Quality of Life

Samuel Charache

Formerly Johns Hopkins, Baltimore, MA, USA

On reading the paper from Belo Horizonte entitled "Sickle Cell Disease: Quality of Life in patients with hemoglobin SS and SC disorders" published in this edition of the *Revista Brasileira de Hematologia e Hemoterapia*, I had no hesitation in accepting the authors' thesis that the quality of life is diminished in patients with sickle cell disease (SCD) both Hb SS and Hb SC[1]. Two questions did come to mind: are Hb SS and HB SC patients in Minas Gerais different from those in Maryland, and whether or not they are different, what can we (in Brazil and in the United States) do to improve the situation?

The SCD patients at the Fundação HEMOMINAS are different from those at Johns Hopkins Hospital: I think that overall, more of our patients do not work, their incomes are lower, and fewer are heads of households. If the numbers in this study reflect the make-up of the HEMOMINAS clinic population, the foundation sees more Hb SC patients than we do, and those we see are not as sick as those in Minas Gerais (as mentioned in the discussion of this paper). Maybe the frequency of the hemoglobin C gene is higher in Minas Gerais, but maybe the 'climate' in the Belo Horizonte clinic is more welcoming and Hb SC patients whose quality of life is only moderately impaired stay home in Baltimore but attend the clinic in Belo Horizonte. The majority of our patients do not travel an average of 254 km to come to the clinic; they can take the city bus, and the rigors of travel may also bias the Brazilian population toward somewhat sicker Hb SC patients who really have to see a doctor. Whether or not the mix of hemoglobin types is really different, I do not doubt for a moment that the quality of life of SCD patients is diminished in both clinics and elsewhere, also cited in the discussion.

The big question, of course, is 'what can we do to improve the situation'. Treatment of SCD is less than satisfactory. Hydroxyurea therapy, if it is available, affordable, and carefully monitored, can help[2]. Chronic transfusion therapy can help, but the expense, discomfort and inconvenience of transfusions, the development of alloantibodies, and iron overload can make such treatment difficult or impossible[2]. Bone marrow transplantation can be curative, but haploidentical donors can be hard to find, and both costs and risks are significant[3]. Use of related donors who are not haploidentical[4] or use of hematopoietic stem cells[5] are still not widely available, and gene therapy[6] is still further off.

Does that mean we can only wring our hands and say "I'm sorry"? Not quite! The three "C's" of patient care, which should be used for all patients, have been available for a long time. **C**omprehensive, **C**onsistent and **C**ompassionate care may not prevent painful crises, but they can make pain, anemia, decreased financial resources and loneliness more bearable. That kind of care can be very difficult to deliver, but spreading the burden to a specially trained nurse, social worker or lay person in an 'outpatient medical home' near the patient can make the job easier[7,8]. When I visited the Fundação HEMOMINAS some years ago, I think the personnel there were trying to do just that. I know that my hospital and other hospitals and clinics in the United States and elsewhere are trying to do the same. The hope is that early provision of the three "C's" outpatient care to some patients can not only improve the quality of their lives, but may be able to save some of the cost of inpatient admission.

Dr. Santos Pereira et al.[1] have done us all a service by providing us with facts to use when trying to justify the expense of such care.

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Corresponding author:
Samuel Charache
800A Southerly Rd Apt 1224
Baltimore, MD, 21286-8438
Phone: 410 494 6990
scharache@verizon.net

www.rbhh.org or www.scielo.br/rbhh

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