One of the reasons for the success of Jornal de Pediatria is the diversity of the articles published. The topics addressed in our articles range from primary care issues, such as breastfeeding, to translational research, as well as observational studies and clinical randomized trials.[4-7] Our only commitment is to guarantee the scientific quality of the manuscripts accepted for publication. We have achieved the objective set out when I was nominated as editor-in-chief of Jornal de Pediatria, in March 2002. At that time, we decided that Jornal de Pediatria should bridge the gap between basic research and clinical practice in pediatrics, always contributing to spread new knowledge and trying to keep our readers updated about the most recent scientific findings in the field.[8]

Such excellent result is a consequence of the hard work and effort of many people: the associate and executives editors, who have actively participated in the project over the years; the Editorial Board, which has always replied to our requests promptly; the presidents of the Brazilian Society of Pediatrics, Lincoln Marcelo Silveira Freire, Dioclécio Campos Jr., and Eduardo da Silva Vaz, who have always supported our decisions; the board of the Brazilian Society of Pediatrics, which has provided the necessary support for the journal's development; the independent reviewers, who were always ready to collaborate; and, in particular, the investigators, who submitted excellent articles for publication.

In sum, I hope that this is just the beginning of the journey for the scientific growth and international recognition of Jornal de Pediatria.

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Toward high quality medical care for sickle cell disease: are we there yet?

Jane Hankins*

Sickle cell disease (SCD), one of the most common single gene disorders in the world, affects approximately 280,000 live births per year worldwide; SCD and thalassemia syndromes together account for 3.4% of all deaths of children younger than 5 years.[1] Infants with SCD are particularly at risk of premature mortality from SCD because of the early loss of the filtrative function of the spleen.[2,3] This places these vulnerable infants at a high risk of invasive infection by encapsulated organisms, especially pneumococcus. In addition, splenic sequestration is an important SCD-related complication that, if not recognized and treated early in its course, can lead to circulatory collapse and death.[4]

Despite the increased risk of fatal complications in early life, mortality rates of patients with SCD have significantly

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declined over the past 4 decades. The most significant decrease has occurred in the first 5 years of life, with a continued shift toward mortality later in life.[5] This important trend of increased survival among children can be attributed to simple measures such as early diagnosis, followed by prophylactic therapy and anticipatory guidance. The first and most fundamental of these steps is the implementation of screening programs for newborns. Identification of an at-risk young population has allowed targeted use of prophylactic penicillin and special immunization with *Streptococcus pneumoniae*, *Haemophilus influenzae,* and *Meningococcus meningitidis* vaccinations. These basic public health strategies have positively impacted the overall outcome of pediatric patients with SCD not only in the USA but also worldwide.[6,7]

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Recent statistics from a pediatric cohort from Dallas, TX, USA, shows that the chances of children with SCD dying before 5 years of age are less than 1%.[8] This represents a major shift from the 1960s and 1970s, when many children with SCD were not expected to live into adulthood and their risk of sepsis was 20 to 30 times higher than that of the general population.[9,10] In this issue of Jornal de Pediatria, Dr. Fernandes and her colleagues have elegantly demonstrated that implementation of an organized, centralized, and well-designed newborn screening program, followed by diligent employment of basic clinical standards, is able to yet again impact and modify the natural course of SCD among young children. Survival rates of children from Minas Gerais, Brazil, who have SCD and are younger than 5 years, are comparable with those from North America, Europe, and the Caribbean, despite the relatively brief existence of Brazil's hemoglobinopathy newborn screening program (Table 1).[11]

In the future, further improvements are expected in the Brazilian SCD program. Dr. Fernandes's study found that the rate of special immunization (pneumococcal and meningococcal vaccinations) was very low among fatal cases (only 40% of the children who died were immunized), and 31% of patients succumbed to acute splenic sequestration. This underlines the need for a greater effort toward global immunization against encapsulated organisms. In addition, further investment in educating family members about the complications of SCD could have prevented fatal cases of splenic sequestration. A greater emphasis on educating health care providers from primary health care units about SCD is likely to improve the outcome of many more children with SCD. Considering that 58% of the families who lost a child with SCD in Minas Gerais had a monthly income of less than US$ 50.00, the relative low death rate observed in Dr. Fernandes's work shows that adequate diagnosis and medical care can prevent mortality even in the setting of extreme poverty.

Another potential disease modifier that could impact the natural history of SCD and reduce early mortality is hydroxyurea therapy. Hydroxyurea appeared to have prevented deaths among adults with SCD in the United States and Greece.[12,13] Given its ease of administration and low cost, hydroxyurea therapy could be adopted and used in developing nations. The feasibility of its use, including the cost of ongoing monitoring of hydroxyurea therapy's toxicity and efficacy, may pose a problem for resource-poor areas of the world, but with adequate planning and some

Table 1 - Comparison of survival rates of children with sickle cell anemia younger than 5 years from different regions of the world where newborn screening for hemoglobinopathies is available

Region	Under 5-year survival (%)	Disease genotype	Period of analysis	Main cause of death	Number of patients evaluated	Duration of observation (person-years)
Minas Gerais, Brazil[11]	89.4	HbSS and HbSβ^0-thal	1998-2005	Sepsis	764	2,493
Kingston, Jamaica[16]	91	HbSS	1979-1994	ACS	105	Not available
Los Angeles, United States[17]	~ 93	HbSS	1975-2003	Sepsis	172	Not available
Cooperative Study of Sickle Cell Disease*[18]	95	HbSS	1978-1988	Sepsis	427	1,781
Dallas, United States[8]	99.2	HbSS and HbSβ^0-thal	2000-2007	ACS	180†	663.6
London, England[19]	100	HbSS	1982-2005	N/A	180	1,542

ACS = acute chest syndrome; N/A = not applicable.
* Nineteen clinical sickle cell disease centers in the United States.
† These 180 patients represent a subset of the Dallas Newborn Cohort that has a total of 940 patients with a total of 8,857 patient-years of follow-up.

basic infrastructure in place, it is doable. Hydroxyurea therapy has the potential to reduce mortality rates of young children with SCD because of its possible role in organ protection and proven effect in reducing acute vaso-occlusive episodes.[14,15]

Finally, it is important to note that the creation of an enterprise that will be responsible for successfully diagnosing and treating SCD in the context of an emerging economy requires substantial efforts from its organizers and sustained government support. Despite the natural obstacles to establishing such a medical infrastructure, as well as the substantial low socioeconomic status of most patients, deaths can be prevented and outcomes improved with such an undertaking. The experience in Brazil can serve as an example to many other nations with similar socioeconomic circumstances. As caregivers of SCD, we have come a long way, and although we have not yet achieved the goal of excellence in medical care for all patients with SCD, the road ahead may be successfully traveled.

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