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The experience of thalassemic adults with their treatment

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This study, based on medical anthropology and oral reports, analyzes the meanings attributed by thalassemic adults to their experiences with the treatment. Interviews were used to collect data, which were analyzed through inductive thematic analysis. Eleven young adults, six of whom were men, at different ages, with different educational levels and occupations participated in the study. The meanings are discussed through the theme "the lives of patients with thalassemia in relation to their treatment". This core meaning highlights the difference made in their identity by having the disease trait, the recognition of the importance of adhering to their treatment, the difficulties in maintaining their social functions, the patients' irregular treatment adherence and their justifications for non-adherence to their treatment. Thalassemic patients conform to their condition and employ a normalization strategy to control the disease and justify irregular treatment adherence.

Descriptors: Beta-Thalassemia/therapy; Culture; Nursing.

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A experiência dos talassêmicos adultos ao tratamento

O objetivo neste estudo foi analisar os sentidos dados pelos talassêmicos adultos à sua experiência em relação ao tratamento, com base na antropologia médica e no método do relato oral. Para a coleta de dados, usaram-se entrevistas e seguiu-se a análise temática indutiva. Participaram onze adultos jovens, seis do sexo masculino; com idades, níveis educacionais e profissões distintas. Os sentidos são apresentados pelo tema "a vida do portador de talassemia com o tratamento". O tema destacou a identidade de diferença pelo traço da doença, o reconhecimento da importância da adesão ao tratamento, as dificuldades em manterem suas funções sociais, os episódios de irregularidade na terapia e suas justificativas. Apreendeu-se que os portadores estão resignados com sua condição, empregam a estratégia de normalização para o controle da doença e justificaram a irregularidade no tratamento.

Descritores: Talassemia Beta/terapia; Cultura; Enfermagem.

La experiencia de los talasémicos adultos con su tratamiento

El objetivo de este estudio fue analizar los sentidos dados por los talasémicos adultos a su experiencia con el tratamiento, con base en la antropología médica y en el método del relato oral. Para la recolección de datos usamos entrevistas y seguimos el análisis temático inductivo. Participaron once adultos jóvenes, seis del sexo masculino; con edades, niveles educacionales y profesiones, distintas. Los sentidos de la experiencia son presentados por el tema "La vida del portador de talassemia con el tratamiento". El tema destaca la identidad de ser diferente por: el trazo de la enfermedad, el reconocimiento de la importancia de la adhesión al tratamiento, las dificultades en mantener sus funciones sociales y los episodios de irregularidad en la terapia y sus justificaciones. Aprendimos que los portadores están resignados con su condición y emplean la estrategia de normalización para el control de la enfermedad y para justificar la irregularidad en el tratamiento.

Descriptores: Talasemia Beta/terapia; Cultura; Enfermería.

Introduction

Forms of thalassemia belong to a group of genetic diseases called hemoglobinopathies that compromise the normal production of chains of alpha and beta globins, which form hemoglobin. The incidence of thalassemia in Latin America and the Caribbean is heterogeneous, about 1% to 2% of the population. It was introduced by Portuguese, Spanish and Italian immigrants. The prevalence of thalassemia major, which is the most severe clinical form, is greater in the south and southeast of Brazil, affecting 1% of the population⁽¹⁾.

Signs and symptoms of the disease emerge at the end of the first year of life and disappear with the diagnosis and beginning of treatment, and growth goes back to normal. If treatment is not initiated, the child may present severe general complications and die within the first or second decade of life⁽¹⁻²⁾.

Because of advanced knowledge concerning the disease and the effects of therapies, health professionals currently care for patients with thalassemia who have reached adult age. Treatments for thalassemia major include regular, lifetime, transfusions of red blood cells associated with iron chelation therapy. Although repetitive transfusions are a palliative therapy, it enables patients to develop normally, improve their quality of life and reach adult age⁽¹⁾.

The transfusion scheme recommended for individuals with thalassemia major is approximately 100 to 200 ml of concentrated red cells per kg/year aiming to maintain blood hemoglobin between 9.0 g/dl and 10.5 g/dl. Repetitive transfusions cause complications such as: hemolytic reactions, alloimmunization, the possibility of seroconversion, and, mainly, the accumulation of iron in

various organs. This accumulation leads to retardation of growth and sexual maturation, endocrine abnormalities, among other problems, although cardiac complications are more severe and account for 50% of deaths⁽³⁾.

There are three types of iron chelators employed in clinical practice: 1) Desferrioxamine (Desferal) must be subcutaneously or intravenously infused by pump at night on a regular basis: five or more times a week; 2) Deferiprone was the first oral chelator to be licensed; its employed dose is divided into three daily doses; 3) Exjade (Deferasirox) is taken in a daily single dose via various effervescent tablets. These drugs have different actions on the various organs and their combined use may optimize the iron chelating effect without, however, increasing the intensity of side effects. The therapy has to be personalized to each patient according to each one's profile of iron overload and clinical condition⁽³⁾.

The use of oral chelators is recent. Patients using them follow a monthly medical monitoring regime due to resulting organic reactions and have their kidney functions and ferritin assessed⁽¹⁾.

The interruption of transfusions and/or chelator infusions does not immediately induce symptoms. Prolonged interruptions, though, worsen the patient's physical condition and iron concentration in different organs may increase, leading to an irreversible condition and death⁽¹⁾.

For optimum adherence, the patient needs to attend a specialized service at least once a month for screening tests and a subsequent blood transfusion, introduce Desferal subcutaneously by infusion pump for eight hours daily, for a limited period, and/or ingest the appropriate dose of Deferiprone and/or Deferasirox.

Non-adhering patients with thalassemia major are reported by various authors⁽²⁻³⁾ and are a cause for concern. However, no Brazilian study addressing the socio-cultural elements influencing the experience of patients with treatment was found. Such knowledge can facilitate nursing care and also care provided by other professionals, which indicates the importance of this study.

The following questions are expected to be answered in this study: How do patients perceive their treatment? What is it like to be obligated to submit to treatment for life? What are the reasons one does or does not adhere to treatment? What is life like under a therapeutic regime? The meanings held by adult thalassemic patients concerning their experience with the therapeutic regime are analyzed in this study.

The impact of thalassemia major and its treatments on the life of adult patients

A study addressing treatment adherence among 40 American individuals older than 18 years old showed that 37 of them used chelators through infusion; 77% of them had missed at least one dose in the previous month due to its side effects such as pain and irritation at the application site. The authors concluded that the chelator's side effects are responsible for non-adherence to treatment⁽⁴⁾.

The expectations of thalassemic individuals in relation to life composed the topic of a study involving 67 Greek patients aged between 18 and 45 years old. Of these, 75% had a bachelor's degree, 71% worked, 67% had negative treatment adherence and 77.6% had important complications. The results showed that pessimism in relation to the future was predominant among those with complications and lower levels of education; a better expectation in relation to life was observed among men⁽⁵⁾.

The hypothesis that the greater the knowledge one has in relation to a disease, the greater treatment adherence was investigated in a group of 32 patients, aged on average 17 years old. The results confirmed such a hypothesis: 43% had inaccurate knowledge of the disease and treatment and only 48% adhered to the therapeutic regime⁽⁶⁾.

Authors related educational level and occupation with other factors such as age, sexual and ethnical characteristics, therapeutic regime (transfusion and chelation) and clinical complications in a sample of 349 American and Canadian adults with thalassemia major. The results indicated that 70% of adults were employed and 60% had an average educational level. Caucasian individuals worked in better-recognized professional activities; work was more frequent among older women without clinical complications. The authors concluded that neither transfusion nor chelation is a factor that hinders educational and professional development⁽⁷⁾.

One of the themes receiving more focus in studies is quality of life among thalassemic individuals in different age ranges. Unfortunately, these present contradictory results. One of these studies was of special interest because it included individuals from the same facility where this study was conducted and individuals presented similar social characteristics⁽⁸⁾. It involved 10 young adults aged between 22 and 28 years old through semi-structured interviews whose answers

were analyzed according to content. Different scales were used to evaluate the participants' psychological conditions. The comparative analysis of the results on two points of the evaluation suggested that the quality of life of patients going into adult life was harmed and some presented increased psychological symptoms.

The experience of British young adults with thalassemia undergoing chelation therapy was the object of a socio-anthropological study⁽⁹⁻¹⁰⁾. A total of 25 individuals, 12 of which were men and 23 were students, ages ranging from 10 to 19 years old, participated in the study. The authors assert, based on the obtained narratives, that the response of the individuals to the need for chelation therapy represents a rupture in their lives and is part of a broader process in which they give meaning to the treatment by relating it to the body, identity and maintenance of their relationships. The daily experiences of chelation therapy, the patients' family relationships and those held with social networks are all implicated in the issue of adherence.

Papers⁽⁴⁻¹⁰⁾ addressing the disease's psychosocial aspects and treatment regimens for adult patients are not abundant. Therefore, there is limited knowledge concerning this population, especially in the Brazilian context.

Theoretical and methodological framework

The theoretical structure of medical anthropology, the branch of the interpretative anthropology, which connects health, disease and culture was used in this study. Culture is seen as a system of meanings constructed in social life and shared with a group of which one is a part, providing parameters for how one thinks and acts. Medical anthropology views disease as a process characterized by the acknowledgement of its disorders, diagnosis, choice of treatment and evaluation. The disease and treatment are also experiences, because the body for the human being is a symbolic matrix that organizes one's experiences, relating them to the social, natural and cosmological worlds. Both disease and treatment generate bodily changes and these can be seen as a subjective reality where the body, perception, and meanings are united in a single interpretation⁽¹¹⁾.

Medical anthropology distinguishes the constructions of 'disease' –the perspectives of biomedical objectivity and 'illness'—which is the patient's perspective, focused on subjectivity and includes the disease's cultural, social and personal elements⁽¹²⁾. For one to learn the

similarities and differences among the perspectives of patients in the course of a chronic disease and treatment, one should explore the treatment's objectives, strategies, and evaluation criteria. Because explanations involve multiple and often contradictory processes and meanings, they are seen as situational explanations⁽¹³⁾.

This approach is designed to associate the meanings held by thalassemic individuals concerning their therapeutic regime beyond the biomedical model that separates the body from one's socio-cultural life, and its consequences for treatment adherence, integrating values, beliefs, and symbols linked to gender, age, socio-economic class and social roles.

The analysis of individual's treatment experience can be carried out through narratives that allow creations based on circumstances, actions, causes, relations, objectives and means, leading the researcher to an interpretative posture that promotes a reciprocity between popular and scientific thinking, uniting cultural and religious structures, common sense, media and medical services. The narratives can be obtained through the oral report technique and interviews. An oral report is an instrument to understand the meaning of human action, a person's social relationships that compose the environment within which individuals move⁽¹⁴⁾.

Study procedures

The study was developed at the Hematology Center of Ribeirão Preto linked to the *Hospital das Clínicas* at University of São Paulo at Ribeirão Preto, Medical School. Eleven patients with thalassemia major receiving treatment from this service and who returned for a monthly medical evaluation from January to April, 2010 and met the following inclusion criteria were invited to participate: being 18 years old or older, either gender, with different socio-economic and educational levels.

All the invited individuals met the inclusion criteria and agreed to participate. After the individuals signed free and informed consent forms, the first interview was scheduled for the following return medical visit.

Data were collected through semi-structured interviews guided by an instrument composed of two parts: the first addressed the participants' social characteristics; the second part included questions that facilitated the individuals' narratives: How did the disease start? What do you think about the treatments? How do you deal with them? Have you ever stopped your treatment? Why? What do you think about having to have treatments for life?

The interviews were conducted during blood transfusion sessions, were recorded with the authorization of patients and took between 30 and 40 minutes. Two interviews were conducted with each patient; the second clarified doubts arising from the initial data analysis. Interviews were fully transcribed and texts were submitted to inductive thematic analysis. This type of analysis consists of an interpretative process of analyzing data according to common aspects, relationships and differences among them. A theme is a general aspect that aggregates data. This process involves the following stages: 1) global reading of material; 2) detailed reading of each text to check for similarities, differences and details of narratives, classifying them into codes; 3) delimitation of common and different meanings expressed by the participants, which enabled the construction of descriptive categories concerning the object; 4) discussion of results (themes) based on the participants' contexts of life, the theoretical framework and available literature⁽¹⁵⁾.

Data were classified into descriptive or empirical categories in the first stage of analysis: signs and symptoms of the disease, beginning of treatment, current treatment, repercussions of treatment, adherence and non-adherence, future prospects. From these categories the thematic core emerged: "the life of patients with thalassemia in relation to their treatment".

Among the rigor criteria the following are highlighted: being engaged with patients for a period of four months; considering the patients' different perspectives using their narratives as examples; involvement of patients of different economic classes and educational levels and both genders; and working with another researcher using the same theoretical-methodological approach to construct the categories and theme⁽¹⁵⁾.

The project was submitted to and approved by the Research Ethics Committee at the University of São Paulo at Ribeirão Preto, Medical School, *Hospital das Clínicas* (process No. 11498/2009, November 9th 2009). The patients were ensured of their right to be informed about the study and freely consent to participate in it, as well as being ensured of the confidentiality of provided information and identity. Hence, fictitious names are used throughout the text to name the participants.

Results

- *Identification of participants*: the group of participants was composed of six men and five women aged between 22 to 32 years old; educational level

varying from vocational programs to bachelor's degrees; income between 1.17 to 5.88 times the minimum wage, while one was a student and had no income and another was on sick leave receiving financial support from the government. Their occupations varied: four salesmen, one cashier, a mechanic, a laundry assistant, a musician, a general services assistant, a student, and one unemployed. In relation to religion, ten reported to be Catholic, six of which were non-practicing Catholics. Ten lived in Ribeirão Preto and one in another state. These social characteristics characterize the group as young adults of a productive age. Despite their education – vocational courses and some with a bachelor's degree—their occupations do not receive much social acknowledgement, showing the difficulty of reconciling a professional activity with the therapeutic regime, even though Brazilian laws favor such integration.

- *thematic core "the life of patients with thalassemia in relation to the treatment"*: The participants reported that the disease's symptomatology emerged in their first years of life, and at the time their parents identified it as anemia. The exacerbation of symptoms or the onset of other symptoms, led them to seek out health services. The therapeutic development within the health system was characterized by difficult access and lack of problem-solving capacity. *We lived in Paraná at the time and my father sought out all kinds of physicians to find out what the problem was. My spleen would swell, I was weak and couldn't walk... Since my father and my mother had the same trait, they sent me to Curitiba and they found out I had thalassemia.* (Maria, 27 years old); *According to my mom, I was six months old. Then I had anemia, profound anemia. Then the doctors took too long to figure out the anemia's origin. They sent me to Jaboticabal and then they referred me here. Then, they found out that I had thalassemia major.* (Diego, 28 years old).

When the participants talk about the disease, they use appropriate medical vocabulary that gives meaning to the disease and treatment: *I have the trait, anemia, thalassemia major, separation of leukocytes, red blood cell concentrate, my ferritin.* This use denotes they have learned and applied the information provided by the service professionals.

Among the meanings given to the experience of many years with a therapeutic regime implemented for a genetic and chronic disease, there is a relationship with the concept *normal*, presented by most patients; for others the relationship was presented as resignation. *Normal! Because I already had in my mind that I had a problem and would have to do the treatment. So, for me it was normal, I got it alright.* (Maristela); *For me, it was like... normal! When*

you're born with something, it's easier. I've always been ok with it, never had many problems. (Joana); [...] I know I need the treatment to get well. So, there's not much I can do. I know it will be like this my entire life, you get used to it. I need the treatment to live. (Rita); We have to conform! What can we do? I have to do it! (Diego).

Nicolas, 25 years old, makes clear that the experience is only suffered, *Today, for me it's normal! To tell the truth, there're times I get fed up! Having to come here [...]. (Nicolas).*

The difficulties in dealing with the therapeutic regime are varied. Considering the social characteristics of the participants, work, leisure, body image and the maintenance of loving relationships are the most negatively impacted. *[...] it was difficult to find a job because of it. Most jobs do not accept it when you miss work, you know. (Maria); I'm coming here every two weeks because my hemoglobin is too low. It is difficult, my boss doesn't like it, I take the doctor's attestation but I know he doesn't like it. I told him about the disease when I accepted the job, that I had to have transfusions. He got a little uneasy, you know [...] the worst is not getting the treatment... is the fact that I have to come here! I lose the entire day. Now it is even worse because I have to come twice a week. (Rita); In the summer, swimming suits, swimming pool and all, and my body is all bruised, there're hematomas everywhere, the skin is red, purple. Sometimes, there're lumps, visible ones! If you look carefully, you'll see lumps coming out of the skin! (Joana).*

In addition to the difficulties in reconciling social life and treatment, the medication needs to be appropriate to the clinical condition of each individual. It implies periodically trying new drugs, dealing with their reactions and setbacks: *I had a lot of iron impregnated in my heart. Then the doctor said for me to come here at least two months in a row to have the pulse therapy! I came every day in a month. Then I asked him to give me 15 days to rest. My body couldn't take it anymore! The body and mind! Then, I took Deferiprone, and it gave me a lot of stomachaches! So I had to stop it. Now, I've started with Exjade since last month, but I had an allergic reaction to it and I went back to the Desferal back, for now (Diego).*

Some also expressed reactions to the severe consequences of interrupting medication. *Ah, I didn't do the treatment right. Every type of chelator you can imagine I've already taken! But I didn't accept it at the time! I'd go away and not have to have pulse therapies; I'd go long periods without treatment and then I had heart failure and cardiac arrest! Then I realized what was happening (Leonardo); I've seen many friends my age with thalassemia and who unfortunately died over the course of their treatment because they didn't take the treatment seriously [...]. (Miguel).*

Hence, the reports showed that the patients acknowledge the importance of maintaining regular treatment for their survival.

The patients reported experiences with social stigma given the disease and especially for having to periodically attend return visits and undergoing transfusions. Consequently, they do not always reveal that they have the disease. *Ah, there's still much prejudice! People are afraid! I've heard people saying I'm a risk. (Leonardo); [...] I don't tell people I do transfusions! I don't like to talk about it. (Igor).*

When the patients were asked if they ever interrupted their treatment, most denied doing so but made clear that this practice occurred in different situations, which are exemplified in the followed reports: *Today, I didn't take the chelator because it gives me diarrhea and I was afraid of having it on the way here. But my ferritin is 900. I know that if I don't take the drug, it may worsen [...] (Rita); [...] I used to say that I was a normal person, only had to have the transfusion once a month. I went one week without Desferal and my ferritin went up. It's so tiresome! Every day you get pricked, gets lumps and you don't manage to do it. You get home late, tired... So, I'd say: today I won't do it! (Francisco); Now, my ferritin increased because I forgot to take the medication (Livia).*

Some have hope and expectations for the future that less aggressive therapies will emerge; others have only strength to keep surviving: *I wish they'd invent some other treatment to increase hemoglobin, that I didn't have to have these blood transfusions. I wish there was a pill so I didn't have to have transfusions. Who knows, perhaps some day, right! (Rita); I hope that I have to come here less often from now on. (Francisco); I expect that in the future... I'll develop professionally, I'd be better! In relation to the treatment, a tiny pill with a blood bag (Nicolas); I don't stop to think about the future! (Diego).*

Discussion

The reports show that being an adult with thalassemia major is a difficult experience due to the frequency of return medical visits, periodic transfusions, the daily use of medication, side effects caused by the transfusions and medications, and social stigma, that alter daily life, as revealed in the literature⁽⁴⁻⁸⁾.

The facility's professionals constantly inform the patients about the need to continue their treatment to ensure their survival, and these are aware of their "genetic trait" (being an individual with thalassemia). This acknowledgment leads them to conform to their situation, *we have to conform to our situation*. It implies in not fighting reality, but trying to live with

it⁽¹⁶⁾. Additionally, the way they approach their clinical conditions – *my ferritin*, and the use of medications – *I'm using Exjade*, are evidence of the fact that they have incorporated their disease and treatment⁽¹⁷⁾. That is, disease, treatment and body begin to constitute a single identity.

Symbolically, every person has an individual body (physical and psychological) that is acquired at birth and a social body that is necessary to live in a given cultural group. The social body is the essence of body image and self-image, because it provides a frame through which the person perceives and interprets physical and psychological experiences. It is also a means through which society controls behavior in disease and health, in working and leisure activities⁽¹⁸⁾. The idea that the sick body is a mediator of social relations has meaning in the case of chronic diseases given these diseases' characteristics. The disease does not exclude the patient from daily life but is at the center of one's social relationships, that is why one conforms to the situation; one of the patient's objectives is to reduce derangement caused by such an experience. For that, thalassemic patients employ the strategy of normalizing body and life to control the disease, the treatment and social reinsertion. Normalizing life does not mean to return to life without the disease, but it is a set of actions and interpretations that allows one to construct a natural attitude, a new way to live life, at least in part, according to the social values and standards of the individuals involved⁽¹⁶⁾.

Hence, interrupting treatment is not an attitude of defiance or ignorance, but an expression of the need to normalize one's body and life, though it does bring severe consequences and pose risks to the survival of patients, who at the same time acknowledge such consequences.

In this context, treatment adherence is not a matter of practical difficulty; it symbolizes a threat to the normality of life –one of the processes through which power is manifested in the field of identity and difference; normalizing means attributing to identity all the possible positive characteristics, because a normal identity is desirable⁽¹⁹⁾.

The process of normalization as a strategy to cope with the disease is culturally learned and valued. It enables one to deal with anguish, uncertainty and suffering, and is a common view of the chronic condition as shown in Brazilian papers addressing the experience of adult oncologic patients undergoing therapy⁽²⁰⁻²¹⁾.

When discussing the challenges associated with prolonging the survival of patients with thalassemia, from childhood to adulthood, authors⁽²²⁾ emphasize that adult patients spend on average 271 hours per month in care (transfusions, medication therapy, medical consultation and travels), which illustrates what treatment adherence really means.

The comparison of the meanings held by young adult individuals in this study with those of the British study⁽⁹⁻¹⁰⁾ with the same theoretical-methodological approach revealed they are similar and we agree with the authors who highlight emotional and social concerns and explain why chelation therapy adherence is irregular and varies among individuals over time.

The overlap of different meanings concerning the care process is worth noting in relation to the survival of thalassemic individuals. Chronic transfusion patients consider their problem constructed and structured by the desired to be normal (without the need to have transfusions and take Desferrioxamine). For the health professionals, the matter is to keep patients alive through a therapeutic regime based on medical knowledge. While all are focused on the same problem, each has a different emphasis concerning its meanings. It generates oppositions and different priorities. But what matters most is to recognize that patients respond differently to therapies⁽²³⁾.

We agree with a hematologist who says that even when there is success in maintaining chelation therapy, the patient is always between two polarized situations, that is, *between the devil and the deep sea* –toxicity by iron and toxicity by chelation. The patient does not directly feel the effects of iron excess or those of chelation; he can only imagine how harmful it can be to not regularly adhere to therapy⁽¹⁹⁾.

Hence, we see that therapy adherence is a process through which patients follow alternative paths that enable the feeling of satisfaction; s/he absorbs, analyzes, interprets, and reacts according to his/her knowledge, values and beliefs (culture), and decides whether to continue following a given treatment or not. Family and friends, rarely health professionals, help the patient with this decision. This influence demonstrates the power of the social network in one's decision-making process concerning care provided to health and disease⁽¹⁹⁾.

Final considerations

The thematic core indicates that the 11 young adults with thalassemia major, regardless of gender,

age, religion and social economic level, narrated their experiences with treatment, mediating their bodies and emotions so that their histories give meaning to their limited bodies, to experience some degree of normality in life. That is why they irregularly follow the chelation treatment, contrary to the regularity of therapeutic adherence expected in the medical model. They justify such irregularity given the limitations imposed on social functions, exposing themselves to risks that threaten their survival.

In this context, we assert that nurses need to understand the difficulties faced by patients, acknowledge their cultural values and support them in their therapy's different phases while working jointly with family members. A support group during the outpatient's return visits can be an intervention strategy used by patients to express and discuss their feelings concerning the disease, treatment and how these impact their lives.

We suggest that this topic be further explored through prolonged data collection, with a larger number of participants, employing participant observation in the family context. Such strategies can enable the collection of narratives with a greater variability of experiences with therapeutic regimes.

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