Comprehensive care for patients with sickle cell disease in Cuba

Haematologica 2008; 93:e20 DOI: 10.3324/haematol.11986

Sickle cell disease (SCD) is the most frequent hereditary disorder in Cuba. The incidence of sickle cell trait is 13, 25 in blacks, 0, 65 in whites and 3, 085 for the general population.[1] There are 4000 sickle cell patients distributed throughout the country, with a greater prevalence in Havana city and in the South-Western provinces.[2] In spite of the fact that Cuba is a country with limited resources: our national health system allows a holistic attention to patients with SCD. A nationwide programme for the prevention of sickle cell disorders was initiated in our country in 1983. Couples at risk have been identified by screening pregnant women and the partner of those who carry the S or C haemoglobin, followed by prenatal diagnosis and genetic counselling.[3] There is no newborn screening up to now but all the babies with SCD are closely monitored from the first months of their life. There is no data regarding the number of children born with SCD before the implementation of the prenatal diagnosis plan.

Sickle cell patients are treated via specialized consultation by haematologists in all 14 provinces of the country and they have easy access to all kinds of health services including hospitalization and emergency services. They have access to narcotic analgesics in the health care institutions, but they do not habitually take them home. Cienfuegos is a central province of Cuba with about 400,000 inhabitants, 96 of whom suffer with SCD at this moment. Throughout the last 10 years 5 patients have died, all adults, three men of acute chest syndrome, one of hepatic complications and one woman died of pulmonary thromboembolism right after a delivery. The Institute of Haematology and Immunology in Havana city is our referral centre for complex cases. The guidelines for management and treatment as well as many investigations of this disease have been developed there.[4-7] In a research done in that centre, it was established that the global survival estimates were 53 years in sickle cell anaemia, 59 years in SC haemoglobinopathy and 48 in Sβ thalassemia. The most frequent cases of death were hepatic complications, stroke and infections.[8] This study was performed in 397 adult patients who had been followed during 25 years; the survival was calculated using the Kaplan-Meier curves. This data was not compared to the Cooperative Study for SCD in the USA and it is not sufficient to support a decrease in the mortality rate in our patients due to the fact that there are no previous statistics. The decrease in infancy mortality has been obtained due to early attention and proper treatment from the first post-partum months, prevention of infections by *S. pneumoniae* with prophylactic use of penicillin in all the children of the country and patient/family orientation with active involvement in the early diagnosis of complications.[8] The all-embracing assistance plan in the patients was carried out with the aid of the *Centro Informazione e Educazione allo Sviluppo (CIES)* from Italy. Today, transcranial Doppler ultrasound is being realized in children and in adults, screening for pulmonary hypertension.

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Key words: sickle cell disease, management, health services

References

1. Colombo B, Martínez G. Hemoglobin variants in Cuba. Hemoglobin 1985;9:415-22.
2. Svarch E, Hernández P y Ballester JM. La drepanocitosis en Cuba. *Rev Cubana Hematol Inmunol Hemoter*. [online]. Mayo-ago. 2004, vol.20, no.2 [citado 11 Julio 2007], p.0-0. Disponible en: www: <http://scielo.sld.cu/scielo.php?script=ci_arttext&pid=S0864-02892004000200009&lng=es&nrm=iso>. ISSN 0864-0289.
3. Granda H, Gispert S, Martínez G, Gomez M, Ferreira R, Collazo T, et al. Results from a reference laboratory for prenatal diagnosis of sickle cell disease in Cuba. Prenat Diagn 1994;14:659-62.
4. Svarch E, Nordet I, Valdés J, González A, Machín S, de la Torres E. Partial splenectomy in children with sickle cell disease. Haematologica 2003;88:222-3.
5. Martínez G, Muñiz A, Svarch E, Espinosa E, Nagel RL. Age dependence of the gene frequency of alpha/talasemia in sickle cell anaemia in Cuba. Blood 1996;88:1898-9.
6. Muñiz A, Corral L, Alaez C, Svarch E, Espinosa E, Carbonell N, et al. Sickle cell anemia and beta-gene cluster haplotypes in Cuba. Am J Hematol 1995;49:163-4.
7. Svarch E, Machín S, Nieves RM, Marcia de Reyes AG, Navarrete M, Rodríguez H. Hydroxyurea treatment in children with sickle cell anemia in Central America and the Caribbean countries. Pediatr Blood Cancer 2006;47:111-2.
8. Machín S, Guerra T, Svarch E, Espinosa E, Mesa Cuervo JR, Dorticós E et al. Morbiletalidad en pacientes adultos con drepanocitosis. Rev Cubana Hematol Inmunol Hemoter [online] 2004;20 [citado 11 Julio 2007], p.0-0. Disponible en: www: <http://scielo.sld.cu.scielo.php?script=sci_arttext&pid=S086402892004000200004&lng=es&nrm=iso>. ISSN 0864-0289.
9. Svarch E, Nordet I, Machín S, Fernandez L, Muñiz A, Wade M. Sickle-cell anemia in the 1st 5 years of life. Sangre (Barc) 1996;41:43-6.