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Original Article

A Young Woman's Experience of Living with Hemophilia: A Qualitative Case Study

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Abstract

Introduction: Hemophilia is a genetic disorder inherited in an X-linked fashion and accompanied with dysfunctional coagulation factors 8 (hemophilia type A) or 9 (hemophilia type B). Hemophilia has a dramatic effect on the quality of life of patients. The objective of this study was to describe a young woman's experience of living with hemophilia from childhood to motherhood.

Methods: This qualitative study explored a 32-year-old woman's experience of living with hemophilia. Data were collected using interactive semi-structured interviews. Date analysis was performed using a phenomenological approach. Member check and researcher triangulation were used in data analysis to ensure the accuracy of the study. All ethical principles were considered in this research.

Results: Data analysis revealed four main themes, including "being involved in treatment", "suffering", "fear and worry", and "acceptance of being different", and eight subthemes, including "pity and humiliation", "social stigma", "fear of marriage and motherhood", "fear of continuing education", "limiting one's relationships", "getting to know others with the same disease", "accepting fate", and "independence".

Conclusion: This study presented the point of view of a young woman with hemophilia regarding her challenges as well as physical, psychological, and social sufferings from childhood to adulthood

Keywords: Hemophilia A, Qualitative research, Quality of life, Social stigma

Introduction

Hemophilia is a genetic disease associated with the x chromosome, which is caused by a disorder in coagulation factors 8 (hemophilia type A) and 9 (hemophilia type B) (1). The disease can also be caused by spontaneous genetic mutations as some patients with hemophilia have no family history of the disease. Hemophilia is a disease characterized by frequent bleeding in joints, especially in knees, ankles, elbows, and soft tissues, including skin and muscles causing hematomas. Patients may experience joint pain, swelling, stiffness, functional limitations, and inflammation which worsen over time if left untreated.

Depending on the severity of the disease, bleeding may have physical or psychological impacts. Lifetime treatment includes intravenous administration of coagulation factors that can be done in emergency

rooms, hemophilia centers, or at home. There are two methods of coagulation factor administration in hemophilia. One is to provide coagulation factors when needed, for instance after each bleeding, during interventions, and when evaluating the symptoms, while the second method is to provide bleeding prophylaxis. In the case of prophylaxis, coagulation factors are prescribed two to three times a week, even in the absence of bleeding. The second method reduces anxiety but can induce psychological and organizational burden on patients and caregivers due to its aggressiveness (2). Previous studies showed that the quality of life of hemophilia patients has been improved from the time alternative factors were administered (3). Bleeding disorder has a significant effect on various aspects of life among hemophilia patients. These aspects may include education, work, social activities, and family life (4). Based on the findings of a study that examined the point of views of hemophilia patients, both the disease symptoms and treatment had affected the quality of life of hemophilia patients (5). Besides hemophilia patients, their families often face psychological challenges (3, 6). Limited quantitative studies have been conducted on hemophilia patients. However, deep understanding of the experience of people with hemophilia and the way they adapt to their living conditions is lacking. This knowledge is important for health professionals and all those who are involved in hemophilia patient care. The objective of this study was to explain a young woman's life challenges with hemophilia from childhood to motherhood.

Methods

The present case study was carried out using a qualitative method. In-depth semi-structured face-to-face interviews were used to collect data. The interviews were conducted at the Charity Foundation for Special Diseases in a quiet and appropriate place. Each interview began with an open-ended question: *"What comes to your mind when you hear the word hemophilia?"* Probing questions such as *"How did you feel? How was it? Give an example, ..."* were also asked to continue the interview and enrich the data. After transcribing each recorded interview, the text of the interview was read several times and the meaningful sentences and themes that described the phenomenon were extracted. The criteria suggested by Lincoln and Guba were used to ensure the accuracy of findings (7). Long-term engagement, integration in research and member check were used to assess the credibility of the findings. In terms of dependability of the findings, the steps and process of the research were recorded and reported consecutively and as accurately as possible. For the confirmability, the researcher expressed the results of the study based on interviews by quoting the participants sentences without his personal assumptions. Van Manen's Hermeneutic phenomenological approach was used to analyze the data (8). A total of 92 thematic phrases, sentences, or paragraphs were extracted from the interviews. Finally, four main themes and 8 subthemes were obtained after combining and comparing the commonalities of the themes and subthemes.

Case report

This study was conducted on a 32-year-old woman with hemophilia A. According to the patient's mother, her first symptom was bleeding gums, which occurred at the age of three months. Her disease was not diagnosed until she visited a tertiary medical center by performing specialized laboratory tests.

The bleeding incidents from childhood caused people around the patient to feel that her disease was infectious due to their lack of information about hemophilia. As a result, they limited or even cut contact with her and her family. Another reason for communication restriction and social isolation was the experience of pity and being humiliated in relationships with others. The participant described the first experiences of the disease in her childhood, when she was banned from doing activities due to prolonged and uncontrollable bleeding. She was banned from doing exercise at school because she had bleeding episodes with the slightest injury. This condition had a great psychological and financial burden on her family. The participant lived in a large family with seven children. The economic status of her family was unsatisfactory. Her father was a worker and two other children in the family had hemophilia. Epistaxis was her most common type of bleeding before puberty. At the age of 11, following the onset of puberty and menstruation, she experienced severe vaginal bleeding with hemorrhagic and polymenorrhea. The bleeding was so severe that she received three units of packed cell to compensate for the bleeding.

Due to lack of access to coagulation factors, she received alternative therapies such as tranexamic acid and blood products including cryoprecipitate, which caused allergic reactions in the patient. Finally, to control the bleeding during menstruation, she started taking low-dose estrogen contraceptive pills from the age of 11. Severe and frequent bleeding affected her living conditions, so that she had repeated absentees from school during her studies, which caused her to drop out of school despite her strong interest in studying. Therefore,

she was forced to choose a field of study which was not her favorite.

At the age of 17, she married but was not accepted by her husband's family due to her condition. Her condition exacerbated as the result of the psychological pressures after marriage. She had informed her spouse about her condition before marriage and her spouse agreed to marry her. The participant voluntarily discontinued the use of LD pills at the age of 19, assuming that her fertility had been lost due to long-term use of contraceptives. She became pregnant one month after discontinuing LD pills. She experienced a lot of anxiety and fear as her doctor strictly prohibited her from getting pregnant. Therefore, she went to a medical center in Tehran, the capital of Iran, to perform accurate and specialized genetic tests. She decided to continue her pregnancy after confirming that the fetus was healthy. She delivered her child through emergency cesarean section due to severe bleeding during vaginal delivery. Her delivery was accompanied by many complications such as recurrent infections and prolonged postpartum hemorrhage, which continued for seventy days after delivery.

Results

The analysis of the data revealed four main themes including "being involved in treatment", "suffering", "fear and worry", and "acceptance of being different" as well as eight subthemes including "pity and humiliation", "social stigma", "fear of marriage and motherhood", "fear of continuing education", "limiting one's relationships", "getting to know others with the same disease", "accepting fate", and "independence". Explicit quotes from the participant were used to explain the experiences.

Being involved in treatment

The experiences of the participant showed that she has been engaged with the disease, its symptoms, complications, and treatment all her life. For example, when asked to talk about her childhood, she said:

"I was a child; I was three months old. My mother said when you began teething, the saliva that came out of your mouth was bloody. I was taken to Mashhad, then to Tehran for treatment. My mother often said that I used to bleed from my nose and gums…".

The first days of school are exciting and memorable days for students. Hemophilia affects childhood memories and experiences. The participant stated one of these different experiences:

"*I was in the fifth grade (primary school), I received three bags of blood. The bleeding continued despite attempts to stop the bleeding …"*

The physical and mood changes in puberty and menstruation make puberty a crisis that may cause anxiety and depression in girls. Menstruation begins with the onset of vaginal bleeding at the age of 11 to 13 years old. The presence of hemophilia, as defined by deficiency in coagulation factors, can increase the severity of bleeding during menstruation. The participant expressed her experiences from the beginning of her menstruation and controlling the severity of bleeding as follows:

"I was in the first grade of middle school when my period started. It was too long, forty days! I was clean for one week and then forty days of bleeding… severe bleeding… [so severe that] I had to have blood transfusion…"

Menstrual disorders, including severe bleeding (menorrhagia) and the short interval between periods (polymenorrhea) require treatment. The patient explained her experience of treating menstrual disorders following a lack of coagulation factors as follows:

"I fainted two or three times. I got a bruised eye. I suffered from heavy bleeding, the bleeding did not stop. I was told [by the physicians] that I had no choice except taking contraceptive pills (LD). I took LD pills from the age of 11 till I was nearly 22 years old".

Long-term use of LD pills can be accompanied with side effects. The patient's experiences of these complications are as follows:

"*I weighed 40 or 48kg, my weight suddenly increased to 55kg and 60kg after that, and now I am 70kg! I can't lose weight, I tried a lot to do exercises, but I take LD pills all the time".*

Hemophilia patients need special attention at different stages of life, including pregnancy. One of the problems hemophilia patients face during pregnancy is the lack of treatment facilities in small towns and villages. Accordingly, the participant in this study also had to travel to larger cities to continue her treatment:

"When I was pregnant, each month I had to go to Tehran by bus while my belly was big!"

Preparing for childbirth, especially in first pregnancy, is a major concern for women with special diseases.

"... I was very annoyed after giving birth due to 70 days of bleeding. They didn't clean the inside of my uterus. It was very difficult. I was so scared that I said I didn't want a baby at all …"

Suffering

Another theme identified in this study was suffering with two subthemes of "pity and humiliation" and "social stigma".

Pity and humiliation

One of the causes of psychological suffering in people is the feeling of pity and humiliation. Feeling

pity is a kind of compassion towards a person in a situation that causes others intentionally do something that makes the person feel bad or makes them have a bad view of the person. The study participant faced humiliation and pity from close family members and strangers many times at different stages of life. This feeling even made her limit her relationships with others.

"... I was blamed a lot by my family. I mean I was really blamed because I was a girl".

"Nothing but suffering and sorrow, nothing but blame" (The participant started to cry).

"Your family looks at you differently. The first time you are looked down by someone, they always look at you the same way".

"Somehow, they look at me with humiliation (crying), so I don't accompany them so often ..."

Social Stigma

Stigma is a form of dissatisfaction and embarrassment because of the misconceptions by the society. These misconceptions may be related to one's racial identity, gender, social status, physical appearance, illness or disability, or other personal characteristics. Many examples of social stigma have been identified in the patient's experiences:

"They said she is sick, don't be with her, her disease is contagious. In a way, I couldn't go to the camp [with other students]. My life was completely different from other students (crying)".

"In general, those who did not know us well used to say that I was sick. Someone came to our house and said our house was unclean. Their house is always bloody, their clothes are bloody, and so they avoided us".

Fear and worry

Feeling worried is a mental state regarding the issues that may occur in the future over which the individual has no control. Hemophilia complications can be an important source of concern for these patients and bleeding is the most important complication that affects important life events, including marriage and motherhood. The participant in this study recounted her experiences of these two important phenomena as follows:

Fear of marriage and motherhood

Hemophilia can be transmitted genetically. Hemophilia patients can get married. As women are carriers of hemophilia, it can be transmitted from mother to child. Patients with hemophilia have specific concerns about marriage and motherhood which can affect the sweetest memories of their life.

"*I was told to inform whoever proposes to marry me about the disease. You should say that you have this disease; maybe if you don't say it, you will run into trouble".*

"As I knew that the baby I was going to have might have the disease like me, I told my husband as soon as he met me. I even kept the written document proving his agreement. I said I couldn't have a baby".

"... when I was banned from pregnancy, someone told me don't take contraceptive pills because some men don't like their wife to take these pills [OCP]. I thought I couldn't get pregnant anymore so I stopped taking OCP. But I became pregnant as soon as I stopped taking the pills ".

Fear of continuing education

Complications of hemophilia, including bleeding, create limitations for patients and may interrupt their normal course of study:

"The school year lasts 9 months, but I only went to school for three months. During the three months, I studied hard to catch up with other students, but in general, the illness made me fall behind. I could not go to school, my nose would start bleeding and sometimes the bleeding continued for a whole week".

"When I was in high school, I was very interested in art... but they told me that studying art is very expensive, and I may not afford to attend its practical classes. Maybe you won't be able to go to class; maybe you can't continue, they said. [Therefore,] I had to study humanities".

Acceptance of being different

Hemophilia is a chronic disease that affects the whole life of the patient. Acceptance of and adaptation to chronic illness depends on several factors. The participant in this study acknowledged that hemophilia had limited her. Accordingly, "acceptance of being different" was identified as another theme of this study with four subthemes, including "limiting one's relationships", "getting to know others with the same disease", "accepting fate", and "independence".

Limiting one's relationships

The patient's experiences of limitations in relationship with others were as follows:

"... For example, they would take students to excursions, but my parents would not agree. If they agreed, the school would say, 'We can't. We don't dare to take your child with us".

Getting to know others with the same disease

One of the ways to cope with the disease is to get acquainted with other patients who similarly have to cope with the difficult conditions of the disease. In

this way, the introduction of other patients by the medical staff was found to be effective in accepting the disease:

"... I had my own problems, but when I saw their problems, I forgot my own pain and I prayed to God for my health!".

Accepting fate

Belief in divine destiny calms the individual and can help the individual accept the existing conditions. The participant in this study explained situations that depicted her acceptance of destiny:

"The reality is that all husbands and wives have problems with each other, but I'm not one of those adulterers who have a problem".

"...we finally made it together. My husband says even I fight with you or beat you, you won't leave me, you won't leave my house, you won't let me go. ... Divorce is not a good thing, I didn't go back to my parents, good or bad, after all, this was my life, it was my destiny ... "

Independence

One of the components of social development is independence. However, sometimes independence is formed as a result of the lack of family support. An example of quotes from the participant that depicts independence is as follows:

"My father took us, me and my sister, to Tehran. We always went to Tehran with my father. After a while, my father said, 'Look, my daughter, I really can't take you anymore. You have to go yourself.' So I went to Tehran by myself!"

"I went to the doctor with my mother once or twice. I went to the doctor with my husband or sister for a while, but now I go alone, which means I don't need anyone else"....

Discussion

This study revealed the sufferings and challenges of living with hemophilia in a young woman from childhood to motherhood. One of the main themes identified in this study was being involved in treatment that severely affected the quality of life of the patient. This finding was in line with the findings of the study by Rambod et al. that showed hemophilia treatment affected various aspects of life, including education (9).

Another main theme identified in this study was suffering due to several reasons that led to limitations, including avoiding others. This finding was consistent with the findings of the study by Limperg et al. that reported patients experienced abandoning their hobbies and school because they felt they had to bear the consequences (for example injury and pain) and be responsible for the treatment of these consequences (10). In the present study, the participant was forced to travel alone to a specialized medical center in a bigger city to continue her treatment from the age of 17 due to the chronic nature of the disease and the poor economic status of her family. The study by Limperg et al. showed the importance of parental support for vulnerable patients and emphasized that this disease has a great burden of responsibility on parents (10).

Continuing education was one of the causes of suffering and concern in the participant in this study. In the study by Breakey et al., the daily life experience of patients with hemophilia was found to interfere with their continuing education (11). Hemophilia is a chronic disease that affects the patient's whole life. Prolonged exposure to the disease often makes the patient accept the disease. Similar findings were reported in a study by Stam et al., on young adults with chronic diseases. The negative perceptions of the disease or refusing to accept the conditions were associated with poor quality of life and feelings of anxiety and depression (12).

The participant in this study experienced severe side effects from long-term use of LD pills, which was administered to control heavy menstrual bleeding. Similar results were reported in the study by Santer et al. regarding the unpleasant experiences of women with heavy menstrual bleeding who had to take contraceptive pills (13).

Since this study is a report of the lived experiences of a female patient, in generalizing the results to other patients, cultural, social, and economic context should be considered.

Conclusion

Patients with hemophilia experience a great deal of physical, psychological, and social stress in their lives, while the community and health care staff are not well aware of them or are unable to effectively support hemophilia patients. This case study illustrated a snapshot of the challenges patients with hemophilia face from childhood to adulthood. The results of this study can help officials and medical and health staff know the patients and realize their need for a comprehensive support in different stages of life and also the need to improve their quality of life.

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Conflict of Interest

The authors declare that there is no conflict of interest.

References

1. White GC, Rosendaal F, Aledort LM, Lusher JM, Rothschild C, Ingerslev J, et al. Definitions in hemophilia. Recommendation of the scientific subcommittee on factor VIII and factor IX of the scientific and standardization committee of the International Society on Thrombosis and Haemostasis. Thromb Haemost. 2001; 85(3):560. PMID: 11307831.

2. Palareti L, Poti S, Cassis F, Emiliani F, Matino D, Iorio A. Shared topics on the experience of people with haemophilia living in the UK and the USA and the influence of individual and contextual variables: Results from the HERO qualitative study. Int J Qual Stud Health Well-being. 2015; 10:28915. doi: 10.3402/qhw.v10.28915.

3. Beeton K, Neal D, Lee C. An exploration of health-related quality of life in adults with haemophilia-a qualitative perspective. Haemophilia. 2005; 11(2):123-32. doi: 10.1111/j.1365-2516.2005.01077.x.

4. Barlow JH, Stapley J, Ellard DR. Living with haemophilia and von Willebrand's: A descriptive qualitative study. Patient Educ Couns. 2007; 68(3):235-42. doi: 10.1016/j.pec.2007.06.006.

5. Flood E, Pocoski J, Michaels LA, Bell JA, Valluri S, Sasanè R. Illustrating the impact of mild/moderate and severe haemophilia on health-related quality of life: Hypothesised conceptual models. Eur J Haematol. 2014; 75:9-18. doi: 10.1111/ejh.12328.

6. Cassis FR, Querol F, Forsyth A, Iorio A, HERO International Advisory Board. Psychosocial aspects of haemophilia: A systematic review of methodologies and findings. Haemophilia. 2012; 18(3):e101-14. doi: 10.1111/j.1365-2516.2011.02683.x.

7. Rezapoor Esfahani M, Jouybari L, Sanagoo A, Araghian Mojarad F. Experiences of living with neurofibromatosis in a young woman: A case report. The Neuroscience Journal of Shefaye Khatam. 2017; 5(3):44-50. [In Persian] doi:10.18869/acadpub.shefa.5.3.44..

8. Van Manen M. Researching lived experience: Human science for an action sensitive pedagogy. England, UK: Routledge; 2016.

9. Rambod M, Sharif F, Molazem Z, Khair K. Pain experience in hemophilia patients: A hermeneutic phenomenological study. Int J Community Based Nurs Midwifery. 2016; 4(4):309-319. PMID: 27713894.

10. Limperg P, Peters M, Gibbons E, Coppens M, Valk C, Grootenhuis M, et al. Themes in daily life of adolescents and young adults with congenital bleeding disorders: A qualitative study. Haemophilia. 2016; 22(4):e330-3. doi: 10.1111/hae.12961.

11. Breakey VR, Blanchette VS, Bolton-Maggs PH. Towards comprehensive care in transition for young people with haemophilia. Haemophilia. 2010; 16(6):848-57. doi: 10.1111/j.1365-2516.2010.02249.x.

12. Stam H, Hartman EE, Deurloo JA, Groothoff J, Grootenhuis MA. Young adult patients with a history of pediatric disease: Impact on course of life and transition into adulthood. J Adolesc Health. 2006; 39(1):4-13. doi: 10.1016/j.jadohealth.2005.03.011.

13. Santer M, Wyke S, Warner P. What aspects of periods are most bothersome for women reporting heavy menstrual bleeding? Community survey and qualitative study. BMC Womens Health. 2007; 7:8. doi: 10.1186/1472-6874-7-8.