



Difficulties in facing alone the demands of treatment: experiences of the adolescent hemophiliac*

Dificuldades para enfrentar sozinho as demandas do tratamento: vivências do adolescente hemofílico

Dificultades para enfrentar solo las demandas del tratamiento: vivencias del adolescente hemofílico

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ABSTRACT

Objective: To understand the reasons why the adolescent hemophiliac does not adhere to hemophilia treatment. **Methods:** A qualitative study, conducted with seven adolescents, whose data were collected through semistructured interviews, using *symbolic interactionism* as the theoretical reference and *interpretive interactionism* as the methodology. **Results:** Adolescents can not meet the demands and difficulties of the recommended treatment alone, they do not assume responsibilities inherent in this, and at the same time, judge what is appropriate treatment, while not valuing as essential the preventive aspects and feeling disbelief of its effectiveness against complications, as well as being dissatisfied with the care received in the service. **Conclusions:** The results reinforced the importance of family support to guarantee continuity of treatment for the adolescent hemophiliac and to provide additional subsidies to rethink care provided to them in specialized services, in order to facilitate the delivery of assistance geared to the demands of this population.

Keywords: Hemophilia A; Adolescent health; Adolescent health services; Patient dropout; Pediatric nursing.

RESUMO

Objetivo: Compreender as razões pelas quais o adolescente hemofílico não adere ao tratamento de hemofilia. **Métodos:** Estudo qualitativo, realizado com sete adolescentes, cujos dados foram coletados por entrevistas semiestruturadas, sendo o Interacionismo Simbólico o referencial teórico e o Interacionismo Interpretativo, o metodológico. **Resultados:** os adolescentes não conseguem enfrentar sozinho as demandas e dificuldades do tratamento preconizado, por não assumirem as responsabilidades inerentes ao mesmo, além de julgarem que fazem o tratamento adequado, mesmo não valorizando os aspectos preventivos essenciais e perceberem-se descrentes de sua efetividade contra as complicações além de insatisfeitos com o atendimento recebido no serviço. **Conclusões:** os resultados reforçaram a importância de apoio da família para garantir a continuidade do tratamento ao adolescente hemofílico e oferecem subsídios para repensar a assistência prestada a eles nos serviços especializados, no sentido de favorecer a condução de um atendimento voltado às demandas dessa população.

Descritores: Hemofilia A; Saúde do adolescente; Serviços de saúde para adolescentes; Pacientes desistentes do tratamento; Enfermagem pediátrica

RESUMEN

Objetivo: Comprender las razones por las cuales el adolescente hemofílico no se adhiere al tratamiento de hemofilia. **Métodos:** Estudio cualitativo, realizado con siete adolescentes, cuyos datos fueron recolectados por entrevistas semiestructuradas, siendo el Interaccionismo Simbólico el referencial teórico y el Interaccionismo Interpretativo, el metodológico. **Resultados:** los adolescentes no consiguen enfrentar solos las demandas y dificultades del tratamiento preconizado, por no asumir las responsabilidades inherentes al mismo, además de juzgar que hacen el tratamiento adecuado, aunque no valorizando los aspectos preventivos esenciales y percibirse descreyentes de su efectividad contra las complicaciones además de insatisfechos con la atención recibida en el servicio. **Conclusiones:** los resultados refuerzan la importancia del apoyo de la familia para garantizar la continuidad del tratamiento al adolescente hemofílico y ofrecen subsidios para repensar la asistencia prestada a ellos en los servicios especializados, en el sentido de favorecer la conducción de una atención volcada a las demandas de esa población.

Descriptor: Hemofilia A; Salud del adolescente; Servicios de salud para adolescentes; Pacientes desistentes del tratamiento; Enfermería pediátrica

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INTRODUCTION

Hemophilia is a hereditary disease, genetically transmitted by the X chromosome, characterized by a deficiency of coagulation factors VIII and IX circulating in the plasma⁽¹⁾, which manifests almost exclusively in males⁽²⁾. The condition of hemophilia in women is rare, because their other X chromosome, inherited from the father, will produce the necessary coagulation factors, but men present a defect in their only X chromosome that, therefore, develops into a disease⁽³⁾.

A deficiency of factor VIII is called hemophilia A or classic, and of factor IX, Hemophilia B, or Christmas disease⁽⁴⁾ which determine the clinical characteristics. Depending on the plasma levels of the deficient factor, hemophilias are classified as severe when the level of the circulating factor is less than 1%; moderate when it is between 1% and 5%; and, mild, when above 5%, compared to the normal level that is 100%, that is, the same as one unit of factor per milliliter of blood⁽⁵⁾.

The clinical manifestations of hemophiliac patients are bleeding that can occur anywhere in the body, most frequently in the musculoskeletal system, and that may cause sequelae, such as hemophiliac arthropathy. In severe hemophilia, there is spontaneous bleeding, after minor traumas that are not always recognized; in moderate, bleeding can be observed during surgical procedures or minor traumas and, in mild, bleeding is common during surgical procedures⁽⁶⁾.

In order to prevent or delay the onset of sequelae you need to minimize bleeding time, promoting hemostasis as quickly as possible by means of intravenous administration of coagulation factors which are insufficient in the plasma⁽⁷⁾.

The treatment of the disease requires the participation of a multidisciplinary team, so that one can have therapeutic success, because children and adolescents with hemophilia will undergo treatment throughout their entire lives, since it is a chronic disease, and it is only possible to control it⁽⁸⁾.

In our practice, we have observed that non-adherence to treatment, as commonly occurs in the adolescent hemophilia population, involves not only lack of success of prophylactic and therapeutic measures, but also strains the staff-patient relationship.

The characteristics and peculiarities of this age group should be regarded because, without question, they will determine their ways of acting with the world, their disease and treatment. In this way, we believe that better understanding can enable adequate care and direct the conduct of care, through the planning of an effective intervention. Concerned with the issue of nonadherence and with the necessity to listen to the adolescent, because we did not identify studies on these points in the

literature, we set out to develop this research in order to understand the reasons why the adolescent hemophiliac does not adhere to the treatment of hemophilia.

METHODS

To develop this study, we opted for a qualitative approach, which works with the motives, aspirations, beliefs, values and attitudes of the actors, going deeply into the world of meaning of human actions and relationships⁽⁹⁾.

Among the numerous possibilities for qualitative research, one option is the Symbolic Interactionism (SI) as a theoretical reference and Interpretive Interactionism (II) as a methodological reference, which were considered pertinent to the objective of this investigation.

The SI is a perspective of analysis of human experiences, whose focus of study is the nature of interaction, that is, the activities of social dynamics that occur between people^(10; 11). According to this perspective, people perform an action based on the meaning that the situation represents for them, and it can be redefined, that is, with the passage of time the individual may give new meaning to the facts⁽¹¹⁾.

Interpretive interactionism is a methodology designed by Norman K. Denzin, that recognizes that the meanings of actions are in the lived experience. It focuses on the experiences that, radically, affect and mold the meaning people give to themselves and their life projects, leaving marks and changing the direction of life of individuals in ways that are called epiphanies⁽¹²⁾.

The study was conducted at the Hemophilia and Other Hereditary Coagulopathies Service of a university hospital in São Paulo. Prior to data collection, the project was approved by the Ethics Committee of the Federal University of São Paulo under Protocol n° 2.057/07. Adolescents who agreed to participate in the study signed a form of Free and Informed Consent, as did their guardians when they were younger than 18 years, as recommended by Resolution n° 196/96⁽¹³⁾.

The subjects were seven adolescents living with hemophilia, enrolled in the hemophilia service of the research institution, who had not followed the treatment recommended by the multidisciplinary team. The number of individuals complied with the criterion of saturation, i.e., data collection ceased when there was repetition of the data observed and an absence of new data, permitting a growing understanding of the concepts identified⁽¹¹⁾.

Data were collected between October 2008 and February 2009, through individual semi-structured interviews, conducted on previously scheduled dates and times with adolescents, in a place reserved in the service itself, and which began with the following guiding question: "Tell me, what leads you to not follow the recommended treatment for hemophilia?"

Throughout their interview other questions were formulated to deepen the understanding of the concepts expressed by the adolescents interviewed.

The interviews were taped and transcribed verbatim, so that the narratives could be subjected to analysis of the data that occurred simultaneously with data collection, involving the phases of bracketing, constructing, and contextualizing the phenomenon, as proposed by the adopted methodological approach⁽¹²⁾.

In the bracketing phase, the narrated stories were analyzed intensively, with the identification of constituent elements and key configurations of discourses and their respective extraction of the experiential units. In the constructing phase, the attribution map was elaborated for thematic categories, by reconstructing, based on isolated elements in individual speeches, the collective subject discourse, within the representative theme. The contextualizing sought to connect the phenomenon to the life of adolescents, developing its meaning and permitting the interpretive process and its understanding.

RESULTS

The data that emerged from the narratives of the adolescents interviewed made it possible to understand the motives that lead to non-adherence to recommended treatment for hemophilia. Their experiences, related to the interaction that they established with their body, with the fact of hemophilia and the treatment experienced, showed to be significant and relevant, for the conduct of that behavior, and revealed four epiphanies, which are described below and exemplified with excerpts of the narratives of the adolescents, identified in the text as A1, A2 to A7.

The first epiphany, *Having to Assume Responsibility for the Treatment*, refers to the moment when the hemophiliac, when he became a teenager, started to assume responsibility for his treatment. It reveals the difficulties that he had in taking responsibility for the treatment, to comply with the demands required, independently, and to live with differences in lifestyle due to the fact of being a hemophiliac.

This caused him to be perceived differently and be excluded from his group of friends, because he ceased to perform many activities of his age, because if he did so he would have to go to the hospital. Because he judged that the other adolescents in the group would consider him differently, he did not tell his friends about the disease.

My brother and I are different from other people who do not have hemophilia. We cannot do whatever the other boys do that will begin the bleeding, like playing football and cycling. (A2)

My friends do not know that I am a hemophiliac. They would keep asking what it is, why I was not going to play ball. They would find me different from them. (A5).

According to the account of the teenager when he was a child, his mother made note of the exams and took him to appointments without losing dates and times; it was obligatory. When he began to assume responsibility for his treatment, he had difficulty to commit to it, with forgetfulness and absences to scheduled appointments. He began to come only when he was injured, he said he was tired of the routine treatment and referred to growing lazy of attending the service as scheduled, preferring to do other things like staying home, going to school or hanging out with friends for fun.

It's inconvenient to have to come here when I could be doing something else ... like reading at home or at school. It is very inconvenient to come here directly. It's like a routine to come to the hospital and then the routine ends up getting tiring. Sometimes, we end up missing other things to come here. (A2)

I'm too lazy to come here! I'd rather stay home and do something there, without having to keep driving here. (A1)

At the time, you think: I have to leave home early. I'm tired, then I say: not today! (A6)

Even when he had some help from his mother, to schedule the exams, he did not attend; he prioritized other activities, exercising the right to do only what he wants.

Before my mother brought me, and I had to come, now I kind of do what I want. (A2)

Before, my mother brought me and waited. Now I have to come alone [...] My mother marked the exam, and I forgot, sometimes because of personal motives I was not there. (A6)

The second epiphany, *Facing Difficulties in Implementing the Treatment*, highlights the difficulties experienced by the adolescent to successfully fulfill the demands required by the recommended treatment for hemophilia. He faced difficulties with different aspects: related to the transportation, the distance from where he lives, the lack of money to come from home to the service, the need to miss school and work, both for him and his family and / or guardians, as well as the unavailability of people to bring him to the service of hemophilia, even when there is some acute hemorrhage and he is with much pain.

I caught three connections to get here. So, I leave to come here when I need to take factor or pick up my dose. (A1)

I live very far from here; it is very difficult to come! Sometimes, my mother has no money. (A3)

It is my mother and me alone! When I'm in pain, I have to wait for her to come from work to bring me here. (A5)

I have to miss class to come and take factor. Then, on the day of collection I also cannot go to class. [...]. When I have to keep coming here, I have problems at work. [...] Once, I was sent away, because I was a few days without work because of bleeding. (A2)

Besides these, there is the difficulty for the adolescent in performing the injection of the household dose of clotting factor, where he or a family member does not have the courage to do the injection or the availability of time to do the training.

I do not want to do it, only if my mother does it for me. But she also cannot come here to learn. So, it is difficult, right? (A3)

They (parents) have no courage, and I also do not! It causes me stress to put the needle into my vein. (A7)

The third epiphany, *Thinking that he has Adequate Treatment*, refers to the fact of the adolescent considering that he is doing the adequate treatment for hemophilia, although he does not completely follow it, he knows it can hurt him and he can assess that, if this happens, he can use the household dose to initiate treatment. As a result, he will feel less pain and could be programmed to come to the service in order to continue the treatment, missing less work and school. He defines what is good for himself, and does only what he evaluates is important for him.

But, in my view, I follow the treatment correctly: at the time I'm feeling a little pain, I've come. If it starts bleeding, I already take the factor and I do not have those pains; after I come here to replenish the dose [...]. (A5)

I have had greater understanding that I had to stop fighting, playing ball. I almost have no more bleeding. [...] And, out of vanity, also my body is changing. (A5)

Whenever I need to take factor, I do it at home or come here to take it. For physical therapy, I do not, because I do not need it. I only take factor when I hurt and pronto! I take the factor immediately when I am bleeding and feel less pain. (A4)

The fourth and final epiphany, *Discrediting the Efficacy of Treatment and Efficiency of the Service*, revealed the disbelief of the adolescent hemophilic about treatment and the care of the service. It was perceived they were unsatisfied with some aspects of this service, such as the delay on weekends, when there is only one doctor to serve various sectors of the hospital. They also indicated lack of satisfaction with the care provided by the physical therapy service, by comparing it to what was offered by other institutions that serve hemophiliacs - better physical space, equipment, prostheses and treatment by the multidisciplinary team.

The schedule on the weekend is very bad, people come and have no doctor, we've been injured and in pain and there is delay in waiting to be attended. I know that the doctor has to stay in the infirmary, the emergency room, here, but at least they should have a doctor here. (A6)

How many times I came here with pain on the weekend, but had no one to meet me [...]. I've done physical therapy here, but there (another service) is better, it has more devices, and the girl is over there, seeing if it is right. (A5)

There (another service) is better, it has more equipment and more things for doing exercises. They gave me three machines and two boots and insoles also. There it has a psychologist, social worker, dentist, occupational therapist and nutrition. It has more professionals and here they do not have them. (A1)

Moreover, the adolescent was found to be skeptical about the importance of some recommendations

for the prevention of disease complications, such as collecting annual exams, and did not believe that the treatment offered any prospect of improvement for his body and the pain that accompanies the bleeding, which he referred to as, "hurting like death" and, faced with which, he felt powerless.

Why keep doing a lot of tests every year? You know what I have. [...] My leg is now all screwed up and even physical therapy failed to improve anything. (A3)

Physical therapy, I do not do it, because I do not need it. (A4)

My elbow is already impaired; I cannot stretch it wide. (A7)

At the time that the bleeding hurts you, it is like death! (A5)

DISCUSSION

This study allowed us to understand the experiences and rationales of adolescents with hemophilia related to the behavior of not following the recommended treatment for hemophilia in its entirety, because they could not face on their own the demands and difficulties inherent in it.

One of the aspects apprehended, relates to the fact that adolescents need to take responsibility for their own treatment and self-care, which proved difficult for these youth. According to the literature, adolescence is the period in which there is greater freedom for these patients in following their own treatment, but it is also shown to be the moment at which the adherence to the same, frequently, declines⁽¹⁴⁾.

Another relevant point was they perceived they were different from other teenagers and did not disclose to their peers that they were hemophiliacs. It is known that the interaction with colleagues can make teenage hemophiliacs perceive themselves differently; while recognizing that they require periodic treatments, with some restrictions, they wanted to adapt and be like their peers. But as hemophilia excessively emphasizes the differences, altering their identity, causing a disturbance in their self-image, they proceeded to conceal this health condition⁽¹⁴⁾.

In this phase, the opinions of friends were important. They may not understand and accept the treatments or the reasons for their non-participation in certain activities. In our professional experience, we have observed that for these young people, having friends within the hemophilia community, providing their participation in events for people with hemophilia helps them feel less different from the others, and inserts them in leisure activities. In this regard, we have participated in meetings among hemophiliacs so that they experience the exchange of ideas, have fun, and receive educational opportunities related to this period of life and to hemophilia.

The literature on this topic emphasizes that adolescents themselves should decide when and to whom

they will tell about their hemophilia, knowing that the differences make them unique and special and should be respected ⁽¹⁴⁾.

The adolescents in this study reported that they were lacking follow-up treatment; they claimed weariness of the routine and that they were too lazy to come to the service. They revealed that before they felt "compelled" to come up with their mothers, not missing appointments and following recommended treatment, whereas now they prefer to do other activities common in everyday life of teenagers, or even stay at home.

Other experiences which proved significant to their nonadherence were difficulties relating to: transportation and the distance from home to services for hemophilia, the promptness of the hematologist to attend him, and financial problems. The indefinite waiting time and, many times, elevated costs are obstacles to the treatment of chronic diseases ⁽¹⁵⁾.

A study about chronic diseases and their consequent necessity for monitoring, highlighted the commitment of the family budget during the course of disease in relation to the cost of treatment, making the attendance of outpatient return visits very difficult for patients, mainly because they lived in places far from treatment centers, necessitating the use of two or more types of transport to the hospital, which leads to interruption in therapy ⁽¹⁶⁾.

It is estimated that 85% to 90% of people with hemophilia in Brazil are poor and must deal with the lack of money for transport to specialized services, with difficult access, high dropout rates and unemployment ⁽¹⁴⁾.

Equally relevant is the fact pointed out by the adolescents in this study that they depend on the family to bring them to the service, especially in acute cases of hemorrhage and in the presence of much pain. In periods of hemorrhage, the hemophiliac often needs to rest and carry out the injection of anti-hemophilic factor and so, being alone brings physical adjustment problems and harms their safety in cases of difficulties ⁽¹⁷⁾, which has been advised already for few decades.

Apart from questions related to the difficulties of meeting the demands of treatment, which are determined by the characteristics of adolescence itself, as in this study, the literature highlights that hemophilia has an impact on work, education, social activities and family life of the hemophiliac. In particular, they feel stigmatized and discriminated against at work and experience significantly higher unemployment than people with normal health, because of the fact that employers take into consideration the need for frequent absences, because if they have a hemorrhage they may stay out for days, weeks and even months ⁽¹⁷⁾.

There is also an important association between the frequency of bleeding episodes and absences

for academic activities in adolescents with severe hemophilia ⁽¹⁸⁾, since the chronic disease leads to difficulties in school related to transport, to the presence of accessibility, inclusion, to absences resulting from the crises of acute pain and the need for visits to treatment centers and hospitalization ⁽¹⁶⁾.

In this scenario of difficulties, there is evidence of a therapy that offers advantages and consequent improvement in quality of life of adolescents with hemophilia, the access to the household dose of the concentrated coagulation factor, which must be administered at the beginning of bleeding events.

A study about the social and psychological responses arising from the use of this therapy in individuals with hemophilia demonstrates its positive impact, enabling them to plan their activities and minimize the demands of friends and family in assisting with the treatment, decreasing financial costs of transport and loss of time at work and / or at school, improving the ability of the hemophiliac to organize his time and plan his activities, increasing the availability and willingness to engage in social activities, including allowing them to travel to distant locations for a treatment center and reducing the risk of bleeding associated with joint dysfunction ⁽¹⁹⁾.

However, this study revealed that adolescents interacted with difficulties for the use of the household dose, for even recognizing and realizing their benefits from its advantages, they were not always able to manage it for lack of personal conditions of their own or of their parents.

The access to concentrated factor is not, however, the only way to receive care related to hemophilia, there is a need for psychological care, family planning, physiotherapy and others ⁽¹⁴⁾, which were not as valued by the adolescents surveyed, because they did not believe in the benefits of these other interventions which comprise the treatment.

Research about attitudes and behavior of young American hemophiliacs also found that 36% of participants believed that the damage to joints cannot be avoided, 60% dealt with hemophilia by avoiding physical activity, and only 31% treated bleeding episodes within 1 hour, with the first reason for why they were slow to start treatment being the failure to recognize their need. It emphasized that young people with chronic disorders need help to understand how they can prevent complications. The main points for the campaigns of prevention of complications of hemophilia should include physical activity to ensure healthy joints and treating of hemorrhagic episodes early and adequately ⁽²⁰⁾.

In addition to failing to obtain regular treatment, for not believing in the possibility of improvement, the

adolescents surveyed also considered that they were not being adequately attended, either because they had to wait for service on weekends, when they go to service with pain or because some visits did not meet their expectation, such as physical therapy, leading them to seek these in other institutions.

Also, they believed they following the treatment adequately, since they took precautions to avoid hemorrhage, not doing physical activities that might harm them, as a strategy for the prevention of hemorrhaging. For them, this attitude meant they were following the treatment.

According to other research, when questioned about the treatment of hemophilia or how to prevent its complications, 60% of hemophilia patients responded to avoid or limit physical activities that may compromise them, while only 27% did exercise as a preventive measure, and 24% said they make use of prophylactic factor concentrate, before physical activities, sports or vigorous exercise⁽²¹⁾. This is corroborated by data from this study that showed how the adolescents surveyed did not value the physiotherapy service or the collection of annual exams, being concerned only with what may have direct effect on their wellbeing and the occurrence of pain.

Additionally, proving relevant for nonadherence to treatment, the discrediting by the adolescents of their prospect of improvement, particularly with regard to pain that occurred in the presence of a hemorrhage that “when it hurts, it hurts like death”.

The fact is reinforced by the literature, according to which the agonizing physical suffering of bleeding of a joint, the emotional pain of being isolated, and the possibility of the weight of additional chronic diseases are summarized in a set of characteristics that lead to the disbelief of adolescent hemophiliacs regarding the already established improvements for the condition⁽¹⁴⁾.

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CONCLUSIONS

The findings of this study may contribute to rethinking some actions to be developed in hemophilia services in order to promote adolescents' adherence to treatment, such as: helping families understand the importance of continuing support and attention to their health; not leaving them alone to confront the demands and difficulties of treatment; reinforcing continuing education regarding the relevance of preventive visits; promoting the development of group activities for social integration and discussion of questions related to adolescence; helping to identify systematically the available or potential support network, which may assist the adolescent in cases of need, especially in emergencies; establishing a communication system before the hemophiliacs come to the services, in cases of emergency; seeking to reduce of the waiting time; acquiring more modern physiotherapy equipment and in sufficient numbers for the care; having a multidisciplinary team able to deal with the adolescent and family, among others.

The establishment of these actions is important, so that the adolescent receives quality care and for services to be a reference to support him and his family, and not just a place to seek pain relief and supply of the medication.

We also emphasize that other aspects, such as the effects of having experienced the death of a family member with hemophilia and issues related to spirituality and religion as sources of support, could be investigated in further studies with adolescent hemophiliacs, in order to seek grants to assist them in coping with this condition.

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