



## Revista Brasileira de Hematologia e Hemoterapia Brazilian Journal of Hematology and Hemotherapy

[www.rbhh.org](http://www.rbhh.org)



### Scientific Comment

# Comment on sickle cell disease and left ventricular hypertrophy<sup>☆</sup>



Monica Pinheiro de Almeida Verissimo

Centro Infantil de Investigações Hematológicas Dr. Domingos A. Boldrini, Campinas, SP, Brazil

Sickle cell disease (SCD) is a monogenic disease that causes a significant reduction in life expectancy due to the presence of chronic anemia, acute painful episodes and organic injuries. The most common chronic injuries are cardiopulmonary complications, such as pulmonary hypertension (HP), chronic lung disease, diastolic dysfunction and congestive heart failure (CHF), which are manifested mostly in adulthood.

These complications are associated with early mortality. With the improvement of the healthcare system and greater access to proper treatment, there is a growing interest in identifying complications as early as possible by evaluating patients in childhood and adolescence in order to improve morbidity and mortality rates.<sup>1</sup>

Patients tend to have increased left ventricular (LV) stiffness with consequent left ventricular hypertrophy (LVH) and progression to increased high pulmonary blood pressure. Another complication due to LVH is diastolic dysfunction, an early marker of cardiac impairment preceding CHF that is frequently found in adults with SCD.<sup>2</sup>

The pathophysiologic mechanism of HP in individuals with SCD has not been well defined but is probably multifactorial. Potential etiologic factors include hemolysis that interferes with nitric oxide-mediated vasodilatation, LV dysfunction, pulmonary thromboembolism, airway hyperreactivity, and sleep-disordered breathing.<sup>3</sup> A high cardiac output and high pulmonary blood flow, combined with intravascular hemolysis promote these structural changes.<sup>4</sup>

The incidence of HP in the pediatric population ranges from 11% to 30%. In a paper published by Colombatti et al., there is confirmation that HP starts early in children showing the

need for early and systematic assessments in this population.<sup>5</sup> This paper also showed that diastolic dysfunction occurred in children with SCD.

In the current issue of the Revista Brasileira de Hematologia e Hemoterapia, there is an article entitled "Left ventricular hypertrophy in children, adolescents and young adults with sickle cell anemia" by Faro et al. In this article the authors aimed to estimate the frequency of LVH in children, adolescents and young adults with sickle cell anemia.<sup>6</sup>

Of the 109 patients enrolled in this study, 37.6% had LVH. SCD patients had high rates of LV mass as seen in other populations. The size of the left atrium, another aspect of cardiac complications and a potential marker of diastolic dysfunction, was significantly larger in patients with LVH. The authors showed an association between this condition and low hemoglobin and hematocrit levels, a low reticulocyte index and a higher albumin:creatinine ratio. Hemolysis markers (lactate dehydrogenase, indirect bilirubin and ferritin) were statistically similar with high values in both groups.<sup>6</sup>

Unfortunately, this paper did not confirm any protective effect of hydroxyurea, but the authors hypothesized that this may be related to the small number of patients who were taking the drug. Clinical trials with larger numbers of patients may help to clarify the role of hydroxyurea in the treatment of these complications.

This paper demonstrates that the population of patients with SCD presented cardiac complications at an earlier age and that the association of clinical signs and symptoms may guide us.<sup>6</sup>

DOI of original article: <http://dx.doi.org/10.1016/j.bjhh.2015.07.001>.

<sup>☆</sup> See paper by Faro et al. in Rev Bras Hematol Hemoter. 2015;37(5):324-8.

Corresponding author at: Centro Infantil de Investigações Hematológicas Dr. Domingos A. Boldrini, Departamento de Hematologia, Rua Gabriel Porto, 1270, Cidade Universitária, 13083-210 Campinas, SP, Brazil.

E-mail address: [verissimo.monica@gmail.com](mailto:verissimo.monica@gmail.com)

<http://dx.doi.org/10.1016/j.bjhh.2015.08.005>

1516-8484/© 2015 Associação Brasileira de Hematologia, Hemoterapia e Terapia Celular. Published by Elsevier Editora Ltda. All rights reserved.

Early diagnosis and appropriate treatment could impact on the morbidity and mortality of these patients by changing the natural history of the disease.

---

### Conflicts of interest

The author declares no conflicts of interest.

### REFERENCES

---

1. Liem RI, Nevin MA, Prestridge A, Young LT, Thompson AA. Functional capacity in children and young adults with sickle cell disease undergoing evaluation for cardiopulmonary disease. *Am J Hematol.* 2009;84(10):645-9.
2. Zilberman MV, Du W, Das S, Sarnaik SA. Evaluation of left ventricular diastolic function in pediatric sickle cell disease patients. *Am J Hematol.* 2007;82(6):433-8.
3. Johnson MC, Kirkham FJ, Redline S, Rosen CL, Yan Y, Roberts I, et al. Left ventricular hypertrophy and diastolic dysfunction in children with sickle cell disease are related to asleep and waking oxygen desaturation. *Blood.* 2010;116(1):16-21.
4. Miller AC, Gladwin MT. Pulmonary complications of sickle cell disease. *Am J Respir Crit Care Med.* 2012;185(11):1154-65.
5. Colombatti R, Maschietto N, Varotto E, Grison A, Grazzina N, Meneghello L, et al. Pulmonary hypertension in sickle cell disease children under 10 years of age. *Br J Haematol.* 2010;150(5):601-9.
6. Faro GB, Menezes-Neto OA, Batista GS, Silva-Neto AP, Cipolotti R. Left ventricular hypertrophy in children, adolescents and young adults with sickle cell anemia. *Rev Bras Hematol Hemoter.* 2015;37(5):324-8.