

Sexuality and sickle cell disease

Maria Stella Figueiredo

Universidade Federal de São Paulo –
UNIFESP, São Paulo, SP, Brazil

For decades, sickle cell disease (SCD) was mainly considered a childhood disease, since until 1973 life expectancy was 14 years⁽¹⁾. However, most children with SCD outlive this barrier⁽²⁾. Newborn screening, prophylactic penicillin, and effective vaccinations against *Haemophilus influenzae* type b and *Streptococcus pneumoniae* are some of the factors responsible for this marked decrease in mortality⁽³⁻⁵⁾. Other contributory factors may include chronic transfusion to prevent stroke and the use of hydroxyurea^(6,7).

However, data from the National Association for Sickle Cell Disease in the USA show that 70% of SCD patients are either partially or totally dependent on financial support⁽⁸⁾. Adolescents may be challenged by issues related to body image due to delayed growth and late sexual maturation⁽⁹⁻¹²⁾, and many adults have difficulty sustaining marital relationships^(8,13).

The above motifs could explain, but not justify, the fact that sexuality is not a common subject in SCD. There are few reports about this theme in SCD, many of them associated with priapism events⁽¹⁴⁻¹⁶⁾. All these facts reinforce the importance of the paper from Cõbo et al. published in this issue⁽¹⁷⁾.

Information is a key factor in reported sexual adjustment and satisfaction⁽⁸⁾. Cõbo et al.⁽¹⁷⁾ observed that individuals with SCD presented a limited knowledge of sexual facts, which may adversely contribute to sexual adjustment and to low self-esteem.

These patients need to become independent and responsible adults, with productive and sexually fulfilling lives. There is an urgent necessity of giving adequate and complete information to these individuals and, also, adopting procedures that allow individuals to feel they have some control over their treatment, so that they can improve their self-esteem and achieve a better sexual adjustment. These efforts will help SCD patients to normalize their experiences and increase their chances of having a normal life⁽⁸⁾.

References

1. Diggs LM. Anatomic lesions in sickle cell disease. In: Abramson H, Bertles JF, Wethers DL, editors. Sickle cell disease: diagnosis, management, education, and research. St Louis: C. V. Mosby; 1973. p. 189-229.
2. Quinn CT, Rogers ZR, McCavit TL, Buchanan GR. Improved survival of children and adolescents with sickle cell disease. *Blood*. 2010;115(17):3447-52.
3. Vichinsky E, Hurst D, Earles A, Kleman K, Lubin B. Newborn screening for sickle cell disease: effect on mortality. *Pediatrics*. 1988;81(6):749-55.
4. Gaston MH, Verter JI, Woods G, Pegelow C, Kelleher J, Presbury G, et al. Prophylaxis with oral penicillin in children with sickle cell anemia. A randomized trial. *N Engl J Med*. 1986;314(25):1593-9.
5. Nuorti JP, Whitney CG; Centers for Disease Control and Prevention (CDC). Prevention of pneumococcal disease among infants and children - use of 13-valent pneumococcal conjugate vaccine and 23-valent pneumococcal polysaccharide vaccine - recommendations of the Advisory Committee on Immunization Practices (ACIP). *MMWR Recomm Rep*. 2010;59(RR-11):1-18.
6. Miller ST, Wright E, Abboud M, Berman B, Files B, Scher CD, Sytles L, Adams RJ; STOP Investigators. Impact of chronic transfusion on incidence of pain and acute chest syndrome during the Stroke Prevention Trial (STOP) in sickle-cell anemia. *J Pediatr*. 2001;139(6):785-9. Comment in: *J Pediatr*. 2002;141(5):742-3; author reply 743.
7. Steinberg MH, Barton F, Castro O, Pegelow CH, Ballas SK, Kutlar A, et al. Effect of hydroxyurea on mortality and morbidity in adult sickle cell anemia: risks and benefits up to 9 years of treatment. *JAMA*. 2003;289(13):1645-51. Erratum in: *JAMA*. 2003;290(6):756. Comment in: *JAMA*. 2003;289(13):1692-4; *JAMA*. 2003;290(6):752-3; author reply 754.
8. Chavis WM, Norman GS. Sexuality and sickle cell disease. *J Natl Med Assoc*. 1993;85(2):113-6.
9. Cepeda ML, Allen FH, Cepeda NJ, Yang YM. Physical growth, sexual maturation, body image and sickle cell disease. *J Natl Med Assoc*. 2000;92(1):10-4.
10. M'Pemba-Loufoua AB, Nzingoula S, Moubouh-Akouala F, Oba A. [Pubertal development in girls with homozygote sickle cell disease. Apropos of 72 cases]. *Bull Soc Pathol Exot*. 2001;94(4):326-9. French.
11. Zemel BS, Kawchak DA, Ohene-Frempong K, Schall JI, Stallings VA. Effects of delayed pubertal development, nutritional status, and disease severity on longitudinal patterns of growth failure in children with sickle cell disease. *Pediatr Res*. 2007;61(5 Pt 1):607-13.

Conflict-of-interest disclosure:
The author declares no competing financial interest

Submitted: 12/21/2012
Accepted: 12/23/2012

Corresponding author:
Maria Stella Figueiredo
Disciplina de Hematologia e Hemoterapia
Escola Paulista de Medicina - UNIFESP
Rua Dr Diogo de Faria, 824, 3º andar
Vila Clementino
04037-002 São Paulo, SP, Brazil
stella.figueiredo@unifesp.br

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

DOI: 10.5581/1516-8484.20130021

12. Uchendu UO, Nwokocha AR, Ikefuna AN, Emodi II, Onwasigwe CN. Evaluation of sexual maturity among adolescent male sickle cell anaemia patients: the usefulness of testicular volume estimation. *South Afr J Child Health*. 2010;4:11-5.
13. Jenerette CM, Brewer C. Health-related stigma in young adults with sickle cell disease. *J Natl Med Assoc*. 2010;102(11):1050-5.
14. Wallois A, Vian E, Loko G, Blanchet P. [Evaluation of sexuality among Afro-Caribbean patients homozygous SS sickle cell disease followed in Martinique]. *Prog Urol*. 2012;22(5):301-6. French.
15. Saad ST, Lajolo C, Gilli S, Marques Junior JF, Lima CS, Costa FF, et al. Follow-up of sickle cell disease patients with priapism treated by hydroxyurea. *Am J Hematol*. 2004;77(1):45-9.
16. Burnett AL. Sexual health outcomes improvement in sickle cell disease: a matter of health policy? *J Sex Med*. 2012;9(1):104-13.
17. Cobo VA, Chapadeiro CA, Ribeiro JB, Moraes-Souza H, Martins PR. Sexuality and sickle cell anemia. *Rev Bras Hematol Hemoter*. 2013;35(2):89-93.

XXX
