Original Article

Unmet needs and perceived shortcomings due to covid-19 in hemophilia patients

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Abstract.

Abstract: The large allocation of human and economic resources to face the pandemic and the lockdown mobility restrictions led the community of persons with haemophilia to be resilient and adaptable, engaging in adopting new methods to ensure their continuity of care. This study aimed to evaluate the emotions which have affected the haemophilia adult patient's daily life, during the first year of pandemic and what behaviors have been adopted to face the socio-economic re-strictions including lockdown, social distancing and self-isolation, as well as the limited access to Hemophilia Treatment Centres (HCT). An 18-item anonymous online survey for adult PWH was translated into four languages and was conducted in 2021, from February to April. The survey revealed that in a condition of social isolation, it is the experience of a sense of uncertainty that prevails, which, although it is not possible to eliminate completely, can however be managed, if continuity in relationships and support is guaranteed even at a distance. The results emerging from our work will be useful in the near future in order to improve professional performance in similar situations, and to optimize the synergy between the HTC doctor and the patient..

Keywords: COVID-19; Hemophilia; pandemic; lockdown; psychological impact

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Introduction

Coronavirus disease 2019 (COVID-19) has made a significant impact on the world's health, economic and political systems; as of October 1, 2022, more than 618 million people have been infected worldwide, with over 6.6 million deaths [1] since December 2019, when a series of cases of severe pneumonia was first described in Wuhan-China. The large allocation of human and economic resources to face the pandemic and the lockdown mobility restrictions led to developing strategies to minimize risks of inadequate access to the standard medical care for patients with chronic diseases and/or requiring regular clinical monitoring, like those with congenital bleeding disorders [2]. Redirection of re-sources towards the pandemic and measures aimed at reducing infection risk have limited access to in-person care at Hemophilia Treatment Centres (HTC)[3]. Although home delivery of the clotting factor for self-infusion is standard practice in some countries, in most of them, people must visit pharmacies or treatment centres to pick it up, when available. During the COVID-19 pandemic era this has been more difficult even if there are governments which have adopted factors concentrates home delivery to assure continuity of treatment for people with bleeding disorders [4]. Treatment centres have also had to adapt quickly to prevent disruption in providing consultations, laboratory tests and other services. Telemedicine has been used to maintain contact with patients in some countries and where this was not available other methods have been adopted often thanks to the help of the local bleeding disorder organizations [5,6]. Although the community of per-sons with haemophilia (PWH) has shown to be resilient and adaptable, engaging in adopting new methods to ensure their continuity of care, a large proportion of adults have faced financial struggles due to job losses or suspensions, while children and adolescents have experienced changes in their education and limits on their social interactions. These multiple challenges have likely had a negative impact not only on PWH's physical health but also on their psycho-social well-being [7]. This study aimed to evaluate the emotions which have affected the haemophilia adult patient's daily life, from different countries, representing a variety of cultural and economic settings, and different health care systems, during the first year of pandemic and what behaviors have been adopted to face the socio-economic restrictions including lockdown, social distancing and self-isolation, as well as the limited access to HTC. Another purpose was to find out possible unmet needs or perceived shortcomings due to Covid-19 in the quality of medical care services, with the aim of understanding how to improve PWH global assistance.

Materials and Methods

An 18-item anonymous online survey for adult PWH was carried out using Google Forms application. It was translated into four languages (English, Italian, Portuguese and Spanish) and was conducted in 2021, from February to April. The study was promoted online using social networks, messaging apps and emails to hemophilia associations world-wide, asking patients to participate voluntarily. The survey consisted of multiple-choice responses and checkboxes. The participants of the study were fully informed about the aim, procedure, and privacy considerations, and an online informed consent was obtained. Data were collected through an 18-question online individual interview composed of five questions about nationality, age, employment, number of inhabitants of residence city, education level, diagnosis, treatment regime, and 12 questions regarding the worries, fears and questions one year after the onset of covid-19 pandemic. The questions on the psychological reaction of patients with hemophilia during lockdowns and possible unmet needs or perceived shortcomings due to covid-19 restrictions, as well as the evaluation about how much and in what way emotions have affected the hemophilia patient's daily life were: “When you learnt of the covid pandemic, were you afraid that your hemophilic condition could somehow expose you to infection more than people without hemophilia?” “What effect did the information provided by the government, health care and local authorities have on you? “Many hemophilia dedicated websites published information and reassurances about hemophilia management during the covid-19 pandemic. Did they help you?” “Did you/do you need medical/psychological support to better address the current situation? If yes, who did you/would you contact?” “In your opinion, may the current situation have psychological consequences in the future, even when the pandemic will be over?” “During the last year, did you happen to experience particular sensations that worried you or made you feel uncomfortable?” “Do you recognize as yours one or more of the following fears?” “Did you find it difficult to manage the relationships with the professionals you need for your hemophilia?” “Which aspect of the current situation has the greatest impact on your health?” “What kind of help do/did you need?” “Have the check-ups and the planned activities at your Hemophilia Center been affected by the pan-demic?” The first question was the agreement to the use of the answers for scientific purposes. The last question was whether or not the person had already had the COVID-19 infection.

Results

Of the 209 PWH who completed the questionnaire, 5 did not provide consent to the use of their responses, so the participating patients were 204 from 24 different countries (36 Spanish-speaking, 47 English-speaking, 40 Portuguese-speaking and 81 Italian-speaking). The age was between 18-30 in 29.8% of cases (63), between 31-50 in 45.5% of cases (96), between 51 and 70 in 22.8% of cases (48) and over 70 in 1.9% of cases (4). About the geo-graphical distribution of the participants: 84 patients were European, 36 Asian, 81 American (Northern: 17, Central: 13, Southern: 51), 3 African. The education level of the participants was: elementary school 9 patients, junior high school 16 patients, high school 90, university degree 86, PhD 3. 17.6% of them said they did not work because unemployed, 18.1 % were students, 30.4% of them worked as clerks, 12.8 % were retired from work, the remaining 21.1 % were freelancers. 64 PWH (31.4%) declared to live in a large city (> 500.000 inhabitants), 83 (40.7%) in a city (> 15.000 inhabitants), 46 (22.5%) in a town

(<15.000 inhabitants) and 11 (5.4%) in a small village (<200 inhabitants). The demographic characteristics are summarized in table 1

Age			Education level			Employment	
18-30	63 (28.8%)		Elementary school	9 (4.4%)		Unemployed/ unoccupied	36 (17.6%)
31-50	96 (45.5%)		Junior high school	16 (7.8%)		Free-lance	43 (21.1%)
51-70	48 (22.8%)		High school	90 (44.1%)		Clerk	62 (30.4%)
>70	4 (1.9%)		University Degree	86 (42.2%)		Retired	26 (12.8%)
			PhD	3 (1.5%)		Student	37 (18.1%)

Table 1: Demographics characteristics of the survey participants (n=204)

With regard to the clinical characteristics: 178 patients reported having haemophilia A (87.3%) and 26 haemophilia B (12.7%), with a prevalence of cases of severe hemophilia in both groups A and B; of these, 12 had inhibitors against FVIII; in relation to the treatments: 121 patients (59.3%) underwent prophylaxis and 83 (40.7%) were treated on-demand. The percentages for prophylaxis patients were different according to the geographical origin: in Italy, for example, 70.4% of the interviewed sample were on prophylaxis, in Central America 44.4% received prophylaxis, while in Africa only 33%. The clinical characteristics of the patients, divided by geographical areas, are summarized in table 2.

Haemophilia type	Total 204	Africa	Asia	Europe	Central America	North America	South America
A severe	117 (57.4%)	3	19	48	6	8	33
A moderate	29 (14.3%)		7	12	2		8
A mild	20 (9.8%)		5	9	1	4	1
B severe	19 (9.3%)		1	7	2	4	6
B moderate	5 (2.4%)		1		2	1	1
B mild	2 (0.9%)		1	1			
A with inhibitors	12 (5.9%)		1	7	1		3
B with inhibitors	0						

Table 2: Health-related and characteristics and provenience of the survey participants (n=204)

Treatment	Total	Africa	Asia	Europe	Central America	North America	South America
Prophylaxis	121 (59.3%)	1	3	59	4	11	43
On demand	83 (40.7%)	2	33	25	9	6	8

Table 3: Anti-haemorrhagic treatment of participants (n=204)

The type of treatment is resumed in table 3. Regarding the questions about emotions and feelings in relation to the pandemic, to question 8, which asked if PWH were afraid that haemophilia makes you more exposed to COVID-19, the sample of people interviewed replied as follows: 47 "not at all" (23.1%), 93 "a little bit" (45.6%), 34 "moderately" (16.6%), 22 "quite a bit" (10.8%), 8 "extremely" (3.9%). About question 9 on the information provided by political and health institutions for which each participant was allowed to give several answers (multiple answers): 42 answered reassured, 76 confused, 15 answered angry, 27 were scared, 9 felt lonely, 35 gave other or combined answers. To question 10, regarding the usefulness of the information obtained from websites dedicated to the haemophilic patient, (also in this case multiple answers were allowed): 62 PWHs answered “yes they made me understand more”, 23 "yes but the fears remained", 8 "yes I understood and I felt reassured”, 97 replied that they had not consulted any site, 14 gave other answers. To question 11 regarding the need for medical / psychological support to deal with the social restrictions caused by the Pandemic, and what type of support was preferred, the sample of people interviewed replied as follows (multiple choice): 115 who did not need it, 24 replied that they had contacted the family doctor, 5 contacted the public psychology service, 6 contacted a private doctor, 36 contacted their HTC, 18 gave other or combined answers. Regarding question 12, if it was believed that the current pandemic may have psychological consequences in the future, even when this is over, almost half of the people interviewed, 93 (45.6%) answered "yes surely", 32.4% "maybe yes, I don't know", while only 20.1% answered “I don't think so” and 1.9% “Absolutely not”. To question 13, if in the last year they had experienced particular sensations that worried or made them feel uncomfortable, and if they recognized they had experienced one or more fears, (multiple choice allowed), the sample of people interviewed answered thus: 72 experienced "generalized anxiety", 58 "great stress", 69 "Worry about not doing enough to protect myself and my loved ones", 8 "guilt", 20 "excessive anger", 53 "sense of powerlessness", 29 "Intolerance toward people, including relatives", 60 "fear of visiting my Haemophilia Center because it is inside a hospital", 47 "fear to be without my medication", 59 "Staying apart from my loved ones", 39 "Job loss", 53 "Staying apart and isolated from other people", 42 "Fear of relating to people outside the family", 30 participants gave other answers . To question 14 "Did you find it difficult to manage the relationships with the professionals you need for your haemophilia?" the people interviewed replied: 99 (48.5%) that nothing had changed, 33 (16.2%) found no difficulty, 40 (19.6%) answered that it was difficult, 15 (7.3%) that it was very difficult and PWH would have managed it online, 17 participants (8.4%) gave other answers. To question 15 about which aspect of the current situation has had the greatest impact on their own life, the sample of people interviewed replied as follows: 22 had “difficulties related to the condition" (10.8%), 17 "difficulties related to the job (8.3%), 24 "the obligation to stay at home" (11.8%), 48 "adapting to new habits and lifestyles" (23.5%), 21 "obligation to use face masks, keep the distance and wash hands frequently" (10.4%), 26 "fear for my own and other people's health "(12.7%), for 13 it was the economic impact (6.4%), 11 replied “the restrictions when traveling” (5.4%), 16 "inability to meet the loved ones" (7.8%), 6 gave other answers (2.9%). To question 16 about whether and what kind of help the interviewee would like or need, 43 (21.1%) replied that "the support of friends or family, even if virtual", 96 (47.1%) replied that they needed more information on both Covid-19 and the Covid 19 vaccine; 58 (28.4%) reported "psychological help" both for the management of their emotional component and for the management of family relationships. 16 (7.8%) said they needed help in managing daily expenses, and wanting financial help from the government. 32 (15.7%) reported needing a greater presence of the Haemophilia Center, 13 (6.4%) by the general practitioner, and 52 (25.5%) said they did not need help. Question 17 asked whether the check-ups and the planned activities at their Haemophilia Center had been affected by the pandemic: for 82 participants (40.2%) there was no change; for 49 of them (24%) there was a reduction in all activities; 56 (27.4%) were delayed only in programmable activities such as check-ups; in 10 cases (4.9%) the pandemic caused a postponement of the switch to a new drug, while 7 gave other responses (3.5%). The last question, 18, asked if, at the date of the survey (January-April 2021), the participant had contracted Covid-19: 156 (76.4%) answered no, 24 (11.8%) answered yes, and 24 (11.8%) said they were unsure. The participants answers related to the unmet needs and perceived shortcomings are resumed in table 4. The answers to questions 13 and 16 were not shown in table 4 as they are difficult to summarize due to the high number of multiple answers

8) Were you afraid that haemophilia could expose you to COVID-19 more?	9) Information provided by the Government, health and local authorities has… (multiple choice)	10) Did haemophilia and COVID-19 dedicated websites help you? (Multiple choice)	11) Did you need medical or psychological support? (Multiple choice)	12) The current situation may have psychological consequences in the future?	14) Did you find it difficult to manage the relationships with the haemophilia professionals?	15)Which aspect of the current situation has the greatest impact on your health?	17) Have the check-ups been affected by the pandemic?	18) Did you have COVID-19?
A little bit 93/204 (45,6%)	Confused me. 76/204 (37,2%)	No, I didn't visit any website 97/204 (47,6%)	No, I think I don't need it 115/204 (56,4%)	Yes, surely 93/204 (45,6%)	No, nothing has changed 99/204 (48,5%)	Adapting to new habits and lifestyles 48/204 (23,5 %)	No 82/204 (40,2%)	No 156/204 (76,4%)
Not at all 47/204 (23,1%)	Reassured me 42/204 (20.6%)	Yes, they made me understand more 62/204 (30,4%)	Yes, I would contact/contacted my haemophilia center 36/204 (17,7%)	Maybe yes, I don't know 66/204 (32,4%)	It's been hard: I was afraid, but I haven't had problems 40/204 (19,6%)	Fear for my own and other people's health 26/204 (12,7%)	Only on planned check-ups, but not on treatment plans and urgent visits 56/204 (27,4%)	Yes 24/204 (11,8%)
Moderately 34/204 (16,6%)	Scared me 27/204 (13,2%)	Yes, but fear and worry are still the same 23/204 (11,3%)	Yes, I would contact/contacted my family doctor 24/204 (11,8%)	I don't think so 41/204 (20,1%)	No, I didn't find difficulties 33/204 (16,2%)	The obligation to stay at home 24/208 (11,8%)	Yes, on all aspects. 49/204 (24%)	I don't know 24/204 (11,8%)
Quite a bit 22/204 (10,8%)	Made me angry, they didn't and aren't doing enough 15/204 (7,4%)	Yes, I understood and I felt reassured 8/204 (3,9%)	Yes, I would contact/contacted private professionals 6/204 (2,9%)	No, absolutely not 4/204 (1,9%)	It's been hard, I would have managed it online 15/204 (7,3%)	Difficulties related to my condition 22/204 (10,8%)	I need to switch my treatment but I couldn't 10/204 (4,9%)	
Extremely 8/204 (3,9%)	Made me feel lonely 9/204 (4,4%)	Other and multiple combined answers 14/204 (6,8%)	Yes, I would contact/contacted the psychological public service 5/204 (2,5%)		Other and multiple combined answers 17/204 (8,4%)	Obligation to use face mask, keep the distance, wash your hands frequently 21/204 (10,4 %)	Other 7/204 (3,5%)	

Table 4: Patient responses from the part of the questionnaire related to the needs and shortcomings

	Other and multiple combined answers 35/204 (17.2%)		Other and multiple combined answers 18/204 (8.7%)			Difficulties related to the job 17/204 (8,3%)		
						The impossibility of meeting the loved ones 16/204 (7,8%)		
						The economic impact 13/204 (6,4%)		
						Travel restrictions 11/204 (5,4%)		
						Other 6/204 (2,9%)		

Discussion

We are certain that our findings offer a contribution to our understanding of the psycho-logical reaction of PWH during lockdowns and subsequent phases of a pandemic by de-scribing the emotions, sensations, and level of distress experienced, as well as the impact of this on compliance with disease management requirements.

The impact of the pandemic has certainly raised concerns related to the PWH condition, manifesting more or less strongly a concern of being more exposed to the infection.

The answers show a significant need for correct information, the only thing in these cases that can indicate more suitable behaviors to face the moment. It is noted, in fact, that the information provided by the various governments, but also by the media, has not had the effect of reassuring, indeed it has often caused further confusion and a sense of uncertain-ty.

On the other hand, those who consulted the sites dedicated to the haemophilic patient, where the various federations, foundations, associations have promoted initiatives aimed at providing information about the management of the pandemic in relation to haemorrhagic diseases, were able to obtain reasons for reassurance and clarity. However, the data emerging from our enquiry reveal that almost half (47.6%) of the population interviewed, despite experiencing uncertain and insecure feelings, did not consult any site. The reasons may be various but it is certainly a fact that invites us to increase and disseminate effective and above all certified online spaces from the point of view of scientific reliability, where the haemophilic patient can get specific information for his pathology and management of social, and health situations related to the pandemic.

The results regarding the question about medical/psychological support are interesting: a high percentage (56.4%) reveals that they do not need it despite being aware that this pandemic will leave psychological repercussions (78%). Some of the patients seem to feel that correct information and good continuity in terms of communication with the teams of the HCT are already supportive and that requests for support and psychological help are more linked to a subjective management of emotions depending on the individual, and that stress related to job loss also has an impact, for those concerned.

Multiple answers were given to the question of emotions, as emotions are never unique. However, it is possible to detect emotions common to world standards and more subjective emotions linked to the social and cultural contingencies of individual local and national contexts. This indicates that some patients require more careful treatment of the emotional aspect and must always be listened to, and even more so if the patient presents fragility that can affect the quality of his life. The answers show the importance of a sup-port that is not only medical, but one which takes care of the person also from an emotional point of view, in order to create greater awareness and rationalization of their emotions; today a patient is increasingly a protagonist of his own life even in the presence of critical and uncertain events.

Regarding the patient relationship with HCT, it seems that despite the pandemic, the patient found continuity, even a strengthening of confidence. Only a small percentage were afraid to go to the Centre for fear of a possible infection and therefore preferred online management.

With regard to the management of check-ups and planned activities, a good percentage reveal continuity, and despite the fact that the check-ups were often postponed, urgent vis-its and treatment plans were always offered. However, a percentage reported inconvenience relating to difficulties in reaching the Centre and fulfilling the needs of their therapeutic plan, both for fear of contracting Covid-19 (27.9%), and for a reduction or blocking of visits to the HCT (56.3%).

But overall, the patients reported that they had no difficulty in contacting the HCT staff (64.7%), thus confirming the availability of the staff of the Centre and the adaptability of patients suffering from rare haemorrhagic diseases, that has always characterized this type of therapeutic management. It is also true, however, that by analyzing the responses of patients mainly from Asia and Africa, this percentage drops to 38.1%.

Conclusions

The survey reveals that in a condition of social isolation, it is the experience of a sense of uncertainty that prevails, which, although it is not possible to eliminate completely, can however be managed, if continuity in relationships and support is guaranteed even at a distance. This is true for all human relationships and in particular for those with chronic pathologies who find themselves experiencing a stronger sense of uncertainty. We believe that the results emerging from our work will be useful in the near future in order to im-prove professional performance in similar situations, and to optimize the synergy be-tween the HTC doctor and the patient.

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Conflicts of Interest

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