

Research Article

Representation of illness in Familial Amyloidotic Polyneuropathy Portuguese Association newspaper: A documental study

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Abstract

This study explores illness representations within Familial Amyloidotic Polyneuropathy Portuguese Association newspaper. A content analysis was performed of the issue data using provisional coding related to the conceptual framework of the study. All dimensions of illness representation in Leventhal's Common Sense Model of illness cognitions and behaviors are present in the data and reflect the experience of living with this disease. Understanding how a person living with an hereditary, rare, neurodegenerative illness is important for developing community nursing interventions. In conclusion, we suggest an integration of common sense knowledge with other approaches for designing an intervention program centered on people living with an hereditary neurodegenerative illness, such as familial amyloidotic polyneuropathy.

Key words

documental study, familial amyloidotic polyneuropathy, hereditary disease, illness representation, Portugal.

INTRODUCTION

Familial amyloidotic polyneuropathy (FAP) is a rare, hereditary, irreversible, and fatal neurodegenerative disease resulting from a genetic mutation (Ando *et al.*, 2013). FAP mainly affects the peripheral nervous system at a late age of onset, between 25–35 years, and causes high dependency levels, suffering, and premature death (Conceição, 2006). The extracellular deposition along the peripheral nervous system of insoluble amyloid protein fibers (transthyretin) and amorphous compounds is reflected by a sensory loss that starts in the feet, accompanied in some cases by pain in the lower limbs, in contrast to the loss of thermal sensitivity that arises in others (Said, 2003). Diagnosis can be difficult, especially if there is no previous family history. Although it endemically originates on the northwest coast of Portugal, multiple endemic foci have been discovered at a high prevalence in different countries, such as Sweden and Japan (Sousa & Saraiva, 2003; Ohmori *et al.*, 2004; Zaros *et al.*, 2008). Considered a rare disease, molecular testing has proved that it has

spread worldwide and outside the well-defined and identified endemic foci (Fleming *et al.*, 2009). The Portuguese mutation, the most prevalent, is also accompanied by a more severe symptoms, an earlier onset, and an anticipation phenomenon of symptoms dependent on gender (Lemos *et al.*, 2014). Treatment options for patients with FAP are very limited and focus on palliation of symptoms to halt disease progression or to stabilize the tetramer (Suhr *et al.*, 2000; Coelho *et al.*, 2013). Symptomatic treatment can improve the quality of life of these patients, and is addressed primarily to peripheral neuropathy (Ando *et al.*, 2013).

Literature review

In daily life we construct beliefs and representations of the world that surround us. Similarly, when someone experiences a symptom, like a headache, or receives some health/illness information, they seek meaning for this new information (Weinman & Petrie, 1997). Disease perception, beliefs, and representations influence coping responses and adaptation to a new health situation (Petrie & Weinman, 2006). The Common Sense Model of illness cognitions and behaviors provides a framework to assist in understanding the experience of patients with any chronic illness (Baines & Wittkowski, 2013). This model is organized in four stages: (i) the moment an individual perceives a stimulus, symptom, or result of a health test; (ii) identification, which corresponds to the construction of the illness representation and emotional responses based on the stimuli previously perceived; (iii)

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coping behaviors, which emerge from the previous stage and direct themselves to the illness and emotional control responses; and (iv) appraisal, which requires the evaluation of coping outcomes (Leventhal *et al.*, 1997). The illness representation formation is an active process that involves emotional and cognitive proceedings, recursive stages of appraisal and representation, and a hierarchical process of concrete and abstract thinking (Leventhal *et al.*, 1997). For the construction of illness representation, this model presents five components or dimensions that relate to each other and contribute to the building of a mental model: identity of the illness, causal beliefs, timeline beliefs, cure or controllability beliefs, and consequences of illness (Petrie & Weinman, 2006). Inherited genetic diseases pose a challenge to illness representations, because, as shown in some studies, people emphasize the characteristics inherited from parents, but do not realize the relationship with genes (Marteau & Senior, 1997). FAP is an inherited neurodegenerative disease, in which the age of onset of the disabling symptoms occurs in young adults. In other neurodegenerative and disabling diseases, the role of illness representation is well known, and mediates prognosis and quality of life (Vaughan *et al.*, 2003; Graves *et al.*, 2009; Ferreira *et al.*, 2010). Media represent an important means to disseminate information about diseases, treatments, and available resources. However, not all images displayed by the media are correct, often inducing erroneous and stigmatizing beliefs in the community about people with certain diseases and influencing the illness representation of the person living with the disease (Collin & Hughes, 2011; Drazic & Caltabiano, 2012; Holland, 2012; Rukavina *et al.*, 2012). In the case of FAP, as an hereditary disease with a strong psychosocial impact, it is important to perform an analysis of the news published by the association representing the interests of the persons living with it. Is it important in order to understand more about people's lived experience with the disease in order to deliver patient centered nursing interventions

Study aim

This study explores the illness representations expressed in the newspapers published by the Familial Amyloidotic Polyneuropathy Portuguese Association.

METHODS

The primary source of this qualitative study was data collected from the newspapers published by the Familial Amyloidotic Polyneuropathy Portuguese Association since December 1980. All issues published from 1980–2014 were analyzed. A total of 44 issues were obtained from the Familial Amyloidotic Polyneuropathy Portuguese Association and from municipal libraries. Content analysis was performed using Milles *et al.*'s (2014) qualitative data analysis method. The authors were guided by the Common Sense Model, a conceptual framework (Leventhal *et al.*, 1997). Content analysis was performed using provisional coding related to the study's conceptual framework (Saldaña, 2009). All articles published in the journal were screened, and news related to the illness or the experience of living with FAP were

included in the study. News of funding events, association activities, and financial reports were excluded because they were not directly connected to the object of the study. The representativeness of this study was achieved because we used all of the issues published to date. Because of the small volume of data and categories, we chose to perform a manual content analysis without recourse to a computer program for data processing. Different colors were assigned to the categories and a deep reading of all articles was performed. A database with the different categories was created a priori in which relevant registration units were transcribed and coded.

RESULTS

The authors analyzed the 44 issues published; as this newspaper was not regularly published until 1999, there were some years in which only an annual issue was published and there were also years in which no issue was published. Since 1996, only one issue has been published annually, usually in December. Five categories emerged that were then included into a core category – illness representation.

Identity

The identity domain includes disease diagnosis or the label attached to the disease and associated symptoms. The identity of the illness is related to the patients' beliefs about possible or actual labels for the condition and the symptoms experienced. The most common diagnostic designation was *Paramiloidose*, a popular term designating the disease. In more recent news, we note the use of the term “familial amyloidotic polyneuropathy” or “FAP.” There are two ways to identify an individual diagnosed with FAP: the first is by using the term “*Paramiloidótico*,” identifying the person with the illness; the second is to designate them as an FAP patient or a person with FAP. We found a relationship between the signs and symptoms of the disease, which are used as designators, such as *doença dos pezinhos*, an idiomatic expression that cannot be translated into English but relates to the most visible symptom, that is, impaired walking.

Cause

Beliefs about the causes of a disease are important in building a representation of illness. After reading the news, we have identified two causes for FAP: biochemical and genetic/hereditary causes. Biochemically, the cause is attributed to a disorder in the accumulation of amyloid substance. The exemplar used within the journal to express biochemical causes are:

The importance of the foreign substance, amyloid, that accumulates in nerves and other organs of patients. J.2,3,4

It has been shown that the protein that becomes abnormal substance gives the name of the disease: amyloid J.18

... in FAP a protein that normally circulates in the blood has a conformation which leads to abnormalities deposit[ed] in various locations in the body including peripheral nerve, in the form of a substance called amyloid J.20

This causal belief is present in the first combination of issues, as well as in the most recent issues, and is particularly associated with the appearance of tafamidis. The assignment of the disease to hereditary/genetic causes has family, emotional and social implications, as can be verified by the excerpts presented:

Patients are carriers of a genetic abnormality that they transmit to a part of their children this ability to manufacture the abnormal protein J.2,3,4

I know it is a hereditary disease that is transmitted from parents to children J.7,8,9,10

Where (...) is discovered that I had "inherited" my father this strange disease J.11

It is known today the responsible gene, its chromosomal location, the mutation suffered and mutant protein that results from it. J.18

Some expressions reinforce or affirm this genetic cause, such as:

Genetic defect J.23

The origin is genetic J.24

Timeline beliefs

From our data, we can see that perceptions of the natural course of the disease are related to the age of the onset of symptoms and the progression of severity of symptoms to disability and death:

One of the earliest symptoms in Paramiloidose is the digestive disorders, often accompanied by poor appetite and marked weight loss J.1

Early in the disease Paramiloidóticos begin to have digestive complaints – Constipation, bouts of diarrhea, prolonged and difficult digestion, morning vomiting J.13

The first symptoms appeared in the form of bites, lack of muscle strength, lack of sensitivity to hot and cold and also pain, mainly in the lower limbs J.43

Through these examples, we understand the natural course of the disease and the expected severity in the progression of symptoms. Representations submitted in the newspapers show a serious, progressive illness, which, until very recently, led to death involving great suffering:

Terrible disease (...) that is chronic, progressive and disabling J.11

The outcome is always fatal (...) No cure J.23

I have to live with (...) it is chronic J.28

... A disease that has no cure and inevitably (...) led patients to death prematurely J.41

Cure or controllability beliefs

Beliefs about disease control emerged from the news analysis, related with timeline beliefs of chronicity. One

transmitted belief is the possibility to control or eradicate the disease in progeny, either by refusing to have children, the use of prenatal diagnosis, or preimplantation diagnosis. The achievement of presymptomatic diagnosis in the offspring of patients with FAP was discussed in the first issue. More recent articles introduced the notion of self-care management and its influence in health decisions:

... The only way to wipe out the disease would be that individuals at risk (...) decided, knowing its mode of transmission not [to] have children J.7,8,9,10

Early diagnosis of (...) Paramiloidose is the most effective means of disease containment (...) [and] will allow accurate genetic counseling J.11

Do some physical activity. J.43

Beliefs of the role of health professionals in the control or cure of this disease mainly focus on the need for specific training to care for patients, the need for proximity care services, and research of new treatments. For some patients, the lack of effective treatment leads to a sense of hopelessness and a loss of confidence in healthcare professionals. Poor information and the detachment of some health professionals toward this disease, lead to a loss of confidence in disease control:

... An evil that no one deigns resolve after so many years it exists in our country (...) and others (...) will suffer, waiting for someone of medical profession should say that this is in the way to being cured; it has now gone many years of fruitless studies. J.7,8,9,10

Liver transplantation (...) changed the situation until then incurable disease and has changed the way patients live and regard disease. J.37

Since I started taking the medication, the changes have been slow but positive. J.41

Consequences

Representations of the consequences of the disease on patients allow us to evaluate its impact in the short and long term at physical, emotional, social, and economic levels (Baines & Wittkowski, 2013). This dimension emphasizes individual subjectivity in perceptions of disease severity. Perception of the physical consequences relates to the impact that the symptoms have on everyday activities. These representations of FAP are conceptualized by personal experience of the disease, but also by past experience of parents or other relatives:

Disease (...) which causes muscle weakness of the limbs (...) Begin to lose strength in the feet, legs and hands (...) Often (...) the patient is unable to move. J.18

Patient is (...) often in pain, unable to walk normally. J.24

When I was seven I realized that my father was not like other parents, was very thin, walked with difficulty, what

little he ate did him harm, he had constant diarrhea, was incontinent, and he did not distinguish heat and cold in his hands and feet J.36

Perception of emotional consequences relates to beliefs connected with the progressive evolution of symptoms, the suffering associated with the disease, and social isolation that the disease leads to. Previous experiences with parents' illness construct a perception of the disease, associated with suffering and hopelessness:

I went through severe depression, having come to think the worst (. . .) What else has cost me so far is not exactly physical suffering itself, but rather accepting the disabilities that this disease has brought me. J.11

We feel that in the skin, we all live fast, because we say from past experience that the time of the great forces will be short and the failure is coming. J.24

When I look in the mirror it happens to me to see my father's face (. . .) I realize there's a bit of his suffering now in my body. J.36

Perceived economic and social consequences go hand in hand. The economic consequences discussed initially focused mainly on the cost and access to medicine in relation to those with low or non-existent incomes, and frequently regarding large families with small children. In the most recent issues, this social and economic aspect is less frequently mentioned; instead, the difficulty of travel to hospital to acquire liver transplant medication or tafamidis is often discussed:

The ongoing support of the disease requires medications, mostly at prohibitive prices, despite the contribution J.2,3,4

Force (. . .) the individual to leave his employment too early, causing a reduction in household income J.23

Others have (. . .) increasingly difficult access to consultations J.42

The news discuss perceived social consequences, referring to social rights, poor housing conditions, and the consequences of diagnosis to decisions regarding choices of employment, marriage, and children:

There are patients living in poor conditions, as all the houses are not architecturally prepared for its shortcomings. J.11

If one has this knowledge it will be more prepared to face your future and may in time make certain choices in your life according to the characteristics of the disease J.11

The diagnosis may influence their career choices (. . .) and can still influence family planning J.24

DISCUSSION

Through our analysis of the FAP Portuguese Association journal articles and use of the Common Sense Model as a conceptual framework, we have identified five dimensions:

identity, causal beliefs, timeline beliefs, cure or controllability beliefs, and the consequences of the illness (Leventhal *et al.*, 1997). The FAP patients' perceived self-care deficit and beliefs about treatments are only superficially discussed in the articles. Identification of this disease is mostly performed by diagnostic label and the carrier of the disease or patient is identified mostly with an adjective derived from it. The label that designates the person that lives with a genetic condition is important because, in this case, the disease is a part of the biological identity of the person (Mendes, 2007). This is more significant, because the FAP is an inherited genetic disorder, whereby from the moment that the patient carries the genetic diagnosis it is registered in his identity (Marteau & Senior, 1997; Mendes, 2007). Identification of the symptoms of the disease can lead the patient to devalue side effects of the treatment or non-adherence behaviors. The popular use of the expression *doença dos pezinhos* used to designate this disease is evidence of this identification of a disease by one visible symptom. (Llewellyn *et al.*, 2007; Baines & Wittkowski, 2013). Beliefs regarding the causes for the appearance of a disease allow us to know the perception of the factors or conditions that led to the emergence of a disease (Llewellyn *et al.*, 2007). It is important to know if the individual's attitude or behavior influences the course of the illness, as well as their emotional responses, including the allocation of responsibility for the disease (Byrne *et al.*, 2005). Biochemical mechanisms or genetic/hereditary mutations are attributed as the causes of this disease. The question of heredity leads us to the social and emotional consequences of the disease, as well as the representations of individual disease control (Marteau & Senior, 1997). Biochemical causes lead us to perceive physical consequences and a natural course of the disease, while representations of disease control signify the importance of health professionals and treatment. Causal belief influences decisionmaking of individual and family aspects of future life, the choice of professional carrier, paternity, marriage, and treatment decisions in FAP carriers (Martinho *et al.*, 2012). Timeline beliefs, related to perceptions of the duration of the disease, particularly in the case of chronic illness, are important because of the close relationship with treatment adherence (Llewellyn *et al.*, 2007). The prognosis and natural course of FAP leaves no room for hope. We know that patients who undergo a liver transplant experience illness progression after 10 years, and we know that the efficacy of the transthyretin tetramer stabilization drug is 60% (Obayashi & Ando, 2012; Coelho *et al.*, 2013; Oshima *et al.*, 2014). If the timeline were prolonged, a person with FAP could hope to live longer with a better quality of life than their parent, even when the anticipation phenomenon is present (Lemos *et al.*, 2014). These beliefs are related to the ideation of by cure or control dependent on personal resources, treatments, or by the action of health professionals on the illness (Ferreira *et al.*, 2010). Until 1990, there was no hope of control over disease progression; support and palliative treatments were the only available options. Liver transplant was the first effective treatment to slow down disease progression; however, many patients died during the procedure, and the later a patient underwent a transplant, the lower quality of life they could expect because

of remaining symptoms (Suhr *et al.*, 2005). There is currently no curable treatment; the disease can be controlled by treatment for a period of time, but the carrier will still transmit the mutation to the next generation. There are three sources of information to build a representation of the disease according to the Common Sense Model of Illness, namely, physical experience of the illness, information provided by an agent (healthcare professional, media, family or friends, patient associations), and information based on past experience of the disease (Hagger & Orbell, 2003; Baines & Wittkowski, 2013). Studies conducted within this conceptual framework of patients with FAP determined that these are also the funding sources for obtaining information about the disease (Jonsèn *et al.*, 1998; 2000; Martinho & Martins, 2011).

Limitations

This study was conducted with the limitation of having a unique source, the newspaper published by the Familial Amyloidotic Polyneuropathy Portuguese Association. Determining the existence of other similar associations and their focus on the disease, would be beneficial in order to study local illness representation. The perception of a serious, progressive, and fatal disease that affects an individual at a young and active age has consequences that influence life choices of patients. However, articles alone cannot demonstrate the influence of family history on previous beliefs and perceptions of the disease and its impact on self-care of the patient (Leventhal *et al.*, 1998). It is also difficult to understand the relationship between disease stage and illness representation of the patient and their family (Arran *et al.*, 2013).

CONCLUSION

The illness representation of a disease is a very important part of a system of beliefs, affecting a healthy transition. This representation is built not only on an individual's previous illness experience, but also by the images the media conveys of that specific disease, which constitutes a version of the social representation of that illness. In order to promote healthier transitions and to deconstruct incorrect beliefs, it is important for nurses to be aware of the social representation of an illness. The choice of the Common Sense Model in this study was helpful in achieving the proposed goal. It was possible to identify all the dimensions proposed by the model, and to establish tenuous links between these dimensions. Although this is a rare disease worldwide, endemic foci affects a significant number of families. New treatment possibilities launch important data that needs to be studied in the light of this model, namely the introduction and cost of orphan drugs used, as well as any changes that result from the stabilization of disease progression that can, in turn, bring a new representation of the disease.

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CONTRIBUTIONS

Study Design: SN, FM.
Data Collection: SN.
Data Analysis: SN, FM.
Manuscript Writing: SN, FM.

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