Sickle cell disease: the family perspective

Doença falciforme: a perspectiva da família

Flavio A. Naoum

Sickle cell disease (SCD) is a chronic disorder that brings suffering to patients and their families. Despite of the considerable progress in the understanding of the physiopathology of this disease, especially in the genetic and molecular fields, there have been no major clinical improvements or therapeutic benefits for these patients so far. Except for bone marrow transplantation, treatment for SCD is largely palliative and good clinical practice should include efforts that improve the quality of life of patients including family based interventions.

By quality of life one should include housing, schooling, work, transportation, social activities, medical care and family support. Due to its historical background, SCD affects ethnical minorities, a common worldwide problem but particularly in Brazil, as most patients come from low income families, living in poor areas, frequently neglected by government policies that are supposed to improve primary healthcare, education, transportation and housing.[1] All these problems can add to the known biological complications of SCD and affect the quality of life by generating anxiety and a considerable level of uncertainty about the future of both the patient and parents.[2] In this setting, family support is critical for successful treatment in SCD.

Hence, parents usually go through different emotional phases when they are told that their child has SCD and may feel confused, anxious, depressed, or even frightened because they do not know how this condition is going to affect their child.[3] Despite of its importance, this subject is rarely discussed in full and parents may find it very stressful to provide care to an affected child. Moreover, when the family fails to cope with this condition, it is not unusual for SCD patients to experience more physical and emotional problems.[4,5]

In this issue of the RBHH, Guimaraes *et al*.[6] report important insights based on interviews with relatives of 10 Brazilian SCD patients. Among the topics addressed, some results are particularly concerning such as maternal overburden and isolation due to dedication to an affected child, non-acceptance of the disease, the feeling of guilt, fear of death specially during episodes of pain, misinformation about the disease and the perception that, in many aspects, the public health services are not prepared to meet their needs.

This work also emphasizes the need for a specific family based program to provide information and support (professional, emotional and even financial) on a regular basis as well as evaluation of feedback. However, it is sad that even nowadays many families are still misinformed or being treated in an unsympathetic manner by health professionals or stigmatized by the general population. In our view, participation of family members in treatment decisions, patient associations and group meetings should be encouraged and information using appropriate language and means should be made available at any time.

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Coordenador do Instituto Naoum de Hematologia e diretor clínico da Academia de Ciência e Tecnologia de São José do Rio Preto.

Correspondência: *Flávio A. Naoum*
Academia de Ciência e Tecnologia
Rua Bonfá Natale, 1860 – Santos Dumont
15020-130 – São José do Rio Preto-SP – Brasil
Tel.: (+55 17) 3233-4490
Email: a.c.t@terra.com.br