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# Retinal Damage in Sickle Cell Disease in Abidjan (Ivory Coast)

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**Abstract:** The objective is to describe the epidemiological, clinical and therapeutic aspects of retinal damage in sickle cell disease. This was a cross-sectional, descriptive and analytical study, conducted from November 2022 to February 2023, i. e. for a period of 4 months in a private ophthalmology centre in Abidjan and at the University Hospital of Cocody (Côte d'Ivoire). It focused on sickle cell patients seen in consultation. A total of 50 patients, i. e. 100 eyes were included, corresponding to the sample. The data were entered and analysed using SPSS software. The mean age of the patients was 30 years  $\pm$  10.495. Sickle cell disease SC was the most frequent form (52%). Half of the eyes (50%) had retinal damage with 34% of proliferative retinopathy (34 eyes). The most common Goldberg angiographic stage was stage III (73.53%). Laser photocoagulation was performed in stage III eyes. Retinal damage in sickle cell disease is common and is most often seen in SC patients. Stage III neovascularisation is the barrier that must not be crossed, as the consequences are dramatic with irreversible blindness. Laser photocoagulation is necessary to prevent the formation or to promote the regression of neovessels. Screening should be annual and especially with fluorescein retinal angiography.

**Keywords:** Retinal Neovascularisation, Sickle Cell, Epidemiology

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## 1. Introduction

Sickle cell disease is an inherited haemoglobin disorder. It results from a genetic error in the synthesis of the  $\beta$  chain, which gives rise to an abnormal structure and function of the haemoglobin molecule [1]. It is linked to a qualitative disorder related to a substitution of a structural gene. The  $\beta$  chain of the 6th amino acid, glutamic acid, is replaced by valine.

In Côte d'Ivoire, the prevalence of the S gene is between 12 and 14% [2]. It is characterised by polymerised haemoglobin (HbS) and sickle cell disease of the red blood cells, leading to chronic haemolytic anaemia and recurrent vaso-occlusive crises (CVO) in many organs responsible for vascular complications such as sickle cell retinopathy [3, 4]. Several studies have been carried out on this subject [5, 6]. In Côte d'Ivoire, the most recent was carried out by Fanny et al in 2004 [7].

In order to update the data on this subject, we propose to describe the epidemiological, clinical and therapeutic aspects of retinal damage in sickle cell disease at the University Hospital Centre (CHU) of Cocody and in a private ophthalmology centre in Abidjan, which specialises in the management of retinal pathologies.

## 2. Methods

### 2.1. Study Setting, Type, Period of Study and Sampling

The study was carried out in a private ophthalmology centre in Abidjan and at the University Hospital Centre (CHU) in Cocody (Côte d'Ivoire).

It was a cross-sectional, descriptive and analytical study, conducted from November 2022 to February 2023, i. e. over a period of 4 months.

It focused on sickle cell patients seen for consultation during the study period in the aforementioned departments.

All patients with sickle cell disease aged over 7 years were included in the study. Patients with other retinal pathologies and those with anterior segment damage preventing retinal analysis were not included.

A total of 50 patients, or 100 eyes, were included, corresponding to the sample or survey population. We registered 15 patients in the private centre and 35 patients in the ophthalmology department of the CHU de Cocody.

## 2.2. Patient Selection Method

We systematically registered sickle cell patients who had consulted the aforementioned departments during the study period. These were patients referred by a haematologist for ophthalmological follow-up or patients who came for an ophthalmological consultation for any visual problem. The patients were selected on the basis of convenience, i. e. no random selection was involved.

A complete ophthalmological examination was carried out, including measurement of distance visual acuity, biomicroscopic examination of the anterior segment and fundus examination using a Goldmann three-mirror lens or an aspheric Volk lens after pupillary dilation.

In sickle cell disease, 5 phenotypes are frequently encountered. These are: homozygous sickle cell disease: SS; single heterozygous sickle cell disease or sickle cell trait: AS; double heterozygous sickle cell disease: SC; beta-thalassa-sickle cell disease with 2 forms: S  $\beta$  thalassaemia: SFA2; S  $\beta$ + thalassaemia: SAFA2. The SSFA2, SC, SFA2 and SAFA2 phenotypes determine major forms and the AS phenotype, an asymptomatic form.

Using the Goldberg classification [8], we were able to identify 2 types of retinopathy. Non-proliferative retinopathy was evoked in the presence of ophthalmoscopic lesions such as pressureless whites, salmon haemorrhages and black sunburst spots corresponding to retinal pigmented lesions caused by proliferation of the pigment epithelium in response to intraretinal haemorrhage. Proliferative retinopathy was classified into 5 progressive stages, namely stage I peripheral arteriolar occlusion (PAO), stage II arteriolo-venular anastomosis (AAV), stage III peripheral preretinal neovascularisation (PVN) or sea-fan, stage IV intravitreal haemorrhage (IVH) and stage V retinal detachment (RD). The therapeutic course of action depended on the classification.

We monitored for non-proliferative lesions. Laser photocoagulation was used to treat proliferative retinopathy. Cases of detached retina were referred to the retinal surgeon at the private ophthalmology centre.

From a paraclinical point of view, fluorescein retinal angiography was performed at the private centre, as the CHU de Cocody did not have a retinal angiography machine. This is an invasive examination, enabling a haemodynamic study of retinal vascularisation. Optical coherence tomography (OCT) was used to carry out a structural analysis of the macular region. An angiographic OCT was not performed, as this would have revealed macular ischaemia.

Data collection was recorded on a survey form, including socio-demographic data, in particular age and sex; clinical aspects such as distance visual acuity and fundus. Paraclinical aspects such as fluorescein retinal angiography, optical coherence tomography (OCT) and type of treatment were reported.

The variables studied were age, sex, distance visual acuity and fundus. For the paraclinical aspect, the variables taken into account were fluorescein retinal angiography and optical coherence tomography (OCT) in the case of associated maculopathy.

The nature of the treatment carried out was also studied.

## 2.3. Ethical Considerations

Patient data was collected anonymously and in strict confidence. All participants had given their free and informed consent prior to enrolment. An individual identification number was used for each survey form and for computer data entry.

## 2.4. Data Analysis

The data were entered and analysed using SPSS software. Descriptive statistical methods were used to estimate numbers, percentages and means. The Pearson Chi-square test was used to assess the existence or otherwise of a relationship between the different qualitative variables. The likelihood ratio was used when the Chi-2 was not applicable (theoretical number of employees less than 5). The confidence level was 95% ( $\alpha=5\%$ ).

## 2.5. Limitations of the Study

The absence of retinal angiography at the CHU de Cocody obliged the ophthalmologist to refer patients to a private centre for the examination. Patients were most often referred at the stage of proliferating lesions.

# 3. Results

## 3.1. Epidemiological Aspects

The mean age of the patients was  $30 \pm 10.495$  years, with extremes ranging from 9 to 49 years. The most common age group was 18 to 35 years. Females predominated in 62% of cases (62 eyes) with a sex ratio of 0.6. Sickle cell disease SC was the most frequent form (52%), followed by the SS form (20%) (table 1).

*Table 1. Distribution of Eyes by type of Sickle Cell Disease.*

Type of sickle cell disease	Number (n)	Percentage (%)
SS	20	20
SC	52	52
SFA2	10	10
SAFA2	4	4
AS	14	14
Total	100	100

## 3.2. Clinical and Paraclinical Aspects

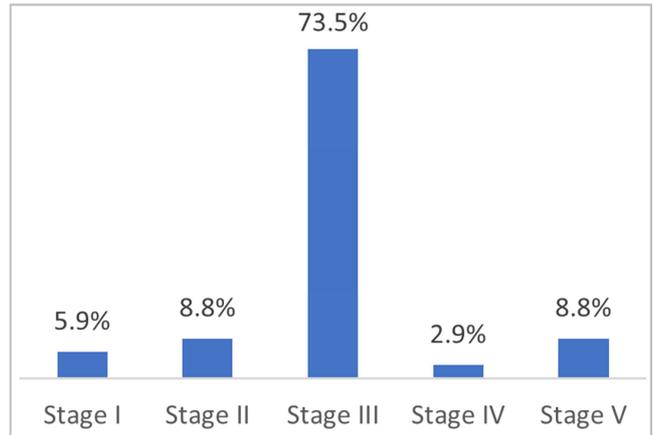
A decrease in monocular visual acuity,  $<1/20$ , was observed

in 5% of cases (5 eyes). Half of the eyes (50%) showed retinal damage, with 34% of proliferative retinopathy (34 eyes) and 16% of non-proliferative retinopathy (16 eyes). The most common non-proliferative lesions were pressureless whites (37.9%), followed by solar black spots (34.5%) (Table 2).

Of the 100 eyes, fluorescein retinal angiography was performed in 48 cases with 70.83% proliferating lesions (34 eyes) and 16.67% non-proliferating lesions (8 eyes). It was normal in 12.50% of cases (6 eyes). The most frequent Goldberg angiographic stage was stage III (73.5%), corresponding to the neovessel stage (Figure 1).

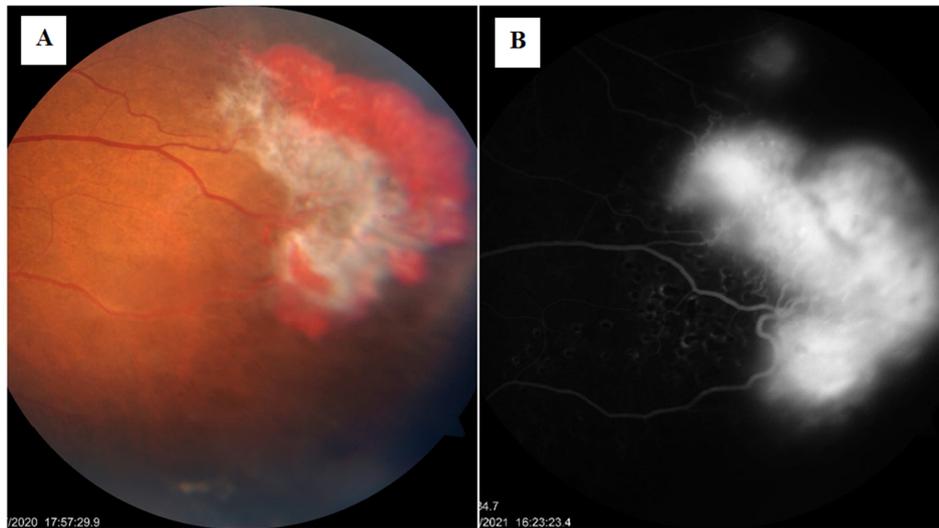
**Table 2.** Distribution of Eyes According to Non-Proliferative Lesions.

Proliferative lesions	Effectifs (n)	Pourcentage (%)
Haemorrhage	8	13, 8
Black sun spots	20	34, 5
Pigmented lumps	8	13, 8
Whites without pressions	22	37, 9
Total	58	100



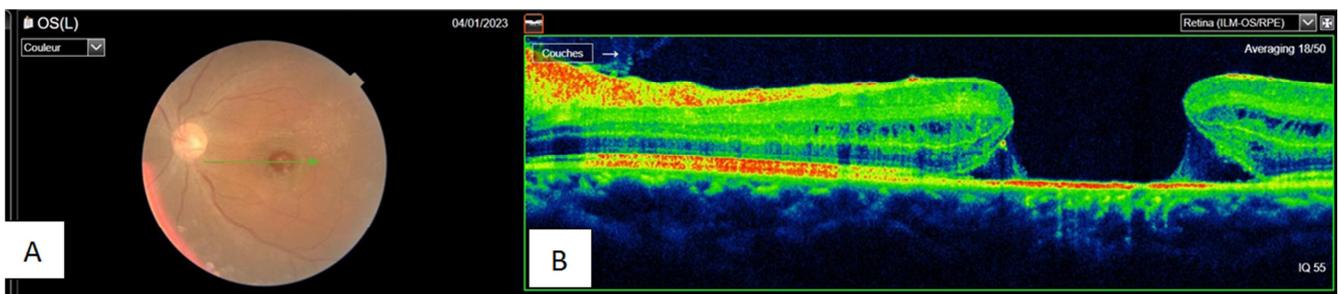
**Figure 1.** Distribution of Eyes According to Angiographic Stage.

These peripheral neovessels have a structure resembling a sea fan (Figures 2, 3).



**Figure 2.** Sea fan Neovessel Characteristic of Sickle Cell Retinopathy. (A) Retinophotography; (B) Fluorescein Angiography Diffusion.

Optical coherence tomography (OCT) revealed 1 case of maculopathy (macular hole) (figure 3).



**Figure 3.** Retinal Detachment Associated with Maculopathy. (A) Inferior Retinal Detachment with Inferior Retinal Tears, Superior Laser Scarring and Macular Hole; (B) Tomographic Appearance of Macular Hole.

### 3.3. Therapeutic Aspects

Laser photocoagulation was performed in all stage III patients. Patients with detached retina were referred to the clinic's retinal surgeon. Monitoring was instituted in the eye

presenting the intravitreal haemorrhage with a view to performing surgery in the absence of resorption.

### 3.4. Analytical Study

There was a significant association between age ( $p=0.00$ ),

sex ( $p=0.03$ ), angiographic examination ( $p=0.000$ ) and retinal damage. There was also a correlation between type of sickle cell disease ( $p=0.001$ ), age group ( $p=0.000$ ) and type of sickle cell retinopathy. There was no significant association between the different stages of proliferative lesions on angiography and the type of sickle cell disease ( $p=0.73$ ).

## 4. Discussion

The frequency of retinal damage in sickle cell disease varies from study to study.

In this series, it represented 50% of eyes on fundus examination, with 36% in SC patients, 6% in SS patients, 4% in AS patients, 2% in SAFA2 patients and 2% in SFA2 patients. This frequency was similar to that of Odoulami-Yehouessi [9] *et al.*, in Benin, in 2009, who noted 50.9% retinal damage. It was lower than that of Fanny [7, 10], who reported 72% retinal lesions in two different studies in 2004 and 2005. This variation in frequency is thought to depend on the type of sickle cell disease and the examination method used to screen for retinal damage. In contrast to this work, in Fanny's study all patients underwent fluorescein retinal angiography. And her study sample only included sickle cell AS patients in 2004 and sickle cell beta-thalassemia patients in 2005.

The mean age of the patients was  $30 \pm 10.49$  years, with a higher proportion in the 18 to 35 age group. This mean age was similar to that of Fanny [10], in 2004, which was 32.6 years. It was also close to that of Conaré *et al.* [11], in 2018, who reported a mean age of 27.84 years. In all cases, the population was young and active. Thus, in accordance with the literature [12, 13], a significant link between age and retinal damage ( $p= 0.00$ ) was demonstrated, with a high frequency of damage from the age of 30. The importance of retinopathy in young subjects could be explained by the high frequency and severity of vaso-occlusive attacks.

The predominance of women has also been observed by Odoulami Yehouessi [9]. This finding could be explained by the sampling selection method or by chance.

A unilateral decrease in visual acuity,  $<1/20$ , was observed in 5% of eyes (5 eyes). Sickle cell retinopathy is a peripheral retinal ischaemia that can sometimes be associated with maculopathies, in particular hole and macular ischaemia. These maculopathies and stage IV and V proliferating lesions are responsible for blindness.

Retinal damage was characterised by 34% proliferative retinopathy compared with 16% non-proliferative lesions. A correlation was reported between the type of sickle cell disease and the type of retinal lesions ( $p = 0.01$ ), with 29% of proliferative lesions in sickle cell disease type SC, 2% in SS, 2% in sickle cell disease type Thalassaemia and 1% in AS. Contrary to the study by Odoulami Yehouessi [9], most authors, in particular Balo [14], Conaré [11] and Diallo [15] were unanimous on the high frequency of retinal damage in SC sickle cell disease. The high proportion of proliferative retinopathy in this series is thought to be related to the greater number of SC sickle cell patients (26 patients, *i. e.* 52% of eyes) referred by haematologists for ophthalmological follow-up. According to the

literature, retinal damage is more frequent in SC sickle cell patients. However, it is essential to institute ophthalmological monitoring in all sickle cell patients, as Boni reported 72% retinal damage with 22.7% proliferative lesions in AS sickle cell patients. No link was found between the type of sickle cell disease and the type of proliferative lesion.

Some authors believe that neovessels are more frequent in HC patients and that the risk exists during the second and third decades [13]. On the other hand, several authors [7, 14] are unanimous about the frequency and severity of retinal damage in SC patients, as confirmed by this study in which severe cases, *i. e.* intravitreal haemorrhage and detachment, were only recorded in SC patients. There was also no association between the stage of proliferative lesion and age group ( $p = 0.12$ ).

From a paraclinical point of view, fluorescein retinal angiography was performed in 48% of cases. It showed 87.5% retinal damage, with 70.83% proliferative lesions and 16.67% non-proliferative lesions. According to Goldberg, the most frequent angiographic stage was stage III (73.5%, *i. e.* 25 eyes). The frequency of retinal damage on angiography (87.5%) was higher than that obtained on fundus examination (50%). Angiography is the best way of detecting and monitoring sickle cell retinopathy. It enables lesions to be classified, their topography to be determined and therapeutic management to be guided. The failure to perform angiography on all patients in this study is thought to be related to the lack of retinal angiography equipment in public health facilities in Côte d'Ivoire, such as ours, forcing ophthalmologists to refer patients to private centres. The cost of retinal angiography in these centres was 80,000 CFA francs. It is relatively expensive for patients who generally have no social security.

The macular hole observed on OCT is most often secondary to retinal vascular pathologies such as proliferative sickle cell retinopathy. Macular ischaemia, which could not be studied in this study, could be analysed by OCT-angiography, which was not performed in this study. The search for macular ischaemia in sickle cell patients will be the subject of another study. As for the therapeutic aspects, all the patients, stage III, *i. e.* 73.53%, were treated by argon laser photocoagulation, guided by retinal angiography with fluorescein. This treatment was carried out in the private centre, which has a laser. Cases of detached retina were referred to the retinal surgery department at the private centre for endo-ocular surgery. The eye with the intravitreal haemorrhage was monitored with a view to performing surgery if the intravitreal haemorrhage persisted for more than 6 months. If patients with retinal vascular pathology are to be treated efficiently, a retina centre must be set up in every public hospital, particularly at the CHU de Cocody. We would like to take this opportunity to remind the relevant authorities that our referral ophthalmology facilities need to be equipped with a technical platform suitable for posterior segment surgery.

## 5. Conclusion

Sickle cell disease is frequently associated with retinal disorders. They are most often found in type C sickle cell patients. It is therefore necessary to carry out a systematic

ophthalmological assessment to look for retinopathy in all sickle cell patients, particularly those with type C, in order to detect lesions at an early stage and avoid blindness. Monitoring should be carried out annually, and fluorescein retinal angiography is the gold standard for detecting these lesions.

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