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CHARACTERIZATION OF PERSONS WITH FALCIFORM DISEASE IN A CITY OF THE STATE OF BAHIA

CARACTERIZAÇÃO DAS PESSOAS COM DOENÇA FALCIFORME EM UMA CIDADE DO ESTADO DA BAHIA

CARACTERIZACIÓN DE LAS PERSONAS CON LA ENFERMEDAD FALCIFORME EN UNA CIUDAD DEL ESTADO DE BAHIA

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Objective: to characterize people assisted in a sickle cell disease referral center in a city in the state of Bahia. Method: quantitative, descriptive and retrospective study, performed between August 2016 and February 2017. Secondary data collected in 326 medical records were processed in STATA, version 12.0. Results: prevalence of young adults, female, single, black, with income of up to one minimum wage, non-literate, Protestant, with up to three children. The prevalent complications were pain, jaundice, and spleen changes. The most commonly used medications were folic acid, hydroxyurea, ibuprofen and dipyrone. 67.79% of the patients remained in treatment, while 4.60% died. Conclusion: people with sickle cell disease attended at a referral center had a high degree of vulnerability and were subject to clinical variability.

Descriptors: Sickle cell diseases; Health profile; Epidemiology.

Objetivo: caracterizar as pessoas assistidas em um Centro de Referência em doença falciforme em uma cidade do estado da Bahia. Método: estudo quantitativo, descritivo e retrospectivo, realizado entre agosto de 2016 e fevereiro de 2017. Os dados secundários coletados em 326 prontuários foram processados no STATA, versão 12.0. Resultados: prevalência de adultos jovens, do sexo feminino, solteiras, pretas, com renda de até um salário mínimo, não alfabetizadas, protestantes, com até três filhos. As complicações prevalentes foram crise álgica, icterícia e alterações do baço. As medicações mais utilizadas foram ácido fólico, hidroxiureia, ibuprofeno e dipirona. Permaneceram em tratamento 67,79% das pessoas, enquanto 4,60% foram a óbito. Conclusão: pessoas com doença falciforme assistidas em um centro de referência possuíam elevado grau de vulnerabilidade e estavam sujeitas à variabilidade clínica.

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Descritores: Doenças de células falciformes. Perfil de saúde. Epidemiologia.

Objetivo: caracterizar a las personas asistidas en un Centro de Referencia tratamiento de la enfermedad falciforme en una ciudad del estado de Bahía. Método: estudio cuantitativo, descriptivo y retrospectivo, realizado entre agosto de 2016 y febrero de 2017. Los datos secundarios colectados en 326 prontuarios fueron procesados en el STATA, versión 12.0. Resultados: prevalencia de adultos jóvenes, del sexo femenino, solteras, negras, con renta de hasta un salario mínimo, no alfabetizadas, protestantes, con hasta tres hijos. Las complicaciones prevalentes fueron crisis álgicas, ictericia y alteraciones en el bazo. Los medicamentos más utilizados fueron el ácido fólico, hidroxiurea, ibuprofeno y dipirona. Permanecieron en tratamiento 67,79% de las personas, y 4,60% terminaron en óbito. Conclusión: las personas con enfermedad falciforme asistidas en un centro de referencia tenían un grado elevado de vulnerabilidad y estaban sujetas a la variabilidad clínica.

Descriptores: Enfermedades de células falciformes. Perfil de salud. Epidemiología.

Introduction

Sickle cell disease (DF) is a chronic, hematological and hereditary pathology that promotes the falcization of red blood cells, causing vaso-occlusion. People with FD may present several complications, such as recurrent infections, splenic sequestration syndrome, pain crises, acute chest syndrome (ACS), leg ulcers, stroke, priapism, among others[1].

Early diagnosis is an important factor for the prevention of FD-related morbidity and mortality. Thus, the FD test in the Foot Test was included in 2001 by the Brazilian National Neonatal Screening Program (PNTN – *Programa Nacional de Triagem Neonatal*)[2].

In the state of Bahia, between 2007 and 2009, it was found that 23.3% of the children in the capital presented a positive result for FD in the Newborn Bloodspot Screening Test[3]. In turn, the second largest city in Bahia presents a prevalence of four cases per 10,000 inhabitants, and approximately 81% of this total resides in urban areas[4].

In the capital of Bahia, the population with FD receives medical and hospital assistance at the Hematology and Hemotherapy Foundation of Bahia (HEMOBA – *Fundação de Hematologia e Hemoterapia da Bahia*) and the Association of Parents and Friends of the Disabled (APAE – *Associação de Pais e Amigos dos Excepcionais*); however, people from other cities of the state need to move to the capital in order to receive specialized treatment, which causes physical and financial wear and tear. In view of this, Bahia's second largest city implemented a Municipal Reference Center for People with Sickle Cell Disease (CRMPDF), in order to meet the specificities that this population presents in the treatment of the Federal District and to prevent them from continuing to have problems regarding geographical access to receive specialized assistance[5].

The characterization of people with sickle cell disease in the second largest city in Bahia proves to be extremely important, since such knowledge will allow identifying who the assisted people are, guiding therapeutic interventions necessary to control the conditions that endanger their life. In addition, it can contribute to the elaboration of evaluation and planning of the health service aimed at this population.

In this expectation, it was delimited as object of study the "Characterization of the people assisted in a Sickle Cell Disease Reference Center in a city in the state of Bahia". The present study should answer the following research problem: What is the socioeconomic, demographic and clinical characterization of the people assisted in the Municipal Reference Center to the Person with Sickle Cell Disease in a city in the state of Bahia?

The results of the study will contribute so that the health professionals of the city know the characteristics of this population and, thus, can act in the prevention of diseases and in the health promotion of people with FD. In addition, they will allow managers to evaluate and plan health

services, supporting intervention policies and coping with this problem. It is also hoped that the data can serve as a basis for further research.

The objective of the study was to characterize people assisted at a Reference Center on sickle cell disease in a city in the state of Bahia.

Method

This is a descriptive study, of a quantitative nature, developed with secondary data. The retrospective method was used to collect professional records in records of people assisted in a CRMPDF from a municipality in the state of Bahia, which assists people with FD and their families since 2012.

The said municipality is located in the countryside in Bahia, 108 km distant from Salvador, capital of the state. It has a total land area of 1,304,425 km^2, an estimated population of 622,639 inhabitants and a population density of 416.03 inhabitants per sq. km. It is the second largest municipality in the state[3].

The CRMPDF was implemented in 2012 and offers several services that include: immunization, health education, specialized care for people with FD and leg ulcers through nursing consultations, consultations with physiotherapist, nutritionist, hematologist, social worker, neurologist and accomplishment of the transcranial Doppler. In addition, through the reference and counter-referral service, it conducts referrals to the various medical specialties, as well as laboratory tests and the treatment of associated diseases.

The data from this study are secondary, and the sample consisted of 326 records of a total of 363, referring to people assisted in the CRMPDF between August 2012 and February 2017. Of the 363 medical records, seven were excluded from the study because they referred to people with Sickle cell trait. Of the remaining 356, 30 were excluded due to incomplete and/or unconfirmed diagnoses, leaving 326 records to be analyzed. Of these, not all of them contained complete information about all the observed variables. Medical records with incomplete information were excluded from the sample at the time of analysis.

Inclusion criteria were: people of both sexes, of any age group, diagnosed with sickle cell disease, who had been assisted in the CRMPDF of a municipality in the interior of Bahia, from its implantation in August 2012 until February 2017. The exclusion criteria were: people with sickle cell trait, diagnosis not confirmed and/or who were not registered in the CRMPDF of a municipality of the countryside of Bahia, from August 2012 to February 2017.

For the recording of these data, a form composed of several variables was elaborated solely to meet the objectives of the present study. The first part of the instrument investigated the socioeconomic and demographic characteristics of people with FD; the second, addressed the clinical characteristics of these people; the third part dealt with the reproductive history of women with FD who attended the Center; while the fourth part addressed the outcome of these people in relation to treatment.

The data collection in the CRMPDF happened between August 2016 and February 2017. It was performed during the week, during service hours, in an available room, so that the researcher and volunteers could work without interfering with the dynamics of the Unit.

The data were computed in a database built by the Epidata program, version 3.1, and then analyzed and processed electronically using the Data Analysis and Statistical Software program (STATA) version 12.0. The quantitative analysis was then performed through descriptive statistics, which evaluated the simple, absolute and relative frequencies of events.

This research is a cut from the research and extension project titled "IKINI: Care Practices for People with Sickle Disease and Their Families," approved by the Committee on Ethics in Research with Human Beings (CEP) by Opinion no. 1,254,708. The study complied with the principles that guide research with human beings, respecting Resolutions nº. 466/12 and nº. 510/2016 of the National Health Council.

Results

Patients assisted at the CRMPDF were aged between 1 and 69 years, the most frequent age group (139) corresponded to 20 to 59 years, referring to 42.64% of the cases, and the mean age was 19.4 years. Of these, 53.69% (175) were female. Regarding marital status, 85.41% (158) were unmarried. Regarding income, 74.29% (52) reported receiving up to a minimum wage, 24.29% (17) reported earning between two and three monthly minimum wages and only 1.43% (1) claimed to earn income greater than three minimum wages. The average income was R$ 784.76. Regarding schooling, 35.85% (19) were not literate and only 3.77% (2) concluded higher education. As for race/color, only 56.52% (13) declared themselves to be blacks, 34.78% (8) declared themselves to be brown. Regarding religion, 61.11% (11) were Protestants. As for the number of children, 88.37% (38) of the patients revealed that they had up to three children and only 11.63% (5) patients had more than three children (Table 1).

Table 1 – Characterization of people with sickle cell disease attended at the Municipal Reference Center to people with sickle cell disease, according to socioeconomic and demographic data. Feira de Santana, Bahia, Brazil – 2017 (continued)

Characteristics	n	%
Age Group (n=326)		
1 to 9 years	106	32,52
10 to 19 years	78	23,93
20 to 59 years	139	42,64
60 years and over	3	0,92
Sex (n=326)		
Male	151	46,32
Female	175	53,69
Marital Status[1] (n=185)		
Not married	158	85,41
Married	23	12,43
Stable union	2	1,08
Divorced	2	1,08
Income[2] (n=70)		
0 to 1 minimum wage	52	74,29
2 to 3 minimum wages	17	24,29
More than 3 minimum wages	1	1,43
Education[3] (n=53)		
Non-literate	19	35,85
Literate	1	1,89
Incomplete elementary	11	20,76
Complete elementary	2	3,77
Incomplete highschool	4	7,55
Complete highschool	13	24,53
Incomplete higher	1	1,89
Graduated	2	3,77
Race/color[4] (n=23)		
White	1	4,35
Black	13	56,52
Yellow	1	4,35
Brown	8	34,78
Religion[5] (n=18)		
Catholic	7	38,89
Protestant	11	61,11

Table 1 – Characterization of people with sickle cell disease attended at the Municipal Reference Center to people with sickle cell disease, according to socioeconomic and demographic data. Feira de Santana, Bahia, Brazil – 2017 (conclusion)

Characteristics	n	%
Number of children[6] (n=43)		
Up to 3	38	88,37
More than 3	5	11,63

Source: Created by the authors.

Notes: [1] 141 medical records did not contain this information.
[2] 256 medical records did not contain this information.
[3] 273 medical records did not contain this information.
[4] 303 medical records did not contain this information.
[5] 308 medical records did not contain this information.
[6] 181 medical records are not requested for this questioning and 102 medical records did not contain this information.

Table 2 shows that SS hemoglobinopathy (HbSS), also known as Sickle Cell Anemia (SCA), is the most prevalent type of FD, characterizing 50.65% (155) of the cases, and it is followed by SC hemoglobinopathy (HbSC) with 44.77% (137) of the cases. Regarding the age of the diagnosis of FD, 43.11% (97) of the patients were still diagnosed in neonatal screening. With regard to complications from sickle cell disease, an expressive part of the analyzed population, equivalent to 88.70% (259), reported having had complications, while only 11.30% (33) denied any type of complication throughout life.

Among the 292 medical records of people who suffered or suffer complications from FD, only 259 of them contained information on hospitalizations in emergency services due to complications of the disease. Of these 259 people, 60.97% (164) reported that they had already had to be admitted to emergency services and 54,45% (153) had already needed to use red blood cells to treat the complications presented (Table 2).

As a form of prevention of some complications, it is necessary that this population maintain the vaccine calendar updated. Among the patients assisted in the CRMPDF between 2012 and 2017, 78.51% (95) kept the vaccination card updated, while 21.49% (26) of the medical records reported that it was outdated (Table 2).

Table 2 – Characteristics of the clinical conditions of the people assisted in the Municipal Reference Center to the Person with Sickle Cell Disease. Feira de Santana, Bahia, Brazil – 2017 (continued)

Characteristics	n	%
Type of FD [1] (n=306)		
SS hemoglobinopathy	155	50,65
SC hemoglobinopathy	137	44,77
S thalassemia	6	1,96
Hemoglobin variants	8	2,61
Diagnostic period [2] (n=225)		
Neonatal screening	97	43,11
Childhood	71	31,56
Adolescence	20	8,89
Adulthood	37	16,44
Presents complications of the disease[3] (n=292)		
Yes	259	88,70
No	33	11,30
Has already been hospitalized in emergency services due to complications of the disease[4] (n=269)		
Yes	164	60,97
No	105	39,03

Table 2 – Characteristics of the clinical conditions of the people assisted in the Municipal Reference Center to the Person with Sickle Cell Disease. Feira de Santana, Bahia, Brazil – 2017 (conclusion)

Characteristics	n	%
Has already undergone blood transfusion[5] (n=281)		
Yes	153	54,45
No	128	45,55
Is the vaccination card up to date?[6] (n=121)		
Yes	95	78,51
No	26	21,49

Source: Created by the authors.

Notes: [1] 20 medical records did not contain this information.
[2] 101 medical records did not contain this information.
[3] 34 medical records did not contain this information.
[4] 57 medical records did not contain this information.
[5] 45 medical records did not contain this information.
[6] 205 medical records did not contain this information.

Regarding the 175 women assisted in the CRMPDF from its implementation until 2017, 74.86% (131) were of reproductive age until the data collection was finalized. The few data found and analyzed on reproductive history (17) indicated that the contraceptive method most used was the oral or injectable contraceptive, with 70.59% (12) of the cases, and then the Intrauterine Device (IUD), with 17.65% (3) and the condom, with 11.77% (2) of adherents. In relation to the number of pregnancies experienced by 42 women, 35.71% (15) had only one child, 45.24% (19) had between two and three children and only 19.05% (8) had more than three children. Finally, among the 29 medical records containing information on the occurrence or not of induced and spontaneous abortions, the results showed that 58.62% (17) of the women suffered this complication.

Among the various complications presented by the 326 people with FD who attended CRMPDF throughout life, the main ones were: pain crisis, 52.15% (170); jaundice, 26.38% (86); spleen changes, 23.94% (78); complications of the respiratory system, 20.86% (68); recurrent infections, 13.80% (45); and leg ulcers, 11.96% (39). Because of the many complications to which they are susceptible, they need to use various medications.

The medical records showed that the main medications used were folic acid, which was recorded in 91.41% (298) of the medical records, followed by the phenoxymethylpenicillin potassium, with 25.15% (82), and hydroxyurea with 22, 39% (73) of registered cases, as shown in Table 3.

Table 3 – History of non-analgesic medications used by people with sickle cell disease attended at the Municipal Reference Center to the Person with Sickle Cell Disease. Feira de Santana, Bahia, Brazil – 2017 (continued)

Characteristics[1]	n	%
Folic acid (n=326)		
Yes	298	91,41
No	28	8,59
Phenoxymethylpenicillin potassium (n=326)		
Yes	82	25,15
No	244	74,85
Hydroxyurea (n=326)		
Yes	73	22,39
No	253	77,61

Table 3 – History of non-analgesic medications used by people with sickle cell disease attended at the Municipal Reference Center to the Person with Sickle Cell Disease. Feira de Santana, Bahia, Brazil – 2017 (conclusion)

Characteristics[1]	n	%
Antibiotics (n=326)		
Yes	17	5,21
No	309	94,79
Deferasirox (n=326)		
Yes	14	4,29
No	312	95,71

Source: Created by the authors.

Note: [1] 6 medical records were not requested for this questioning and 18 medical records did not contain this information.

During pain crises, the most commonly used painkillers for patients with DKA-assisted CRP were ibuprofen, with 38.34% (125) and dipyrone, with 33.44% (109), as shown in Table 4:

Table 4 – Analgesics used during pain crises by persons with Sickle Cell Disease attended at the Municipal Reference Center to the Person with Sickle Cell Disease. Feira de Santana, Bahia, Brazil – 2017

Characteristics[1]	n	%
Ibuprofen (n=326)		
Yes	125	38,34
No	201	61,66
Dipyrone (n=326)		
Yes	109	33,44
No	217	66,56
Paracetamol (n=326)		
Yes	45	13,80
No	281	86,20
Codeine (n=326)		
Yes	28	8,59
No	298	91,41
Tramadol hydrochloride (n=326)		
Yes	24	7,36
No	302	92,64
Morphine (n=326)		
Yes	15	4,60
No	311	95,40

Source: Created by the authors.

Note: [1] 37 medical records did not apply to this questioning and 104 medical records did not contain this information.

Regarding the situation of the 326 people assisted in the CRMPDF between 2012 and 2017, the results showed that, at the end of the data collection of this research, 67.79% of them continued to be followed regularly, 0.61% were transferred to another service, 26.99% were separated from the service for more than one year, and 4.60% died during that time.

Discussion

The most prevalent age group was that of adults between 20 and 59 years of age, which suggests that the adult population is aware of the importance of specialized follow-up for the treatment of the disease and the prevention of worsening. The high prevalence also in

children under 10 years indicates that the diagnosis made in neonatal screening is being effective in the identification of people with FD and the referral to referral centers in the treatment of the disease contributes positively to increasing the life expectancy of these people[6].

Regarding the low percentage of elderly people found in the study, the literature indicates that the life expectancy in patients with FD is still low, remaining between 40 and 50 years[7].

There was no significant difference between the prevalence of men and women in the results of the present study. However, studies indicate that women often adhere to health care more frequently than men. This is because, due to historical and sociocultural issues, women have always been responsible for providing health care to the family, which brings health services closer to them and ensures greater access to them[8].

Regarding marital status, the high prevalence of singles is supported by the Brazilian literature and suggests fragility in the social support of people with FD. This factor may be related to the stigma of the disease, its repercussions on the body, as well as the limitations it causes[9].

Draft Law no. 7.103/2014 seeks to make it mandatory to fill out the race/color questionnaire in all SUS records, so that this data can be used to substantiate the need to implement public policies aimed at the black population[10]. The scarce record – only 23 charts – of the CRMPDF patients' race/color reveals the carefreeness of the service professionals – nurses, physicians, psychologists, physiotherapists, and nutritionists – with a very important epidemiological aspect, since it is a disease inherited from the black population.

The results on income and schooling are supported in the literature, where it can be observed the predominance of average schooling and income lower than three minimum wages among people with DF[1]. It is known that low level of education brings losses to the individual, both in the aspect of self-care and in adherence to treatment and entry into work life, due to the limitation it causes in terms of reading, writing, speaking and understanding[11]. Thus, the professional life of these subjects is also compromised, as they end up carrying out activities that do not require professional qualification – mostly handicrafts and low-wage jobs – increasing the level of family dependency and social security commitments.

Regarding religion, it was observed the scarce filling of this variable in the charts analyzed. Of the 18 records found, 61.11% (11) of the people declared themselves Protestant and 38.89% (7), Catholic. Faith can be an important support in the lives of people with chronic diseases. In people with FD, for example, religiosity helps in coping with the disease and promotes well-being[12].

The present study also investigated the number of children that people who are assisted in CRMPDF have had throughout life and observed a small prevalence of only 43 people; of these, 88.37% have up to three children and an even smaller number (11.63%) have more than three children. Such results can be explained both by social isolation[7] and by the fear of these people transmitting the sickle cell gene to their heirs[13]. In addition, due to the physiological changes imposed by the disease, people with FD have delayed pubertal development[14]. Therefore, delayed body development and sexual maturation associated with social isolation and fear of transmitting the sickle cell gene to the heirs may negatively influence sexuality and reproductive experiences of both men and women with DF.

It is known that the risk of thromboembolism in women who use oral contraceptives is high and becomes even greater when these women have DF[11]. In view of this fact, the contraceptive method most appropriate for women with FD is the condom, but this was the least used by the women investigated in the study, whereas the oral contraceptive was the most used, which points to the risks of this therapy.

Gestation in women with FD increases the risk of maternal-fetal morbidity[11], as well as the fear of death and the fear of transmitting FD to the child. Faced with these expectations, women with FD experience negative feelings when they become pregnant, and they may often consider abortion the most viable outcome in this type of situation[15].

There is also an increased risk for spontaneous abortions, caused by the clinical condition of women with DF[11]. In addition to confirming the findings about the low prevalence of children with FD, the almost non-existent register of induced and/or spontaneous abortions in the study may be associated with both underreporting by the CRMPDF professionals and their fear of these women verbalizing the experience of abortion and suffer moral, religious and ethical judgments from society.

Regarding the FD type presented, the majority have HbSS, followed by HbSC. The results coincide with the literature, which shows that SCA is the most common and most severe form of FD in the world, and is commonly associated with low life expectancy[7], while HbSC is known as a medium-severity hemoglobinopathy[8].

The early diagnosis is an important factor in the prevention of FD complications, since preventive measures can be instituted and usually interfere positively in the treatment[1-2]. In this perspective, it is considered as a positive factor the fact that most of the diagnoses have yet occurred in neonatal screening, as recommended in Ministerial Decree GM no. 822, of 2001[2]. However, the percentages of people diagnosed in childhood and in adulthood are still high, which points to the need for training of health professionals, so that they are able to correlate clinical findings with pathology.

During vaso-occlusive crises, hemotransfusion is a commonly applied procedure with the objective of reducing the crisis and correcting hypoxia by reducing sickle hemoglobin in the bloodstream[12], which explains the percentage of people who underwent blood transfusion in the study. Vaccination is also an important factor in reducing mortality rates in FD, which points to the importance of keeping the vaccination calendar rigorously updated[13].

Regarding the clinical manifestations of FD, they are quite variable and are closely related to the degree of severity of the disease. The results of the present study corroborate those of other studies regarding the pain crisis[1,6]. Pain crises are very common in people with DF[12]. However, although pain is reported as one of the main complications of DF, the results found in this study were not as significant as expected, as it appeared in only 170 patients, which may suggest underreporting by CRMPDF professionals.

Jaundice also presented high prevalence in the present study. Liver changes are common in people with FD, especially in those with SCA, and appear less frequently in people with HbSC and thalassemia[13].

Other frequent complications identified in the results were changes in the spleen. Splenic dysfunction is one of the main complications of FD known for high mortality, especially among children[1,6].

Due to its important action in the fight against infections, splenic dysfunction makes the person with FD subject to recurrent infections. These infections, in turn, had a frequency of 13.8% in the present study, while in the United States, they account for 35% of the hospitalizations of people with DF aged over 20 years[13].

Known to be also prevalent complications in people with FD[1], STA leg ulcers, cholecystitis stroke and priapism were less frequent in the results.

Constantly, people with FD need to use medications that aid in the prevention and also in the treatment of the complications presented. The high prevalence of folic acid use and potassium phenoxymethylpenicillin in the study is explained by the fact that these medications are part of the routine treatment of FD[2,14].

Hydroxyurea, the third non-analgesic drug most frequent in the results, decreases vaso-occlusive crises and significantly reduces the acute and chronic complications of FD[7]. In view of its proven effect on adults and children, Ordinance no. 55/2010 of the Ministry of Health, approved its use for patients with FD and determined that the Secretariats of Health of the States of the Union and Federal District[7] would be responsible for dispensing the medication.

The measurement of pain is subjective and does not always present physical evidence[14]. Because of this, people living with chronic pain frequently suffer from the stigma caused by the invisibility of pain, such as the absence of vital

signs, which leads to professional discredit and to the simulation claims of the disease[16].

In the present study, the use of non-opioid analgesics/NSAIDs, such as ibuprofen, dipyrone and paracetamol in relation to opioids, such as tramadol hydrochloride, codeine, morphine and fentanyl prevailed; however, it was not possible to identify whether the persons investigated respected the doses and intervals indicated for each drug.

Low adherence to treatment may lead to a reduction in patients' quality of life[17], a fact that reveals the importance of linking patients, families and health professionals. Given this, it is considered a positive factor that most of the population remains in treatment in the CRMPDF through regular consultations, which may explain the small number of deaths that were identified in the study.

It is worth mentioning that there are limitations in the study inherent to its retrospective nature, and the bias may result from the revision of the medical records, the reliability of the data in them, the inadequate filling and even the difficulty found by the researchers to understand the handwriting of the CRMPDF professionals. However, it is important to emphasize that the quality of the records made in medical records is a criterion for the evaluation of the quality of care provided in the health services[18].

In view of this, it is necessary to raise the awareness of the CRMPDF team in order to better fill out the records of the people assisted. The service coordinator must identify the failures and encourage the professionals to modify the inadequate behaviors, always aiming at improving the quality of care.

Conclusion

This study allowed the characterization of people with FD assisted in a referral center in a city in the state of Bahia by means of the analysis of secondary data found in the patient records of the referred center.

The socioeconomic and demographic results of the present study corroborate those found by other authors and reinforce that people with sickle cell disease have a high degree of socioeconomic vulnerability, since they have low expectations in terms of life, income and schooling. Access to specialized treatment is a necessity for this population and the State must guarantee this right as a way to promote health and quality of life.

Clinical results, for the most part, also agree with much of the national and international literature and show that people with sickle cell disease have a wide clinical variability from childhood and may suffer from several lifetime limitations. The use of analgesic and non-analgesic medications is also a necessity that has to be assured to this population.

The results about the reproductive history of the women of childbearing age assisted in the CRMPDF suggest that this theme is little valued by service professionals, since the records on contraceptive methods, gestation and abortion are scarce. Fear and insecurity are common feelings in women with FD who experience the desire to be mothers. It is necessary that professionals understand the importance of reproductive health in people with FD and clarify the doubts of both women and men, so that they can decide on their sexual and reproductive health.

It is necessary to emphasize the expressive absence of records of several variables of the study, especially that related to the race/color issue. This fact configures itself as an important information bias that makes the results found questionable, but also emphasizes the importance of quality records for the evaluation of the care provided. The quality of health records is a challenge to be achieved, given the importance they have for the implementation of safe care for patients and for the multiprofessional team, as well as being important tools in the formulation, implementation, and evaluation of public policies aimed at population health.

It is suggested that the professionals of the reference centers that assist people with sickle cell disease are sensitized and trained – through courses, workshops, and lectures – in a permanent education proposal that emphasizes, among other aspects, the importance of registering

patients' data with quality, aimed at improving the service provided to the target population.

It is concluded that people with sickle cell disease assisted in a referral center have a high degree of vulnerability and are subject to clinical variability.

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