The subjective experience of living with haemophilia in the transition from early adolescence to young adulthood: the effect of age and the therapeutic regimen

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ABSTRACT
The main aim of the research is to study how youths affected by haemophilia, a congenital hemorrhagic chronic disease, make sense of their condition, with particular reference to the transition from early adolescence to early adulthood. We administered face-to-face semi-structured interviews to 20 Italian youths with haemophilia, aged 11–25 years, in on-demand treatment or prophylaxis therapy. A thematic analysis was performed with the help of software for textual data to figure out the main topics and the role of the two selected variables in the emergence of the themes (age and type of therapy). The results highlight how the experience of suffering from haemophilia is organized around five core themes (fragmented body, intimacy, family history, autonomy, dreams), that are more or less typical of some age group or kind of treatment. These results may be useful for designing appropriate and differentiated interventions for psychosocial support.

Introduction
Transition from adolescence to adulthood is a complex process (Gorter, Stewart, & Woodbury-Smith, 2011). For chronically ill youths it is even harder, because of problems imposed by the disease and by the therapies, the assumption of responsibility for care and the progressive awareness that the disorder will accompany them throughout their life (Björquist, Nordmark, & Hallström, 2015; Petrini & Seuser, 2009; Plevinsky, Gumidyala, & Fishman, 2014).

In this research we have studied youths suffering from haemophilia, a rare bleeding disorder that results from reduced levels or lack of clotting factor VIII (haemophilia A) or IX (haemophilia B). Haemophilia A and B are X-chromosomal recessive disorders affecting half the children of carrier females. The frequency and the severity of the haemorrhages that characterize this illness are, in general, proportional to the severity of the clotting factor deficiency. Bleedings can be spontaneous or can follow trauma; they occur in the joints, muscles and soft tissues, causing damage, bruising or swelling, often accompanied by pain and physical restrictions. Treatment consists of the intravenous administration of the deficient clotting factor that can take two forms: on demand or prophylactic. In the former case, the factor is administered after a bleeding episode, requiring timely intervention to avoid long-term damage, and skill in evaluating symptoms. In prophylaxis, the factor is administered regularly even in
the absence of bleeding episodes: this reduces the anxiety over the risk of recurrent haemorrhages, but the 2 or 3 infusions per week can be quite invasive, thus entailing a significant psychological and organizational burden that affects patients’ everyday life. Nowadays, it is a very different disease compared to a few generations ago due to effective and safe clotting factor concentrates available for treatment.

The medical progress of the last few decades has reduced the life-threatening events, improved patients’ lives and prevented chronic complications; nevertheless, there are still some physical and psychosocial issues that impact the quality of life of people with haemophilia (Beaton, Neal, Watson, & Lee, 2007; Cassis, Emiliani, Pasi, Palareti, & Iorio, 2012; Emiliani, Bertocchi, Potì, & Palareti, 2011). These issues are more evident in some age transitions, such as that from early adolescence to young adulthood (Simmons, Frick, Wang, Miller, & Fragueiro, 2014). While in childhood, parents are personally responsible for their child and for all care practices, the patient becomes progressively more independent and needs to transfer personal responsibility from parental to self-care. However, the literature has found that this taking of responsibility proves difficult for young people, confirming, for example, poor adherence to treatment in adolescence, a common issue in haemophilia as well as in other chronic diseases (Boyce, 1997; Dovey-Pearce, Doherty, & May, 2007; Kyngäs & Rissanen, 2001; Lindvall et al., 2006; Mazanderani, Locock, & Powell, 2012; Petrini & Seuser, 2009). To account for these phenomena, most psychological studies investigate the impact of chronic disease in adolescents and young adults focusing on individual variables, such as anxiety, depression, lack of self-esteem, the risk of suicide (Ghanizadeh & Baligh-Jahromi, 2009; Kyngäs & Rissanen, 2001; Litt, Cuskey, & Rosenberg, 1982).

Other authors, in a less individualized perspective, have studied the ideographic character of the illness experience focusing on the complexity of psychosocial aspects, both relational and contextual (Christian & D’Auria, 1997; Goldstein & Kenet, 2002; Markova, 2009; Nilson et al., 2012; Weisner, Matheson, Coots, & Bernheimer, 2005).

Some recent reviews on the psychosocial issues of haemophilia (Breakey, Blanchette, & Bolton-Maggs, 2010; Cassis, Querol, Forsyth, & Iorio, 2012) claim the need for further as well as international studies capable of exploring subjects’ perspectives and giving guidance as to how to improve comprehensive patient care. This is in line with recent guidelines referred to other chronic illness, which emphasize the importance of integrating the psychosocial domain within routine care (Holland, Watson, & Dunn, 2011). Indeed, the literature offering insights into the personal experiences of haemophilia is not vast and the available studies on young people are even less frequent (Khair, Gibson, & Meerabeau, 2011, 2012; Nilson et al., 2012; Simmons et al., 2014). Such studies on the subjective aspects of haemophilia have been carried out almost exclusively in Northern Europe, the USA and Canada. Moreover, the concept of transition has only been studied within a limited timeframe, and referring to the transition from paediatrics to adults services within the health care system, in countries where the ‘Haemophilia Centres’ are organized in this way (Betz, Lobo, Nehring, & Bui, 2013; Sterling, Nyhof-Young, Blanchette, & Breakey, 2013).

To increase our understanding of transitions in haemophilia, we have initiated a research project aimed at understanding how the subjective experience of haemophilia might change from adolescence to adulthood. One of the first specific goals of this study has been to investigate the subjective meanings of living with haemophilia throughout the developmental stages from puberty to adulthood. A further goal was to evaluate whether and how the use of prophylactic or on-demand treatment may impact the disease experience. Such an understanding can guide counselling and education to the patient and the parents by implementing health care practices that are culturally and developmentally appropriate.

The context of this research is Emilia Romagna, a region in northern Italy where Haemophilia Centres located in different cities provide care for patients of all ages, who from diagnosis are always treated in the same clinic by the same team, and where a national Summer Camp for young people with haemorrhagic diseases is located. The treatment is guaranteed to everyone in need by the Italian National Health System.

We carried out interviews with youths aged from 11 to 25 years old. This age range was motivated by the fact that, according to the literature and the indications of the World Health Organization (Gorter et al., 2011), at the present time the transition processes are not well-defined through rites of passage or by clear boundaries. Hence, in this paper we will use the general term ‘youths’ for teenagers and...
young adults from 11 to 25 years, and will divide this range into three subgroups: early adolescence, middle adolescence and young adulthood.

**Method**

**Research design**

In order to understand the subjective meanings by means of which the adolescents and young adults interviewed make sense to their experience of illness, in this study we adopted a qualitative research design with a bottom-up approach (McCarthy, Sullivan, & Wright, 2006). In particular, semi-structural interviews were carried out with youths aged between 11 and 25 years.

All the interviews were recorded and verbatim transcripts were gathered in a single textual corpus (omitting the interviewers' questions). A thematic analysis was performed on the whole text, with the help of a software for textual data, already used in health research (Caputo, 2014; Denzin & Lincoln, 1994; Reinert, 1993). Specifically, we used T-Lab, which was preferred to other software programs like Atlas-Ti or NVivo also because of its rich Italian dictionary, the language of the interviewees. The software analysis was carried out by a trained researcher (Crowley, Harre, & Tagg, 2002; Lancia, 2004).

The research output was interpreted separately by three of the authors, according to an abductive thematic approach which aims to interpret the sense-making process (Rizkallah, 2013) from a psycho-social point of view (Markova, 2009). By comparing the different considerations, we reached a common data interpretation.

**Participants**

All the participants were male and recruited at the Haemophilia Centre of the University Hospital and from a national Summer Camp for young people with haemorrhagic diseases. Patients and parents, in the case of minors, were first contacted by their physician and then received a letter from the University that presented the research aims and methods, offering a coupon for the purchase of books or musical products and asking for parents’ informed consent for underage patients. Twenty of the 35 people contacted agreed to participate and these were then divided into 3 age groups, 11–15 year olds (n = 8), 16–19 year olds (n = 4), 20–25 year olds (n = 8). The therapeutic regimen is equally distributed between each group, for a total of 10 subjects receiving prophylactic therapy and 10 on-demand. Specific characteristics of participants are illustrated in Table 1.

| Interview order | Age groups | Therapeutic regimen | Type and severity of haemophilia |
|-----------------|------------|---------------------|---------------------------------|
| # 3             | 1 (11–15)  | On-demand           | Mild A                          |
| # 13            | 1 (11–15)  | On-demand           | Mild B                          |
| # 11            | 1 (11–15)  | On-demand           | Moderate A                      |
| # 12            | 1 (11–15)  | On-demand           | Severe A                        |
| # 20            | 2 (16–19)  | On-demand           | Moderate A                      |
| # 4             | 2 (16–19)  | On-demand           | Mild A                          |
| # 2             | 3 (20–25)  | On-demand           | Mild A                          |
| # 9             | 3 (20–25)  | On-demand           | Mild A                          |
| # 10            | 3 (20–25)  | On-demand           | Moderate A                      |
| #17             | 3 (20–25)  | On-demand           | Moderate A                      |
| # 1             | 3 (20–25)  | Prophylaxis         | Mild A                          |
| # 15            | 1 (11–15)  | Prophylaxis         | Moderate A                      |
| # 5             | 1 (11–15)  | Prophylaxis         | Severe A                        |
| # 8             | 1 (11–15)  | Prophylaxis         | Severe A                        |
| # 14            | 1 (11–15)  | Prophylaxis         | Severe A                        |
| # 16            | 2 (16–19)  | Prophylaxis         | Severe A                        |
| # 6             | 2 (16–19)  | Prophylaxis         | Severe B                        |
| # 7             | 3 (20–25)  | Prophylaxis         | Severe A                        |
| # 18            | 3 (20–25)  | Prophylaxis         | Severe A                        |
| # 19            | 3 (20–25)  | Prophylaxis         | Severe B                        |
Each participant was interviewed individually face-to-face by a trained psychologist. The semi-structured interviews lasted about 60 min and were divided into three sections: (1) the first section explored self-perception, body image and the organization of daily life; (2) the second investigated the coping strategies, the perceived social support and compliance/non-compliance to therapy; (3) the third part investigated the image of the future and professional and personal plans. The University research ethics committee approved the study.

Data analysis

The analysis: (i) yielded a map of the general meanings by which adolescents and young adults make sense to their life with illness; (ii) allowed us to identify a list of the most specific themes as a form of recognition of word patterns within the data; (iii) provided information about how the themes are particularly relevant for one or more subgroups of respondents.

(i) The analysis graphically shows a map of the whole text: a factorial plane, by extracting number of factors (usually 2 or 3), which best summarize the means of the discourse. These factors represent the axes of the factorial plane and are characterized by dichotomy polarities, i.e. opposite processes of meaning. Each polarity is formed of different keywords that most often co-occur in the same parts of the text ($V$-test $> \pm 2$). After interpreting the keywords that characterize the pole of each factor, the researchers then give them a label.

(ii) The analysis yielded the significant themes as clusters. Through the analysis, we can also infer the degree of relevance for each theme, compared to the entire text (defined by the ratio between the total number of sentences in the text and the number of significant sentences that contribute to the construction of that particular cluster). The software also extracts a list of quotations relevant to each theme and a list of the keywords that are associated most often in these quotations, ordered by value of $\chi^2$. We assigned a label to each cluster according to the quotations and the keywords. Clusters are represented in the factorial space.

(iii) Through the analysis we can consider how some structural variables are related to each theme. This relationship emerges from the research output and is graphically suggested by the position of structural variables on the factorial plane. In this study, two variables were chosen by the researchers: (a) type of therapy (prophylaxis, on demand); and (b) age group (1: 11–15 years; 2: 16–19 years; 3: 20–25 years). At first, the type of haemophilia (A or B) and the level of severity (mild, moderate, severe) were also considered as variables, but the results showed they did not lie in a significant position in the factorial space, and for this reason they are not discussed in this paper. Other researchers have similarly shown that these aspects do not have a significant impact on the patients' subjective experience (Hegeman et al., 2011).

Results

We present below the results of the thematic analysis, illustrating the factors, the clusters, and their relationship with the variables.

The analysis identified two factors that describe the main dichotomous oppositions in which the variability of the whole text can be organized (Figure 1). The first factor (the horizontal axis) shows a process organized around the opposition between body and personality. In the negative polarity (on the left) the factor represent the disease in its medical and physical aspect, with blood and bruises, pain and fear of haemarthroses, and the parts of the body involved in the traumatic episodes. On the positive polarity (on the right) there is no allusion to the disease, replaced instead by reference to leisure and educational activities where personality aspects such as being shy, nice or good are fundamental. The second factor (the vertical axis) instead shows a contrast between a historical perspective of the disease, with hopes and plans for the future (down), and a focus on episodes and limitations in the here and now (up).
The analysis revealed also 5 clusters placed on the factorial plane: (1) Disclosure; (2) Fragmented body; (3) Autonomy; (4) Genetic responsibility; (5) Dreams and passions. The relevance of each cluster in terms of percentage compared to the general richness of the text, the set of meaningful keywords that co-occur, and a set of quotations for each cluster are provided in Tables 2, 3 and 4.

With reference to the variables, the two modes of therapy are opposed in the factorial plane. On-demand therapy is placed in a zone of the space marked by attention to the body and to the symptoms frequently encountered in everyday life. It is associated with a representation of the disease as a traumatic and unpredictable event. Prophylaxis is in a zone of the factorial plane particularly marked by factors and clusters linked to issues like hobbies, personality traits, dreams and interests. It seems that patients in this kind of therapy develop a subjective thinking on personal identity as a whole, rather than just limited to the body.

The first age group (11–15 years) shares a representation of the disease and treatment strongly anchored to the theme of the body, the second (16–19 years) focuses on experience of being ill and questions on how and whether illness can affect their personalities. Finally, the investigation of family history and genetics, the search for explanations that make sense of their condition and the effort to realize personal autonomy, are typical of young adults between 20 and 25 years of age.

**Discussion**

The most representative theme focuses on the issue of disclosure, that is whether or not to hide or to reveal their chronic condition. This is the crucial issue participants have to face, especially in their relationships with friends and teammates. The theme is generally significant for each patient with a chronic illness (Bukowski, Hoza, & Boivin, 1993; Christian & D’Auria, 1997; Damon & Hart, 1988; Hegeman et al., 2011). In haemophilia, previous studies have shown that people tend to hide their disease for as long as possible, in order to avoid being treated differently, or as untouchable at school or at work: they tend to
Table 2. Percentage of sentences for each Cluster compared to the total sentences.

| Cluster | % Cluster sentences | N° Cluster sentences | N° Total sentences |
|---------|---------------------|----------------------|-------------------|
| 1       | 37.65               | 468                  | 1243              |
| 2       | 20.11               | 250                  | 1243              |
| 3       | 18.83               | 234                  | 1243              |
| 4       | 12.23               | 152                  | 1243              |
| 5       | 11.18               | 139                  | 1243              |

Table 3. Main keywords for each Cluster (in order of $\chi^2$).

| Cluster | Keywords and $\chi^2$ |
|---------|------------------------|
| Cl. 1 ‘Disclosure’ | Friends (24,7), tell (14,9), playing football (12,32), know (11,89), team (10,77), city (10,13), group (10), child (9,49) study (9,4), relationship (9,4), fine (7,87), parents (7,68), hyperactive (6,64), convince (6,6), basketball (6,4), together (6,4), week (6), tired (5,8), on-demand (5,57), prophylaxis (5,57), angry (5,3), patience (4,3), participate (4,3) |
| Cl. 2 ‘Fragmented body’ | Days (99,14), bruise (80,33), blood (74,36), factor (62,97), feet (49,78), knee (45,37), ankle (41,96), elbow (41,80), surgical intervention (37,52), clot (34,46), explain (30,32), cut (29,67), strain (27,96), prophylaxis (25,25), attention (22,9), serious (22,27), remember (21,99), haemorrhage (18,46), episode (17,14), age 1 (15,24), doctor (13,99), leg (13,17), medical examination (13,08) |
| Cl. 3 ‘Autonomy’ | Pain (40,37), home (36,43), go (26,71), work (23,82), university (20,88), walk (18,69), Age 3 (15,12), night (14,85), week (14,07), intravenous (14,07), call (12,98), consider (12,98), pill (12,74), infuse (12,74), follow (12,65), decide (11,85), summer (10,28), dad (10,15), difficult (9,38), day (9,26), stop (9,11), need (8,85), trade (8,28), company (8,28), panic (8,28), month (7,90), environment (7,70), grow up (7,14), be (7,03), drug (6,95), crying (6,76), practice (6,58), management (6,42), indifference (6,42), emergency room (6,41), leave (6,41), alone (6,00), daily (5,56), hourly (5,56), available (5,56), worse (4,45) |
| Cl. 4 ‘Genetic responsibility’ | Carrier (92,77), responsible (46,62), healthy (44,48), limit (37,36), accept (34,86), family (32,31), be set aside (22,98), illness (19,86) understand (19,78), grow (17,16), future (16,77), body (16,77), searching (16,64), genetic (16,10), diagnosis (15,85), children (15,00), others (14,60), rationalizing (13,40), possibility (11,60), grandfather (11,41), psychological (10,91), overcome (10,67), mother (8,46), wrong (8,42), be like others (7,24), be different (6,73), pay (6,15), analyze (5,33), affection (4,79) |
| Cl. 5 ‘Dreams and passions’ | Music (120), school (100), character (87), nice (87), becoming (75), shy (74), play (71), choose (54), listen (48), extroverted (31,8), band (31,6), guitar (31,6), swim (31,6), passions (27,26), piano (27,12), tennis (25,72), physical aspect (23,93), good (23,93), write (22,33), interest (20,46), emotions (16,45), sport (13,35), chat (12,04) |

Table 4. Examples of quotations for each cluster.

| Cluster | Quotation |
|---------|-----------|
| Cluster 1 | I play a match with friends who already know that they cannot pull (Age 3, on demand) |
|         | My issue of haemophilia is known by one or two friends, all the others do not know about it, that is my choice, because I do not want my friends, even when you are playing football or basketball […], not to give your best for fear of getting hurt (Age 3, on-demand) |
| Cluster 2 | I need surgery on my foot. For example, I had no knee injury, they are very healthy, the other foot is very healthy, my left elbow is very healthy, in the right I have had several problems, since I started prophylaxis I stopped having ankle problems (Age 3, prophylaxis) |
|         | The bruise lasts much longer: I explained the disease very casually, because I don't want anyone to worry (Age 3, therapy on-demand) |
| Cluster 3 | Everyone had a scooter or a bicycle, they could go out easily, I always had to be ferried around, I was not independent, I had to call to be picked up and be taken home, and so this was a deprivation (Age 3, prophylaxis) |
|         | About autonomy, the nurse once asked me if I wanted to learn how to give myself intravenous injections. I looked at her with my eyes popping out and said: Are you out of your mind? (Age 3, on-demand) |
| Cluster 4 | I was a little angry […]. The question I often asked myself was why it happened to me […] and it's not too easy to understand for a child (Age 3, prophylaxis) |
|         | My girlfriend said: just think if I were a carrier, our child would necessarily be haemophiliac or carrier (Age 3, on demand) |
| Cluster 5 | What is my dream? To do well at school, to become a famous guitarist and not have haemophilia! (Age 1, prophylaxis) |
|         | I have […] a passion for music. The rest … such as physical appearance, it’s a bit … well, it is just not one of my things … (Age 2, prophylaxis) |
be more secretive, isolate themselves because of embarrassment, or due to the fear of being excluded from the group or of losing their job or partner (Beaton et al., 2007; Betz et al., 2013; Bukowski et al., 1993; Damon & Hart, 1988; Markova, 2009). The phenomenological characteristics of this disease and the progress made in the medical treatments tend to make this state of invisibility possible. Furthermore, for adolescents and young adults, as our results highlight, differences from the healthy peers in a period normally marked by competition, can bring out emotions of anger and frustration, due to the feeling of being marginalized with regard to activities, such as sports, important for socialization, self-esteem and group membership. An interesting aspect of our results shows that disclosure is a relevant topic for all the patients, whatever forms of treatment they follow. In fact, in the case of on-demand therapy they may be worried about how to justify their concerns about getting hurt. On the other hand, the same topic also appears to be relevant for patients undergoing prophylaxis for two different reasons: the practice of infusion which sometimes can be visible, and the lack of symptoms which makes more salient the disclosure as an aspect of intimacy in the relationship with friends. Moreover, the words of the cluster reveal that parents are also involved in this topic, and the disclosure plays a key role within the process of negotiation of choices in family life.

The second most relevant theme that emerged is the issue of body-image. From our findings adolescents and young adults represent their body as fragmented and suffering, with bruises and haemarthrosis which makes the disease visible through symptoms (associated with states of alarm and attention), stressing once again the difference from the ‘healthy’ people. As indicated by Khair et al. (2011), the main problem for the patients is not the haemophilia itself, but the bleeding. Our respondents do not manifest an integrated body self-perception, which is described in its most hurt and painful parts, producing a fragmented representation of it (Jodelet, 1997). These youths are indeed quite capable of giving a detailed reconstruction of the visible manifestations of their disease and its treatment, explaining objectively, in scientific terms, bleeding and bruises to the others, acting as expert patients. This expert knowledge requires the doctor (a word that appears in the cluster) as an active interlocutor and relevant point of reference, considered in his/her professional value and without any affective connotation. Finally, compatibly with the advent of puberty and the body changes, this theme is particularly significant for young people between 11 and 15 years (Biolcati, Ghigi, Mameli, & Passini, 2016), and is associated with on-demand therapy, as already shown in our previous studies (Emiliani et al., 2011). Another significant topic refers to autonomy and to the process of constructing an autonomous daily life in contexts outside the family. According to our results, this process requires a set of organizational skills and implies a control of emotions (like panic and fear) related to not being able to cope with the daily grind and unexpected events. As indicated in the literature, the transition towards the autonomy of young to adult age is problematic in the life of every individual (Jacquin & Levine, 2008), but may be more difficult for a young haemophiliac, since he/she has to cope with his/her daily life, with the suffering, the pain and the treatments, elements that keep on reminding him/her of his/her fragility and the condition of being dependent on others. According to Beaton, Neal, and Lee (2005), young adults find it less hard to accept and integrate the illness and treatment in everyday life when they have a job that makes them feel useful in society and attaches less importance to contact sports or other risky activities. Instead, being unemployed (as in the case of our sample) makes the process of a direct responsibility with respect to their disease even harder. In addition, not only everyday life but also pleasant moments such as summer and holidays are problematic, because they might require an emergency room close at hand in order to manage possible traumatic episodes. This theme is particularly characteristic of young adults aged between 20 and 25 years.

Related to this topic, the following in order of importance is the theme of genetic responsibility, typical of young adults between 20 and 25 years. In this theme, the illness is approached from the perspective of hereditary family histories. Young adults seem to try to make sense of their present and future condition by searching in family history and genetics for a rational attribution of cause in a scientific and logical manner. The purpose of this search for meaning is to be able to accept the chronic condition, process it and make sense of the latent feeling of being a victim of a genetic error, different from the others, in an actual frame of ambivalent feelings towards the mother carrier and the
offspring. Young adults also have to face the future and they have to make decisions in respect to the reproductive choices and expectations of having a child.

Lastly, the last theme that emerged, less representative than the others, is linked to passions and personal interests, and mainly characterizes youths undergoing prophylactic therapy. This issue, specific to late adolescents, concerns whether or not haemophilia has influenced their personality, interests or motivations, such as study and work or the kinds of hobbies. It seems that these adolescents are wondering: ‘Does it depend on me or on the illness?’; proving that they have not yet integrated the disease into their own identity. Adolescence is experienced as a time for constructing identity and personality, a time marked by the desire to please others and to be skilled in certain tasks, but also by sadness and regret because of the limits imposed by haemophilia. Although the disease does not emerge explicitly in the keywords of the theme, it seems to be present through the limits it imposes. Indeed, hobbies and passions that appear in our data are only related to possible activities for a haemophiliac, without risks, such as playing the piano, guitar, tennis and swimming.

**Strengths and limitations of the study**

Although it must be borne in mind that haemophilia is a rare disease, the small number of participants can certainly be considered a limitation to our study, so that further studies will be needed to have a more complete understanding of the transition from adolescence to adulthood.

Notwithstanding this limitation, a particular strength is the balanced distribution of the sample with respect to the variables chosen, which has allowed for an adequate interpretation of the data. This condition is particularly meaningful for exploring the differences related to the type of treatment, as studies conducted in other countries – differently influenced by local resources and health care policies – hardly ever have balanced samples.

Another strength is represented by the contemporaneous focus on the subjective experience of youths and the use of statistic software that allows us to preserve the typical richness of qualitative data and to be accurate as well as rigorous.

Finally, our analyses, unlike other thematic research, have enabled us to pay attention not only to the main issues emerging in the patients’ speech, but also to bring out the relationship between the core themes and the structural variables, allowing us to obtain more specific results.

**Issues for clinical practice**

The interpretations of the material analysed can be helpful in structuring educational interventions and/or psychological and social support. These interventions may differ according to the age group so as to accompany their process of transition.

First of all, we think that it could be particularly helpful for children with haemophilia aged from 3 to 10 years to offer psycho-educational actions designed to recover their perception of being ‘lived-in-bodies’, not just ‘bodies under care’. We consider the case of psychomotricity in particular, which facilitates the integration of functions and representations of the body (Aucouturier, 2005; Manuzzi, 2009). Indeed, if the clinical approach, which identifies the body with the organism and offers a fragmented vision of it, becomes internalized by children, this does not help teens to reconstruct a vital and satisfying – albeit bruised – body image. The theme of the body has emerged as especially important for those treated on-demand who could, therefore, be the target for this type of intervention.

For teenagers, peer education groups could be organized, with the aim of reinforcing communication strategies and sharing emotional experiences related to revealing the disease to friends. It is noteworthy that this aspect of our study is significant for patients both undergoing on-demand therapy and prophylaxis.

For young adults psychotherapeutic help may be offered to facilitate the ability to assign new and more adaptive meanings to the illness and family history, redefining expectations, desires, fears, and values, recognizing and encouraging the expression of emotions and not the defence from them (Beaton
et al., 2007), and supporting them in the planning of their future, also with regard to the choice of having a child.

Special training for teachers, coaches and parents can be organized, in order to improve their understanding of actual limits and real possibilities of youths with haemophilia. As concerns parents, our results show that they are often perceived by adolescents as being overprotective and concerned about them, whereas they are increasingly occupied with a process of autonomous self-care and the need to improve their self-efficacy (Khair et al., 2011).

Conclusions
Recalling the statements of the sociologist Frank, Thomas and colleagues (2007) remind us that health is a construct that depends on meanings and experiences lived out by patients, calling into play attitudes, interpretations, beliefs, emotions and subjectivity. While going through the objectivity of the disease, patients develop their own points of view about the condition that they experience intimately and socially.

In this research, we have explored the subjective meanings of living with haemophilia to figure out how the experience changes alongside the development of adolescence, and how the type of therapy can affect this transition.

Our results show that the meanings attributed to the illness go together with representations of the self. This means that for the youngest the centrality of the body (both suffering and undergoing change) highlights images of the self overall as a patient. The image of such a fragmented body suggests how much psycho-educational actions like psychomotricity can prove useful. In mid-adolescence, the problem of disclosure reveals that illness is at the heart of the system of relationship with peers and adults. In search for their personal identity, adolescents question themselves about the impact that haemophilia can have on their attitudes and personality. For such adolescents, peer education groups can be useful to improve communication strategies and to cope with emotions. Young adults, on the other hand, tend to investigate family histories and try to understand the steps in genetic transmission having in mind an own family in the future. Actually, their efforts are oriented to achieving a place in society through high education. They also express their concern for the self-management of treatment over time and in contexts outside of the family, the difficulty of integrating the therapies into everyday life and the need to accept the weight of the physical pain of the disease. For this age group, free access to psychological counselling can be very important.

As regards the types of therapy, we have to consider that, commonly, less severe patients are treated on-demand, while the more severe forms are treated in prophylaxis. Our results evidence that on-demand youths are more concentrated on their body and on the worry about getting hurt, while the others, notwithstanding the burden of the 2 or 3 infusions per week, are keener to talk about interests, hobbies, and dreams and, in this sense, seem to have a better quality of life. In spite of this diversity, the topic of disclosure is common to respondents undergoing both types of therapy.

In conclusion, as we had envisaged, the results of this research have allowed the Haemophilia Center of Bologna to implement some specific and well-directed measures for adolescents and young adults.

Note
1. Generally speaking, due to a combination of social, economic and cultural factors, young people in Italy continue to live with their parents for longer than in other countries. A recent European survey, for example, indicates that 46.7% of Italians between 25 and 31 still live with their parents, as opposed to 1.8% in Denmark and 13.7% in the UK.

Acknowledgements
We wish to thank all the adolescents and young adults who participated in this study. We also wish to acknowledge F. Barosi, G. Bondi, S. Bertocchi professional psychologists, R. Brigati, Professor of moral philosophy at the University of Bologna, and Ch. Biasoli haematologist of the Summer Camp for their contribution in realizing this research.
Disclosure statement
No potential conflict of interest was reported by the authors.

Funding
This work was supported by the Health Committee of the Emilia Romagna Region (Italy).

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