Quality of assistance provided to children with sickle cell disease by primary healthcare services

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Objective: *To evaluate the quality of healthcare provided to sickle cell disease children by primary healthcare services in a region of high prevalence.*

Methods: *A cross-sectional, descriptive study was performed by interviewing members of families with sickle cell disease children. The children had been identified from the Neonatal Screening Program in Minas Gerais state over the last 12 years in towns of the Montes Claros-Bocaiuva microregion. A structured questionnaire specially developed for this study and based on three axes was used: indicators of the child's health (immunization, growth and development, prophylaxis antibiotic therapy), perception of care by the family (health education and accessibility) and knowledge of the family about the disease.*

Results*: Sixty-three of 71 families with children identified as having sickle cell disease were interviewed. The predominant genotypes were Hb SS (44.4%) and Hb SC (41.2%). Adequate monitoring of growth and development was recorded for the first year of life in 23 children (36.6%) and for the second year of life in 18 children (28.6%). The basic vaccination schedule was completed by 44 children (69.8%) but 62 vaccination record cards (98.4%) identified delays of special vaccines. Regular use of prophylactic penicillin was reported by 55 caregivers (87.3%). The family's perception of the care provided suggests poor accessibility to health services and lack of opportunities to answer doubts. The average performance of families in knowledge testing was 59.8%.*

Conclusion: *The quality of healthcare is unsatisfactory. The care provided to children with sickle cell disease in primary healthcare services needs improvements.*

Keywords: *Quality of health care; Primary health care; Anemia, sickle cell; Children*

Introduction

Sickle cell disease is characterized as the commonest hereditary blood disease worldwide. It is a genetic disorder with deformation of red blood cells that assume a sickle shape, which in certain situations, induces polymerization of hemoglobin. This causes vascular obstruction, episodes of pain and injury to organs.[1] In Brazil the prevalence of sickle cell trait in the general population is estimated at between 2% and 8% and there are between 25,000 and 30,000 cases of sickle cell anemia.[2] In Minas Gerais, the incidence of sickle cell disease is 1:1400 live births with a clearly higher incidence in the northern region of the state according to data from the Newborn Screening Program of the State of Minas Gerais (PETN-MG).[3]

As a hereditary and chronic disease, sickle cell disease has a great impact on the entire family and the effects need to be better understood by healthcare services. An adequate approach to the disease results from the interaction of a multidisciplinary team with family involvement. The instability of the clinical manifestations and complications of the disease implies the necessity of assistance provided by the different levels of healthcare services.[4] Unfortunately, the quality of healthcare services is not yet so good in respect to sickle cell disease.[5] Often there is negligence that culminates in a high infant mortality rate.[6] In healthcare at this age, it is extremely important to ensure that effective practices are adopted and integrated to meet the needs of both the children and families.

Historically, healthcare provided to patients with sickle cell disease is considered to be the responsibility of blood centers. However, the entire healthcare network must be able to adequately serve these patients, promoting self-care and anticipating risk situations. Studies evaluating the care provided to sickle cell disease children in primary healthcare services will enable the identification of the needs and weaknesses in the healthcare network. These studies represent an important tool for future interventions. This study aimed to evaluate the quality of care provided to sickle cell disease children in primary healthcare services in the northern region of the state of Minas Gerais, Brazil.

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The authors declare no competing financial interest

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Methods

This is a descriptive, cross-sectional study carried out in the Montes Claros-Bocaiuva microregion in the north of Minas Gerais. The study area covers a population of approximately 450,000 inhabitants and has the second largest group of sickle cell disease patients of the state.

The sample size was not calculated. All families of sickle cell disease children in the microregion were considered eligible for the study. Families were identified from the records of PETN-MG provided by the Diagnosis Support Center (NUPAD-FM-UFMG). Sickle cell disease children are screened and monitored by the NUPAD. Children living in rural areas were excluded for logistic reasons. Families that were not located on three attempts to interview them and those who refused to participate were not included in the study.

A structured questionnaire was prepared for data collection based on the protocols of the Minas Gerais State Health Department[7] and the Brazilian Health Ministry.[8,9] Specially trained interviewers collected data directly from families. Three different axes were evaluated: indicators of the child's health, the family's perception of care and the family's knowledge about the disease. The indicators of child health were measured using an objective interview and analysis of the patient's health records. The family's perception about healthcare was assessed using a questionnaire with Likert scale responses. The family's knowledge was assessed using objective questions with true/false answers.

The Criteria of the Economic Classification in Brazil[10] was used to define the economic status of participating families. Householders were classified according to occupation in a simplified form as top-level professionals, skilled manual workers (technicians), semi-skilled manual workers and unqualified workers.[11]

The data collection instrument was validated by five professionals (two specialists, two pediatricians and a nurse with experience in primary healthcare) in respect to its comprehensiveness, objectivity and relevance.

The data were consolidated using the Statistical Package for Social Sciences (SPSS) software version 17.0 which allowed descriptive analysis.

Ethics guidelines were respected. All participants signed a written consent form before the interview and the study was approved by the Research Ethics Committee of the Universidade Estadual de Montes Claros - Unimontes (No 1517/2009).

Results

Seventy-one sickle cell disease children were identified through the PETN-MG registers for the studied microregion. Four (5.6%) families that lived in rural areas were excluded. Of the 67 children eligible for the study, 63 children (94.0%) were located and their parents or guardians consented to participate in the study. Four families were not located on three home visits at different times and on different days.

According to the Criteria of Economic Classification for Brazil, 45 families (71.4%) belonged to classes D and E. The householder was the father in 34 families (54.0%) and most worked as skilled or semi-skilled manual workers (55.6%). Twenty-two (35.0%) of the mothers had not completed high school. Thirty-three (52.4%) of the children were female. The Hb SS genotype (28; 44.4%) was predominant followed by the Hb SC genotype (26; 41.2%). Other sociodemographic data are presented in Table 1.

Table 1 - Main sociodemographic characteristics of children with sickle cell disease in the Montes Claros-Bocaiuva microregion

Characteristics of children	n	%
Economic Classification		
B2	2	3.2
C	16	24.4
D	39	61.9
E	6	9.5
Mother's age (years)		
≤ 25	16	25.4
26-39	43	68.3
≥ 40	4	6.3
Mother's education (years schooling)		
≤ 8	22	35.0
9 - 11	36	57.2
≥ 12	5	8.0
Occupation of father		
Top-level professional	3	4.8
Skilled and semi-skilled manual worke	33	52.4
Unskilled manual worker	21	33.3
Not working+	6	9.5
Number of brothers and sisters		
None	18	28.6
One	24	38.1
Two or more	21	33.3
Gender	30	47.6
Male	33	52.4
Female		
Age (years)		
≤ 2	11	17.5
3-5	14	22.2
6-10	29	46.0
11-12	9	14.3
Ethnic background		
White	11	17.5
Mulatto	48	76.2
Black	3	4.8
Asian	1	1.6
Genotype		
SS	28	44.4
SC	26	41.2
SD	2	3.2
Sß-Thalassemia*	7	11.2

+ Out of the economically active population
*Unable to distinguish Sß0 -Thalassemia or Sß+-Thalassemia from the information provided by the family

Table 2 - Perception of families of the care provided by healthcare services in the Montes Claros-Bocaiuva microregion

Questions evaluated	Frequency				
	Never n (%)	Rarely n (%)	Sometimes n (%)	Frequently n (%)	Always n (%)
Do the professionals usually advise on nutrition early in life?	26 (41.3)	5 (7.9)	15 (23.8)	6 (9.5)	11 (17.5)
Did the family receive guidance on the need to keep the child hydrated?	9 (14.3)	5 (7.9)	8 (12.7)	14 (22.2)	27 (42.9)
Did the family receive guidance on the need to avoid staying with the child in very hot or very cold environments?	17 (27.0)	4 (6.3)	11 (17.5)	10 (15.9)	21 (33.3)
In the presence of any of the warning signs, is the attendance at the health service prioritized?	19 (30.2)	12 (19.0)	9 (14.3)	5 (7.9)	18 (28.6)
In the health service, did professionals encourage families to make an appointment at the blood bank?	13 (20.6)	6 (9.5)	11 (17.5)	6 (9.5)	27 (42.9)
Did a family member participate in educational support groups related to the disease or child care in the health service?	60 (95.2)	2 (3.2)	1 (1.6)	-	-
When you received any guidance from professionals at the health service, did you have the opportunity to ask questions?	35 (55.6)	2 (3.2)	5 (7.9)	9 (14.3)	12 (19.0)
Are there difficulties in making appointments in the health service when necessary?	17 (27.0)	5 (7.9)	14 (22.2)	4 (6.3)	23 (36.5)

The regular use of penicillin for children with sickle cell disease until the age of five years old was reported by 55 caregivers (87.3%) and the regular use of folic acid was reported by 62 caregivers (98.4%). Within the first year of life, 23 children (36.6%) had 12 or more consultations to monitor their growth and development the results of which were registered on the child's record card.

From the second to sixth years of life, only 18 children (28.6%) had had at least the minimum number of recommended follow up consultations [14] to monitor development registered in their record cards.

Forty-four (69.8%) children were up to date in their routine vaccinations as registered on their record cards. Figure 1 shows the special vaccines recommended for sickle cell disease children; 62 children (98.4%) were behind in their vaccination schedule according to their record cards. The vaccine which was least given was the 7-valent pneumococcal vaccine (8.2%).

The families' perception about the actions of family healthcare teams is presented in Table 2. Twenty-six (41.3%) families reported never having been counseled in respect to the child's diet as a young baby. Sixty interviewees (95.2%) reported that they never participated in educational groups related to sickle cell disease and child care. Doubts were not explained by healthcare teams according to 35 respondents (55.6%). About half of parents reported that treatment was never or rarely prioritized even with the appearance of warning signs (31; 49.2%). In the opinion of 23 respondents (36.5%), there are always difficulties in making appointments in primary healthcare services.

Table 3 shows the actions developed by the family healthcare teams for sickle cell disease children. Among the children studied, 54 (85.7%) were recognized by healthcare teams as children with sickle cell disease. Genetic counseling and guidance on the necessity of spleen palpation were recorded for 56 (88.9%) and 45 (71.4%) children, respectively.

Table 3 - Actions taken by healthcare teams in relation to the care of sickle cell disease children in the Montes Claros-Bocaiuva microregion

Variable	N	%
Referral to the dentist every 6 months		
Yes	19	31.7
No	41	68.3
Total*	60	100.0
Regular referral to an ophthalmologist		
Yes	8	14.0
No	49	86.0
Total*	57	100.0
Do the professionals at the health service know that the child has sickle cell disease?		
Yes	54	85.7
No	9	14.3
Were parents taught how to palpate the spleen?		
Yes	45	71.4
No	18	28.6
Genetic counseling for parents		
Yes	56	88.9
No	7	11.1

(*)When collecting data, children who were too young to receive the vaccine were excluded from the statistics

Forty-two families (66.7%) used the blood center identification card regularly. According to parents, hospitalizations were common in childhood and involved 35 (55.6%) children. The most frequently reported reasons for hospitalization were pneumonia, other infections, painful crises and acute splenic sequestration.

Knowledge of families was tested by asking 20 questions. The mean number of correct answers was 11.95 (59.8%; SD = 2.34). The questions that were least answered correctly were related to genetic aspects of the disease and warning signs.

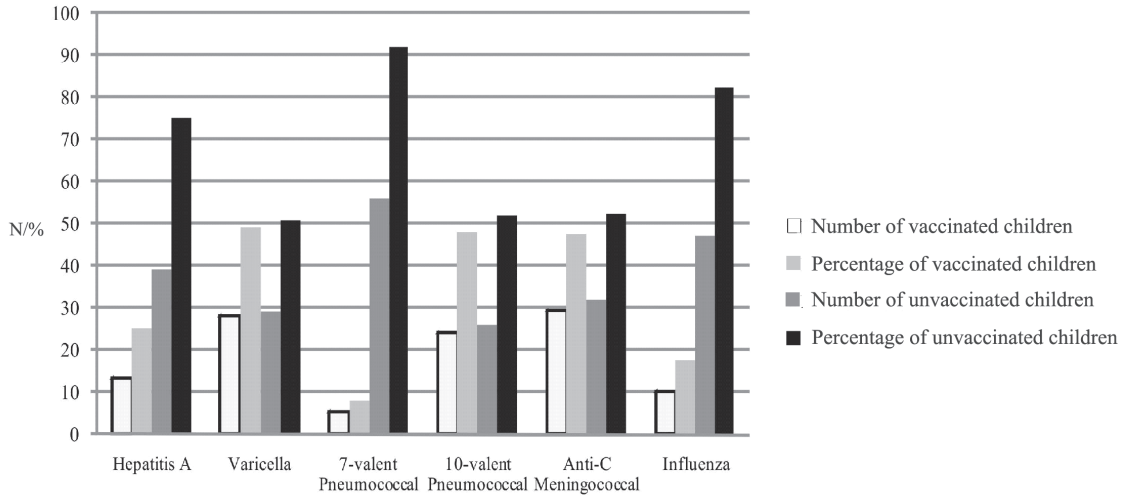

Figure 1 – Use of special vaccines for sickle cell disease children in the Montes Claros-Bocaiuva microregion

Discussion

In this study the quality of care provided to sickle cell disease children in primary healthcare services was assessed in a region of high prevalence. The results show a worrying situation, especially because the issues addressed were based on the guidelines of official institutions.[7,8]

The profile of families is similar to that described in the literature with a predominantly low income population,[12] that is, mainly socioeconomic classes D and E. Most mothers had little schooling and the fathers' occupations generally involved specialized or semi-specialized manual activities. These features denote a needy population, for which the support of health services is essential for disease management. Most children were diagnosed early by neonatal screening, which allows teams to start management early, with guidance on necessary prophylaxis with vaccines and antibiotic therapy.[13]

The actions of the services were below the desirable and necessary level. Only folic acid supplementation and prophylaxis with penicillin were prescribed to high percentages of children. The prescription of folic acid is important because the child's needs are elevated in this period of constant growth.[14] Its supplementation is important for erythropoiesis in the bone marrow, specifically for the maturation of erythroblasts in erythrocytes.[15] According to official guidelines, the prescription of folic acid and prophylaxis with penicillin is a responsibility of blood banks however primary healthcare teams should check that these drugs are prescribed and encourage adherence to treatment.

Adherence has been a problem in monitoring patients with sickle cell disease as highlighted in the literature by other authors.[16,17] The reasons involved can include family beliefs, religious beliefs, parents' concern about the consequences of long-term antibiotic therapy and others reasons. Verification in this study was limited to family reporting. Studies assessing adherence by more specific strategies show failure in continuous administration and low adherence.[18]

The proper growth of the child is a summary measure of healthcare. Nutritional disorders may be present in sickle cell disease children, specially related to the severity of the disease caused by the genotype; it is important to carefully accompany these children.[19,20] Monitoring growth and development by health services should include monthly meetings for under one-year-old children and every three months thereafter until the age of five.[7] It was found that most children do not receive the recommended number of consultations. There is thus a lack of commitment to monitor these children or at least a lack of recording the results on the child's record card.

The basic vaccination coverage was within the levels reported by other authors, between 65 to 100%.[21] This did not occur with special vaccines as almost all vaccination cards showed that the patients were behind with their immunizations. Similar findings were found in the state of Espirito Santo, also in Brazil, where it was found that 50% of sickle cell disease children had incomplete immunization against pneumococcal.[22] A study conducted in London found that vaccinations against encapsulated bacteria and the influenza virus is bad among adults and children with sickle cell disease.[23] However, even developing countries can have a satisfactory coverage as was shown by a study conducted in northern of Africa.[24] It was impossible to define all the reasons for the delayed vaccination schedules. The authors believe that the bureaucratic processes involved in providing special vaccines and the lack of knowledge of professionals about the vaccination calendar for sickle cell disease children are two determinants.

The perception of care by the family was also evaluated in this study. Health education in primary care with promotion of health has been rarely evaluated for families of sickle cell

disease children. It would be desirable that healthcare teams provide necessary knowledge about the disease, enabling decision-making and promoting self-care, demystifying the disease and allowing the development of citizenship.[14]

The rare family participation in educational groups related to the disease and child care reflect the lack of health education through support groups. It is necessary that professionals improve the care provided for the most common genetic disease, which is a public health problem in Brazil and worldwide. It is believed that due to the small number of patients in coverage areas, educational activities can be carried out in consultations with health professionals. However, it is desirable that teams articulate and promote the integration of patients so that they can share positive and negative experiences and learn more about the disease with each other. The establishment of educational groups is very important to guide and educate caregivers and reduce complications with the institution of preventive measures.

Some aspects, such as the necessity to keep the child hydrated and to avoid too hot and too cold environments for most children were observed by primary healthcare professionals. These guidelines are important and prevent vaso-occlusive crises, but do not seem to be affordable for all families. Additionally, according to the families, few were counseled on nutritional needs of very young children with sickle cell disease, a responsibility of primary healthcare services.

Forty-five parents and guardians (71.4%) were instructed about the technique of spleen palpation in order to identify early acute splenic sequestration. Teaching spleen palpation to parents and caregivers can help to reduce mortality as it is an effective way of preventing splenic sequestration crises. Another issue is the teaching of warning signs to parents so that they can demand immediate healthcare attention at the level of complexity required. It is essential that families receive such guidance because this can significantly reduce the mortality of these children.[25]

Genetic counseling was provided to 56 families (88.9%) when genetic implications on future pregnancies were discussed.[26] It was impossible to determine if information relating to guidance on the spleen palpation technique and guidance regarding the genetic counseling were provided by the blood center or by primary healthcare services.

Regarding access to primary healthcare services, a non-negligible percentage of families (36.5%) reported that it is always difficult to schedule appointments. Another worrying aspect is that, in the presence of warning signs, attendance of the children was always given priority in only 28.6% of the cases. Accessibility for children with sickle cell disease in primary care services needs improvement, according to the perception of families.

Most caregivers reported that they never had an opportunity to clarify their doubts with the professionals in primary healthcare services. To clarify questions, it is necessary to build a relationship of trust between healthcare professionals, patients and family.[27] It was observed that primary healthcare services are not fulfilling one of their principles: longitudinality.

Pneumonia was the most common reason for hospitalization. Pneumonia caused by pneumococcus and Haemophilus influenzae are common in these patients.[28] It must be remembered that there may be a possible correlation between the number of hospitalizations for pneumonia and the low rate of vaccination against pneumococcus; this should be investigated by primary healthcare services.

Painful crises and other infections were the second most common cause of hospitalization; this differs from other studies in which the majority of admissions were due to episodes of pain.[12,29] Being a frequent complication, family members should be taught to recognize the origin and intensity of pain in order to take appropriate measures.[25] Another important cause of hospitalization in this study was acute splenic sequestration. This is usually listed as the second leading cause of death among sickle cell disease children and also a common clinical complication in the evolution of children.[6,25,29]

Families showed poor performance in a test about their knowledge on the disease. This may reflect the poor performance of primary healthcare services to educate families. Ideally, after diagnosis, families should be included in healthcare and health education programs to promote better disease management and improve the normality of the family, despite of the effect of the disease on the familiar unit.[28]

The results of this study should be interpreted considering its limitations. This is a study in a restricted region of the state of Minas Gerais and, although it is the second largest center for medical care of sickle cell disease patients in the state, it has special features, which differ from the rest of the country. One aspect to note is that there is no specific instrument to assess the quality of care provided to sickle cell disease patients. The family's perception in relation to healthcare also may just reflect an unsatisfactory relationship with professionals. Another issue is related to the study being based on a surviving population. It is well-known that these children have a high mortality rate. The effort spent gathering information on three different axes give a general idea of the care provided to sickle cell disease children. Unfortunately, this is a critical care that needs improvement in many respects, in particular accessibility, immunization, monitoring of growth and development, and health education. As there are few national studies on the subject it is desirable that further research should be undertaken to address the quality of care provided to sickle cell disease children in order to encourage discussion of this theme in respect to the knowledge of professionals and to promote improvements in care.

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