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ORIGINAL ARTICLE

EVALUATION AND INTERVENTION IN THE FAMILY OF ADOLESCENTS WITH SICKLE CELL DISEASE

AVALIAÇÃO E INTERVENÇÃO NA FAMÍLIA DE ADOLESCENTES COM DOENÇA FALCIFORME EVALUACIÓN E INTERVENCIÓN EN LA FAMILIA DE ADOLESCENTES CON ENFERMEDAD FALCIFORME

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ABSTRACT

Objectives: to evaluate the structure/functionality/development of the family of adolescents with sickle cell disease and implement interventions after evidence identified. **Method:** it is a case study with a qualitative approach. The Calgary model of evaluation and family intervention was used as theoretical reference and the case study as a methodological strategy. Data were collected from two families. **Results:** It was found that one family was single-parent and the other was extended. The application of the theoretical reference allowed to know aspects related to the structure/functioning/development of the family. From this integral evaluation, it was possible, in partnership with its members, to propose interventions such as guidelines on sickle cell disease/treatment and music therapy sessions, contributing to socialization and improvement in the family environment, as well as relaxation and reduction of anxiety. **Conclusion:** home care could provide support and strengthen each family, within its specificity. **Descriptors:** Family; Anemia, Sickle cell; Adolescent health; Nursing; Adolescent.

RESUMO

Objetivos: avaliar a estrutura/funcionalidade/desenvolvimento da família de adolescentes com doença falciforme e implementar intervenções após evidências identificadas. **Método:** estudo de caso com abordagem qualitativa. Utilizou-se como referencial teórico o modelo Calgary de avaliação e intervenção familiar e como estratégia metodológica o estudo de caso. Os dados foram coletados em duas famílias. **Resultados:** verificou-se que uma família era monoparental e a outra era estendida. A aplicação do referencial teórico permitiu conhecer aspectos relacionados à estrutura/funcionamento/desenvolvimento da família. A partir dessa avaliação integral foi possível, em parceria com seus integrantes, propor intervenções, como orientações sobre a doença falciforme/tratamento e sessões de musicoterapia, contribuindo, dessa forma, para socialização e melhora no ambiente familiar, além de relaxamento e redução da ansiedade. **Conclusão:** a atenção domiciliar foi capaz de proporcionar suporte e de fortalecer cada família, dentro de sua especificidade. **Descritores:** Família; Anemia Falciforme; Saúde do Adolescente; Enfermagem; Adolescente.

RESUMEN

Objetivos: evaluar la estructura/funcionalidad/desarrollo de la familia de adolescentes con enfermedad falciforme e implementar intervenciones después de evidencias identificadas. **Método:** estudio de caso con enfoque cualitativo. Se utilizó como referencial teórico el modelo Calgary de evaluación e intervención familiar y como estrategia metodológica el estudio de caso. Los datos fueron recogidos junto a dos familias. **Resultados:** se verificó que una familia era monoparental y la otra era estendida. La aplicación del referencial teórico permitió conocer aspectos relacionados a la estructura/funcionamiento/desarrollo de la familia. A partir de esa evaluación integral fue posible, junto con sus integrantes, proponer intervenciones como orientaciones sobre la enfermedad falciforme/tratamiento y sesiones de musicoterapia, contribuyendo para socialización y mejoría en el ambiente familiar, además de relajamiento y reducción de la ansiedad. **Conclusión:** la atención domiciliar fue capaz de proporcionar soporte y de fortalecer cada familia, dentro de su especificidad. **Descriptor:** Familia; Anemia de Células Falciformes; Salud del Adolescente, Enfermería; Adolescente.

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INTRODUCTION

Sickle cell disease (SCD) is a more common genetic disease in the world,¹ coming from a mutation in the hemoglobin beta globin gene, giving rise to an abnormal hemoglobin, designated as hemoglobin S (HbS), which replaces normal hemoglobin A (HbA).¹⁻² Minas Gerais is the third state in Brazil to have sickle cell disease, followed by the states of Bahia and Rio de Janeiro.^{1,3} An estimated of 3,500 children are born each year in the country with sickle cell disease and 200,000 have this trace.²

The clinical manifestations of SCD in adolescents include chronic anemia, painful crises, infections, stroke, ocular complications, biliary calculus, lower limb ulcer, delayed growth, delayed secondary sexual characteristics, menarche, and the first late ejaculation. Adverse psychological manifestations typical of chronic diseases, such as low self-esteem, are also frequent, aggravated by the underprivileged socioeconomic situation of most of the patients and, with frequent difficulties, especially at school and at work.⁴⁻⁵

The association with a chronic disease in adolescents who are already in a phase of intense transformations can be overwhelming, with the corporeal, emotional and cultural transformations, besides contributing to the disorganization of the family structure. A successful passage will surely be a young man well prepared to deal with predictable and unpredictable situations that the future holds for him.⁶

Individuals with sickle cell disease and their family members should be aware of possible clinical events. Because it is a chronic disease, the treatment will be lifelong and, to be successful since the diagnosis, family members will need to learn about the signs of complications, as well as to act correctly in the different intercurrents.⁷⁻⁸

Also, young people are still not able to deal with information as an adult due to several factors, such as information complexity, less ability to deal with uncertainties, and often little social and affective support.^{6,9} Health, who care for adolescents, should aim to contribute to the acquiescence of their limits, potential and the ability to dare to live. To do this, they must have knowledge of their histories, experiences, the meaning they give to health and disease, and their connection with lifestyles.⁹

It is essential that the adolescent be supported by the family to face up to this moment, which comprises a series of semantic

variants that, even with some disagreements among them, are not directly related to their originality.⁹⁻¹⁰ The family should be an integral part of intervention in health at any stage of the disease and in all care settings, which is relevant in the relationship between care to the individual and their family context, as a necessary factor to the integral care of the patient's person.¹¹⁻²

Based on the information obtained, health professionals need to use their knowledge about each family to ensure holistic care to relate biological, social and spiritual factors, elaborating the best care.

In this context, it was chosen to use the Calgary Family Assessment and Intervention Model (MCAIF), which allows an extended family spectrum, including internal and external relationships, weaknesses and strengths, as a possibility to life and health of people. For this model, the family is considered from its existence as a long-term relationship commitment, in which people are organized in mutual relationships, and the different family conformations must be understood in the health care of the population.¹³

The Calgary Family Assessment Model is a multidimensional structure formed by three fundamental categories: structural, developmental, functional and its multiple subcategories that allow joining elements to give subsidy and direct care with the family.^{10,13} Its use promotes understanding dynamics and family functioning in an interactional way and providing the evaluation of its elements and the observation of changes in its dynamics.^{10,14}

The family systems approach has been used to advise on the understanding of the family as a unit of care and not only as the sum of the individuality of each family element, in different contexts.¹³⁻⁴ In Brazil, MCAIF has been little used in families of adolescents with chronic disease. The use of this model allows the nurse to know the family in their context and identify their needs, as well as care alternatives specific to their condition.¹⁴

The motivation of this research was due to the fact that it was a matter of major importance, a disease that requires scientifically based care, as well as the delicacy due to the chronic nature. Thus, the interest of knowing, in a profound way, the daily life of families of adolescents with sickle cell disease that had undergone significant transformations, in this case, a family went from nuclear to single parent due to the matriarch's death with a malignant neoplasm of the breast and the other extended nuclear

family related to the entrance of a pregnant teenage member.

Because it is believed that chronic disease management, evaluation and care should cover the family and not only the affected person,³ it is considered to be paramount to make public this experience, since it can corroborate the practice of professionals working in this context.

From these considerations, the following questions arose: how is the structure and functioning of the families of the adolescent with sickle cell disease that have undergone significant changes? What is the affective bond between the members of these families? Are there interventions to be undertaken with these families?

Therefore, this study aimed:

- To evaluate the structure/functionality/development of the family of adolescents with sickle cell disease.
- To implement interventions following evidence identified.

THEORETICAL REFERENCE

♦ Calgary Family Assessment and Intervention Model

The Calgary Family Assessment and Intervention Model (MCAIF) is an instrument for nursing actions, with a susceptible use by an interdisciplinary team. It aims to improve living conditions and the relationship between family members defined as a multidimensional structure, integrated, based on the theory of systems and cybernetics and consists of three broad categories: structural, developmental and functional, represented in Figure 1.

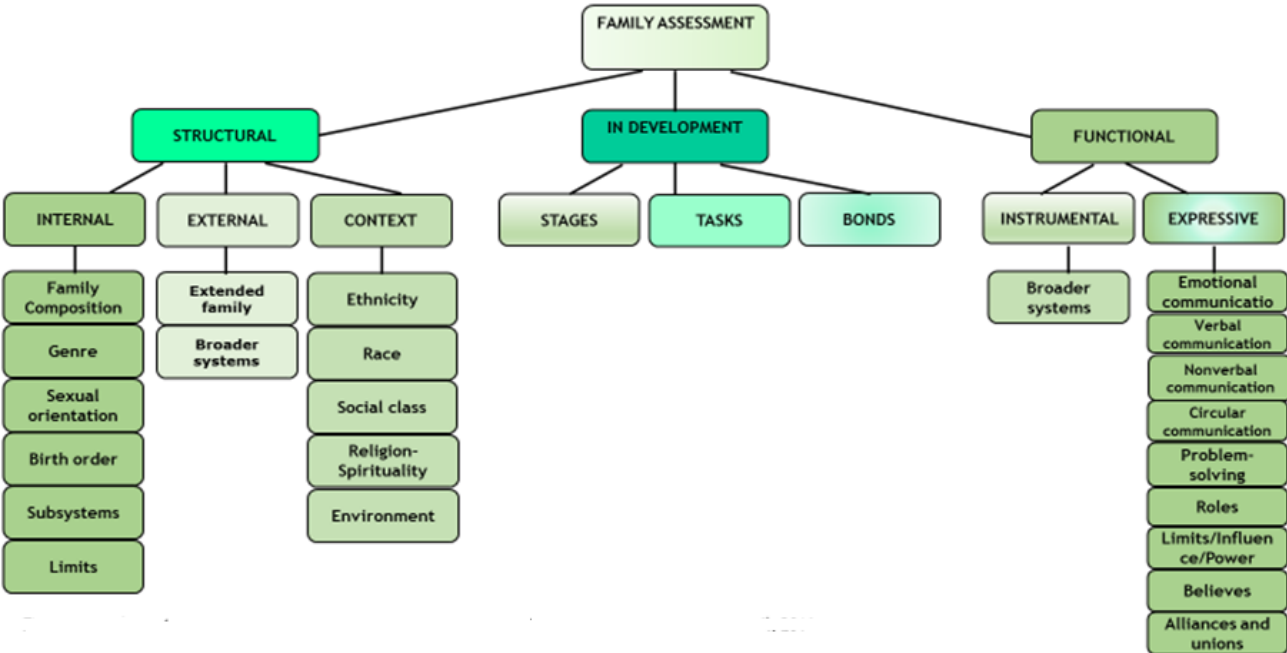


Figure 1. Schematic representation of the Calgary Family Evolution Model.¹³ Divinópolis (MG), Brazil, 2016.

Structural (it refers to family composition, affective bonds between its members compared to individuals outside and its context), **in development** (it emphasizes the unique trajectory built by a family and modeled by predictable and unpredictable events) and **functional** (it details on how individuals behave with each other) organized in assessment (MCAF) and intervention (MCIF).¹³⁻⁴

The instrument allows visualizing how the individual, the family, and the community, in an amplified way, are building their “web of interactions,” which are the networks of family support, bonds, conflicts and communication.¹⁰ These categories are divided into subcategories, which integrate complex concepts that require a thorough discussion, in a perspective of transdisciplinary knowledge. In primary care, the adoption of this model is indicated as

beneficial to the promotion of interaction with families and better care planning. The family’s difficulties in providing care are related mainly to the lack of knowledge, resources, and social support. Often, even with knowledge and skills that would enable them to provide the necessary care, the family shows difficulties in carrying them out, for reasons as different as the lack of internal resources and the lack of external resources that could be mobilized.^{10,14}

It is the job of each nurse to select the subcategories of this model to be explored. Thus, not all subcategories are evaluated at a first encounter, and some may never be explored.¹⁴

Two instruments are used to outline the internal and external structures of the family: the genogram and the ecomap.^{10,13} The genogram is a graphical representation of the internal family structure. The fundamental

scope of the genogram is to assist in the evaluation, planning and family intervention.¹³ Symbols and codes that follow a pattern are used in its elaboration. As soon as it is ready, it allows a clear verification of which members make up the family, and it provides basis for the analysis and discussion of family interactions.¹⁴ It also allows the family to identify which elements integrate it and the relationships established among these members.¹⁰

The ecomap is a diagram of the living or non-living relationships between the community and the family and allows estimation of social supports and available networks, as well as their use by the family.¹³ The ecomap is dynamic since it represents the presence or absence of economic, social and cultural resources during a certain period of the family life cycle, which can be modified over time.^{10,13}

The category of development refers to the progressive change in family history during the phases of the life cycle: its history, the flow of life, family growth, birth, and death.^{10,13-4}

Regarding the functional category, it refers to the way people in the family interact. It can be explored the aspect of instrumental functioning, which is related to the activities of daily life, and the aspect of expressive functioning, referring to modes of communication, problem-solving, beliefs, roles, rules, and alliances.¹³⁻⁴

When considering the family of the adolescent with sickle cell disease, it contributed to the support of the operationalization of an integral family assessment, which recognizes its structure, the development of its tasks and its links throughout the life cycle and the functioning of its activities and communication styles.

METHOD

This study is a subproject of an “umbrella” project titled “Integrity of care for patients with sickle-cell disease: home care and the strengthening of the support network of a hemonucleus located in the state of Minas Gerais.”

This is a case study with a qualitative approach. It refers to a broad method that allows being applied to a multiplicity of problems and can be used in different areas of research to provide a greater involvement and knowledge of the researcher with an observed real conjuncture.¹⁵

As a theoretical reference, the Calgary Family Assessment and Intervention Model was used, consisting of open questions, focusing on the family structural evaluation (with the

construction of the genogram and the ecomap), evaluation of the family development throughout the life cycle with their tasks and links, and evaluation of instrumental functioning (activities of daily living) and expressive functioning (communication styles, roles, influence, beliefs, family alliances).¹³⁻⁴

The study scenario was the home of the families of the adolescents with sickle cell disease, registered in a Hemonucleus located in the State of Minas Gerais, Brazil. The families lived in different municipalities. Home visits were from December 2015 to May 2016.

These families were intentionally chosen because the first family consisted of the young woman with sickle cell disease and the elderly father, besides the life trajectory and the rite of passage for adolescents accompanied by the inexorable, that is, the death of the matriarch due to malignant neoplasm of Breast to provide her and her father with a better adaptation and quality of life. The second family due to a teenager with sickle cell disease had become pregnant and, before her daughter was born, they went to live with her mother-in-law's wife and sister-in-law.

The study was performed through six home visits with each family studied. In all the meetings had at least two researchers. Firstly, informal contact was made to verify the availability of family members to be part of the research. Later, each interview was marked at the residence of each family, with a date and time chosen by the participants.

At the first home visit (HM), the ethical and legal aspects were presented to carry out the research and invited them to participate in it. During the second visit, interviews were conducted with semi-structured questions, guided by the MCAIF, were recorded after authorization, through a digital device, with an average duration of 60 minutes. The genogram and ecomap were constructed, with the participation of the family to allow analysis and meet the objectives of the study. On the third visit, discussions, and considerations about the genogram and the ecomap and specific guidelines were made, when necessary, for each situation experienced by the family. In the other three visits, interventions were implemented and evaluated together with families.

After collecting the information and transcribing the interviews in their entirety, the analysis of the elements was used as the strategy, thematic analysis, form of recognition of patterns within the data, in which the themes that emerge are formed

into categories.¹⁶⁻¹⁷ In this evaluation, there are different forms of approach, such as deductive, based on previously determined template models; and the inductive, driven by the data. In this study, the chosen method was a hybrid model, which groups both the deductive and the inductive.¹⁶

In this way, at first the data are analyzed inductively, originating codes and initial themes, and later the template is applied. In this research, the Calgary Model was used with the goal of identifying meaningful units of text also in a deductive way.¹⁶ Finally, data were obtained within the structural, developmental, functional and intervention categories of the families proposed by the MCAIF.

The anonymity of the participants was maintained through the adoption of the letter E, followed by the sequential number of the interviews, besides the letter F, accompanied by the family number. The participants were six members of the two families that accepted to participate in the research: a father, mother-in-law, a partner, a sister-in-law and two adolescents with sickle cell disease. All participants were invited to observe their family, as recommended by the MCAIF reference.¹³

The Project was approved by the Ethics and Research Committee of the Federal University of São João del Rei, with the Hemominas Foundation as co-participant, according to

CAAE: 13201013.6.0000.5545, opinion number 599.680-0. The participants of the study accepted voluntarily to participate. The members of families over 18 and the legally emancipated adolescent, signed the Informed Consent Form (TCLE), one of the adolescents signed the Free and Informed Consent Form and Responsible for signing the Informed Consent Form for Minors. They were identified with fictitious names to preserve their identity.

RESULTS

The following will be presented under the Calgary Family Assessment and Intervention Model categories.

♦ Structural Assessment

Regarding the internal structure, it was observed that family 1 was of the mononuclear type, composed by the 60-year-old father (Lírio), retired, a self-employed cobbler in his residence and his 16-year-old daughter (Dália), single, student of the ninth grade of elementary school, in public school. Margarida, Dália's mother, and Lírio's wife, died in 2014, at the age of 50, due to malignant neoplasm of the breast, she was a housewife and resided with Lírio and Dália, married for 25 years. In Figure 2, there is the genogram of family 1, and in Figure 3, there is the ecomap of family 1. It is emphasized that they were built next to Lírio and her daughter Dália.

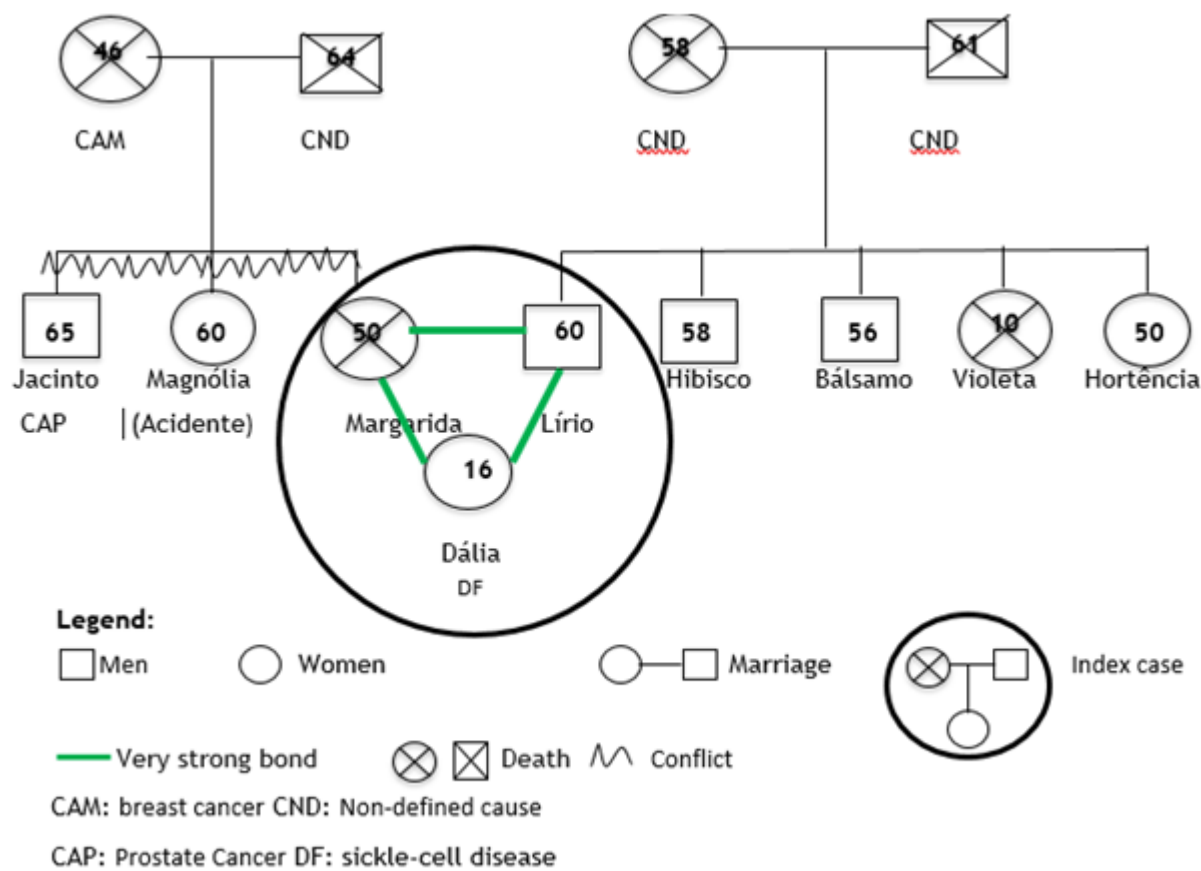


Figure 2. Genogram of the Family 1 of the family of the adolescent with sickle cell disease. Divinópolis (MG), Brazil, 2016.

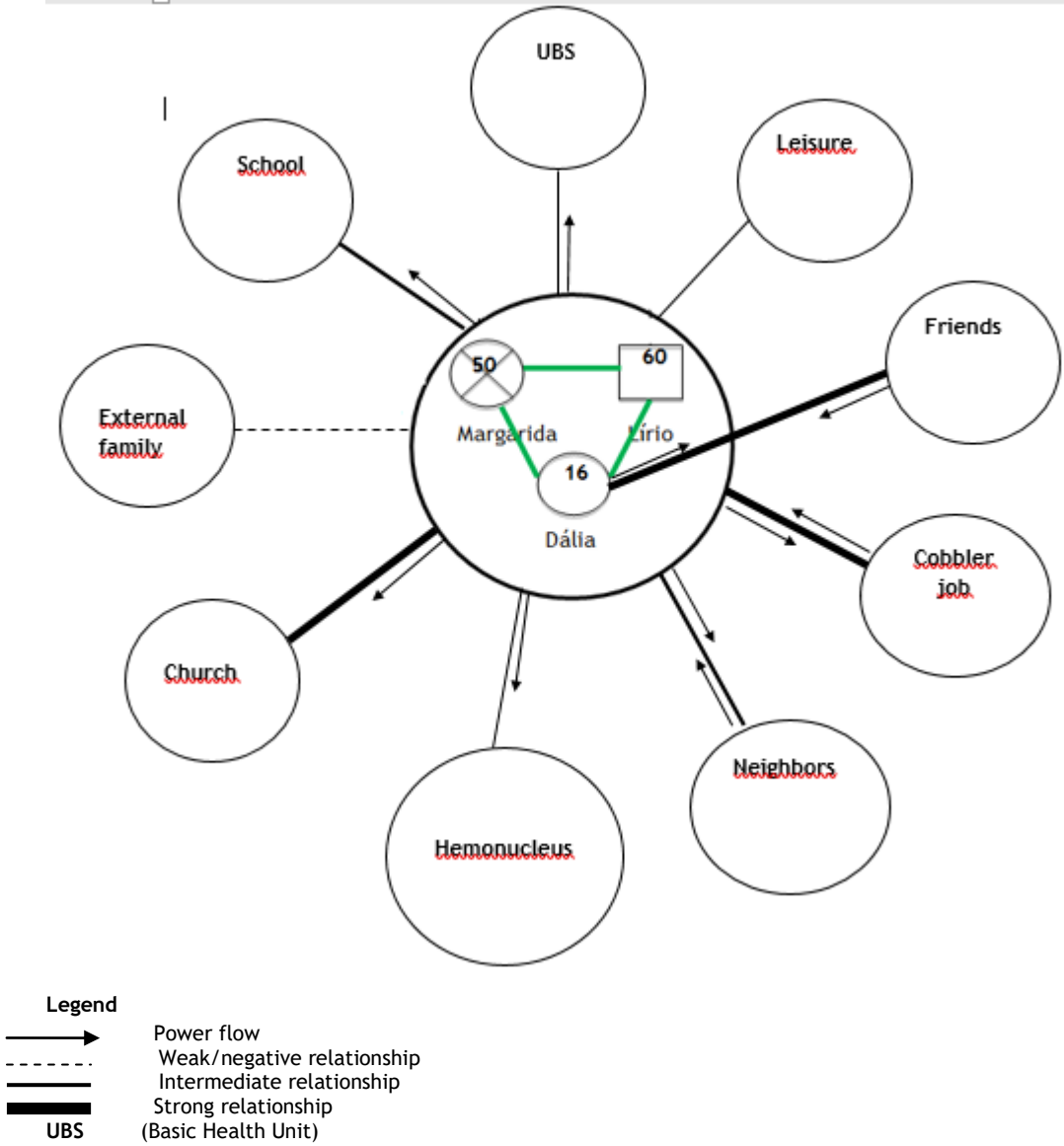


Figure 3. Ecomap of the family of the adolescent with sickle cell disease. Divinópolis (MG), Brazil, 2016.

Family 2 was of the extended type, comprised of Bia, a 17-year-old adolescent, with sickle-cell disease; her partner (Rui), 19 years old, the mechanic at a motorcycle workshop, her daughter (Wal), 15 months old; her 23-year-old sister-in-law (Ziva), working in a clothing trade and her 45-year-old divorced mother-in-law (Lena), working as a hairdresser

in a salon next to her residency. Bia lived with Rui’s family after being legally emancipated in the year 2014, when she was 30 weeks pregnant, her only daughter, by her parents. Figure 4 shows the genogram of family 2, and in Figure 5, there is the ecomap of family 2. It was observed that they were built together with Lena, Ziva, Rui, and Bia.

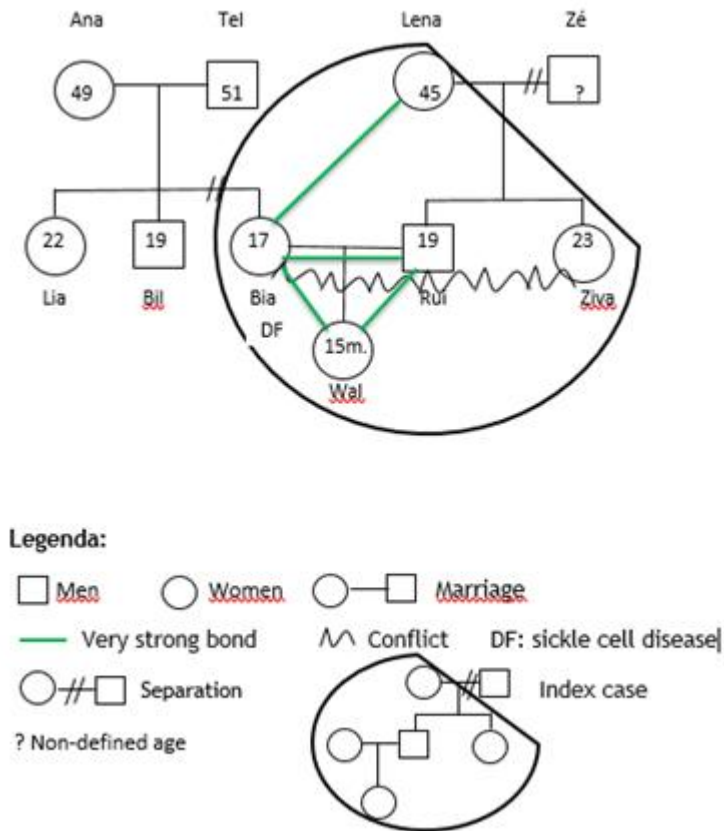


Figure 4. Genogram of the Family of the family of the adolescent with sickle cell disease. Divinópolis (MG), Brazil, 2016.

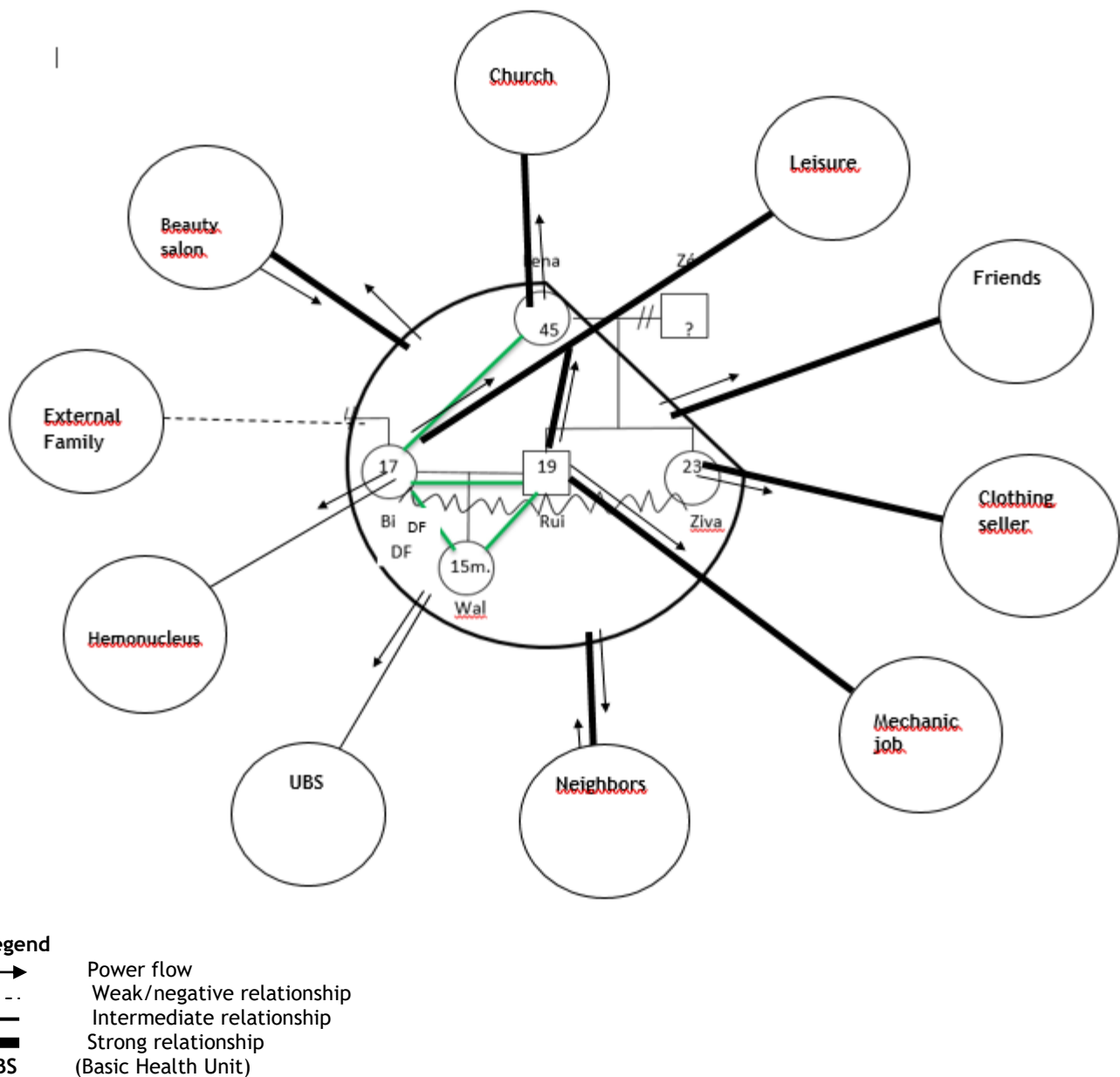


Figure 5. Ecomap of the family of the adolescent with sickle cell disease. Divinópolis (MG), Brazil, 2016.

The **subsystems** identified in the evaluation of the internal structure regarding relationships, in family 1, Lírio (father) and Dália (adolescent daughter with SCD) have infrequent communication, however, show affection and concern for each other. Conflict

subsystem was present in family 2, at many times, they exposed a conflicting relationship between Bia and sister-in-law Ziva, due to the difference in personality and way of living life. Ziva reported that Bia goes out a lot at night to have fun and leaves her daughter (Wal) with her mother Lena (Wal's grandfather) overloading her who needs to open the salon early and works late on weekends. In moments of stress, Rui and Lena try to mediate the conflict between Bia and Ziva. The subsystem of the couple (Bia and Rui) was articulated, with expressions of affection and complicity during home visits.

Among the **limits** observed in the internal structural evaluation, the diagnosis of SCD in the Bia Neonatal Screening, being that at the time she lived with the mother, the father, a two-year-old sister with sickle cell disease, and a healthy brother of five years old was overwhelming, and more recently, the early pregnancy of the Bia teenager was devastating to her as her parents did not accept the pregnancy and put her out of residence before the child's birth, and the concern and fear of having a child with malformation or with sickle cell disease, since she (Bia) and her sister (Lia) have SCD. However, Ms. Lena welcomed her like a daughter (Bia's report), which provoked jealousy in Ziva (daughter of Lena and sister of Rui, Bia's partner and father of his daughter Wal).

In family 1, the **limits** verified were related to the diagnosis of breast cancer of Margarida (mother of the young Dália) and the treatment of this malignant neoplasm, which lasted two years culminating in her death, felt by both the adolescent (Dália) and By Lírío (Dália's father and Margarida's husband), not only because of the severity of the disease, but because of all that they experienced; With this, Dália and Lírío said to hide their feelings, cries, fears, uncertainties to prove themselves strong, avoided talking about death, showing dismay and externalizing emotions. It is worth noting, even within the **limits**, the daily fatigue and pain episodes reported by the adolescents with SCD of the two families.

Regarding the **external structure**, it was verified that the families had multiple elements of the supra-family system, that is, friends, neighbors, church, work environment, health services. When it is necessary, Dália and Bia are supported by the Hemonucleus, setting the social and support network of these families. They affirmed that they do not correctly follow all the guidelines of the health professionals and the appointments when they can, are time-consuming and of low quality, mainly for the adolescents with

SCD, since many health professionals are not aware of the illness. The two families followed health care through the Unified Health System (SUS).

Regarding structural **context** assessment, it was observed that **social class** and financial condition influenced the quality of care provided to adolescents with SCD. The income of family 1 came from the retirement and work of Lírío (complete elementary school) as a self-employed cobbler at his residence, totaling a monthly income of approximately three minimum wages, his daughter (Dália), a student in the morning, helping with house chores. Family 2 had a salary of six minimum wages from the works of Rui, Lena, and Ziva.

As for the **occupation**, it was observed that a teenager with sickle cell disease was a student, the other girl was responsible for household tasks and took care of her 15-month-old son, the rest of the family members performed work activities. Regarding the education of the family members, it was found that a teenager studied until the eighth year of elementary school, a young woman attended elementary school, the other members reported having a complete elementary education.

Regarding the subcategory **religion and spirituality**, the search for spiritual support has been reported by all. Spiritual support and faith gave strength to overcome obstacles and comfort by maintaining hope and helping to promote the well-being of families. Dália, an adolescent in the family, attended the youth group at the nearby church.

In the subcategory **environment**, it was verified that the two families lived in neighborhoods with satisfactory sanitary conditions. About the dwelling, the two families lived in their houses of masonry, both with five rooms, being room, two rooms, kitchen, and bathroom. They had piped water, garbage collection, sewage, and electricity. Household hygiene was satisfactory. The families' meals consisted of four daily meals, rich in carbohydrates and lipids. Regarding the **ethnicity/race** subcategory, family 1 was brown, and in the family 2, the teenager was considered black, and the other interviewed members were white.

◆ Development Assessment

Regarding the **stages** of family life, in this research, one family was in the "family with a teenage son" (F1) stage, and the other family had a teenage partner of a young adult, a child, a young single adult and a divorced adult (F2).

Also within the **stages** of life, in family 1, Dália was diagnosed with sickle cell disease only at two years old, symptoms began at eleven months, the diagnosis was late because of her birthplace. The sickness and later death of Dália's mother and Lily's wife was an overwhelming milestone in this family. In family 2, teenage Bia said that her parents' refusal to accept her early pregnancy provoked a feeling of strong sadness and left deep marks in her "heart," "impossible to heal" according to her report.

In this study, there were two to three generations living, so the phases interacted, and families often performed developmental **tasks**, providing support for the family promoting arrangements for the performance of family roles corresponding to more than one phase.

Regarding the bonds, the families reported strong ties, even if there were conflicts at times, due to different generations, different personalities and jealousies.

◆ **Functional assessment**

Within the functional evaluation process, the **instrumental** evaluation showed that families had support from neighbors and friends during the interurrences, and the distribution of daily tasks facilitated coping and family reorganization even with chronic illness, the birth of a new member, change of residence and death of the matriarch.

In the activities of daily living, both adolescents with SCD stated that they did not correctly follow all the recommendations of the health professionals regarding the care with their chronic condition, and reported that they did not provide guidance. They reported daily use of folic acid and hydroxyurea from time to time, due to nausea and often they vomit after ingesting the drug, as well as non-steroidal anti-inflammatory drugs (NSAIDs) when they present an allergic crisis. They do not perform any physical activity, as it triggers painful episodes. Dália reported that in the last twelve months she had not been hospitalized at one time and Bia had hospitalized for the last time for the birth of her daughter, that is, 15 months ago.

Regarding **expressive** functioning, specifically religious beliefs and spirituality, these have been facilitating elements in the process of accepting losses and coping with sickle cell disease. It was observed in the expressive evaluation that the families developed an effective **communication** between their elements, that is, each understood and considered the message of the other. In family 1, Lírio plays the **role** of father, responsible for the home, trying to be

a partner and companion of his daughter. In family 2, Lena is the matriarch, leading the family calmly and wisely.

In the **problem solving**, the families evaluated demonstrated empowerment in the face of the disease/treatment due to previous experiences, presented a dynamic and effective problem-solving capacity, using the available resources in each family and social context.

◆ **Interventions established from identified evidence**

In MCAIF, the interventions consisted of promoting, increasing or sustaining the functioning of the family regarding their cognitive, affective and behavioral aspects, as well as helping the family to discover new solutions, considering the fragilities and strengths, and aiming to reduce or alleviate the suffering. The behaviors adopted by the studied families were the establishment of relational ties of proximity to therapeutic listening, welcoming, building bonds of mutual trust, permeating the awakening of the resilience of families in search of facing the adversities experienced, and the sickle cell disease in adolescents.

Guidelines on SCD and treatment for both families were made, which allowed the adherence to the hydroxyurea treatment of adolescents resistant to medical prescription due to nausea and vomiting.

There were also sessions of music therapy, three sessions with each of the families, on different days, lasting 50 minutes each, playing Symphony N° 41 "Jupiter" by Wolfgang Amadeus Mozart; "Jesus Joy of Men/Jesus bleibet meine Freude" by Johann Sebastian Bach and the "Four Seasons" by Antônio Lucio Vivaldi; a radio with a CD player in a moderate volume was used (40 to 60 decibels). It is emphasized that one of the researchers is qualified to perform therapy with the use of music.

DISCUSSION

One family was a the single-parent typology, represented by the father and the adolescent daughter (Dália). The health-disease-illness process of Dália with sickle cell disease began at eleven months old, and since then, several confrontations have been experienced by her and her family. One of them came from the access to the health care. Only in 2001, the Ministry of Health was committed to the re-evaluation of the Neonatal Screening in the Unified Health System through the Department of Health Care, which culminated in the publication of the ministerial order (Ordinance GM/MS

number 822, dated June 6, 2001), creating the National Neonatal Screening Program (PNTN).^{1,3} However, the consolidation of the PNTN for hemoglobinopathies was only in 2005 throughout Brazil.³

Another confrontation was the every day of the family, each family is a unit, and acting with it, it is necessary to focus our eyes on the interaction between its members, and not only on the person. Therefore, in the management of chronic disease, evaluation and care should include the focus on the family and not only on the patient,³ which was verified in both families.

During the research, after trying to understand the daily life experienced by the families, it was verified that the treatment of Dália and her mother (family 1), with constant hospitalizations, caused not only an impact for the adolescent but extended to the family context, social context, and group of friends. The early pregnancy of the teenage Bia caused significant changes in her life, before living in a nuclear family, after living with the family of her daughter's father (family 2). Studies have shown several causes involved with the event of teenage pregnancy, especially the unplanned one,¹⁸⁻¹⁹ as the case of one of the adolescents in this study. Among them, clinical, social, cultural and emotional factors stand out. As an implication, there are changes in the project of the adolescent's life, limiting the probability of engagement of these young people in society.¹⁹

The disease affects the interpersonal relationships in the family, since the alterations the woman is passing of so much physical, emotional and social order extend to the relatives,^{3,14} as the case of the malignant neoplasia of the mother of one of the adolescents. The family is not prepared to face health-disease-illness and death of a loved one.²⁰⁻²¹ The death of those who love brings deep ruptures, requesting transformations and adaptations in the attitude of understanding the world and making plans for continuing living in it. However, reactions to the process of loss are processed differently among people and depends on different circumstances that pervade death, such as age, relationship that existed, chronic illness or not, faith, personality, and culture.²¹

Studies showed that rites of passage in a family could be an opportunity for both psychological and spiritual changes, as well as changes in the routine of people.^{14,21}

In this study, it was observed that for the reduction of pain, fatigue, and admissions of the adolescents, multiple factors were

primordial such as coexistence with friends and neighbors, reception of the pregnant adolescent by family², and adherence to drug treatment, since Hydroxyurea is currently the largest polymerization inhibitor of known deoxy-HbS and prevents erythrocyte finalization, chronic hemolytic anemia, painful vessel-occlusive crises, necrosis in various organs, and improves the clinic and life expectancy of people with sickle cell disease.^{3,7,12}

The benefits of the correct use of hydroxyurea were explained in an accessible manner by the researcher. New studies that endorse these improvements, aimed at ingesting the medication with a little cold water to avoid nausea; prefer foods served cold or at room temperature, avoid foods that increase nausea such as fried foods, strong condiments, very sweet foods, alcoholic drinks, and coffee; avoid lying down right after meals. Prefer meals with high protein content and poor carbohydrates and fat, as they reduce nausea due to the reduction of gastric dysrhythmias.²²⁻²³

There is a follow-up of patients using hydroxyurea for 17 years and a half in the United States of America, showing that the drug is safe and that its use also seems to be related to the reduction of mortality. There was also a reduction in the incidence of painful events and hospitalizations, improving the clinical and life expectancy of people with sickle cell disease.¹²

The bond is formed through the affective and closeness bonds that, most of the time, are present in the expressions of affection, feeling of will to remain together, admiration and respect, which impress feelings of happiness.^{2,14} In the case of this study, even fatigue or pain was prominent, follow-up, support, and complicity made a difference in the control of sickle cell disease in both adolescents. Friends and neighbors played a key role in family life 1.

Conflicts in relationships in both families, in the different lifestyle, personality, only affected the connection between the sister-in-law and the adolescent of the family 2. However, whenever necessary, the matriarch Lena, managed the conflicts, demonstrating to be an assertive leader. The affective bonds can also be formed with those who do not have consanguineous bonds. It is worth emphasizing that each person reacts differently with chronic disease and the links do not settle in an egalitarian way.³

Their families are the fundamental source of help and support of the person suffering from a chronic illness like the SCD and being

followed-up most of the time. However, the social network, consisting of individuals who can support the person, such as friends, neighbors and the host family, are also shown as fundamental and essential to overcome difficulties.^{3,14}

Regarding the spirituality/religion, faith assists the person to trust in a superior force and helps to maintain hope, and constitutes a constructive way of thinking; it is a feeling of confidence that the best will always happen.^{11,14,16} Indeed, faith is a feeling rooted in our culture, and it is as indispensable as other forms of coping with a chronic condition.^{14,16} Beliefs, values, and behaviors assimilated in social life conjecture the experiences of life that the person and the families are acquiring in the health-disease-illness process and in self-care,¹ as reported by the members of the families participating in this study.

Based on these assumptions, the nurse needs to understand the family, welcoming and recognizing their experiences to mobilize the investigation of new knowledge and forms of learning to exercise care with the sick family member, highlighting the importance of the needs and priorities of the person and his family, and not the nurse, seeking to qualify and humanize the care given.^{3,11,14}

The resilience of these families to the adversities of the chronic disease and the way they act to confront the challenges and threats, the anguish, the suffering, the loss, the change, attest to the capacity of the human spirit to understand and resign in the face of situations imposed by life and are profoundly inspiring attitudes. Thus, despite the difficulties faced by the chronic diseases that afflict a member of each of the participating families, we can consider them healthy families, because they can take care of each other and manage the adversities imposed by the disease.

Family health needs were evidenced in the biopsychosocial aspects, which demanded diversified behaviors involving mainly orientations and music therapy, as well as support by attentive listening of the difficulties they faced in the experience of the crises and, in this context, the strengths were emphasized in the satisfactory results that were not always possible due to the complexity of the situation.

It is noteworthy that the sessions of music therapy allowed a better family environment, socialization among the members of the families; relaxation and amelioration of everyone's anxiety, and reducing the fatigue of adolescents.

Studies show that music has physiological effects involving sensory, hormonal and psychomotor reactions, such as changes in metabolism, release of adrenaline, regulation of respiratory rate, changes in blood pressure, reduction of fatigue and muscle tone, and increase of sensory stimulus, improving attention and concentration, as well as stimulating sensitivity, placing the person in direct contact with their emotions and establishing a bond that leads to better integration and group coexistence.²⁴⁻²⁵

The coexistence with these families provided the opportunity to share the anguish and difficulties lived by these relatives, the opportunity to listen to them, to be closer to their daily life and better understand how it is to live daily with sickle cell disease, highlighting the adversities, conflicts, and interurrences that arise during the process of living with the family.

CONCLUSION

MCAIF proved to be an effective instrument for dealing with families, as it provided a broad view of its conditions, internal and external relationships, and the conditions of each of its members. The identification of the strengths existing in the family relationship and in each member, allowed to act in a more consistent way in the fragilities and to establish health actions with greater balance between both.

The experience provided in caring for these families has enabled to experience the emptiness of the process of death and dying, situations of conflict and approximations, constituting a lesson so that we can analyze our values and prejudices, which, once they were alien to the true reality of being human, today they fit a little closer to this real and not the ideal.

Despite agreeing with the criteria stipulated in the methodology, one of the limitations of the study was to be carried out with two families of adolescents with sickle cell disease, hindering to generalize the results. However, this limitation did not invalidate the study and satisfactorily responded to the research propositions. The results stimulate the continuity of this type of clinical study with cases not described in the literature, because, in this way, it will provide a continuous, updated and quality care with a focus on the person's well-being and the reach of its autonomy of health.

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