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Impact of Haemophilia -A Disease on Social Life, Education and Employment of Caregivers and Patients

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ABSTRACT

Objective: To determine the impact of Haemophilia-A disease on social life, education and employment of caregivers and patients

Study Design: Cross-sectional study

Place and Duration of Study: Hemophilia Patients Welfare Society Centre Rawalpindi, Pakistan from Jan 2020 to Jan 2022.

Methodology: A total of 80 couples who were parents to Haemophilia -A patients were recruited as subjects. After an informed consent, the couples were interviewed utilizing predesigned questionnaire comprising questions on basic demographic variables, education, monthly income, social life, employment and child education. The data were analyzed using Statistical Package for Social Sciences (SPSS) version 22.0.

Results: A total of 55(68.75%) male parents had minimum qualification of matric or higher education in comparison to 49(61.25%) female parents who have achieved similar qualification. Families having Monthly income below 20000 rupees, between 20000-40000 rupees and above 40000 rupees were 24(30%), 48(60%) and 8(10%) respectively. A total of 74(95%) families believe that they are experiencing severe financial burden due to disease of their child. The family life of couples was found to be mildly affected in 4(5%) families, moderately affected in 11(13.75%) families and severely affected in 65(81.25%) families. None of parents were found to be satisfied with their child's educational performance.

Conclusion: The Haemophilia-A disease has a negative impact on the education, social life and employment of caregivers and patients. Moreover, Haemophilia-A disease adds to financial burdens of parents due to additional financial expenses on management of disease.

Keywords: Employment, Haemophilia-A, Social life

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INTRODUCTION

Haemophilia-A disease is a X- linked recessive disorder and among the commonest coagulation factor deficiency. The incidence of the disease is 1 in 5000 live births approximately.[1] This disease is transmitted by the mothers provided they are carriers of disease to their male off springs. The management of the disease is difficult, costly and lifelong. The disease leads to many complications and has significant morbidity and mortality due to arthropathy and life threatening bleeds.[2]

Parents or caregivers have to face many challenges during management and care of their child with Haemophilia. This puts both parents and patient under multiple psychosocial strains that compromise their education, employment, monthly income, social life and family life.[3-4] Ultimately both parents and patient have compromised quality of life. Those patients who have developed complications of the disease or inhibitors to replacement therapy experience more difficulties and compromise in all walks of life. According to world federation of Haemophilia, a total of 195263 cases have been reported till 2019 from 115 countries across the globe and 2233 cases have been registered from Pakistan. According to study conducted by cutter et al.,[5] on American population of Haemophilics, 95% and 94% Haemophilics had a negative impact of disease on their education and employment respectively.

Pakistan is a developing country where Haemophilia–A disease can have severe financial and social impact on the parents and caregivers. This study aimed to analyze the effects of disease on education, social life, family life, and employment of parents and patients.

METHODOLOGY

This cross sectional study was conducted at Hemophilia Patients Welfare Society Center

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Rawalpindi, Pakistan from Jan 2020 to Jan 2022. A sample size of 73 was calculated by using WHO sample size calculator keeping confidence level of 95%, absolute precision of 0.05 and anticipated population proportion of 0.95 of Haemophilic patients.[5] However, it was decided to include 80 Haemophilic families. Haemophilic patients were enrolled at Hemophilia Patients Welfare Society (Rawalpindi chapter) after confirmation of their coagulation results and factor VIII levels. They were enrolled with Hemophilia Patients welfare society for provision of recombinant factor VIII concentrates, management of disease and regular follow up. Ethical approval was granted by institutional ethical review committee of Riphah International University, Islamabad, Pakistan (Ref # Riphah/IRC/19/0362). Sampling technique employed was non probability purposive sampling.

Inclusion Criteria: Male Children irrespective of age group, who were diagnosed cases of Haemophilia–A disease on coagulation assay and factor VIII levels were enrolled. Couples who were parents to a child suffering from Haemophilia-A were also enrolled in the study.

Exclusion Criteria: Patients having bleeding disorder other than Haemophilia-A were excluded. As Haemophilia-A is a rare bleeding disorder, we took all the available patients in our study duration.

All the eighty enrolled couples were requested for participation in a survey. They were assured about confidentiality of their personal views and not to disclose their identity. The interview was carried out by a qualified doctor in a separate room. The national language "Urdu" was medium of conversation for conducting interviews but the responses were noted in English on survey form by the doctor. The participants were encouraged to ask any relevant questions during survey to address ambiguity regarding questions of survey, the disease or any other reservations. They were also given choice to refuse to answer any survey question and could show their unwillingness for participation in survey at any stage of interview.

The participants were interviewed on a predesigned questionnaire comprising questions on basic demographic variables, education, monthly income, social life, effect of disease on their family life, employment, effect of disease on their financial status, patient education and effect of disease on patient education. The questionnaire of the study was pilot tested and face validated before commencing the actual data collection procedure. All those participant families who had not received genetic counseling in the past were considered as Group-A and those with previous genetic counseling were regarded as Group-B for statistical analysis of the survey.

Statistical Package for Social Sciences (SPSS) version 22.0 was used to analyze the data. Frequencies were calculated for categorical data while mean and standard deviation (SD) were calculated for continuous normally distributed data. To compare qualitative variables, Chi-square test was applied and *p*-value of 0.05 was considered significant.

RESULTS

A total of 80 families were studied. The number of children per family ranged from two to seven. Each family had three children on an average. The mean age of the Haemophilic child was 9.23±3.76 years and it ranged from three to 18 years. There were three families who had more than one Haemophilic child. A total of 59(73.75%) families (Group-A) had not received genetic counseling in the past regarding the disease whereas 21(26.25%) families (Group-B) had received genetic counseling in the past. The educational, family income, social life and job employment analysis of studied population are shown in Table-I.

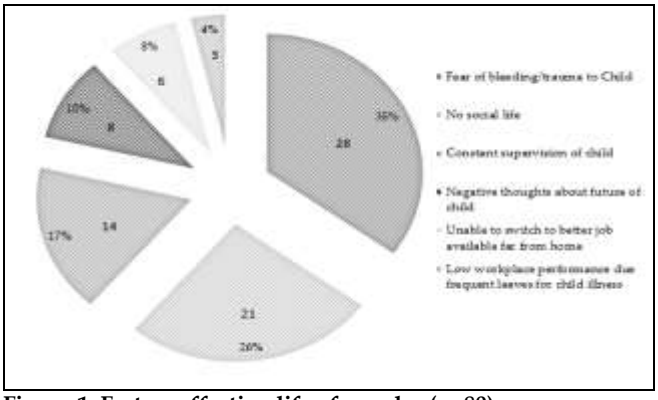

Figure-1: Factors affecting life of couples (n=80)

A total of 4(5%) families reported that they sometimes feel financially hard pressed due to additional financial expenditure on their affected child care. Whereas 74(95%) believe that they are experiencing severe financial burden due to medical management and care of their child. The family life of couples was found to be mildly affected in 4(5%) families, moderately affected in 11(13.75%) families and severely affected in 65(81.25%) families. Factors affecting life of couples due to Haemophilic child were

also evaluated. The results are shown in figure-1. Out of 80 Haemophilia-A patients, 5(6.25%) were in preschool age and remaining 75(93.75%) were studying in different grades in school and colleges. Parents were asked about that educational performance of their child. None of parents was found satisfied with their son's performance. They quoted different reasons for low educational performance which are summarized in Figure-2. Parents also reported low educational performance of their Haemophilic child in comparison to normal siblings of the patient.

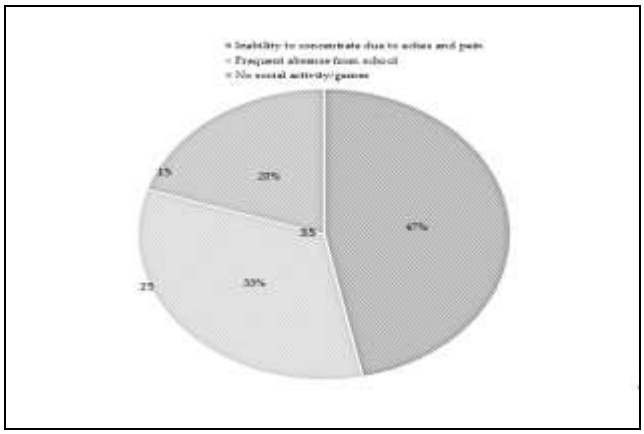

Figure-2: Factors affecting Educational Performance in Haemophilic Patients (n=75)

DISCUSSION

Haemophilia-A is one of the commonest inherited bleeding disorders. It is a single gene disorder resulting in deficiency or reduced plasma level of factor VIII. Phenotypic severity is characterized by factor VIII plasma level: severe phenotype having plasma levels of <1%, whereas moderate and mild phenotypes having plasma levels of 1-5% and 5-30% respectively.[6]

In this study, the fathers of Haemophilia-A patient were found to be more educated in comparison to mothers of Haemophilic patients. A total of 55(68.75%) fathers have minimum completed their Matric or higher education in comparison to 49(61.25%) mothers who have achieved similar qualification. The majority of families belong to low socioeconomic status. The monthly income was found to be in upper middle economy poverty scale (US $ 5.5 per day) in 48(60%) families. Similarly, 24(30%) families were found to be falling in Lower middle economy poverty scale (US $ 3.2 per day). Haemophilia -A disease requires high cost on the management of the patients. Moreover, marked inflation during and post COVID19 pandemic has created additional financial difficulties for all and especially for Haemophilia-A families in lower or upper middle economy poverty scale. This additional financial burden due to Haemophilic patients' management on parents was found to have a severe effect on financial situation in 76(95%) families in this study. Transportation cost was also found as a major non-medical expense in Haemophilia -A management in studies conducted in Turkey and Iran.[7,8]

Table-I: Education, Family income, Social life and Employment Analysis of Studied Population (n=80)

Parameters	Study Groups		*p*- value
	Group A (Without Genetic Counseling) n=59 n(%)	Group B (With Genetic Counseling) n=21 n(%)	
Education of Mother			
Matric & below Intermediate & Graduate	43(72.88) 16(27.12)	14(66.66) 7(33.33)	0.29
Education of Father			
Matric & below Intermediate & Graduate	41(69.5) 18(30.5)	14(66.66) 7(33.33)	0.057
Family Income			
Below Rs.40,000 Above Rs.40,000	52(88.13) 7(11.87)	20(95.23) 1(4.67)	0.86
Family & Social Life			
Mild - moderately effected Severely effected	12(20.34%) 47(79.66%)	5(23.80%) 16(76.20%)	0.11
Employment			
Negative impact on employment No impact employment	55(93.22%) 4(6.78%)	20(95.24%) 1(4.76%)	0.10

Haemophilia -A disease also has an impact on parents or caregivers' life which can result in low quality of life due to stress, anxiety, financial strains, cancellation of social plans, mental and physical readiness for bleeds. They face challenging task to give their child normal life experience but at same time minimizing risks of harmful bleeds.[9-12]

In this study having a child with Haemophilia severely affected family life of 63 (78.7%) families. The most frequent factor reported by 28(35%) families which has affected their family life was the fear or anxiety of spontaneous bleed or trauma to their child. In our cohort, the mothers were the primary Caretaker of child since they spent more time with the child. This helped mother to gain experience regarding how much activity her child could do. But this experience and confidence quickly vanishes as soon as bleed occurs. The risk of a bleeding episode will remain forever and each episode of bleeding will have a setback for parental confidence. To cope up with this stress/challenge, support from family, friends, employers and Haemophilia societies is utmost important.[13]

Parents of Haemophilia patients have to restrict their lifestyle and activities. This modification of lifestyle change is greater for male parents in comparison to female parents.[14] According to a study conducted by Recht et al. bleeding episodes can disrupt planned activities of parents in 40% of the affected days.[15] The interference with planned activities of family is greatly increased with duration and number of bleeding episodes, joint bleeds and in families having Haemophilic child with inhibitors. In our study around 21(26%) of families feel socially isolated and have limited social life. In a study conducted by Recht *et al*.,[15] social isolation was also reported as one of the major impact factors of caregivers/parent's burden and around 23.3% families socialize less with friends due to Haemophilia.

Haemophilia can also have an impact on unaffected siblings in addition to parents as they receive less time and attention from parents. At the same time, some parents feel that Haemophilia has positive impact on siblings, and they have become more responsible and mature. In addition, this disease has brought their family closer and are satisfied from their family support.[16] A study from United States of America concluded that in 43.3% versus 8.8 % caregivers of patients with and without inhibitor respectively believe that they had frequent to nearly always no time to relax, socialize and look after themselves.[17] In our study 14(17%) families report that their personal relaxation time is affected due to constant supervision of child especially during his infancy and early childhood.

Around 8(10%) families in our study, especially mothers have negative feeling and thoughts about future of their child. In a study conducted by Wiedebusch *et al*.,[18] it was reported that having concerns and thoughts about the future can negatively affect quality of life of parents.

In our study 9(11.25%) couples face difficulties in switching to better job available in other city or in managing good workplace performance. According to study conducted by Forsyth *et al*.,[19] around 63% parents had negative impact on their job experience due to Haemophilia. Around 29% parents selected their job taking into consideration disease of their child, whereas 24% opt for a job with flexible working hours. Other studies reported that employment of caregivers/parents can be affected by missing working hours due to bleed in patient, working less than full time and demanding concession at work due to condition of their child.[20] In our study 75(93.75%) patients with Haemophilia were attending school and college according to their age. The most frequent reason for low educational performance reported by parents is the inability to concentrate due to aches and pain. This problem is more pronounced in patients having arthropathy. In a study done by Cutter al.[5] it was reported that 69% of Haemophilic students had a negative impact on their education owing to inability to concentrate due to pains and bleed.

In this study frequent absence from school was reported as second most common factor of low educational performance in patients. In 25(33.3%) families, absence from school was either due to spontaneous bleed, follow up visits to treatment centre or due to severe pain. Another reason for negative impact on education is no co-curricular, social activity and games. Since most parents don't allow their child to have physical activity and contact sports both at home and in school to avoid trauma. Moreover, children with arthropathies are also unable to participate in physical activities. The school staff should be educated about Haemophilia and be informed about immediate care if child develops bleed.

There is a strong need of an integrated and multidisciplinary approach for care of the patient of Haemophilia and support to families of patients. During management of the patient, parents have to undergo a lot of stress and it necessitates the importance of provision of psycho-educational help to cope up with stress and improve quality of life. Government and non-governmental organizations should formulate a policy for financial help and

provision of jobs to Haemophilic families on priority basis.

LIMITATION OF STUDY

The participants of the study were resident of the Punjab province only.

CONCLUSION

The Haemophilia-A disease has a negative impact on the education, social life and employment of caregivers and patients. Moreover, Haemophilia-A disease adds to financial constraints of parents due to additional financial expenses on management of disease.

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Authors' Contribution

Following authors have made substantial contributions to the manuscript as under:

MAS & SA: Conception, study design, drafting the manuscript, approval of the final version to be published.

SA & ST: Data acquisition, data analysis, data interpretation, critical review, approval of the final version to be published.

AE: Conception, data acquisition, drafting the manuscript, approval of the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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