PATIENTS’ PERSPECTIVES ON ADHERENCE TO PROPHYLAXIS IN HAEMOPHILIA:

A GROUNDED THEORY APPROACH

*Master Thesis - Nursing Science*

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Status: Definitief

Datum: 05-07-2013

Opleiding: Universiteit Utrecht, Masteropleiding Klinische Gezondheidswetenschappen, Masterprogramma Verplegingswetenschap, UMC Utrecht

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Journal: Haemophilia

Referentiestijl: Haemophilia

Aantal woorden manuscript: 3388 woorden (max 3500, main body)

Aantal woorden abstract (Eng): 269 woorden (max 300)

Aantal woorden abstract (Ned): 296 woorden (max 300)

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**BACKGROUND**

Haemophilia is a X-linked inherited bleeding disorder characterized by a deficiency of clotting factor VIII (haemophilia A) or IX (haemophilia B) [1,2]. The severity of the disease is graded according to the level of clotting factor present (mild 6-40% IU/dl, moderate 1-5% IU/dl, and severe <1% IU/dl) [3]. In the Netherlands, there are currently 1600 patients with haemophilia, including 700 with severe haemophilia [4]. Especially these patients with severe haemophilia are at risk of spontaneous bleeds in muscles and joints. Repeated joint bleeds eventually result in arthropathy, leading to severe disabilities in daily life [5]. Since 45 years, the missing clotting factor can be injected intravenously to treat bleeds and prophylactic to prevent spontaneous bleeds (± 3 times per week) [6,7]. Patients learned to inject themselves, to eventually practise this at home [8]. Home treatment with prophylaxis greatly improved quality of life and autonomy in patients with severe haemophilia [9,10]. In Europe, the majority of patients with severe haemophilia practise home treatment [11].

Prophylactic therapy is initiated around the age of two and is intended to be continued for life. Its regular application is the crucial factor in preventing bleeds, because one (large) bleed can lead to irreversible damage, especially in a joint or the central nervous system. In order to reduce the risk of bleeds the treatment should be continued indefinitely, without interrupting or discontinuation [12,13]. Adherence is defined as the extent to which a person’s behaviour corresponds with agreed recommendations from a healthcare provider [14]. Levels of adherence in severe haemophilia are not established yet (ranges adherent 44% - 87% [15,16]); however from clinical experience we believe that adherence could be improved. Furthermore, given the fact that the current treatment is expensive (€80.000 per patient/year), discontinuation can lead to decreased short term costs. However, economic aspects go beyond the short term; indirect costs, including hospital admissions, joint surgery and permanent joint damage leading to deteriorated participation on the labour market, lead to a greater impact on the society [17,18].

There is a lack of well-performed studies in the field of adherence and haemophilia [19]. Especially studies in understanding the processes which explain (non) adherence behaviour from a patient perspective are missing. However, a limited number of studies observed associations between a decreased adherence and negative perception towards prophylaxis [16], increasing age and fewer symptoms [15]. Furthermore, there are a number of theories describing the processes influencing health behaviour in chronic illnesses, yet the applicability for an effective intervention is unknown [20]. Until now, these theories have not been applied in haemophilia.

**PROBLEM STATEMENT, AIM, RESEARCH QUESTIONS**

Little is known about the determinants underlying adherence to prophylactic treatment in haemophilia, from the patient perspective. Insight in these determinants may help to design a nursing intervention to promote adherence. In haemophilia care, the haemophilia nurse has a key role in training and supporting patients with prophylaxis. Understanding the determinants in adherence helps the haemophilia nurse to provide more tailored care and support to overcome barriers. A high adherence is expected to lead to a reduced bleeding risk, a higher degree of self-management and eventually improve quality of life. The aim of this study is to explore and clarify the factors underlying adherence to prophylactic treatment in severe haemophilia, from the patients’ perspective. This study is a starting point for a nursing intervention to improve adherence and subsequent self-management skills.

The following research questions will guide the process to find the core component:

1. How do patients with severe haemophilia experience their task to administering prophylactic treatment at home in the practice of daily life?
2. How do patients with severe haemophilia experience to adhere to prophylactic home treatment?
3. What are the barriers and facilitators influencing (non) adherence behaviour in patients with severe haemophilia using prophylactic home treatment?

**METHODS**

A qualitative study was performed, based on the grounded theory approach [21,22]. This involves a process to develop theoretical understanding of subjective processes. It was chosen, due to the following arguments; 1. A qualitative design is very suitable to explore a new (subjective) emerging field, including adherence and self-management in haemophilia 2. There is a need for a detailed understanding of this complex issue 3. The existing literature was limited and not explorative [23]. This study was approved by the ethical committee UMC Utrecht (nr 12/643).

*Sampling*

During the period of February 2013 until June 2013, patients using prophylaxis were purposefully selected from two haemophilia treatment centres (Amsterdam and Utrecht, covering 50% the total of Dutch patients with severe haemophilia). Eligible patients were 18 years or older and had prescribed prophylactic home treatment with a minimum frequency of two infusions per week, for at least an uninterrupted period of two years. Patients were selected to obtain maximum variation in adherence levels, bleeding frequency and age. These constant variables were in accordance with previous literature [24] and pre-specified definitions were used (Box 1). After the initial sample, theoretical sampling was used as much as possible; new respondents selected with regard to the emerging theory.

Selected patients received a telephone call by the researcher (LS and MB) to explain the purpose of the study and to inform whether they were interested in receiving additional information. After a week for consideration, patients decided on their participation. Written informed consent was obtained prior to interviews. The proper sample size is based on theoretical saturation [21].

*Data collection*

Semi structured in-depth interviews were undertaken to unravel experiences, perceptions and beliefs concerning adherence of prophylaxis. A topic list (Box 2) was translated into an interview guide and provided direction for the interviews.

The single session interviews took approximately 45-100 minutes. The interviews were conducted by two haemophilia nurses (LS and MB), both trained in interviewing. Respondents were interviewed by the nurse from a different haemophilia treatment centre, to avoid a potential conflict of interest. The interviewers disregarded their own experiences and were open and objective towards the respondent. During the interviews they emphasized the need to learn from the participant in a non-judgemental atmosphere. Interviews were recorded at home, to create a comfortable atmosphere.

*Data analysis*

All interviews were audio taped, transcribed verbatim and anonymised. The grounded theory approach is based on a cyclic process, where established items were compared with the input of new material. All interviews were reread and summarized. An expert team of five members, including both interviewers, were involved in the entire process of analysis. The first researcher coded the interviews and discussed the results with two experts (KF, MK) [25]. During monthly meetings, all experts worked to consensus on the existing material and new material, eventually leading to the core component. During the analysis the software program nVivo 10® (QSR International Pty Ltd 2012) was used.

*Validity*

During the different stages of the study, reliability and validity criteria were embedded in the study procedures. Internal validity of the data collection was established by literally transcribing the interviews and experts who provided feedback on the interview style of both interviewers. The use of researcher triangulation in all phases of the study enhances the validity of the interpretation. External validity was increased through a maximum variation sample.

**FINDINGS**

Fourteen of the sixteen selected patients participated in the study. In Table 1 the demographic and background characteristics of the respondents are shown. All respondents were men, as haemophilia is a X-chromosomal inherited disorder. Their ages varied from 21 to 63 years. Also in adherence levels, bleeding frequency, living arrangement and educational level maximum variation was obtained. Saturation was reached at the main themes; no new information was found on the main components after 14 interviews [21].

This study revealed that ‘*the position of haemophilia in patients’ life’*, was the core concept influencing adherence to prophylaxis in haemophilia. Table 3 provides illustrative quotes supporting the present findings. Through the complexity of the core component, it was not possible to answer each research question separately and therefore was chosen for a coherent display of the findings.

**Adherence behaviour**

Some patients judged themselves to be adherent, while their reported behaviour showed that they created their own regimen. Patients seemed to judge their adherence by their own standards, which differed from the prescribed regimen. Forms of non-adherence were; stopping/ decreasing prophylaxis with a (minimum) of one infuse per week, postponing treatment, using on demand treatment or over treating (increasing the dose or frequency).

Mostly this ‘created’ regimen was not in accordance with the prescribed regimen. However, patients were persistent in their behaviour for years; therefore some healthcare providers permitted the patient initiated treatment changes. Most patients were satisfied with their own regimen and were convinced they made the right choice. Respondents mentioned that they were prepared to take risk on severe bleeding and subsequent consequences when they were non adherent. It was unclear if the respondents were fully aware of the risk on traumatic or intracranial bleeds.

Respondents, who followed the regimen correctly, found prophylactic treatment it easy to follow. Especially, experiencing a ‘natural reminder’ (symptoms/ bleeds) was a good motivator for taking the treatment. Sometimes they postponed the prophylaxis until the afternoon, due to time pressure in daily life or forgetfulness. Usually this behaviour was unconsciously and when they became aware of this, the treatment was administered as soon as possible. Furthermore, often respondents mentioned that taking prophylaxis gave a pleasant feeling, physical or psychological.

**Handling during the occurrence of a bleed**

Self-monitoring of the body during the occurrence of a bleed, was experienced as difficult, symptoms differed in each person. Furthermore, it was difficult judging whether to treat or not, especially distinguishing between haemophilic arthropathy and a joint bleed was difficult. Sometimes this led to over treatment, due to fear for bleeding. There were various descriptions of experiencing a bleed, there was no key factor for treating or not.

Respondents who followed the regimen correctly found that treating bleed was not an issue. The patient mostly chose to treat directly, or discuss the situation with the treatment centre. In addition to infusion of clotting factor concentrates (CFC), most of the respondents used crutches (when it concerned a bleed in the lower extremities) and tried to take some rest. However, respondents who experienced difficulties with following the regimen, only administered CFC in case of severe bleeding with pain. Small bleeds, especially in muscles were not treated at all and they found follow-up treatment unnecessary.

**The position of haemophilia in patients’ life**

The underlying reasons for differences in adherence behaviour seemed to be influenced by the position of the haemophilia patients gave in their life.

The patient can adopt one of the two basic positions: ‘acknowledge the illness and the influence on their life’ or ‘not being prepared to let haemophilia influence their life’. This basic position determined the way patient see their disease and subsequent consequences of the disease. This was the major determinant influencing adherence.

Acknowledge their illness and the influence on their life, must be understood as acceptation of haemophilia and its necessary adjustments in their life. Goals in life were adapted to the haemophilia, e.g. finding suitable education, work and hobbies. Most respondents experienced bleeds and some had serious joint limitations. Prophylaxis was a condition to have a normal life. Positivity about prophylaxis dominated: ‘It gives me a regular life’, ‘I can work’ or ‘It gives me freedom’.

Not being prepared to let haemophilia influence their life, must be understood as: ‘I am not sick’ and ‘I don’t want to adapt my activities to haemophilia’. Individual obstacles against administering treatment, e.g. needle-phobia, fear of HIV or confrontation with haemophilia, were reasons for not taking the treatment as prescribed. They experienced that haemophilia and prophylaxis restricted their life. Furthermore, it seemed that these respondents also had or experienced less bleeds, which made it easier for them to avoid prophylactic treatment.

**Barriers and facilitators leading to a theoretical model**

These findings led to the development of a model (Figure 1), which combines the barriers and facilitators leading to the actual adherence behaviour in the patient. Each factor can be explained as both a barrier and a facilitator; extremes are described below to show the differences.

The basic position towards the haemophilia was strengthened by the following determinants: *attitude towards prophylaxis, willingness to take risks, bleeding frequency and knowledge.* Adherent patients had a positive attitude towards the medication and had confidence in the prescribed regimen, in contrast to the non-adherent respondents whom were negative or ambivalent towards prophylaxis. Furthermore, adherent respondents stated that they had a healthy fear to risk moments and often avoid them. On the other hand, the non-adherent respondents accepted the fact that took a risk to experience severe bleeds. A separate group was identified here, the group of respondents who treated more than prescribed. This group was not prepared to take any risks at all, seemed to overestimate symptoms of bleeding or were afraid they would regret when they not treated (extra). It was unclear whether this group actually experienced more bleeds. Another influencing determinant was the bleeding frequency; adherent patients stated they often experienced bleeds. The non-adherent respondents stated that they experienced less bleeding and therefore they found it legitimate to delay or avoid the treatment. Knowledge influenced patients’ behaviour; adherent respondents had a good knowledge and followed the instructions carefully. The knowledge of non-adherent respondents was adjusted to their own behaviour and they had an own interpretation of the prescribed regimen. It was insufficiently clear whether factors concerning disclosure of the illness, social support/ family relations, relationship with the health care provider and age were influencing adherence.

**DISCUSSION**

This is the first qualitative study explaining the factors of (non) adherence behaviour in severe haemophilia. The ‘position of haemophilia in patients’ life’ was defined as a new framing concept which provides insight in the factors of being adherent or not. It describes the difference between adherent and non-adherent patients through the basic position: acknowledge or not acknowledge the illness. Adherent patients were prepared to acknowledge their haemophilia and its influence on their life. In contrast, non-adherent patients were not prepared to let haemophilia influence their life. This process was influenced by four major determinants: 1) the attitude towards prophylaxis, 2) the willingness to take risks, 3) the bleeding frequency and 4) level of knowledge.

Some limitations of this qualitative study should be discussed. First, saturation was reached on the main themes; due limited time, minor determinants could not be further investigated. However, maximum variation in respondents was followed accurately, which contributed to the internal validity of this study. Second, existing literature demonstrated age/ puberty as an important determinant of adherence. Age could not be demonstrated as an influencing factor in this study, possibly due to ethical restrictions of including respondents below eighteen years. Finally, there were some limitations concerning the external validity. Only patients could be included who were willing to talk about their haemophilia. Patients, who cannot face the confrontation with their illness, are missing in this sample. Furthermore, cultural factors could influence the attitude towards haemophilia, therefore a larger sample is needed to identify these factors.

Greater understanding of the processes concerning adherence could be obtained from a literature review in haemophilia and from studies in other chronic illnesses. A systematic review about barriers and motivators towards adherence in haemophilia was recently performed in this group [19]. Despite the limited number of studies available, some significant factors were defined. Factors extracted from the literature were: belief in in necessity of treatment, absence/ presence symptoms, age and relationship with the health care provider. The first two determinants are in accordance with our findings. Belief in necessity of treatment may correspond with the (positive) perception towards prophylaxis. The third and fourth determinant could not be demonstrated yet in this study. A possible explanation could be that the data on these specific subject was insufficiently saturated and possibly to age limitations (>18 years).

In the absence of other data in haemophilia, other conditions, especially those occurring requiring life-long continued treatment, were studied. Vervoort et al. [26] performed a comparable study in patients affected with HIV. She stated that acceptance or non-acceptance of HIV lead to two basic stances towards adherence. These basic stances are in accordance with our finding. Influencing determinants in HIV (ambivalence towards medication, HIV and medication are part of their life or not, positive experiences after (non) adherence behaviour and knowledge) show major similarities with determinants in haemophilia.

Furthermore, Hortensius et al. [27] studied this phenomenon in Diabetes Mellitus type 1 and insulin-treated type 2, using a focus group method. Hortensius et al. described the core concept ‘perception of self-monitoring: friend or foe’. Respondents assigned to be adherent perceived the self-monitoring in a positive way (as a friend). This is comparable to adherent haemophilia patients who had a positive perception towards prophylaxis. The non-adherent respondents, experienced difficulties with the procedure and never feel free. This is also seen in our non-adherent population who has a negative attitude towards prophylaxis.

The ‘position of haemophilia in patients’ life’ and its determinants is akin to theories explaining adherence behaviour, like the health belief model or social cognitive theory [28,29]. The attitude towards medication or illness, outcome expectancies, perceived threat and expected controllability, play an important role in the intention/ motivation to perform the actual behaviour. Different is, as mentioned before that social influence could not be identified (yet) in haemophilia. Self-efficacy seems a promising determinant to improve adherence, however this could not been demonstrated in other studies [20,30] as well in our study.

*Clinical implications*

The present findings have both clinical and research implications. Being non adherent as a consequence of not acknowledge their disease, could have a large impact on patients’ quality of life. From a clinical perspective, it is important to identify patients’ perception towards haemophilia and the position the patient give in their life. These identified determinants in Figure 1 could be used as a ‘benchmark’ during the consultation. In the group who don’t prioritize their haemophilia, there is a need for development of psychological intervention focussing on acceptance of their haemophilia. More research is needed to define which psychological method could be successful in this group.

Both groups, adherent and non adherence patient can benefit from ongoing support and positive feedback. Supporting skills of self-management and self-monitoring as health care provider, but also peer-to-peer, is a need that patients indicated. The development of structured program with (e-)learning components and peer-to-peer support could be a helpful tool to support the life-long dedication.

Patients who experience difficulties with forgetfulness a real time medication reminder could be a useful tool. Patients receive a SMS reminder when they forgot to open their (‘smart’) pillbox. Studies in diabetes care have shown that real time medication reminders improve adherence rates with 11% (80% test-group vs 71% control-group) [31]. Therefore, this tool could be useful in this specific group, however no studies has proven this effect yet in haemophilia. This reminder could be used with other forms as well, like appointment-adherence, ordering medication and keeping their logs.

The present study was targeting adults only. To define adherence issues in parents administering their child, adolescents and young adults, it is useful to repeat this study in patients/ care givers below 18 years.

**CONCLUSION AND RECOMMENDATIONS**

This study explored the experience of adherence to prophylaxis in patients with haemophilia. The concept of ‘the position of haemophilia in patients’ life’ is described as a core concept which provides insight in the process of being adherent or not. It means: in adherent patients that they acknowledge their illness and the influence on their life. In contrast to non-adherent patients, who were not prepared to let haemophilia influence their life. This opinion was formed by four major determinants: 1) the perception towards prophylaxis, 2) the willingness to take risks,3) the bleeding frequency and 4) level of knowledge.

Obtained from this conclusion, the following recommendations were formulated:

* Identify non adherent patients using this model as a benchmark.
* Development of a psychological intervention to help patients with accepting/ acknowledging their illness and eventually increase treatment success.
* Development of a structured self-management program to provide ongoing support for each patient, to enhance or continue the life dedication. Peer-to-peer support could be a helpful method in this program.
* Providing patients who have issues with forgetfulness a real time medication reminder.
* Repeat this study in children/ parents and adolescents to gain a broader patient perspective on this subject.

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**ABSTRACT**

*‘Patients perspectives on adherence to prophylaxis in haemophilia: a grounded theory approach’*

**Background:** Haemophilia is an inherited bleeding disorder, were especially patients with severe haemophilia are at risk for spontaneous bleeds. These bleeds can be prevented with prophylactic treatment; however a lifelong, high level of adherence is needed. To provide adequate support and individual recommendations as haemophilia nurses, it is important to understand the processes underlying adherence in patients with severe haemophilia.

**Aim:** The aim of this study is to explore and clarify the factors underlying adherence to prophylactic treatment in severe haemophilia, from the patients’ perspective.

**Method:** A grounded theory study was undertaken and involved one-time individual in-depth interviews with 14 adult patients with severe haemophilia using prophylaxis. This prescribed prophylactic treatment consists of an intravenous injection ± 3 times per week, performed by the patient self (at home).

**Findings:** ‘The position of haemophilia in patients’ life’ was identified as the core concept. It means: in adherent patients that they acknowledge their illness and the influence on their life. This is contrast to non-adherent patients, who were not prepared to let haemophilia influence their life. The core concept was formed through four major determinants: the perception towards prophylaxis, the willingness to take risks, the bleeding frequency and level of knowledge.

**Conclusion:** This concept provides a framework for the main reason for being adherent or not. The core concept and its determinants clarify adherence behaviour, anticipation during the occurrence of a bleed and the basic position towards their illness.

**Recommendations:** Identification of non-adherent patients, using this concept as a benchmark for risk assessment. For this specific group, development of a (psychological) intervention to change behaviour could increase the treatment success.

**Keywords:** Adherence, compliance, qualitative study, determinants, barrier, facilitator.

**ABSTRACT**

*‘Patients perspectives on adherence to prophylaxis in haemophilia: a grounded theory approach’*

**Achtergrond:** Hemofilie is erfelijke bloedstoornis, waarbij patiënten met ernstige hemofilie risico hebben op spontane bloedingen. Deze bloedingen kunnen worden voorkomen door het nemen van profylactische behandeling, echter hierbij is adequate therapietrouw van groot belang. De hemofilie verpleegkundige speelt hierbij een belangrijke rol in de ondersteuning van de behandeling. Deze studie kan hier aan bijdragen door inzicht te geven in de processen rondom therapietrouw bij patiënten met ernstige hemofilie.

**Doel:** Inzicht krijgen in de processen die een rol spelen bij het volgen van de profylactische thuisbehandeling bij patiënten met ernstige hemofilie.

**Methode:** Een ‘grounded theory’ studie, waarbij gebruik gemaakt werd van eenmalige individuele diepte interviews. Deze werden afgenomen bij patiënten met ernstige hemofilie die ongeveer 3 per week profylaxe thuis zelf toedienen.

**Resultaten:** ‘De plaats van hemofilie in het leven van de patiënt’ was geïdentificeerd als het hoofdcomponent. Dit betekend dat trouwe patiënten de ziekte erkennen en ook bereid zijn om zich hier aan te passen. Ontrouwe patiënten erkennen de ziekte minder en zijn ook minder bereid om het in te passen in het leven. Deze bevinding werd ondersteund door vier determinanten: perceptie over profylaxe, risico bereidheid, bloedings-frequentie en kennis niveau.

**Conclusie:** Dit concept biedt een kader voor de hoofdreden om de behandeling correct te volgen of niet. Het verklaart therapietrouw gedrag, de handelingswijze tijdens een bloeding en de ideeën over hun ziekte.

**Aanbevelingen:** Door gebruik te maken van dit ontwikkelde concept kunnen ontrouwe patiënten geïdentificeerd worden. Voor deze specifieke groep is ontwikkeling van een mogelijke (psychologische) interventie nodig, om zo het succes van de behandeling te vergroten.

**Sleutelwoorden:** Therapietrouw, naleving, kwalitatieve studie, determinanten, barrière, facilitator.

Box 1. Definitions used to create a purposeful sample.

|  |  |
| --- | --- |
|  | Definition: |
| *Adherence level* | Adherent: followed the prescribed regimen for at least 80%-100% of the time;  Non adherent: followed not the prescribed regimen or for less than 80% of the time. |
| *Bleeding frequency* | Low: less than 5 bleeds per year;  Moderate: between 5-15 bleeds per year;  High: more than 15 bleeds per year. |

Box 2. Topic list

|  |
| --- |
| * Experiences with prophylaxis   + Integration of treatment in daily life   + Impact of prophylaxis   + Personal and medical history * Beliefs and perceptions concerning prophylaxis * Acceptance * Adherence   + Barriers   + Facilitators   + Self-monitoring of symptoms * Knowledge * Social influences   + Disclosure * New treatment developments * Patient characteristics |

Table 1. Demographic and background characteristics.

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| ID number | Adherence level\* | Age category | Bleeding frequency† | Living arrangement‡ | Education§ |
| 1 | Adherent | 60 | Moderate | F | High |
| 2 | Adherent | 20 | Low | M | High |
| 3 | Adherent | 40 | High | M | Low |
| 4 | Adherent | 50 | Moderate | A | Middle |
| 5 | Non adherent | 20 | Low | P | High |
| 6 | Non adherent | 20 | Moderate | P | Low |
| 7 | Non adherent | 50 | Low | M | Middle |
| 8 | Non adherent | 50 | Low | F | Middle |
| 9 | Non adherent | 50 | High | F | Low |
| 10 | Adherent | 60 | Moderate | F | Middle |
| 11 | Adherent | 20 | Low | A | High |
| 12 | Non adherent | 40 | Moderate | F | High |
| 13 | Non adherent | 20 | Low | P | Middle |
| 14 | Non adherent | 30 | Low | P | Low |

\* Adherent: followed the prescribed regimen for at least 80%-100% of the time;

Non adherent: followed not the prescribed regimen or for less than 80% of the time.

† Low: less than 5 bleeds per year, Moderate: between 5-15 bleeds per year, High: more than 15 bleeds per year.

‡ Living with parents (P), Alone (A), Married or cohabiting (M), Family with Children (F).

§ Low: primary school, lower secondary general, Middle: higher secondary general education, intermediate vocational education, High: higher vocational education, university.

Table 3. Core component and factors influencing adherence, including illustrating quotes.

|  |  |  |
| --- | --- | --- |
| Component | Examples | Illustrative quotes (in Dutch) |
| Actual adherence behaviour | Non adherent:  ‘Prepared to take the risk’  Adherent:  ‘Treatment is easy to follow’  ‘Prophylaxis gives a pleasant feeling’ | *‘And in former times, when I thought ‘ I will do some risky things today’ (motorracing), I took something medication precaution. But nowadays, I won’t do it anymore, because I prefer to not inject at all.’*  *‘If you do it twice per week, then it’s just like eating. Yes, it’s easy to remember it.’*  *‘I always found it difficult to draw a firm conclusion, because it can also be psychological, but after prophylaxis I walk better. My knees, it seems that my knees find it pleasant, they feel smooth.’* |
| Anticipation during the occurrence of a bleed | Non adherent:  ‘Each injection was considered carefully’  ‘Only treat when it is really necessary’  Adherent:  ‘Treating bleeds is not an issue’ | *‘When I have overcome my resistance, ‘yes, now it is really necessary’, than I inject. The cognitive process to do it is difficult.’*  *‘I have a great resistance towards injecting. I always had a great resistance. There’s something in my head that says no I don’t want it, I won’t need it.’*  *‘When it’s a small bleed in the muscle, I skip the injection. Only, when it’s really painfull, in my knees for example, than I will.’*  *‘When I have a bleed, I treat immediately. And I also use cruthes.’* |
| **Core component:**  The position of haemophilia in patients’ life | Non adherent:  ‘I don’t want haemophilia to guide my life’  ‘Haemophilia limited my freedom’  Adherent:  ‘I only have a normal life with prophylaxis’ | *‘I have a very strong motto: I’m not listening to my body, my body is supposed to listen to me.’*  *‘When I had decided to go out on Saturday night, than I just went, because I didn’t want to lay on the couch due to a bleed. That’s not possible.’*  *‘Prophylaxis has a very positive influence on my life. I don’t think I could manage without it, knowing from my own experience that it’s impossible.’* |

Figure 1: Model explaining being (non) adherent to prophylactic treatment in severe haemophilia.

|  |  |  |
| --- | --- | --- |
| *Influencing determinants:* | Adherent patients | Non adherent patients |
| **Position of haemophilia in**  **patients’ life** | Acknowledge/ Accept their illness and the influence on their life | Not being prepared to let haemophilia influence their life |
| * *Perception towards prophylaxis* | Positive attitude towards prophylaxis | Negative or ambivalent attitude towards prophylaxis |
| * *Willingness to take risks* | Healthy fear of traumas | Prepared to take the risk  Or no risk at all = over treatment |
| * *Bleeding frequency* | High or moderate bleeding frequency | Low or decreased bleeding frequency |
| * *Knowledge* | Knowledge in accordance with provided education and following the instructions carefully | Knowledge adjusted to own behaviour and own interpretation of the prescribed regimen |