Purpose: To characterize full-field electroretinogram (ffERG) in patients with early sickle-cell retinopathy according to hemoglobin type.

Methods: Retrospective study, in Centre hospitalier intercommunal de Créteil, France

3 groups: SS, SC and control

Patient inclusion criteria
- Non proliferative sickle cell retinopathy (stage 1/2) (fig 2)
- Preserved VA

Patient exclusion criteria
- Other ophthalmological issue, laser treatment history

Control inclusion criteria:
- Aged matched, no ophthalmological history

Performed: VA, fundus, FA, OCT and ffERG

Results: Twenty-four eyes from 12 patients (6 HbSS and 6 HbSC) and twelve eyes from six controls were included.

ffERG results are visible in tables 1 (amplitudes) and in figure 3.

Significant alterations were found for amplitudes between the 3 groups:
- **Patients from HbSS group** showed a dramatic decrease of b-wave amplitudes for all dark-adapted ffERG responses (\( \text{HbSS HOMOZYGOUS} \))

- Patients from the HbSC group showed reduced a-wave amplitudes for all dark-adapted and light-adapted ffERG responses, compared with the control group.

- Patients from the HbSS+HbSC group exhibited reduced a-wave amplitudes for all dark-adapted and light-adapted ffERG responses, reduced flicker 30Hz amplitudes and reduced b-wave amplitudes for DA 0.01 and DA 10.0 responses compared with the control group.

Discussion:

This is the first report on eletrophysiological alterations in HbSS and HbSC patients, occurring so early in the evolution of sickle cell retinopathy

In 1987, Peachy et al.\(^2\) studied retinal function in sickle cell disease patients and could not find any ERG modifications in sickle cell patients without proliferative lesions; however, they did not differentiate between SS and SC patients and the methods preceded the ISCEV standards era.

In this study:
- In HbSC patients: The significantly decreased a-wave amplitude found for ffERG responses are likely related to outer retina damage, as seen in chronic retinal ischemia.\(^3\)
- In HbSS patients: The significant decrease of the dark-adapted b-wave amplitudes was not associated with a significant a-wave reduction, suggesting inner retina dysfunction.\(^4\) The trend toward reduced OPs is another strong argument for this latter hypothesis in HbSS patients.\(^6\)

Recent imaging studies demonstrated significant thinning of the inner retina and vascular abnormalities in the superficial and deep capillary plexus to be common and early features in sickle cells patients; even before severe peripheral retinopathy.\(^7,8,9\)

Conclusions:

The main limitation of this study is the small number of patients included. Of course, to be able to support our hypothesis, larger studies are needed. It would also be of interest to compare ffERG results to OCT-Angiography findings; however, current systems mostly cover the area of the central retina, while ffERG collects responses from the whole retina.

Evaluating ffERG responses of sickle cell patients would help determining possible correlations between global retinal function and the severity of vascular systemic complications

Bibliography: