CARTA AL EDITOR

Program for comprehensive sickle cell disease care in Cuba

Programa de atención a la drepanocitosis en Cuba

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Cuba has a population of 11 million inhabitants. The percentage of blacks and mixed race is about 30 %. The prevalence of AS trait is 3.04 % in the general population [(1)]. A prenatal diagnosis has been carried out by the *National Center for Medical Genetics* since 1986. Until 2012; 4 127 986 pregnant women have been screened: 144 229 had AS or AC trait, and 6 507 were couples at risk. In 5 042 pregnancies, molecular prenatal diagnosis was performed and 992 foetuses were found with sicklecelldisease(SCD) (personal communication). Since 1990, a program for comprehensive SCD care was developed atInstitute of Hematology and Immunology, the leading center for SCD care in Cuba. In the setting of this program, guidelines and a booklet for patients and relatives were written. The comprehensive care includes: 1) prenatal diagnosis, 2) haematological follow up since the first months of life, 3) prophylactic administration of penicillin in the first five years of age, 4) folic acid supplementationand 5) growth and development surveillance. We use a conservative transfusional regime and in patients receiving transfusions, HIV and hepatitis, B and C were tested periodically. Transcranial Doppler ultrasonography was performed in 107 children with sickle cell anemia (SCA). The velocity in the median cerebral arteries was conditional (170-199 cm/s) in six patients and abnormal (≥200 cm/s) in seven. In these patients we indicated continuous treatment with hydroxyurea 25 mg/kg/day [(2)]. In patients with microalbuminuria (>30 mg/L), angiotensin converting enzymes inhibitor treatment was administered. Some outstanding results must be pointed out: partial splenectomy was performed for the treatment of acute splenic sequestration crisis and after this procedure no recurrences occurred [(3)] and only one patient had overwhelming septicaemia with complete recovery [(4)]. Children with severe disease were treated with hydroxyureaat a fixed dose of 15 mg/kg/day. In addition, in 51 children from Central America and the Caribbean countries treated according to our

program guidelines, a significant decrease of painful vaso-occlusive crisis, acute chest syndrome, transfusions and admissions occurred[5]. In 87 patients a neurocognitive study with the *Wecshler* intelligence scale was performed. It was observed a decrease of ratios of full scale intelligence, performance scale, order figures and block design [6]. The comprehensive care of SCD in Cuba provided a better quality of life. Psychosocial support including genetic counselling and educational and labour orientations are given to all the patients.

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