OCULAR FINDINGS IN CHILDREN WITH HOMOZYGOUS SICKLE CELL DISEASE IN THE DEMOCRATIC REPUBLIC OF CONGO

KAIMBO WA KAIMBO D.[1],
NGIYULU MAKUALA R.[2],
DRALANDS L.[3], *MISSOTTEN L.*[3]

ABSTRACT

Objective: The purpose of the study is to determine the frequency of ocular manifestations in Congolese children with homozygous sickle cell disease.

Methods: Sixty-six children with homozygous sickle cell disease were examined for ocular abnormalities between March 1 and August 31, 1998. The ages of children ranged from two to 18 years. Routine ophthalmic examination included measurement of visual acuity, inspection of the adnexa and cornea, refraction, slit-lamp examination and dilated ophthalmoscopy.

Results: Ocular abnormalities were found in 47 (71%) children of 66 subjects. Ophthalmologic abnormalities included conjunctival signs (32%), retinal vascular tortuosity (29%) and dilatation (26%), AV crossing (29%).

Conclusions: Ocular findings in this study were similar to those previously published in Africa, which showed a low frequency of retinal changes.

RESUME

But: Déterminer la fréquence des anomalies oculaires chez les enfants congolais drépanocytaires homozygotes.

Méthodes: 66 enfants drépanocytaires homozygotes ont été examinés du 1 Mars au 31 Août 1998 pour déterminer les anomalies oculaires. Leurs âges ont varié de 2 à 18 ans. Un examen ophtalmologique de routine a été réalisé comprennant une mesure de l'acuité visuelle, l'inspection des annexes, la réfraction, un examen à la lampe à fente et un fond d'oeil dilaté.

Résultats: Des anomalies oculaires ont été trouvées chez 47 de 66 enfants (71%). Les anomalies oculaires comprennaient le signe conjonctival (32%), la tortuosité vasculaire rétinienne (29%) et dilatation des vaisseaux (26%) et le croisement artérioveineux (29%).

Conclusion: Les anomalies oculaires trouvées dans cette étude sont comparables à celles publiées en Afrique qui montrent une fréquence moindre des lésions rétiniennes.

KEYS-WORDS

Sickle cell disease. Ocular findings.

MOTS-CLÉS

Hémoglobinopathie SS. Anomalies oculaires.

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[1] *Department of Ophthalmology,*
[2] *Department of Paediatrics, University of Kinshasa*
[3] *Department of Ophthalmology, Catholic University of Leuven*

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INTRODUCTION

Sickle-cell disease is a family of haemoglobin disorders in which the sickle beta globin gene (beta S) is inherited. The gene for sickle haemoglobin (HbS) results in the substitution of valine for the glutamic acid normally present at the sixth position from the amino terminus of the b chain of haemoglobin (16). The most common type of sickle cell disease is homozygous sickle cell anaemia (haemoglobin SS). The sickle beta globin gene is spread widely throughout Central Africa (the Democratic Republic of Congo).

Ocular findings of homozygous sickle cell disease have been well documented (2,6,18). All segments of the eye can be involved. The conjunctiva may show the sickling sign (4,14,18); the anterior segment may show iris atrophy with anterior or posterior synechiae secondary to occlusions in the iris circulation (5). The posterior segment may show optic, macular, nonproliferative and proliferative retinal changes (5).

Most of the work on the ocular manifestations of homozygous sickle cell disease was done in Jamaica (1,2,17), in the U.S.A. (5,7,12,18), in the United Kingdom (9) and in France (8).

Although ocular changes in sickle cell disease have been studied extensively in this part of the world, few studies have been performed in Africa (3,10,13).

We report on the ocular complications of homozygous sickle cell disease in Congolese children. It is the first work in the Democratic Republic of Congo on this disease.

MATERIAL AND METHODS

Study design.

As part of the multidisciplinary clinical study of the sickle cell disease conducted at the University Hospital of the University of Kinshasa, a prospective, cross-sectional study of sickle cell children was performed to determine the relative frequency of ocular manifestations in the Democratic Republic of Congo. The study was performed at the department of Ophthalmology of the University of Kinshasa between March 1 and August 31, 1998.

Patient selection.

Children with homozygous sickle cell disease were referred to us by the Department of Paediatrics and the "Sickle cell anaemia center" of the county of Yolo in the city of Kinshasa. A total of 78 children were examined. Their ages ranged from 1 to 18 years. Of 78 children examined, 12 were excluded because of poor collaboration at eye examination. For this study, the records of 66 children were considered.

Ocular examination.

All patients underwent measurement of visual acuity, refraction, slit lamp examination and dilated fundus examination. Visual acuity was estimated by using illiterate E, or Snellen visual acuity charts among older children. The ability to fixate and follow a light, and a judgement made as to whether the child was sighted or not was used for other children. All children were dilated with 0.5 or 1% tropicamide collyrium and were examined once. Fluorescein angiography was not performed.

On ocular examination, the presence or absence of the conjunctival sign, anterior or posterior segment changes was considered. The conjunctival sign consisted of the presence of regular venous dilatations, or other signs such as capillary microaneurysms, telangiectasic dilatations and comma-shaped vessels. Retinal findings were classified in nonproliferative retinopathy and proliferative retinopathy. Nonproliferative retinopathy consisted of retinal venous tortuosity (occasionally arteriolar tortuosity), a silver wire appearance of the retinal arterioles, CRAO or CRVO with haemorrhages, angioid streaks, salmon-colored patches. Proliferative retinopathy consisted of retinal artery occlusions, choroidal vascular occlusions, retinal haemorrhages, neovascularization.

Statistical analysis.

Chi-square and Fisher's exact test were performed for non-parametric data and Student's *t*-test for parametric data. The frequency of a specific type of lesion was defined (was determined) by the development of such a lesion in one or both eyes. When two eyes of the child were discrepant in the presence of a lesion, the frequency (grade) assigned for the child was that of the more severely involved eye.

RESULTS

Age and sex distribution.
Details of age and sex distribution are shown in Table 1. Of the 66 children examined, 38 (58%) were boys (mean age±SD, 8 years±4) and 28 (42%) were girls (mean age±SD, 8 years±4).

Subjective ocular symptoms.
None of 66 patients presented with subjective ocular symptoms. Sixty-one children had normal vision and five had mild ametropia.

Ocular findings.
Ocular abnormalities were found in 47 (71%) children of 66 subjects. Conjunctival signs were present in 32% of children (21 out of 66 children), retinal vascular tortuosity in 29% (19 out of 66 children), retinal vascular dilatation in 26% (17 out of 66 children), arteriovenous crossing in 29% (19 out of 66 children). One child presented arteriolar narrowing and none presented an arteriolar wire silver aspect. None of the ocular abnormalities was associated with gender except the retinal vascular tortuosity which showed a higher frequency in girls (12 out of 28 girls, 43%) than in boys (7 out of 38 boys, 18%) (chi-square = 4.70, $P = 0.03$). All lesions increased with age.

DISCUSSION

Sickle cell disease causes severe systemic complications such as painful crises and severe haemolytic anaemia. Ocular complications, however, are usually mild and asymptomatic. A variety of ocular changes have been described in patients with sickle cell disease. These changes result from vasoocclusion (vascular occlusions caused by sickled erythrocytes and from the increased adhesion of these cells to the vascular endothelium) and secondary tissue changes in all the vascular structures of the eye, including the conjunctiva (conjunctival sickling sign) (1,12,14,15), the iris (iris atrophy), the choroid (occlusion of the posterior ciliary vessels), the optic disc, and the retina (5). The constellation of retinal abnormalities observed in these patients constitutes the retinopathy of sickle cell disease. The retinal findings are usually classified as proliferative or nonproliferative (5). Nonproliferative retinal changes consist of retinal venous tortuosity, occasionally arteriolar tortuosity, a silver wire appearance of the retinal arterioles, CRAO or CVO with retinal haemorrhages, angioid streaks (11), salmon-colored patches, refractile iridescent deposits or yellow deposits within the retina, disc-shaped hyperpigmented lesions, often with bone spicule or stellate pigment configuration at the borders, and optic atrophy (5).
In this study, ocular abnormalities were found in 47 (71%) children of 66 subjects. Ophthalmologic abnormalities included conjunctival signs (32%), retinal vascular tortuosity (29%) and dilatation (26%) and AV crossing (29%). The proportions of ocular manifestations in children with homozygous sickle cell disease found in our study support the findings of other African studies (3,10,13). These studies have identified a low proportion of retinal changes when compared to those of other studies in Jamaica (1,2,17) and the U.S.A. (5,7,12,18). Studies in Africa didn't describe proliferative retinal changes. It is possible that ocular findings in homozygous sickle cell disease are different in different populations. There is a need for larger studies to further clarify the ocular abnormalities in sickle cell disease. It is already planned to study this question.

Table 1: *Age and sex distribution of 66 children with homozygous sickle cell disease.*

Age (years)	Number of children		
	Boys	Girls	Total (%)
2 - 10	23	18	41 (62)
11 - 18	15	10	25 (38)
Total	38	28	66 (100))

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Request for reprints: Kaimbo Wa Kaimbo D.,
BP 16540 Kinshasa 1
Democratic Republic of Congo