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Pregnancy Surveillance in Sickle Cell Disease Patients: A Cohort Study in an African Country - Angola

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Abstract

Sickle Cell Disease (SCD) is a severe monogenic disease caused by the inherence of a recessive mutation in the β-globin gene, with an especially high prevalence in sub-Saharan Africa. In Angola, the prevalence of the disease is almost 2%, and the carriers reach 21% of the population. Although its presentation tends to be very heterogeneous, chronic hemolytic anemia, frequent painful crisis and extensive organ damage are common features of these patients. Pregnancy in SCD patients is associated with an increase in severe outcomes, namely, high risk of eclampsia and pre-eclampsia, stroke and even death. Therefore, it is crucial to maintain continuous medical surveillance during pregnancy, especially in women with previous strokes. Moreover, health services in low- and middle-income countries are generally not prepared to follow these patients.

The present cohort study, conducted at the Lucrecia Paim Maternity Hospital (Luanda, Angola), aims to determine pregnancy complications in SCD women, especially those responsible for maternal death, and, by supporting the obstetric consultations in this hospital, contribute to the reduction of mortality and morbidity rates. Pregnancy monitoring includes analysis of clinical history and incidents (number of hospitalizations, blood transfusions, strokes and other clinical complications), hematological and biochemical analysis, transcranial doppler to assess cerebral hemodynamics and genetic analysis (confirmation of the diagnosis, genotyping of four SNPs in the β-cluster to assess the haplotype, and evaluation of the presence of the 3.7kb deletion of the

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To date, 61 women are being followed in the obstetric consultations, with ages from 18 to 40 years old (mean 26.1±5.4). There are no records of previous strokes, although 83.9% of the patients have been previously transfused (47 out of 56), 98.2% have been hospitalized (55 out of 56) due to SCD complications and 19.6% (10 out of 54) had at least one miscarriage. At the first appointment, total hemoglobin values ranged from 4.70 to 10.40 g/dL (n=52, mean 7.18±1.30), erythrocytes from 1.46 to 5.42 $x10^{12}$/L (n=52, mean 2.46±0,72), white blood cells count from 1.67 to 61.88 $x10^{9}$/L (n=51, mean 12.20±8.69), platelets from 24.2 to 710.0 $x10^{9}$/L (n=52, mean 272.2±155.9), and lactate dehydrogenase (LDH) from 263.3 to 2836.7 (n=50, mean 708.1±450.46).

The CAR/CAR haplotype, which is usually associated with a more severe prognosis, is the most prevalent in this population (57.7%, 30 out of 52), followed by the CAR/SEN haplotype (25.0%, 13 out of 52). In this population, 17.3% (9 out of 52) are homozygous for the 3.7kb α-thalassemia deletion and 44.2% (23 out of 52) are carriers. This deletion influences hematological and clinical aspects of sickle cell disease patients, resulting in less severe phenotypes.

TCD time-averaged mean of the maximum velocity (TAMMx) at the middle cerebral arteries ranged between 41 to 132 cm/s (n=61, mean 84cm/s) and peak systolic velocity (PSV) from 61 to 180 cm/s (mean 129 cm/s). At the basilar artery level, TAMMx obtained were between 29 to 102 cm/s (n=60, mean 52 cm/s) and PSV ranged from 43 to 141 cm/s (mean 78 cm/s).

The main goal of this project is to study pregnancy-related complications and outcomes by giving support to an Angolan cohort of SCD pregnant women, seeking to reduce them and improve these women and neonates' future quality of life. By the end of the project, we also intend to obtain TCD reference values of cerebral blood flow velocities in pregnant women with SCD as a risk predictor of vascular events as there are no values in the literature for this specific population.

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Disclosures

No relevant conflicts of interest to declare.

Author notes

* Asterisk with author names denotes non-ASH members.