

ORIGINAL ARTICLE *Clinical haemophilia*Parents of children with haemophilia at an early age:
assessment of perceived stress and family functioning

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Summary. Haemophilia is a chronic disease that requires a multidisciplinary approach for proper management and control of its clinical manifestations. The perception and management of parents of children with haemophilia can be affected by stressful situations as a result of treatment or disease progression. The aim of this study was to evaluate the perception of stress and family functioning in parents of children with haemophilia 1–7 years. This is an observational clinical study involving 49 parents of children with haemophilia 1–7 years who attended the VIII Workshop for Parents of Children with haemophilia, organized by the Spanish Federation of Hemophilia in La Charca, Murcia (Spain). After obtaining parental consent, the questionnaires was applied to them, FACES III (family functioning) and Pediatric Inventory

for Parents (perceived stress), and a record of data on the clinical characteristics and treatment. Significant differences in the perception of stressors by gender of parents were found. A family history of haemophilia, the use of port-a-cath, inhibitor development and gender of the parents were the descriptive variables most correlated with dependents variables. These variables, together with the type of haemophilia affect significantly in the parental stress and family functioning. Parents have difficulty adjusting to disease management, perceiving many stressors. Gender and family history, can hinder the proper compliance with treatments, reducing its effectiveness.

Keywords: early age, family functioning, haemophilia, parents, stress

Introduction

Haemophilia is a congenital, hereditary disease in which there is a deficit of clotting factor VIII (haemophilia A) or IX (haemophilia B) [1]. It is recessive and is transmitted from mothers to sons.

Prophylactic treatment of these patients, especially in children, has been shown to be effective in preventing or delaying of haemophilic arthropathy [2,3]. However, pharmacological treatment of haemophilia is a necessary, but not sufficient condition for ensuring the patient's well-being. A comprehensive approach that includes the family is essential, as diagnosis is early and the patient cannot take control of the disease. The study of family factors and of styles of coping with the disease is of special interest for a mul-

tidisciplinary approach to patients with haemophilia (PwH) [4,5].

Several studies [6–8] have examined the quality of life as perceived by young and adult PWH, but hardly any published studies [9,10] have investigated how the disease affects the parents of children with haemophilia (CwH), especially when recently diagnosed.

Haemophilia, such as chronic disease, can affect the emotional state of parents [6,11] as it is an incurable disease which manifests itself at an early age, and is genetically transmitted. This latter characteristic can cause feelings of guilt, especially in mothers [9,12]. In addition, many of its clinical signs and symptoms are spontaneous and cause pain and suffering, thus resulting in some disruption to the family environment and the child's routines, impacting on the whole family's quality of life [13]. The fact that infectious diseases such as HIV and HCV were transmitted to PWH in the 1970s and 1980s generates anxiety and fear in relatives who fear that similar situations may arise [14].

One of the first studies of families with haemophilia, Saviolo-Negrin *et al.* [10] observed stress, tendency to depression, feelings of guilt and social maladjustment

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in a group of 42 parents of CwH. Other studies [15,16], observed these feelings and gender differences: how mothers of children with disabilities had high levels of depression and also more difficult to adapt to the situation than their spouses.

Other studies [17–19] have evaluated the perception of disease and the stress suffered by adult PWH, but few articles have given much weight to how haemophilia affects parents' family functioning after the birth of a child with this disease [9,13].

A proper assessment of family functioning and the stress perceived by parents can promote more effective adaptive coping strategies, early intervention to reduce the complications of the disease and better adherence to treatment. The aim of this study was therefore to assess the perception of family functioning (through the degree of cohesion and adaptability) and the degree of stress experienced by parents of CwH aged between 1 and 7.

Material and method

This descriptive and correlational study was carried out during the VIII Workshop for parents of CwH an early age (from 1 to 7 years), organized by the Spanish Federation of Haemophilia at the 'La Charca' Lifelong Learning Centre, in Murcia. The number of parents attending the Workshop was 49 and 26 children: 26 mothers (three attended alone: one was divorced and two their husbands for work they could not attend), eight were potential carriers, the rest were cases *de novo*, and 23 fathers. The researchers informed about the aims of the study, and the participants signed the informed consent document, in accordance with the Helsinki standards (1975).

The sample was made up of 26 women (53.1%) and 23 men (46.9%). The average age was 38.86 (SD 3.26).

The average age of the children was 4.02 (SD 2.01). Of the children of the subjects in the sample, 87.75% had severe haemophilia; 81.63% had haemophilia type A; 30.61% had a family history of haemophilia; 16.3% had inhibitors; 55.1% had a history of haemarthrosis; and port-a-cath had been implanted in 30.61% of them.

Table 1 lists all the descriptive variables for the subjects in the sample.

To assess the parents' family functioning and their perception of stress, we used the Spanish versions of the following questionnaires:

1. Family Adaptability and Cohesion Evaluation Scales (FACES III) [20,21]. This questionnaire is a scale developed to assess family functioning. It consists of two dimensions: cohesion and family adaptation. The final result is a total family functioning score. Olson [20] provides 16 types of family, based on the possible combinations of four types of cohesion (detached, separated, connected and enmeshed) and four types of adaptability (chaotic, flexible, structured and rigid). These types can be grouped into three ranges: optimal family functioning, family functioning intermediate and extreme or dysfunctional family functioning)
2. Pediatric Inventory for Parents (PIP) [22]. This questionnaire was used to assess the parents' stress levels through eight subscales that measure the frequency of stressors and the intensity of the stress caused by communicating, medical care, emotional distress and family role.

In addition, a total score was obtained for the frequency of stressors and a total score for the intensity of stressors.

Statistical analysis

The statistical analysis was performed using the Statistical Package for Social Sciences (SPSS) programme, version 19.0 for Windows (Brush Prairie, WA, USA). A descriptive analysis was made of the subjects in the study. Student's *t*-test for related data was used for analysing gender-based differences, and Pearson's correlation coefficient was used to evaluate correlations between the variables in the study. Linear regression analysis was performed to assess the influence of descriptive variables on the set of dependent variables.

Results

Perception of stress and family functioning

According to Olson's model [20], our sample is within the intermediate range of family functioning, which implies a cohesion enmeshed and flexible adaptation. This leads a dependency between their members, lack of autonomy of its members, loss of resources and capacity of reaction to stressful situations.

Significant gender-based differences were found for the following dependent variables: the frequency of stressors in communication ($P = 0.035$), in emotional distress ($P = 0.040$), total stressors ($P = 0.033$) and

Table 1. Descriptive characteristics of participants ($n = 49$) of the study.

Variables	Mean \pm SD (range)
Age of father/mother (years)	38.86 \pm 3.26 (32–46)
Age of the child (years)	4.02 \pm 2.01 (1–7)
Number of children	1.32 \pm 0.47 (1–2)
	<i>n</i> %
Gender of parents (Male/Female)	(23/26) (46.9/53.1)
Type of haemophilia (A/B)	(40/9) (81.6/18.4)
Severity of haemophilia (Severe/Moderate/Mild)	(43/1/5) (87.8/2/10.2)
Type of treatment (Prophylaxis/On demand)	(26/23) 53.1/46.9)
Inhibitor (Yes/No)	(8/41) (16.3/83.7)
Port-a-cath (Yes/No)	(15/34) (30.6/69.4)
Family history of haemophilia (Yes/No)	(15/34) (30.6/69.4)
Previous haemarthrosis (Yes/No)	(27/22) (55.1/44.9)

%, percent; *n*, number of parents; SD, standard deviation.

the effort to cope with stress in medical care ($P = 0.035$).

Mothers scored higher and presented more stress overall. However, no significant differences were observed between fathers and mothers for the family functioning variables.

Table 2 shows the results obtained from the questionnaires for the whole of the sample of parents and also the differences between mothers and fathers.

Correlations analysis

Table 3 shows the correlations obtained for the whole of the sample of parents. The gender variable was correlated with the frequency of stressors in communication, in emotional distress, in total stressors and with the effort to cope with stress caused by medical care.

Family history was correlated with the effort to cope with stress caused by medical care and with frequency of stressors in the family role.

The presence of inhibitors correlated with the effort to cope with stress in emotional distress. Finally, the presence of a port-a-cath correlated with family adaptation and overall family functioning, and with the effort to cope with stress in medical care (control and management disease).

Regression analysis

Table 4 shows the results of the regression analysis. The gender variable influences in the frequency of

stressors in communication ($P = 0.026$), in emotional distress ($P = 0.040$), in total frequency of stressors ($P = 0.033$), and in the effort to cope with stress caused by medical care ($P = 0.023$) and in emotional distress ($P = 0.036$).

The child's type of haemophilia influences the frequency of stressors in communication ($P = 0.037$), whereas the presence of an inhibitor affects the effort to cope with stress caused by emotional distress ($P = 0.019$). Family history influences the frequency of stressors in the family role ($P = 0.043$) and the effort to cope with stress caused by medical care ($P = 0.033$). The use of a port-a-cath influences the effort to cope with stress caused by medical care ($P = 0.038$).

Discussion

From an early age, PWH face multiple stress-producing situations [22] such as intravenous medical treatment, the pain, the development of physical sequelae, school absences and uncertainty about possible bleeding.

The importance of an integrated and multidisciplinary approach to haemophilia implies that not only the patients but also their family environment must be cared for, as it is initially the family that faces up to and resolves all the issues raised by the disease. Saviolo-Negrin *et al.* [10] indicated that a specific knowledge of the parents' stress level will make it possible to adapt effective and efficient psycho-educational

Table 2. Mean \pm standard deviation (and range) of the variables of family functioning and stress perception of the 49 parents who participated in the study and differences in the variables according to gender.

Variable	Males ($n = 23$)	Females ($n = 26$)	All parents ($n = 49$)	Range/ Percentile	Sig.
Family cohesion	41.29 \pm 4.92 (33–48)	43.88 \pm 3.05 (39–48)	42.33 \pm 4.76 (25–50)	36–40	0.160
Family adaptation	26.57 \pm 5.36 (15–37)	28.06 \pm 4.99 (21–40)	27.14 \pm 5.30 (15–40)	25–32	0.659
Family functioning	67.86 \pm 7.69 (50–79)	72.53 \pm 6.20 (63–83)	69.67 \pm 8.15 (43–85)	60–80	0.203
Subvariables					
Relationships with children	12.13 \pm 3.50 (2–18)	12.12 \pm 2.73 (7–20)	12.12 \pm 3.08 (2–20)	10–18	0.987
Emotional unit	13.13 \pm 1.74 (9–15)	13.31 \pm 1.82 (9–15)	13.22 \pm 1.77 (9–15)	12–15	0.731
Family commitment	18.61 \pm 2.55 (10–22)	19.81 \pm 3.00 (12–25)	19.24 \pm 2.84 (10–25)	15–21	0.142
Family creativity	12.09 \pm 1.75 (9–15)	11.88 \pm 2.28 (7–15)	11.98 \pm 2.03 (7–15)	8–14	0.732
Responsibility	4.91 \pm 2.29 (2–8)	5.00 \pm 2.40 (1–10)	4.96 \pm 2.32 (1–10)	3–7	0.898
Adaptability to problems	8.04 \pm 1.43 (6–11)	8.54 \pm 2.26 (3–14)	8.31 \pm 1.91 (3–14)	6–12	0.373
Frequency of stressors in communication	21.36 \pm 3.41 (14–27)	23.24 \pm 4.36 (16–33)	22.12 \pm 3.80 (14–33)	45	0.035
Effort to cope with the stress in communication	18.79 \pm 6.31 (11–29)	20.00 \pm 5.90 (12–30)	19.78 \pm 5.68 (11–30)	70	0.348
Frequency of stressors by medical care	24.86 \pm 7.37 (11–35)	26.59 \pm 4.12 (16–35)	24.61 \pm 6.12 (11–35)	50	0.123
Effort to cope with the stress caused in medical care	14.21 \pm 4.49 (7–23)	17.76 \pm 6.91 (9–34)	16.76 \pm 6.20 (7–34)	70	0.035
Frequency of stressors in emotional distress	35.07 \pm 8.55 (18–53)	37.82 \pm 8.12 (24–58)	37.08 \pm 8.26 (18–58)	45	0.040
Effort to cope with the stress in emotional distress	37.14 \pm 8.74 (25–54)	40.94 \pm 11.26 (20–61)	40.33 \pm 10.77 (20–63)	55	0.068
Frequency of stressors in the family role	22.36 \pm 4.60 (12–30)	22.76 \pm 5.52 (12–33)	22.47 \pm 5.09 (12–33)	60	0.587
Effort to cope with the stress in the family role	22.79 \pm 5.88 (15–33)	22.71 \pm 6.58 (13–35)	22.90 \pm 6.19 (13–35)	55	0.843
Total frequency of stressors	102.43 \pm 18.28 (55–127)	110.41 \pm 20.15 (70–146)	105.94 \pm 18.76 (55–146)	45	0.033
Effort to cope with total stress	93.43 \pm 21.58 (65–136)	100.82 \pm 26.30 (59–154)	99.69 \pm 23.99 (59–154)	55	0.154

n, number of subjects; Sig, signification; Range: measurement of the variables and subvariables of the FACES III questionnaire of family functioning; Percentile: measurement of the variables of the PIP questionnaire of stress perception.

Table 3. Correlations of the dependent variables of the study with the descriptive variables of subjects ($n = 31$).

Variables	Gender	Severity of haemophilia	Family history	Number of children	Presence of an inhibitor	Use of a port-a-cath	History of haemarthrosis
Family cohesion	0.204	0.221	-0.085	0.186	-0.016	0.140	0.120
Family adaptation	0.065	0.098	0.069	0.026	0.075	0.288*	0.038
Family functioning	0.185	0.183	-0.043	0.110	0.051	0.286*	0.072
Frequency of stressors in communication	0.302*	-0.021	0.080	-0.021	-0.249	-0.143	-0.018
Effort to cope with the stress in communication	0.137	-0.062	0.076	0.098	0.198	-0.223	-0.110
Frequency of stressors by medical care	0.223	0.007	0.191	-0.146	-0.247	-0.079	0.105
Effort to cope with the stress in medical care	0.302*	-0.110	0.298*	0.069	-0.063	-0.293*	-0.071
Frequency of stressors in emotional distress	0.294*	0.009	0.142	-0.084	0.058	-0.026	0.021
Effort to cope with the stress in emotional distress	0.263	-0.064	0.091	-0.028	0.303*	-0.096	-0.012
Frequency of stressors in the family role	0.079	-0.120	0.290*	-0.023	0.019	-0.184	-0.092
Effort to cope with the stress in the family role	-0.029	-0.043	0.119	-0.085	0.218	-0.098	-0.192
Total frequency of stressors	0.305*	-0.024	0.208	-0.083	-0.108	-0.088	-0.006
Effort to cope with total stress	0.207	-0.082	0.165	-0.011	0.223	-0.194	-0.103

*Correlation is significant at the 0.05 level (bilateral).

Table 4. Linear regression analyses of the dependent variables assessed in the study with the descriptive variables of the subjects.

Descriptive variables	Dependent variables	P	β
Gender of parents	Frequency of stressors in communication	0.026	0.309
	Effort to cope with the stress in medical care	0.023	0.302
	Frequency of stressors in emotional distress	0.040	0.294
	Effort to cope with the stress in emotional distress	0.036	0.290
	Total frequency of stressors	0.033	0.305
Family history	Effort to cope with the stress in medical care	0.033	0.283
	Frequency of stressors in the family role	0.043	0.290
	Effort to cope with the stress in medical care	0.038	-0.276
Port-a-cath	Effort to cope with the stress in medical care	0.038	-0.276
Presence of an inhibitor	Effort to cope with the stress in emotional distress	0.019	0.328
Type of haemophilia	Frequency of stressors in communication	0.037	-0.289

β , typified coefficient Beta; Sig, significance.

programs to their needs, and consequently improve the quality of life both for the parents and for the children.

Perception of stress

In our study, we observed that parents perceive a degree of stress (both frequency and effort for coping) is within the average population. The majority of the scores on the various subscales and total scores for the PIP were around the 50th percentile, except for aspects of communication and medical care, which were in the 70th percentile.

The descriptive variables that correlated with stress are the gender of the parent, family history, presence of inhibitors and port-a-cath. Mothers of CwH compared with fathers show more often stressors in communication, emotional distress and the total frequency of stressors. Also, they recognize make more effort to cope with the stress in medical care and distress emotional. Family history correlated with the effort to cope stress in medical care and frequency of stressors

in the family role. The presence of inhibitors correlated with effort to cope with the stress on the emotional distress, and the presence of a port-a-cath correlated with the effort to cope with stress in medical care.

Perrin *et al.* [23] noted as the parental stress increases the child's stress, and may affect the frequency of bleeds, especially those associated with accidents, with the risk that this implies for PwH. In our study, we found a high incidence of haemarthrosis (55.1% of the children had already suffered some haemarthrosis), but did not find any stress-related variable that was affected by the number of haemarthroses suffered. In fact, with the exception of the use of a port-a-cath and the presence of inhibitors, which have been significant in the measurement of stress measured, the other variables are no related to clinical aspects. Thus, it seems appropriate that from the beginning of the diagnosis, we dealt with parents not only the clinical aspects but also others aspects that can influence the way to control the disease and cope successfully by these.

Perception of family functioning

As soon as the diagnosis is confirmed, it would be appropriate begin a process of adaptation. Provide parents resources and coping strategies to facilitate a good adjust to the situation of illness [22,23]. Other relevant element is the family functioning (inevitably affected by the presence of a disease). In our study, it was measured by the degree of cohesion and adaptability. Cohesion is the emotional bond between family members and how this affects in family role and decision-making; adaptability is the ability of the family to change under circumstances as they arise along the life cycle. Knowing these dimensions promote their understanding and impact on homeostasis and family functioning [21]. Extreme scores in both dimensions are related to dysfunctional families and moderate levels are related with better family functioning [24].

The results obtained on the FACES III show that family functioning of parents is within the intermediate range, according to the model of Olson. The dimensions assessed: a degree of cohesion (enmeshed) and adaptation (flexible), promotes a stable family functioning because both are offset to make it so. However, we must be cautious; its adaptation seems to be appropriate to adapt to changes and to resolve problems in a satisfactory manner when they arise. However, its cohesion is excessively enmeshed, confusing limits between their members and disconnection with the outside. This leads a waste of resources and ability to react to stressful situations, which can hinder decision-making when it comes to cope the disease. Of the descriptive variables measured, the presence of a port-a-cath was the only one that correlated with adaptability and general family functioning. That is to say, the use of this venous access device helps parents to better manage the treatment and can be tailored more easily to the illness of his son.

Regression analysis

Linear regression analysis clearly showed that there are four descriptive variables that influence the perception of stress in parents of a child with haemophilia: gender, presence of inhibitors, family history and the use of a port-a-cath.

The gender variable influenced up to five dependent variables: the parents perceive many stressors in communication (9.54%), emotional distress (8.64%) and total frequency of stressors (8.3%). Furthermore, they recognize to make a great effort to cope with the stress in emotional distress (8.41%). Mothers suffer more stress than fathers, probably due to the fact that they are who are more involved in the child's disease. Also, the 'feeling of guilt' by the heritability of the disease may influence them feel more responsible and cope with the disease in a different way.

This is consistent with previous studies [9,10,12,25], who reported that mothers are usually concerned with the control and management of the disease, and therefore they suffer more anxiety and depression states than the fathers. We think this is one aspect to consider, and try as far as possible, to make it work more shared by both parents, and thus, to promote family functioning and good adaptation to the situation.

In our study, we observed the appearance of inhibitors only affects the effort to cope with the stress on the emotional distress (10.75%). In the same line, other authors [12,24,26], noted how arthropathy and inhibitors may cause fear, isolation, depression or aggression in the parents.

Another relevant factor is the family history of the disease. They give rise to stress factors in the family role (8.41%) and generate a great effort to cope with the stress in medical care (8%). In our sample, we observed that the percentage of parents with a family history of the disease is small compared to the *de novo* cases (31% vs. 69%). Even so, we found that this circumstances leads to many problems to face the child's medical care and many stress factors in the family role. This could be related to the degree of enmeshed cohesion shown by parents, which affects family functioning. Moreover, it suggests that the prior knowledge of the disease does not provide their coping: is not the same a brother or a father that his own son.

The use of a port-a-cath affects the effort to cope with the stress in medical care (7.61%). Some studies [14,27] have indicated that the use of a port-a-cath is a relief to parents of CwH, reduces dependence on the hospital, decreases the anxiety caused to the child by intravenous infusion of clotting factor, and allows for a faster response to emergencies. Our results, however, are similar to those described by Furmedge *et al.* [14], where parents despite recognizing the benefits it brings, they also admit to be afraid and anxiety of not knowing how to use it and by the risk of infections.

Finally, we noted that family functioning is affected by stress perceived by parent: high frequency of stress factors in the family role and in general, besides a great effort to face it. We noted again, as the degree of cohesion leads to overprotection by parents, dependents and even symbiotic bonds that could hinder the adaptation and disease control. This is similar to what Beckman *et al.* [15] observed in the parents of children with disabilities.

Interest for clinical practice

Authors such as Fisher (29) have mentioned the importance of providing information to the parents of children with chronic illnesses, and reflected the dissatisfaction felt by these parents when their true needs are not taken into consideration.

Adopting a model of coping that increases parents resources and skills and is not only about the disease will thus ensure that children don't feel guilt as a result of their parents' concern and suffering, and this will allow them to satisfactorily accept and integrate the disease [22].

This model must include: (i) detailed and up-to-date information and education on the medical management of haemophilia that will make it easier for families to accept, adapt and cope with their child's disease and improve their quality of life, (ii) an emphasis on teaching parents self-treatment techniques and how to care for the reservoir, making it easier for them to administer the treatment, (iii) detailed information on the advantages and disadvantages of the treatment – which may reduce stress and improve compliance, (iv) support for the patient and family, encouraging CwH to become more integrated into their surroundings and avoid isolation and (v) updating the treatment and giving guarantees of its safety and efficacy, not only to the *de novo* families but also to families with a history of haemophilia.

Limitations of the study

Although the sample obtained in this study is large if one takes into account the low prevalence of the disease, the fact that the parents included in it were not chosen randomly is a limitation of this study.

Comparisons of parents' perception of stress and family functioning after a psychosocial intervention

and after a longer period of time, in which their children's medical treatment has become stabilized, will help to confirm the results obtained.

Conclusions

The results reveal the need to implement measures and strategies to reduce the impact of stress on the parents of CwH and ensure a level of family functioning that is better suited to the situation.

Parents of CwH have difficulties in adapting to managing the disease; they perceive many stressors and have problems coping with them, not only with the clinical aspects but also at a psychosocial level.

The multidisciplinary approach must convince the parents that they are the most important people when it comes to improving their child's health and preventing risk behaviours, and that they need to use self-control strategies to cope with the challenges posed by their child's disease and its clinical signs and symptoms.

Other factors, such as gender and family background, must be taken into account, as these may become barriers or obstacles to properly following the treatments and consequently reduce their effectiveness and negatively affect quality of life.

Disclosures

The authors stated that they had no interests which might be perceived as posing a conflict or bias.

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