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Hemoglobin sickle cell disease in Brazil

We read with great interest the recent paper presented by Lionnet *et al*.[1] and we would like to briefly report our experience with hemoglobin SC (HbSC) disease. Before the institution of neonatal screening in Brazil, HbSC disease was considered a rare manifestation.[2] After 2001, however, the screening program had showed an unexpected high frequency of HbSC disease of 1:3,450.[3-5]

In our clinic, we have 94 children (median age 3 years, range 1-17 years) and 29 adults (median age 35 years, range 19-81 years) with HbSC disease. It is important to point out that 67% of the patients were under ten years of age. We presume that the toddler group had a better follow up because they have been followed since they were born. Otherwise, other patients came to our clinic after a late diagnosis, and perhaps could have undergone selection, and patients with more severe manifestations did not have time to reach our service.

Furthermore, the age distribution observed in our clinic reflects the data from neonatal screening and is in agreement with the point of view of Lionnet *et al*. that Hb SC disease is not a mild form of sickle cell anemia.[1]

Regarding clinical manifestations in adult patients, we observed a higher frequency of leg ulcer (3 of 29, 10.3%), a similar frequency of osteonecrosis (4 of 29, 13.8%), and a lower frequency of vasoocclusive crises, retinopathy and priapism of 8 of 29 (27.6%), 6 of 29 (20.8%), 3 of 29 (10.3%), respectively. No patient had stroke. In addition, we observed a high frequency of gallbladder (9 of 29, 31.0%).

The prevalence of α-thalassemia in our patients was 26.7%. We also accessed the βs haplotype of these patients and observed a high frequency of Benin haplotype (51.7%) in a region where a large proportion of the sickle cell anemia patients (61.7%) come from Bantu ethnic groups.[6,7] This disparity in data obtained on SC patients' haplotype could be related to internal migratory flow, since many individuals from Bahia (were Benin haplotype prevails) come from the southeastern region.[8] However, another possible interpretation could be a modulatory action of Benin haplotype allowing these patients to have a better survival than patients with other βs haplotypes.

Our results reinforce the view that HbSC disease has important comorbidities. In children, for example, we observed 26.5% of acute chest syndrome, a manifestation associated with low HbF levels (*Cabañas-Pedro AC et al., manuscript in preparation*).

We agree with Lionnet *et al*.[1] that specific guidelines for HbSC disease management are needed, but we would like to highlight the importance of clarifying the role of haplotype analysis in these patients.

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