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Letter to the Editor

Terminologies regarding sickle cell retinopathy and maculopathy

Laurence Beral[a,b], Marc Romana[b,c], Nathalie Lemonne[d], Thierry David[a] and Philippe Connes[c,e,*]

[a]*Service d'ophtalmologie, CHU de Pointe-à-Pitre/Abymes, Pointe-à-Pitre, Guadeloupe, France*
[b]*Université des Antilles, CHU de Pointe-à-Pitre, Guadeloupe, Université Sorbonne Paris Cité, Université Paris Diderot, Inserm, INTS, Unité Biologie Intégrée du Globule Rouge UMR_S1134*
[c]*Laboratoire d'Excellence GR-Ex ≪ The red cell : from genesis to death ≫, PRES Sorbonne Paris Cité, Paris, France*
[d]*Unité transversale de la Drépanocytose, CHU de Pointe-à-Pitre/Abymes, Pointe-à-Pitre, Guadeloupe, France*
[e]*Laboratoire Interuniversitaire de Biologie de la Motricité (LIBM), Equipe Biologie Vasculaire et du Globule Rouge, Université Claude Bernard Lyon 1, COMUE Lyon, Villeurbanne, France*

Dear editor,

We read with great interest the letter written by Guerra et al. [1]. To answer their questions, we defined retinopathy as any stage of proliferative sickle retinopathy displayed by our patients (classification of Goldberg ranging from 1 to 5, [2]). It is well known that tractional maculopathy can be related to proliferative Sickle cell retinopathy (SCR), and can be involved in a retinal detachment including the macular area. Maculopathy has also been described in several other proliferative retinopathies such as diabetes retinopathy [3, 4]. Nevertheless, our description of sickle cell maculopathy did not include tractional disease. None of our patients had tractional retinal detachment or tractional complication. As a matter of fact, 15 of our patients displayed maculopathy without retinopathy [5]. In our study, we defined maculopathy as any macular thinning under 226 micron, as proposed by the Copernicus Potopol Spectral Domain Ocular Coherence Tomography guidelines [6].

As ophthalmologists, we thought that we could observe a difference in the onset and risk factors of these two different diseases (i.e., retinopaty and maculopathy). We decided not to perform any invasive examination test, such as fluorescein angiography, if the fundoscopy or wild field retinographies were efficient enough to describe the stage of retinopathy. Angiography is used to evaluate retinal ischemia in order to guide a possible laser treatment when a vitreous hemorrhage is not elective for vitrectomy or in case of extensive sea fans. But no such case was encountered in our patients.

It has been shown that low fetal hemoglobin level could increase the risk for sickle cell patients to develop sickle retinopathy [7, 8]. However, in contrast to Dell' Arti et al., we found no link between fetal hemoglobin expression level and the presence of maculopathy or retinopathy [9].

*Corresponding author: Philippe Connes. E-mail: philippe.connes@univ-lyon1.fr.

This finding could be related to the small number of patients included in our study. Only 22 patients of our study were treated by hydroxyurea and, surprisingly, we found no link between this treatment and the level of fetal hemoglobin. No association was found between hydroxyurea and SCD retinopathy or maculopathy. Further studies based on larger patient cohorts are needed to definitively address the role of biological factors in the onset of retinopathy and maculopathy in sickle cell disease.

We thank again Guerra et al. [1] for their valuable comments, and the editor for giving us the opportunity to answer them.

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