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ORIGINAL ARTICLE

THE THERAPEUTIC ITINERARY OF THE CHILD WITHFALCIFORM DISEASE[1]

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ABSTRACT

Objective: to know the therapeutic itinerary of the child with a sickle cell disease monitored in a hematology clinic of a public children's hospital. **Method**: this is qualitative research carried out in a pediatric hematology clinic of a public hospital in the state of Espírito Santo. Data collection washeld from January to June 2019 through semi-structured interviews with 10 family members of the child with a sickle cell disease. We submitted the data to the Thematic Analysis. **Results**: the therapeutic itinerary begins with the discovery of the disease and referral to specialized health services. The participants reported their knowledge of the crisis and the places of care to which they take the child to be attended. Also, there was a report regarding the frequency of consultations and access to the service, showing that they have no difficulty with it and are satisfied with the service provided. **Final considerations**: the study revealed a therapeutic itinerary in which the child goes through general health services at the time of diagnosis and is referred to the specialized service, in the secondary health care network to monitor his treatment and prevent complications. The health services are also effective, yet centralized.

Keywords: Anemia, sickle cell. Child. Therapeutics. Child health.

INTRODUCTION

Approximately 3,500 children with a sickle cell disease (SCD) are born each year in Brazil and 20% die before the age of 5, especially when they do not receive regular care from the health team[(1)]. This problem is complex in the lives of children and adolescents with sickle cell disease and their families since there is a pilgrimage between health services of different complexities due to their health care needs, interfering in the proper development[(2)].

Bahia is the state of Brazil with the highest incidence of SCD, where for every 650 live births, 1 is diagnosed with the disease. While in others, such as Rio de Janeiro, the ratio is 1: 1,300. The rate in the states of Pernambuco, Maranhão, Minas Gerais, and Goiás is 1:1,400; in Espírito Santo is 1:1,800;in the southern states there is Rio Grande do Sul with 1:11,000; Santa Catarina and Paraná 1:13,500[(1)].

Due to its great epidemiological and clinical relevance, the early diagnosis of SCD in children is essential to start appropriate treatment and appropriate early care. In this sense, for its management, prevention of disabilities and deaths, the Ministry of Health (MS) recommends carrying out the heel prick test, regulated through the National Neonatal Screening Program MS/GM No. 2,048, of September 3, 2009[(1)].

According to a study that analyzed the profile of hospitalizations of patients with SCD in the state of Espírito Santo from 2001 to 2010, 29.7% of hospitalizations were of children aged up to 4 years old. The length of stay in the hospital of up to 4 days was more frequent in children up to 9 years old and more than 4 days for adolescents over 15 years old. Also, the highest frequency of hospitalizations and deaths were in the capital (Vitória), followed by the city of São Mateus[(3)], justifying the data because it is configured in a municipality close to the border with the State of Bahia.

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Thus, from the first signs and symptoms of illness to the early or late diagnosis of the disease, there is a path to be followed by the child and his family. This path is influenced by individual and socio-cultural practices in an attempt to resolve child health impasses and it is called therapeutic itinerary[4].

Research on the therapeutic itinerary in the search for health care of SCD has contributed to the understanding of the performance of this care andhow health services are performed and used[5-6]. However, the paths followed are sometimes different and do not necessarily are similar to schemes or flows predetermined by the health system[4]. The basic guidelines of the line of care for people with SCD establish that Primary Care should be the way to health care and consequently guiding to the appropriate care in the service network[1], according to each child's need.

Thus, when this process is improperly performed, it can lead to a late diagnosis, reducing the chances of recovery and increasing the risks of sequelae and death. Thus, we need to know the journey of families of children with SCD to better assist them, minimizing possible complications, and providing a better quality of life for both. Reflecting on the above, the following guiding question emerged: How did the therapeutic itinerary of the child with sickle cell disease monitored at the hematology outpatient clinic take place? Thus, this study aims to know the therapeutic itinerary of the child with sickle cell disease monitored in a hematology clinic of a public children's hospital.

In this sense, the production of knowledge in this area is important to present the configuration of the service currently provided to children with SCD and to subsidize the construction of integrated practices systematically and longitudinally, providing access and bonding, whose impacts will affect the quality of life of these individuals and their families. Concerning to nursing, it may contribute to guiding health education and professional training actions, improving the flow of care for these children.

METHOD

This is a qualitative study[7] carried out in a pediatric hematology outpatient clinic of a public children's hospital in the state of Espírito Santo. We chose this study scenario because it is a public institution that is a reference in the diagnosis and monitoring of children and adolescents with SCD in the state.

Ten family members of the child with SCD, aged 0 to 12 years old participated in the study such as father, mother, and grandmother. We follow these criteria: family members and/or guardians of the children, present during the child's consultation at that institution; and responsible for the direct care of the child. Those whose child had some clinical complications during data collection would not participate.

The inclusion of new participants was closed according to the criteria of theoretical saturation. It occurs when new elements cease to emerge from the collected data because supposedly, the possible theoretical density has been reached, based on the empirical data available and the analytical and interpretive attributes of the researchers[8]. Thus, it is necessary to comply with methodological rigor in the production of scientific knowledge[9].

We carried out data collection from January to June 2019 through a semi-structured interview script with open questions related to maintaining the health of the child living with SCD. The questions were: What do you know about sickle celldisease? How was the child's diagnosis performed? Talk about the way you took to seek help when you noticed something different from the child and today, where do you look for support to maintain the child's health? What are the facilities and difficulties found to maintain the child's health from diagnosis until now? As we conducted the interview, other questions emerged. Data were collected by a single researcher, in a room provided by the clinic.

We recorded the interviews using a voice recorder on the cell phone of one of the researchers and we later transcribed them in full. The interviews lasted an average of 13 minutes. We submitted the data obtained to the Thematic Analysis technique, which is divided into three stages: pre-analysis; exploration of material and data processing; inference, and interpretation[7]. The thematic unit emerged from the empirical data: Therapeutic itinerary in sickle cell disease: from the discovery of the disease to outpatient follow-up.

This study is part of the project "Formation of the State Nucleus of Permanent Education in Sickle Cell Disease". All ethical aspects of this study were respected following Resolution 466/2012 of the National Health Council and approved by the Research Ethics Committee of the Health Sciences Center of the Federal University of Espírito Santo, under opinion No. 3,073,499 and CAAE No. 03108918.9.00005060. We used the alphanumeric system to guarantee the anonymity of the participants using the letter E for "interviewee", followed by numbering according to the chronological order.

RESULTS

From the disease discovery to the outpatient follow-up

The process of most children with SCD started from their diagnosis through the heel prick test and, after its confirmation, they were referred to the Association of Parents and Friends of Exceptional People (APAE acronyms in Portuguese) and the reference children's hospital, as we can verify through the following statements:

> When he was born, they do the heel prick test at the maternity hospital. Then, when we took him, the girl {doctor} said that he had sickle cell anemia and had already referred him to APAE. **(E4)**

> He did the heel prick test at the maternity hospital. After 45 days that we took him to the health center, the girl said that he had sickle cell anemia and had already been referred to APAE and from there they referred him to the children's hospital. **(E8)**

> There was a change in his heel prick test. Getting there {APAE} she said she had a disease called sickle cell anemia, we had never heard of it.**(E1)**

One participant reported that the diagnosis in his child was only when hewas 8 months because he had not yet done the heel prick test and that it was performed after presenting a complication for the first time.

> At 8 months old, when he was sick for the first time. When he was 8 months old, he didn't have a heel prick test. Another heel prick test was performed and it was found that he had sickle cell anemia.**(E6)**

Another participant reported that the heel prick test did not diagnose SCD, only the sickle cell trait. However, after the child was hospitalized due to some major complication, a new investigation was carried out at the hospital, and the disease was confirmed:

> The feet prick test was done and it showed that she had the sickle cell trait. However, she was very sick. Here {hospital} they did several tests and found that she has sickle cell anemia and not the trait.**(E10)**

With the confirmation of sickle cell disease, most participants reported following the child's follow-up without difficulties. Also, they reported that all the child's health needs were adequately met, and were well met in the health services they went through:

> No! I've never had anything difficult. I've always been very well assisted, his things very easy here. I have nothing to complain about. **(E6)**

> No, he has had a gall bladder operation, he has had a head exam. Everything here at the children's hospital. **(E5)**

> We were very well received both at APAE and here at the hospital. All our doubts have always been answered. We have nothing to complain about. **(E7)**

One participant reported difficulty accessing the specialized health service before the diagnosis because the health team did not know what their child had. However, when receiving the diagnosis and they were referred to the reference service, the participant reported having received the necessary care:

> At first, yes, because they didn't know what she had. [...], but after we entered the children's hospital it was wonderful. Here {hospital} we have always had all the assistance. I have nothing to complain about.**(E10)**

Regarding the frequency of consultations, the study participants referred from biweekly to half-yearly consultations. However, after some time, the consultations are becoming more spaced out:

> Every month. Sometimes every 15 days. **(E4)**

> At first, it was every 30 days. The consultations were spaced, 2 months, 3 months. Today I come here in about 5, 6 months. At most, I come in

about 4 months.**(E2)**

On the other hand, children also need to access the health services during the complications from the sickle cell disease and, according to the study participants, pain is the main factor that makes this search happen. Also, paleness, tiredness, fever, swollen hands, and feet stand out:

> The symptoms are a pain. I remember the pain more. **(E10)**
>
> Pain in the wrists because of the passage of blood. **(E9)**
>
> He went pale, very tired. **(E6)**
>
> Fever, tired, pain in the body. **(E4)**
>
> Swollen feet, the little hand. **(E1)**

In this context of complications, the participants informed the places where they usually go in case the children have a sickle cell crisis that is the service where they perform follow-up, the location of this study. However, sometimes, they also went to hospitals that attended the family health plan.

> Children's Hospital! **(E3, E10)**
>
> The first time I took him to the children's hospital. The other times we use health insurance. **(E4)**

For participants who do not live in the state capital, they seek to take their child to the referral hospital near their city. However, when they have a mild crisis, they do not need to travel to seek care.

> This year, thank God, he had a little crisis. There was no need to go to hospital x {hospital in the neighboring city}, it worked out in Conceição da Barra. **(E5)**

DISCUSSION

In most of the cases of this study, the discovery of the disease was through neonatal screening and the children were soon referred to reference institutions such as APAE and/or the children's hospital in the region. This moment represented the starting point for the search for the maintenance of the child's health in the context of sickle cell disease.

The data presented are in line with a study on the therapeutic itinerary of *quilombola* children with SCD, in which the mothers reported that the diagnosis was based on the heel prick test and that they monitor the disease at the APAE. However, they highlight that some procedures such as blood collection and childcare consultation could be done at the Basic Health Unit because it is closer to their home[5].

The heel prick test became mandatory across the country in 1992. It is a preventive action that allows the diagnosis of asymptomatic genetic and infectious diseases in the neonatal period, allowing the institution of specific early treatment and the monitoring of cases. In 2001, the Ministry of Health created the National Neonatal Screening Program, reaching an average national coverage of 83.6% in 2015[10]. Thus, the earlier the child is diagnosed, the better his prognosis will be since the early diagnosis allows the family to be adequately prepared to manage the disease and prevent its problems.

The literature clarifies that early diagnosis is an important role in preventing morbidity and mortality from sickle cell diseases, confirming the need to strengthen the Neonatal Screening Program so that there is broader coverage throughout the national territory[10].

Even though it was recommended that the heel prick test should be done as close to birth as possible, one participant in this study reported that his child only took the test at 8 months old due to a complication with his son, with a late-diagnosed of SCD. These data are similar to research with adults with SCD about their therapeutic itinerary. One of the participants whose pseudonym was Bemba reported that he was diagnosed only at 3 years and 9 months old, as they thought it was hepatitis[6].

Therefore, the late diagnosis is performed in the postneonatal period and intended for people of any age group, and once confirmed, the person or family (in the case of minors) should receive adequate information from the medical professional who requested the examination. and, then, be referred to the specialized service and maintained thebond with the basic health unit (UBS). At this stage, the doctor will be able to advise on the genetic situation and offer the possibility of diagnosis to the family. The diagnosis has been frequent in adults already in reproductive life, after the greater visibility that

the disease has achieved in recent years[1].

In the context of the diagnosis, the performance of the health professionals is usually highlighted as they must know about the disease and pass on the necessary information clearly to their parents[2].

Another participant reported that his child took the test while still in the hospital, but accused that she had a sickle cell trait and diagnosed with the disease only later when she faced some complications.

Sickle cell trait is a genetic condition in which the carrier inherits the hemoglobin A gene from one parent and hemoglobin S from the other, which makes it genetically HbAS since normal hemoglobin is given by HbA and HbS that is the sickle-shaped abnormal hemoglobin [11]. This means that the people with sickle cell trait do not have the disease but it is essential that they know their genetic condition so that in the future they will be able to make conscious choices regarding their sexual and reproductive health.

In addition to the "heel prick test", the diagnosis for SCD can be performed at a later time through a laboratory test called hemoglobin electrophoresis, in which the type of hemoglobin in the blood is detected[12]. Thus, it is necessary that in the case of suspected SCD, the doctor requests additional tests to confirm the real diagnosis as early as possible.

Participants reported that they did not find it difficult to access health services and still report satisfaction in care, having met all the child's health needs. Only one of them complained that access was difficult when they were still investigating the child's disease, but that after the SCD was confirmed, there were no problems.

In this sense, data showed an effort to comply with the guidelines of the National Policy for Comprehensive Care for People with Sickle Cell Disease and other Hemoglobinopathies regarding comprehensive and multidisciplinary care[13], implemented in practice in the place where the study was conducted. Therefore, we should not generalize these data considering that the other people reference institution for children with sickle cell disease is in the same region as the study place and, in other places, especially in the interior of the state, there is a lack of specialized health services in this area.

Different from these results, a study carried out in southern Brazil revealed children and adolescents facing a very different reality, such as the lack of integration in the health care network[2.)]

The access of people with sickle cell disease to health services should preferably be through the Family Health Strategy, which is considered the priority way for the entire health system, assuming the role of coordinating care and also responsible for the referral of the patient to the other levels of care, ideally organized in integrated care networks[2,14]. However, the access of people with sickle cell disease to primary care is reduced because they seek secondary care directly and in many cases, represented by the blood center[15], or another reference place other than basic care.

The search for secondary care has historically occurred because the treatment of sickle cell disease is perceived as complex and the exclusive competence of blood centers. Thus, professionals at other levels of care may ignore people with the disease, causing the patient to recognize only hematological centers as places to take care of their health[15].

Corroborating with the aforementioned data, there is the invisibility of sickle cell disease for decades that goes further, opening up the fragility of State policies since it puts the lives of people with sickle cell disease at risk by preventing them from being seen as individuals of rights, and not as a sick body to be prevented in a eugenic practice[16].

The participants in this study revealed the frequency of consultations with children with SCD since the beginning of the diagnosis and varying according to their age and health status. Thus, when care starts in the first months of life, the proposed preventive program usually on an outpatient basis includes periodic returns for monitoring growth and development; introduction of family members in educational programs on pathophysiology, consequences and limitations resulting from the disease, therapeutic alternatives, hydration pattern and desirable diet and expectations of growth and development; family coexistence strategies reducing overprotection and stimulating the child's independence[17-18].

When the sickle cell disease is identified in a

specific person, they need regular and adequate health monitoring, according to the guidelines defined by the Ministry of Health. The health team needs knowledge related to the SCD and, based on it, they should guide the person and/or his family to quickly identify the signs of the severity of the disease and seek immediate treatment, andbe informed of day-to-day care and prevention measures[19]. When this does not happen, dissatisfaction and low credibility in the service provided can compromise the process of monitoring the disease and weaken the bond between the patient and service[20].

Thus, we can see that in the case of children, when morbidity and mortality are greater, the period of return to consultations may be shorter. We should consider that each patient reacts to treatment differently, making it necessary to outline a unique therapeutic itinerary for each patient and the knowledge they have about SCD to intervene and prevent its complications and guide the service location.

The most-reported clinical manifestation of SCD by the participants was the pain. However, according to them, the children also showed pallor, tiredness, fever, and swelling in the hands and feet. The pain crisis or vaso-occlusive crisis is the most common demonstration of sickle cell disease and results from the deformation of red blood cells that interferes with the flow of blood causing them to adhere to the blood vessel wall and cause painful crises and tissue damage that can be chronic and progressive[1].

Also, the way to be taken by families to face the complexity of symptoms and complications and to recurrent hospitalizations end up changing the daily lives of children and their families, as the chronic condition often involves prolonged treatments that extend to the home and occur unpredictably, overloading family members and more intensely the caregiver. In this way, each family responds in different ways to chronic disease[21].

A limitation of the study was that it was carried out in just one institution, which did not allow to know the therapeutic itinerary of families of children with SCD assisted in other public and private health settings. In this sense, we recommend other studies to expand and compare the results.

FINAL CONSIDERATIONS

The study showed a therapeutic itinerary in which the child goes through general health services at the time of diagnosis and is referred to the specialized service, in the secondary health care network to monitor his treatment and prevent complications.

The results showed that access to these health services in Espírito Santo is effective and adequate according to the participants, but centralized in Grande Vitoria, which can be a hindrance to access for children living in the countryside but can also mask the reality of other health services. Thus, the study demonstrated the need for investment in the decentralization of care to facilitate faster access for children with a crisis, especially in the countryside.

Also, they recommend that health professionals involved with these people mobilize for comprehensive care with the inclusion of the child and his family in the planning and provision of care since the diagnosis. In this context, we highlight the increasing importance of investment and professional training for the recommended diagnosis since in some cases it is done late, compromising the quality of life.

O ITINERÁRIO TERAPÊUTICO DA CRIANÇA COM DOENÇA FALCIFORME

RESUMO

Objetivo: conhecer o itinerário terapêutico da criança com doença falciforme acompanhada em um ambulatório de hematologia de um hospital público infantil. **Método:** pesquisa qualitativa realizada em um ambulatório de hematologia pediátrica de um hospital público do estado do Espírito Santo. A coleta dos dados ocorreu no período de janeiro a junho de 2019 por meio da entrevista semiestruturada com 10 familiares da criança com doença falciforme. Os dados foram submetidos à Análise Temática. **Resultados:** o itinerário terapêutico se inicia a partir da descoberta da doença e encaminhamento aos serviços de saúde especializados. Os participantes relataram o conhecimento sobre a crise e quais os locais de atendimento que levam a criança para ser atendida. Além disso, houve relato quanto à frequência de consultas e o acesso ao serviço, demostrando não terem dificuldade com ele e estarem satisfeitos com o atendimento prestado. **Considerações finais:**o estudo revelou um itinerário terapêutico no qual a criança perpassa por serviços de saúde gerais no momento do diagnóstico e é encaminhada para o serviço especializado, na rede de atenção à saúde

secundária, para seguimento do seu tratamento e prevenção das complicações. Além disso, os serviços de saúde são eficazes, porém centralizados.

Palavras-chave: Anemia falciforme. Criança. Terapêutica. Saúde da Criança.

EL ITINERARIO TERAPÉUTICO DEL NIÑO CON ANEMIA FALCIFORME

RESUMEN

Objetivo: conocer el itinerario terapéutico del niño con anemia falciforme acompañado en un centro ambulatorio de hematología de un hospital público infantil. **Método:** investigación cualitativa realizada en un centro ambulatorio de hematología pediátrica de un hospital público del estado de Espírito Santo-Brasil. La recolección de los datos ocurrió en el período de enero a junio de 2019 por medio de entrevista semiestructurada con 10 familiares del niño con anemia falciforme. Los datos fueron sometidos al Análisis Temático. **Resultados:** el itinerario terapéutico se inicia a partir del descubrimiento de la enfermedad y el encaminamiento a los servicios de salud especializados. Los participantes relataron el conocimiento sobre la crisis y cuáles los lugares de atención que llevan al niño para ser atendido. Asimismo, hubo relato sobre la frecuencia de consultas y el acceso al servicio, demostrando no tener dificultad con él y que están satisfechos con la atención prestada. **Consideraciones finales:** el estudio reveló un itinerario terapéutico en el cual el niño pasa por servicios de salud generales en el momento del diagnóstico y es encaminado para el servicio especializado, en la red de atención a la salud secundaria, para la continuación de su tratamiento y la prevención de las complicaciones. Además, los servicios de salud son eficaces, pero centralizados.

Palabras clave: Anemia de células falciformes. Niño. Terapéutica. Salud del niño.

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