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Medical doctors and internal medicine students: are they prepared to care for sickle cell disease patients?

Docentes médicos e internos do curso de medicina estão preparados para uma boa prática médica no atendimento aos portadores de doença falciforme?

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ABSTRACT: Sickle cell disease is a denomination given to the hemoglobinopathic group. It is estimated that there are 25,000 to 30,000 patients with sickle cell disease in Brazil and 3,500 new cases occur each year. In the state of São Paulo, there is an incidence of 1:4000 live births. There are no data on sickle cell disease mortality/lethality in Brazil. It is important to emphasize that health professionals are trained so that mortality rates continue to decrease. Therefore, this work aims to analyze the training of internal medicine students, as well as their teachers in relation to sickle cell disease. *Materials and methods:* A quantitative cross-sectional study was carried out with 5th and 6th semester students and medical professors from the *Universidade Cidade de São Paulo*. A questionnaire with 5 questions was applied. *Results and discussion:* The results showed that students and teachers know what sickle cell disease is; with respect to the diagnosis, most of the students had this knowledge and all the teachers would know how to diagnose it; with respect to the clinical manifestations, most of the students knew how to answer the question, and 100% of the teachers answered yes; with respect to the fourth question, both students and teachers were more often aware of the general observed manifestations. Regarding the treatment, it was observed that the students of the 9th and 12th semesters as well as the medical teachers had proper knowledge of the treatment for the acute

manifestations of sickle cell disease, unlike the students of the 10th and 11th semesters, who reported not knowing the treatment of these manifestations. Our studies point to the importance of adequate knowledge of the disease, its diagnosis, its clinical manifestations and treatment in the academic environment to improve the quality of life of patients with this disease, as well as to significantly reduce the mortality rate of patients with sickle cell disease.

Keywords: Anemia, sickle cell; Anemia, sickle cell/epidemiology; Health personnel; Health knowledge, attitudes, practice; Education, medical/manager.

RESUMO: Doença falciforme é uma denominação dada ao conjunto de hemoglobinopatias. Estima-se que existam no Brasil de 25 mil a 30 mil portadores de doença falciforme e que surjam, anualmente, 3.500 novos casos. No estado de São Paulo, temos uma incidência de 1:4000 nascidos vivos. Ainda não há dados consistentes sobre a mortalidade e a letalidade da doença falciforme no Brasil. É importante ressaltar que os profissionais de saúde sejam devidamente treinados para que as taxas de mortalidade continuem a decrescer. Para tanto, este trabalho tem como objetivo analisar a capacitação de estudantes

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internos de medicina, bem como seus docentes em relação a doença falciforme. *Materiais e métodos:* Foi realizado um estudo transversal quantitativo, com alunos do 5º e 6º ano e docentes médicos da Universidade Cidade de São Paulo. Foi aplicado um questionário com 5 perguntas. *Resultados e discussão:* Os resultados mostraram que os alunos e os docentes sabiam o que era doença falciforme; com relação ao diagnóstico a maioria dos estudantes tinha este conhecimento e todos os docentes saberiam diagnosticá-la; com relação as manifestações clínicas, a maioria dos alunos souberam responder à pergunta e 100% dos docentes responderam que sim; com relação a quarta pergunta tanto os alunos quanto os docentes conheciam com maior frequência as manifestações gerais observadas. E com relação ao tratamento, foi observado que os alunos das etapas 9ª e 12ª, bem como os

docentes médicos tinham o devido conhecimento do tratamento das manifestações agudas da doença falciforme, ao contrário dos alunos da 10ª e 11ª etapas, que relataram desconhecer o tratamento destas manifestações. Nossos estudos apontam para a importância do adequado conhecimento da doença, seu diagnóstico, suas manifestações clínicas e tratamento no meio acadêmico para melhorar a qualidade de vida dos pacientes com esta doença, bem como diminuir significativamente a taxa de mortalidade de pacientes portadores de doença falciforme.

Descritores: Anemia falciforme; Anemia falciforme/epidemiologia; Pessoal de saúde; Conhecimentos, atitudes e prática em saúde; Educação médica/recursos humanos.

INTRODUCTION

Sickle cell anemia (SCA) is a genetic disease due to a structural defect in the beta globin chain, causing a change in the hemoglobin molecule and the shape of red blood cells into a sickle shape in the absence of oxygen. It is represented by the homozygous state for hemoglobin S (SS), corresponding to the most severe form of Sickle Syndromes. The term “sickle cell anemia” is reserved for the type of disease that occurs in these SS homozygotes. In addition, the HbS gene can combine with other inherited hemoglobin abnormalities such as hemoglobin C (HbC), hemoglobin D (HbD) and beta-thalassemia, generating combinations that are also symptomatic, which represent the group known as sickle cell disease (SCD)^{1,2,3,4}.

Sickle cell disease is one of the most common genetic and hereditary diseases in Brazil¹. Data provided by the Ministry of Health, referred to by Cançado and Jesus⁵, estimate that there are 25,000 to 30,000 patients with sickle cell disease in Brazil, and that 3,500 new cases emerge annually. In the state of São Paulo, incidence is in every 1:4000 live births⁶.

It is estimated that there are at least two million HbS (heterozygous) carriers in the world⁷. In Brazil, the disease is predominant among black and brown individuals, but also occurring in those with white complexions. In southeastern Brazil, the average prevalence of heterozygotes (carriers) is 2%, a number that rises to about 6-10% among black persons. This percentile rises about 0.1% to 0.3% in this population⁵. Regarding the mortality rate in Brazil, there is still no consistent data on mortality and lethality⁶.

However, it is known that the mortality rates of sickle cell anemia patients have been decreasing significantly over the last decades. This fact is due to the implantation of programs for the early diagnosis and consequent institution of prophylactic measures starting in the neonatal period⁷.

In order for mortality levels to continue to decline, health professionals need greater knowledge about the disease to be able to recognize an affected patient, since they may present different clinical manifestations in

different systems. It is important to emphasize that health professionals should be properly trained so that mortality rates continue to decrease. Thus, this study aims to analyze the training of interning medicine students, as well as their professors in relation to sickle cell disease.

MATERIAL AND METHODS

The present quantitative cross-sectional study was carried out with a questionnaire applied to intern medicine students, in which 38, 49, 47 and 38 of the students were undergoing their 9th, 10th, 11th and 12th semesters of the course, respectively. It should be noted that the 9th and 10th semesters (in Brazil) correspond to the 5th year of the traditional medical course, and the 11th and 12th periods correspond to the 6th year of the course. Medicine professors (22 individuals), regardless of their specialties, participated in the research. The questionnaire was applied within the University of São Paulo (PBL system - Practice based learning) only to students and teachers who attend this university. The applied questionnaire was based on a previously validated questionnaire (Research Ethics Committee of NOVAFAP, CAAE No. 0379.0.043.000-11) applied by Laise Maria Formiga Moura Barroso¹⁰, which has been modified and approved.

Data were collected after Informed Consent was given by participants and only after the approval by the Research Ethics Committee under CAAE number: 56600616.7.0000.0064. Participants were invited by an informal invitation from the researchers, exposing the study objectives and clarifying possible doubts.

The data collected was then separated into graphs according to the semester the student was in. The responses regarding the fourth question “If so, what are they? Name them” were separated by large systems (of the body), considering that the answers given by the students could be related to different systems. Thus, responses such as anemia and hepatosplenomegaly were directed to the **lymphohematopoietic system**. Responses such as bone pain, arthritis, myalgia, limb pain, myasthenia and hand and foot syndrome were included in the **osteoarticular-muscular system**. The **cardiopulmonary system** included

responses such as thrombosis, vaso-occlusive crises, dyspnea, acute chest syndrome, chest pain, respiratory failure, pulmonary thromboembolism. The **urogenital system** included responses such as priapism, renal failure and delayed growth (due to hormonal changes that occur in the reproductive system). Responses such as pallor, cyanosis, leg ulcers and jaundice were included in “cutaneous-mucosa”. The **gastrointestinal system** included only abdominal pain. The **central nervous system** involved ischemic cerebrovascular accident, hemorrhagic encephalic stroke and headaches. Finally, **overall manifestations** included asthenia, painful crises, recurrent infections, fatigue, malaise, dizziness and fever.

Therefore, it is important to emphasize that the students’ responses in relation to exactly which clinical manifestations observed in patients with sickle cell disease (question 4) were grouped according to the systems (of the body) described above. Thus, the total number of students participating in the study does not correspond to the total answers presented.

The following questions seek to evaluate your knowledge about sickle cell disease. Answer what you know.

Question 1. Do you know what sickle cell disease is? 1. Yes 2. No

Question 2. Do you know how to diagnose sickle cell

disease? 1. Yes 2. No

Question 3. Do you know the main clinical manifestations of sickle cell disease patients? 1. Yes 2. No

Question 4. If so, what are they? List them.

Question 5. Do you know how to treat the acute manifestations of the disease? 1. Yes 2. No

RESULTS

Among all students interviewed from the 9th, 10th, 11th and 12th semesters, all responded “yes” to the first question “Do you know what sickle cell disease is?” (data not shown). The same 100% “yes” responses for this question was also observed among the medicine professors (data not shown).

Regarding the second question “**Do you know how to diagnose sickle cell disease?**”, it was observed that the great majority of students knew how to answer the question, regardless of their course semester.

It is interesting to point out that the students of the 10th semester (Figure 1b) were those who most reported being aware of the diagnosis of sickle cell disease (14%). Still in relation to sickle cell disease, all the medicine professors (100%) reported having knowledge on how to diagnose the disease (data not shown).

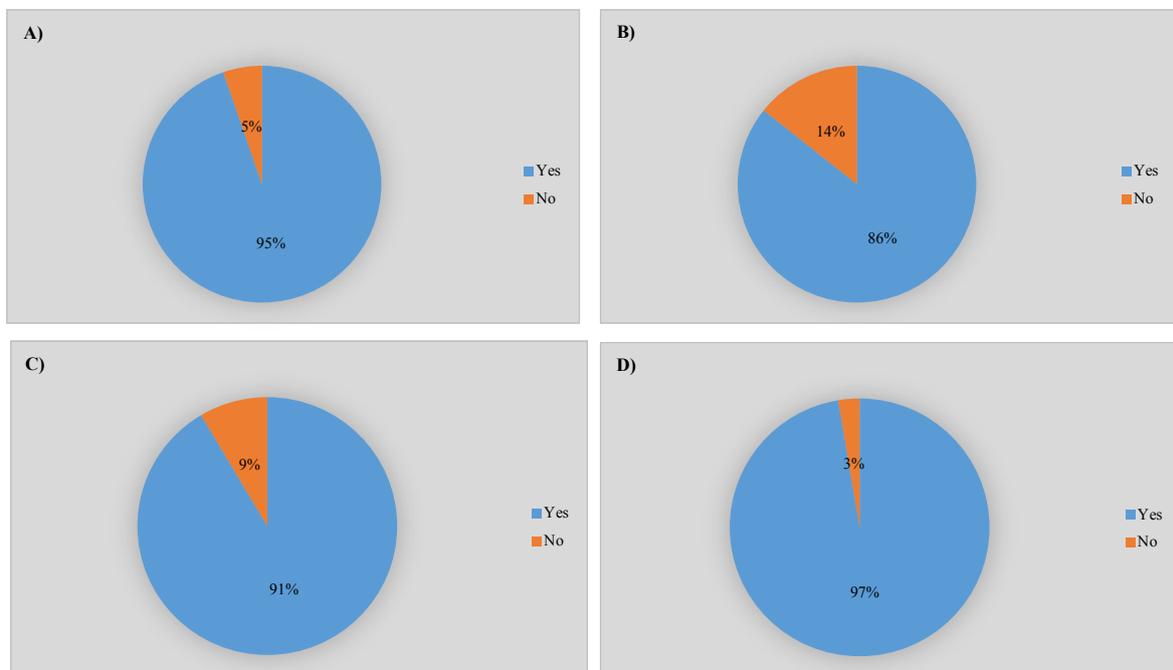


Figure 1. Students from the 9th (A), 10th (B), 11th (C) and 12th semesters (D) who can diagnose sickle cell disease

From the questionnaire it was possible to observe that most of the students knew how to answer the question related to the knowledge of the clinical manifestations of patients with sickle cell disease.

Students from the 11th semester (Figure 2C) were those who most reported being aware of sickle cell disease clinical manifestations (13%).

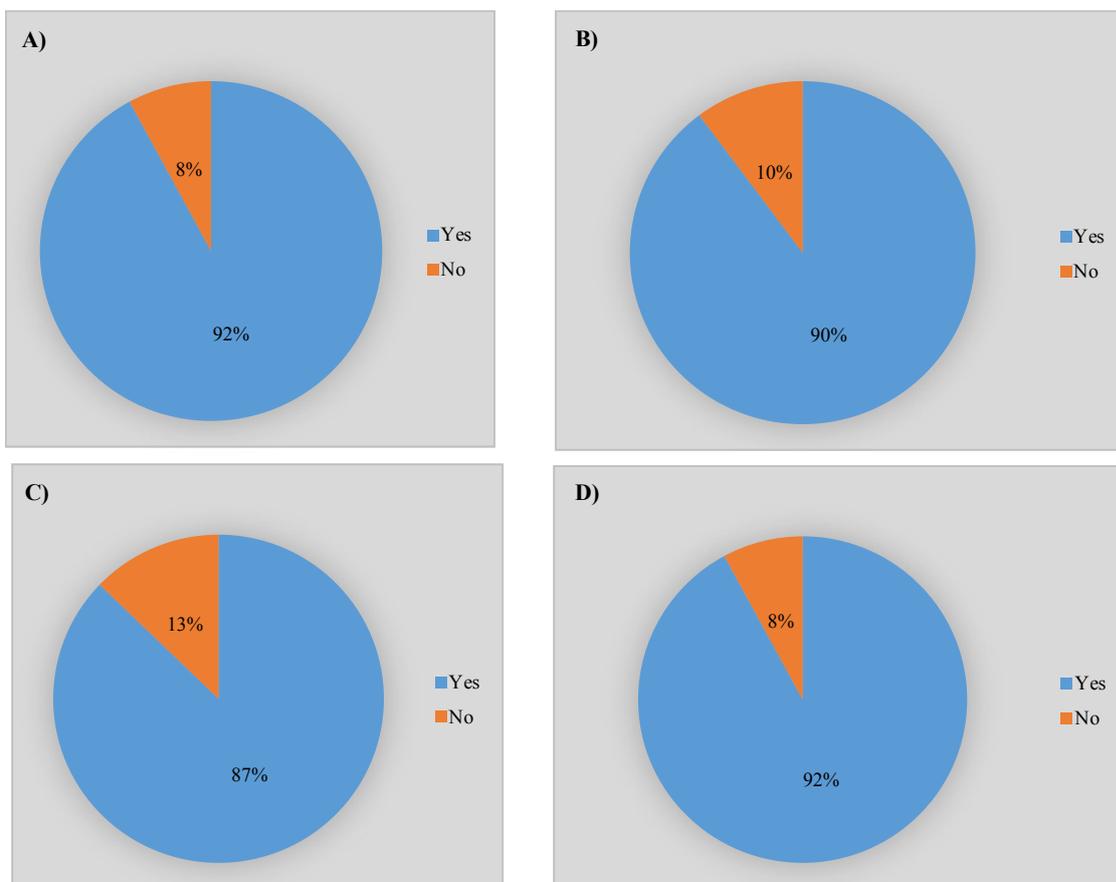


Figure 2. Students from the 9th (A), 10th (B), 11th (C) and 12th semesters (D) who are aware of sickle cell disease clinical manifestations

Similar to the previous question, all the medicine professors (100%) answered “yes” to this question (data not shown).

We also questioned whether students from the 9th, 10th, 11th and 12th periods were aware of what are the exact clinical manifestations of the disease. As previously

described in “Material and Methods” section, the student’s responses related to the clinical manifestations observed in patients with sickle cell disease (question 4) were grouped according to the body systems. Thus, the total number of presented responses does not correspond to the number of students participating in the present study.

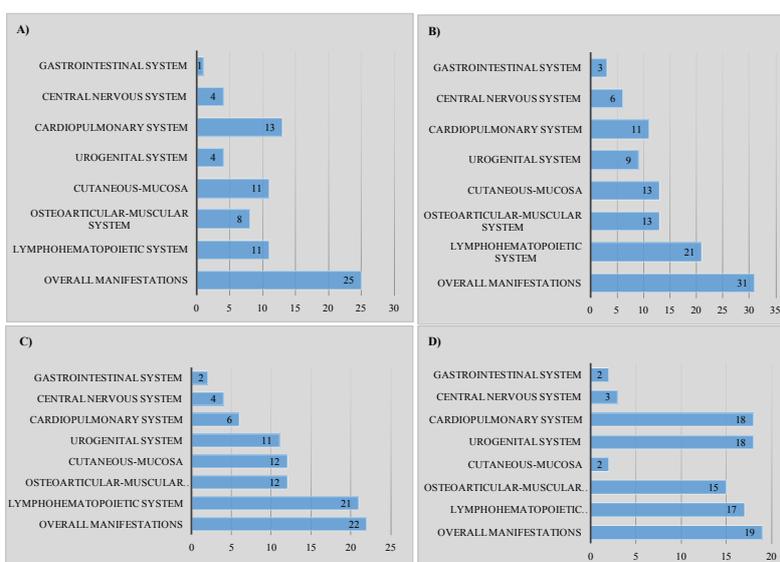


Figure 3. Students from the 9th (A), 10th (B), 11th (C) and 12th semesters (D) who are aware of the clinical manifestations related to sickle cell disease

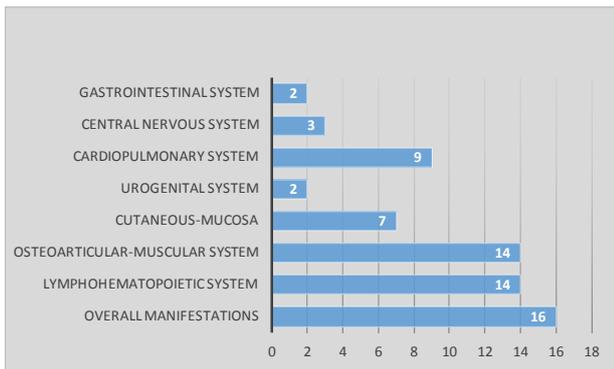


Figure 4. Medicine professors who are aware of the clinical manifestations related to sickle cell disease

The results show that both students and professors were aware of the most frequently observed general manifestations (asthenia, painful crises, recurrent infections, fatigue, malaise, dizziness and fever - Figures 3A, B, C, D and Figure 4).

It is worth pointing out that manifestations related to the gastrointestinal system (abdominal pain) were the least reported (Figures 3A, B, C, D and Figure 4).

With regard to the fifth question “**Do you know how to treat the acute manifestations of the disease?**”, it was observed that the students from the 9th and 12th semesters (Figures 5 A and D), as well as the medicine professors (Figure 6) had proper knowledge related to treating acute manifestations of sickle cell disease; this differed for students from the 10th and 11th semesters (Figures 5 B and C), who reported not being aware of how to treat these manifestations.

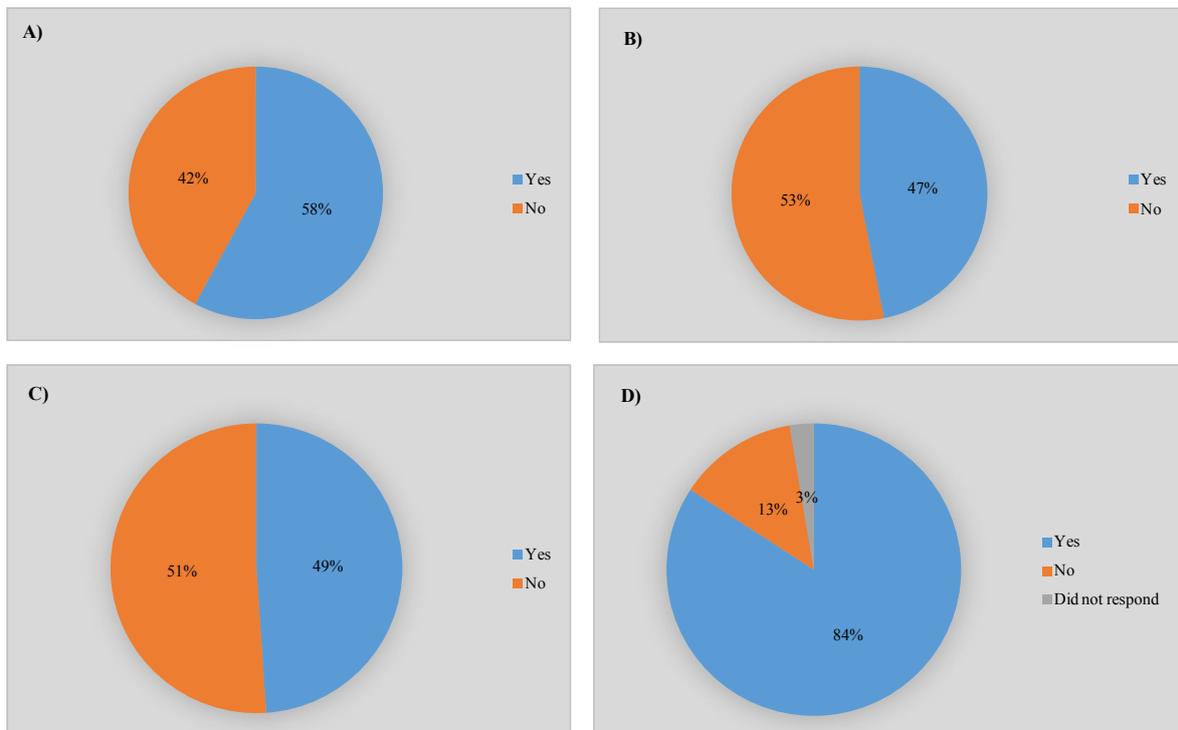


Figure 5. Students from the 9th (A), 10th (B), 11th (C) and 12th semesters (D) who are aware of how to treat acute manifestations of sickle cell disease

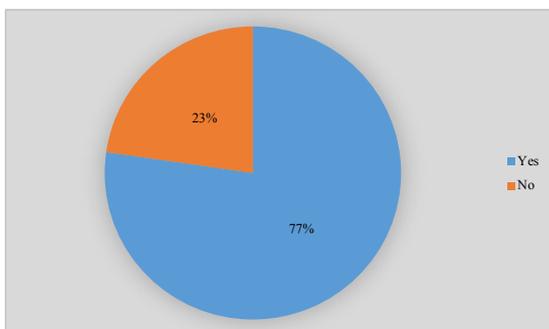


Figure 6. Medicine professors who are aware of how to treat acute manifestations of sickle cell disease

DISCUSSION

Sickle cell anemia is a genetic, hereditary and multisystemic disease characterized by the presence of abnormal erythrocytes due to the presence of HbS. When deoxygenated, the HbS polymerizes, causing loss of cations and water, thus damaging erythrocytes. These damaged cells present physicochemical abnormalities, resulting in hemolytic anemia and vaso-occlusion. Based on this fact, a number of complications can arise: vascular endothelial dysfunction, functional nitric oxide deficiency, inflammation, oxidative stress and reperfusion injury,

hypercoagulability, increased neutrophil adhesion and platelet activation^{3,4,8}.

Although the characterization of sickle cell disease is complex, the present study shows that the vast majority of students and professors are aware of what sickle cell disease is and are able to diagnose it.

The neonatal screening test consists of collecting blood drops from a child's heel (foot test). It is required that this collection happens within 48 hours of the newborn's first feeding, and up to the fifth day of the child's life at the closest health unit. The child should also receive the first vaccines and the initial care needed in the Primary Care Network. After 4 months of age, the diagnosis can be performed by any of the existing methodologies for hemoglobin electrophoresis⁶.

When detection of sickle cell disease is performed in the neonatal screening tests, the diagnosis enables the start of specific required care, considering that precocity and comprehensive care can be determinant for limiting the disease symptoms. Among the early care, primary health care has the responsibility of checking on children for their development and their family: breastfeeding, introduction to food/weaning, hygiene, vaccines, and oral health, as well as other specific indications for the disease. The family should be properly prepared for self-care - and care in this sense needs to be multiprofessional and humanized. It is understood as humanization, in addition to having a comfortable environment for the care of people, taking into consideration their welcoming by health workers must occur in an effective manner, with solidarity and without any prejudice or stigma^{6,9}.

Identification and diagnosis of the disease are fundamental for eliminating the mortality rate of sickle cell patients, since they are susceptible to several serious diseases. As explained in the Booklet "Basic Guidelines for Care in Sickle Cell Disease - *Doença Falciforme Diretrizes Básicas da Linha De Cuidado*"^{7,6}, children with sickle cell disease up to 5 years of age without health care reach 80% mortality (average 8 years of life), in comparison to those with health care with a 1.8% mortality rate (average 45 years of life).

Clinical manifestations appear in patients with sickle cell disease when identification and diagnosis by the health professional are not adequate.

The manifestations are diverse and can affect different systems. In the gastrointestinal and abdominal system they include: painful crises, gallstones, obstructive jaundice, liver disease; in the central nervous system they include: transient ischemic attack, infarction, cerebral hemorrhage; in the cardiopulmonary system: cardiomegaly, heart failure, pulmonary infarction, pneumonia; in the urogenital system: priapism, hypostenuria, chronic renal failure; on the cutaneous-mucosal surface they include:

pallor, jaundice and ulcers; in the osteoarticular system: pain, aseptic necrosis of the femoral head, gonopathy, proliferative retinopathy, glaucoma; in the hematopoietic system: anemia, asplenia, chronic splenomegaly; and the overall manifestations can include: delay of sexual maturation and greater susceptibility to infections^{2,3,4,10}. Acute thoracic syndrome is a particularly dangerous type of vaso-occlusive crisis involving the lungs which typically present fever, cough, chest pain, and pulmonary infiltrates. Pulmonary inflammation (such as those which can be induced by simple infection) cause blood flow to become slower and "spleen-like", promoting sickling and vaso-occlusion. This compromises pulmonary function, creating a potentially fatal circle of worsening of pulmonary and systemic hypoxemia, sickling and vaso-occlusion⁸.

The present study found that Internal Medicine Students, as well as their professors know how to recognize the clinical manifestations that occur when sickle cell disease is not well diagnosed. However, only students from the 9th and the 12th semesters and the professors were aware of how to treat these manifestations.

Contrary to what was shown in our study, data in the literature show a lack of knowledge of health professionals in recognizing clinical manifestations of sickle cell disease patients. In Minas Gerais¹¹ in a study carried out in hemocentres of the Hemominas Foundation, the probability of death at 5 years (10.6%) in children with sickle cell disease was much higher than the overall mortality rate, even though there is a neonatal screening program with strict treatment control. Many deaths were justified by the fact that health professionals have difficulties in recognizing some serious symptoms/conditions of these children affected by the disease. In the Northeast¹², a study applied in Primary Care with the aim of evaluating health professionals showed that these professionals need better technical preparation/training to deal with patients affected by sickle cell disease. It was observed that more than half of the professionals had their knowledge on sickle cell disease classified as inadequate or irregular. Moreover, Gomes et al.¹³ reported that (only) 50% of health professionals can recognize the clinical manifestations and management of sickle cell disease.

Based on all the above, health professionals who are on call on the day a sickle-cell-disease patient seeks the health service with a pain crisis should be prepared to provide care and help the patient¹⁴.

In conclusion, these data reinforce the importance of adequate knowledge about the disease, its diagnosis, its clinical manifestations and treatment in the academic environment in order to improve the quality of life of patients with this disease, as well as to significantly decrease the mortality rate of sickle cell disease patients.

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