
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about haemophilia and practice of management of haemophilia were measured. The inclusion criteria for participation in the study was parents of children with haemophilia A or B aged 2 to 18 years, with an understanding of the Marathi language, with a telephone/cellular/ mobile phone number and providing consent to participate in the study.

4.4 Intervention

The PSS module was delivered to 133 parents who attended the World Haemophilia Day 2012 celebrations organized at the HTCs; at Pune (n=30), Aurangabad (n=16), Kolhapur (n=32), Nagpur (n=37) and Dhule (n=18) in Maharashtra state.

4.5 Data collection tools for measurement of impact

Data was collected from enrolled parents after obtaining informed consent. Four tools were used for data collection, viz. a socio-demographic questionnaire, knowledge questionnaire, a questionnaire to document the practice of management of a bleeding episode and a translated, validated PedsQL™ Family Impact Module (PedsQL™ FIM) tool to measure the HRQOL of parents. The PedsQL™ FIM tool assesses physical and emotional health as well as social and family relationships, cognitive functioning, communication, worry, daily life and family relationships. The tool has a five point Likert scale to document how frequently parents perceive problems in these eight domains.
4.6 Data collection:

Four types of data were collected at baseline prior to intervention; i) socio-demographic and clinical data, ii) data on knowledge of haemophilia, iii) practice of management of a bleeding episode and iv) HRQOL. After intervention, data on HRQOL and practice of management of bleeding episodes was collected twice, at six-months and one year post-intervention. Knowledge on haemophilia was collected immediately after intervention, and at the end of one year post-intervention. Data was entered in Microsoft Office Excel 2007 and analysed using SPSS (Statistical Package for the Social Sciences) software version 17. Paired t-test was used to determine whether the change in the scores was significant. Standardized mean effect size (Cohen’s d score) was calculated to determine the difference between pre- and post-intervention scores. The effect size was considered as small when the score was less than 0.2, medium when it was between 0.2 and 0.5 and large when it was greater than 0.8\textsuperscript{23}.

5 Results

5.1 Psychosocial needs assessment of parents

Content analysis of the in-depth interviews identified the need for PSS. The major reasons for parental frustrations were high costs of haemophilia treatment, debt due to catastrophic expenditures incurred for treatment, concerns about the availability of CFC, and inability to find a healthcare provider who would be able to infuse CFC. The need for increasing awareness about haemophilia amongst medical practitioners was identified from reports of misdiagnosis of the condition by the practitioners, resulting in delayed
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diagnosis. An overarching theme was the need for sensitive medical communication, as insensitive interaction with parents was responsible for causing hurt, which some parents remembered after many years. Another issue emerging from incomplete medical communication was the need for parents to meet an adult patient, to be reassured about the longevity of their child. A lack of access to information was reflected in all interviews. In lieu of medical communication, information was collected from varied sources. Mothers were more overprotective than fathers. In the background of OOP for treatment, there was an emphasis on educating the child, with the hope that the child would be well employed and capable of paying for his treatment. Marriage of the patient was identified as a future support system for patients in the absence of any form of social services for disabled individuals. The issue of marriage raised concerns about disclosure of haemophilia in case of not only the patient, but also on disclosure of the carrier status of the patient’s sister. Maternal guilt was overarching and manifested as a need to demonstrate acceptance by her husband’s family. Marital discord was observed. Maternal stress was observed as mothers reported inability to concentrate on anything other than the affected child.

5.2 Development of PSS intervention module

A PSS intervention module was developed based on a review of existing global modules and Indian studies on haemophilia. The data from the PSS needs assessment study served to prioritize and contextualize the data for constructing the module.
5.3 Impact of Intervention

5.3.1 Socio-demographic characteristics of participants

Of the 133 parents of haemophilic children recruited for testing the impact of the PSS module, 70 were mothers and 63 were fathers. Majority of parents (57% of mothers and 55% of fathers) were between 31-40 years of age. Seventy percent mothers and 69% fathers had completed up to 12 years of schooling. Most fathers (58%) were farmers and unskilled workers, while 57% mothers were housewives. Majority (47%) of participants belonged to the lower middle income group, and 55% of parents lived in nuclear families.

5.3.2 Knowledge on haemophilia

The mean score for knowledge of haemophilia amongst 133 parents at baseline was 47.7±16.7. This score significantly improved to 95.2±6.5 post intervention, e =0.9 and remained higher than baseline score at 78.5±12.3 one year after intervention, e=0.4. The scores of all five domains improved significantly post intervention (p=0.000) but reduced after one year of intervention as compared to post intervention scores. However, the scores remained significantly higher in comparison to the baseline.

5.3.3 Practice of management of bleeding episodes

Practice of management of bleeding episodes was measured using four indicators; number of bleeding episodes arising from trauma related injuries, use of CFC, use of first aid for home management of bleeding episodes, perceived duration of the bleeding episode and number of school days lost. At the end of one year there was no significant change in number of trauma-related bleeding episodes and use of CFC, although the early
initiation of first aid for management of bleeding episodes increased at six months and remained significantly increased after one year of intervention. There was no significant reduction in number of school days lost at the end of one year period.

5.3.4 HRQOL

The PedsQL™ FIM tool was validated before it was used to measure the impact of the PSS intervention on the HRQOL of participants. Data on HRQOL and practice of management of bleeding episodes was collected thrice, at baseline before intervention, six months after intervention and one year after intervention, while data on knowledge about haemophilia was collected before intervention, immediately after intervention and one year after intervention. The mean PedsQL™ FIM score for 133 participants was 52.4±9.6 at baseline. This score significantly improved to 63.6±10.2 (p<0.05) with an effect size of 0.5, six months after intervention. The score significantly reduced to 55.9±10.1 (p<0.05) one year after the intervention.

6 Discussion:

This study is the first from India and from a low and lower middle income country to test the impact of a PSS intervention module on HRQOL, knowledge and management of bleeding episodes by parents. Although several PSS and knowledge modules for parents are available, these are not relevant to the healthcare and socio-cultural milieu of haemophilia in a developing country like India. As numerous studies in other developed nations had shown that PSS can improve the QOL of parents, this study was developed and delivered a PSS intervention to parents of children with haemophilia in Pune, Maharashtra. This study identified that parents from developing countries have similar
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cpsychosocial needs as parents of children with haemophilia from the developed world, possibly arising from the chronic nature of the condition. The health and social support systems were, however, major determinants of parental psychosocial need arising primarily from the lack of financial ability to provide treatment for the bleeding episodes of haemophilia. The study identified the need for increasing awareness amongst parents about the use of CFC and when it could not be purchased, the need for correctly used first aid for managing bleeding episodes. The study also identified the need to increase awareness on haemophilia amongst medical providers, and training of health care providers on the need for sensitive communication.

The study showed that after providing the PSS, the HRQOL improved at the end of six months of intervention, but reduced to baseline level at the end of one year, post-intervention. However, Knowledge on haemophilia, and practice of management of bleeding episodes remained higher than baseline after one year of intervention. The results of the study indicated that PSS is capable of increasing knowledge and improving the practice of home management of bleeding episodes using first aid. PSS was however not effective in improving HRQOL, as possibly the basic issue, that is lack of access to CFC, continued to remain unaddressed during the study.

7 Utility of the study:

The results of this study show that a PSS module contextual to the health and social situation can improve knowledge and empower parents in appropriate usage of first aid till the parents reach the HTC for treatment. This PSS delivered by a counsellor, can be an integral part of a national haemophilia program. The study underlines that although the
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PSS intervention could increase the parental knowledge, the provision of CFC remains the essential element for a public health programme for haemophilia in India.