Dynamics of sickle cell disease as one of the determinants of quality of life

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Sickle cell disease (SCD) is a genetic disorder belonging to a group characterized by the predominance of abnormal hemoglobin S (Hb S). Under conditions of deoxygenation, Hb S polymerizes causing vaso-occlusion and tissue injury. Clinical manifestations occur in the course of the disease and infections can also affect patients throughout their lives[1,2]. Even though the incidence varies, sickle cell disease is present all over the world[3]. In regions where the S gene predominates, such as in sub-Saharan Africa, the birth rate of homozygotes is high. In contrast, the migration phenomenon is responsible for the emergence of the disease in non-endemic regions[4]. Ireland saw an increase in the disease affecting children because of a flow of refugees from Nigeria, Angola and the Congo[5].

In 1993, Cyprus hosted the joint meeting of the World Health Organization and the International Federation of Thalassemia. Recommendations were made on the need for exact and regular updated information and possible strategies to control the situational of thalassemia and sickle cell disease worldwide[4]. Among the countries that attended the meeting, Greece´s response was the creation of a national record of all patients with thalassemia and sickle cell disease, newborns affected and hemoglobinopathy-related deaths[6].

Knowing the dynamics of sickle cell disease and its prevalence among communities poses a major challenge to the effective and equitable management of the disease in view of the scarcity of epidemiological data[7]. With this knowledge it is possible to contribute to the planning of more effective therapeutic regimens[6].

In Brazil, the prevalence of heterozygotes for Hb S is higher in the north and northeast of the country (6% to 10%), while the rates in the south and southeast are 2% and 3%, respectively[8]. In the Midwest, especially in Mato Grosso do Sul, in the period 2000 - 2005, after the introduction of the State Neonatal Screening Program, there has been no record of mortality in children screened for sickle cell disease. In 2005 the coverage was 90.78%[9]. In Bahia, an evaluation of the neonatal screening program in 2003 showed coverage of 71.52%, with an incidence of 1:650 newborns (257 patients) with sickle cell anemia[10]. Another study conducted in Bahia of 99 patients throughout the State, with mean age of 30.2 ± 10.8 years and mean time of diagnosis of 12.7 ± 12.1 years, showed that the majority was diagnosed after an acute event[11].

The article "Demographic aspects and quality of life of patients with sickle cell anemia" provides information on 32 patients living in Salvador, Bahia, members of the Associação Baiana de Portadores de Doenças Falciformes (ABADFAL)[12]. Most of the patients are female, single, mulatto or Black, unemployed, and with incomes up to one and a half minimum wages with 40% completing at least high school. The sociodemographic organization of this sample can direct ABADFAL to host and defend the rights of people with sickle cell anemia.

In the U.S., socioeconomic data of 3358 Black sickle cell patients participating in the Cooperative Study of Sickle Cell Disease were compared with the U.S. black population. The data show similarity as to the ability of both groups to reach the same level of schooling. However, a higher unemployment rate was seen for the group with sickle cell disease with working patients often exerting bureaucratic positions. The findings suggest that vocational advisers may have directed these patients to this type of job[13].

In 2007, Belo Horizonte, Minas Gerais hosted the National Forum for Integrated Policy for Caring for People with Sickle Cell Disease that proposed several resolutions related to welfare, education and work and income generation, which would guarantee that this segment of the population would have social security. In welfare the recommendation was that the concept of "disabled person" should be extended to that of "people with special needs" capable of leading an independent life, including at work. In relation to education, one of the proposed resolutions guaranteed continuity of studies to compensate for classes missed due to crises, with due replacement. And with regard to work and income generation, there was a proposal to qualify and professionally train sickle cell disease patients, respecting the specific characteristics of the infirmity, so as to provide better integration in the job market[14].

According to the aforementioned article, the mean age of the 32 patients involved was 31.9 ± 12.67, comprising a phase of life in which the complications are more serious,

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even leading to early death[12]. It is thus necessary to know the patient's quality of life, understood in this study as the ability to perform daily tasks, something beyond clinical manifestations. This assessment is an important aspect in the management of chronic disease[15].

The questionnaire on the quality of life is a useful tool to assess the overall impact of the disease and medical treatment on the patients from their own point of view. Looking at health from this new perspective, attributing due importance to the patient´s own views, implies changes in daily practice[16].

In order to meet this change, new tools have been developed to measure the patient´s well-being before the disease and his treatment. The generic Medical Outcomes Study 36 - Item Short Form Health Survey questionnaire (SF-36), already translated and adapted to the Brazilian culture, examines components related to functional capability, physical aspects, pain, general health, vitality, and social, emotional and mental aspects. Due to its reproducibility and validity it has proved to be a suitable parameter in the assessment of rheumatoid arthritis[17] and other diseases[18].

The article at issue adequately used the SF-36 questionnaire and demonstrated the negative impact of quality of life in the following dimensions: limitation of physical aspects, functional capability, and emotional and social aspects[12]. The chronic nature of sickle cell anemia represents a highly debilitating potential[15].

The SF-36 was used in a study conducted in Saudi Arabia to compare the health-related quality of life (HRQOL) among two groups of adolescents (14 to 18 years old), one with and the other without sickle cell anemia. Adolescents with the disease and the consequent complications reported having worse experiences in HRQOL in the following domains: functional capabilities, physical aspects, bodily pain, and general health regardless of gender. HRQOL scores were negatively associated with increasing age, female gender (emotional domain), residence, low family income, presence of diseases and complications related to frequent hospitalizations[19].

In the absence of a national record of sickle cell disease in Brazil, contributions like the article "Demographic aspects and quality of life of patients with sickle cell anemia" are important and necessary to provide a continuous record of the dynamics of the disease, as well as to expose the quality of life of this population[12].

References

1. Di Nuzzo DV, Fonseca SF. Anemia falciforme e infecções. J Pediatr. 2004;80(5):347-54. Comment in: J Pediatr (Rio J). 2005;81(1):88.
2. Zago MA, Pinto AC. Fisiopatologia das doenças falciformes: da mutação genética à insuficiência de múltiplos órgãos. Rev Bras Hematol Hemoter. 2007;29(3):207-14.
3. Weatherall DJ. The inherited diseases of hemoglobin are an emerging global health burden. Blood. 2010;115(22): 4331-6.
4. Angastiniotis M, Modell B, Englezos P, Boulyjenkov V. Prevention and control of haemoglobinopathies. Bull World Health Organ. 1995;73(3):375–86.
5. McMahon C, Callaghan CO, O'Brien D, Smith OP. The increasing prevalence of childhood sickle-cell disease in Ireland. Ir J Med Sci. 2001;170(3):183-5.
6. Voskaridou E, Ladis V, Kattamis A, Hassapopoulou E, Economou M, Kourakli A, Maragkos K, Kontogianni K, Lafioniatis S, Vrettou E, Koutsouka F, Papadakis A, Mihos A, Eftihiadis E, Farmaki K, Papageorgiou O, Tapaki G, Maili P, Theohari M, Drosou M, Kartasis Z, Aggelaki M, Basileiadi A, Adamopoulos I, Lafiatis I, Galanopoulos A, Xanthopoulidis G, Dimitriadou E, Mprimi A, Stamatopoulou M, Haile ED, Tsironi M, Anastasiadis A, Kalmanti M, Papadopoulou M, Panori E, Dimoxenou P, Tsirka A, Georgakopoulos D, Drandrakis P, Dionisopoulou D, Ntalamaga A, Davros I, Karagiorga M; Greek Haemoglobinopathies Study Group. A national registry of haemoglobinopathies in Greece: deducted demographics, trends in mortality and affected births. Ann Hematol. 2012;91(9):1451-8.
7. World Health Organization. Fifty-Ninth World Health Assembly. Resolutions and decisions annexes. Resolution WHA59.20. Sickle-cell anaemia [Internet]. Geneva: WHO; 22-27 May 2006 [cited 04 Apr 2013]. Available from: http://apps.who.int/gb/ebwha/pdf_files/WHA59-REC1/e/Intro-en.pdf
8. Cançado RD, Jesus JA. A doença falciforme no Brasil. Rev Bras Hematol Hemoter. 2007;29(3):203-6.
9. Holsbach DR, Ivo ML, Honer MR, Rigo L, Botelho CA. Ocorrência de hemoglobina S no estado de Mato Grosso do Sul, Brasil. J Bras Patol Med Lab. 2008;44(4):277-82.
10. Almeida AM, Godinho TM, Teles MS, Rehem AP, Jalil HM, Fukuda TG. et al. Avaliação do programa de triagem neonatal na Bahia no ano de 2003. Rev Bras Saúde Matern Infant. 2006;6(1):85-91.
11. Zanette AM, Gonçalves MS, Bahia RC, Nogueira LV, Arruda SM. Sickle cell anemia: delayed diagnosis in Bahia, Brazil--a largely Afro-descendant population. Ethn Dis. 2011;21(2):243-7. Erratum in: Ethn Dis. 2011;21(3):ii.
12. Santos JP, Gomes Neto M. Sociodemographic aspects and quality of life of patients with sickle cell anemia. Rev Bras Hematol Hemoter. 2013;35(4):242-5.
13. Farber MD, Koshy M, Kinney TR. Cooperative Study of Sickle Cell Disease: Demographic and socioeconomic characteristics of patients and families with Sickle Cell Disease. J Chronic Dis. 1985;38(6):495-505.
14. Brasil. Ministério da Saúde. Relatório final – Resoluções. In: Fórum Nacional de Políticas Integradas de Atenção às Pessoas com Doença Falciforme, 22 junho. Belo Horizonte (MG); 2007 [cited 2012 Nov 21]. Available from: http://www.cehmob.org.br/downloads/resolucoes.pdf
15. Asnani MR, Reid ME, Ali SB, Lipps G, Williams-Green P. Quality of life in patients with sickle cell disease in Jamaica: rural-urban differences. Rural Remote Health. 2008;8(2):890.
16. Berlim MT, Fleck MP."Quality of life": a brand new concept for research and practice in psychiatry. Rev Bras Psiquiatr. 2003;25(4):249-52.
17. Ciconeli RM, Ferraz MB, Santos WS, Meinão I, Quaresma MR. Tradução para a língua portuguesa e validação do questionário genérico de avaliação de qualidade de vida SF-36 (Brasil SF-36). Rev Bras Reumatol. 1999;39(3):143-50.
18. Vilela RQ, Cavalcante JC, Cavalcante BF, Araújo DL, Lôbo MM, Nunes FA. Quality of life of individuals with sickle cell disease followed at referral centers in Alagoas, Brazil. Rev Bras Hematol Hemoter. 2012;34(6):442-6.
19. Amr MA, Amin TT, Al-Omair OA. Health related quality of life among adolescents with sickle cell disease in Saudi Arabia. Pan Afr Med J. 2011;8:10.