Genetic factor effect on blood rheology in children with sickle cell anemia

Céline RENOUX1, Philippe JOLY1, Camille FAES2, Yves Bertrand3, Nathalie Garnier3, Daniella Cuzzubo3, Kamila Kebaïli3, Cécile Renard3, Arthur Dony3, Giovanna Cannas4, Cyril Martin2, Vincent Pialoux2, Philippe Connes2

1UF Pathologie Moléculaire du globule rouge - Hôpital Edouard Herriot - Hospices civils de Lyon
2Laboratoire LIBM, Equipe “Biologie Vasculaire et du Globule Rouge”, Université de Lyon
3Institut d’Hématologie et d’Oncologie Pédiatrique (IHOP) - Hospices civils de Lyon
4Clinique de Médecine Ambulatoire/Hématologie (Hôpital Edouard Herriot, LYON)

Background: Sickle cell anemia (SCA) is a severe hereditary hemoglobinopathy characterized by abnormal blood rheology, which plays a role in the occurrence of several acute and chronic clinical complications. While beta-haplotypes and alpha-thalassemia modulate SCA clinical severity, their effects on blood rheology have been incompletely described. The aim of this study was to test the effects of these genetic modifiers on the hemorheological properties and frequency of vaso-occlusive crises (VOC) of SCA children.

Materials and methods: Steady-state hemorheological profile (blood viscosity, red blood cell (RBC) deformability and aggregation), biological parameters, beta-haplotype, alpha gene status and VOC frequencies (3 years period) were analyzed in 44 SCA (S/S or S/bêta0-thalassemia) children (aged from 3 to 17).

Results: Alpha-thalassemia patients showed increased RBC deformability and aggregation compared to those without. Mean VOC frequency was higher in patients with 2 deleted alpha-genes compared to those with a normal alpha genotype. The hemorheological profile was not influenced by the beta-haplotype in our cohort.

Conclusions: Our results suggest that alpha-thalassemia increases the risk for VOC events in SCA children through its effects on blood rheology.