OUTER ORBITAL DISTANCE, INNER CANTHAL DISTANCE AND INTERPUPILLARY DISTANCE, PROPTOSIS IN CHILDREN WITH HOMOZYGOUS SICKLE CELL DISEASE

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ABSTRACT

Background: On basis of clinical observation paediatricians in Kinshasa had the impression that children with homozygous sickle cell disease have a special face characterised by hypertelorism.
Objective: The purpose of the study is to determine outer orbital, inner canthal and interpupillary distances as well as proptosis in children with sickle cell disease.
Methods: These measurements were performed on 66 Congolese children with homozygous sickle cell disease, aged from 2 to 18 years. The measurements were performed with the Hertel exophthalmometer for the proptosis and the outer orbital distance, with the pupillometer model PD-2 meter for the interpupillary distance and with a ruler for the inner canthal distance. The results were compared with those of 95 healthy children of similar age.
Results: All measurements were age related. In every age group the values for inner canthal distance were identical to those of healthy children, but the interpupillary, the outer orbital distances and the proptosis were significantly smaller.
Conclusions: A slow growth of orbital tissue in children with homozygous sickle cell was suggested to explain the difference with healthy children. Other biometric studies comparing the orbital measurements with the rest of the body are needed to confirm or refute this hypothesis.

RESUME

Proposition: Sur base de l'impression clinique, à Kinshasa les pédiatres ont eu l'impression que les enfants drépanocytaires homozygotes présentent une physionomie spéciale avec tendance à l'hypertélorisme.
But: La distance intercanthale, interorbitaire et interpupillaire et la saillie oculaire ont été mesurées chez l'enfant drépanocytaire homozygote.
Méthodes: Ces mesures ont été effectuées chez 66 enfants congolais (34 garçons, 32 files) âges de 2 à 18 ans. La distance interorbitaire externe et la saillie oculaire ont été mesurées à l'aide de l'exophtalmomètre de Hertel, la distance intercanthale à l'aide d'une simple réglette graduée et la distance interpupillaire à l'aide du pupilomètre Topcon PD modèle PD-2. Toutes ces mesures ont été comparées à celles de 95 enfants congolais normaux de même âge.
Résultats: Toutes les mensurations augmentent avec l'âge. Pour chaque groupe d'âge, la distance intercanthale montre que les valeurs des enfants drépanocytaires homozygotes sont superposables à celles des sujets normaux. Les valeurs des distances interorbitaires et interpupillaires et de la saillie oculaire des enfants drépanocytaires homozygotes sont inférieures à celles des enfants normaux.

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Conclusion: Une faible croissance de tissu orbitaire est suggérée pour expliquer cette différence entre les enfants drépanocytaires homozygotes et les enfants normaux. Une très large étude basée sur la biométrie de l'orbite et du reste du corps est entreprise pour confirmer ou réfuter cette hypothèse.

KEY WORDS

Sickle cell disease. Outer orbital distance. Inner canthal distance. Interpupillary distance. Proptosis.

MOTS CLES

Hémoglobinopathie SS. Anomalies oculaires. Distance interorbitaire externe. Distance intercanthale. Distance interpupillaire. Saillie oculaire.

INTRODUCTION

Normal values for the proptosis, the outer orbital distance, the interpupillary distance and the inner canthal distance have been previously reported in normal healthy Congolese (1,2). Those studies showed that values of healthy Congolese were higher when compared with those of Caucasian subjects. It was suggested that those values must be taken into account in interpretations of orbital and eye diseases and craniofacial deformities for Congolese people.

On the basis of clinical observations, paediatricians in Kinshasa suspected that children with homozygous sickle cell disease develop hypertelorism. Valid evidence to support this view is lacking. Therefore, a prospective study was undertaken to study biometric parameters in the orbit in children with homozygous sickle cell disease. The purpose of the present article is twofold:

(1) measure the position of the eyes in the orbits in homogenous sickle cell children;
(2) compare these values with those in healthy normal Congolese children.

SUBJECTS AND METHODS

POPULATION

From March 1, 1998 through August 31, 1998, 78 children with homozygous sicklecell disease (ages ranged from 1 to 18 years) were entered into this prospective study. These children were referred to us for eye examination by the department of Paediatrics, University of Kinshasa and the "Sickle Cell anaemia Center" of the county of Yolo in Kinshasa. Twelve children were excluded from the study because of poor collaboration. For this study 66 children were considered. Of the 66 children studied, 34 (51.5%) were boys (mean age ± SD, 8.25 years ± 3.78) and 32 (48.5%) were girls (mean age ± 8.17 years ± 3.98). No statistically significant difference was found between the two sexes for mean age ($P < 0.05$; $t = 0.08$, $df = 64$). Children were divided into four age groups and ranged in age from 2 to 6 years (average age ± SD, 4.07 years ± 1.08) in

Tab. I:*Patient characteristics*

Group	Age group (years)	N	Mean age	Standard deviation
1	2-6	30	4.07	1.08
2	7-10	11	8.18	0.63
3	11-14	17	11.9	0.83
4	15-18	8	16.00	0.75

group 1, from 7 to 10 years (average age ± SD, 8.18 years ± 0.63) in group 2, from 11 to 14 years (average age ± SD, 11.9 years ± 0.83) in group 3, and from 15 to 18 years (average age ± SD, 16 years ± 0.75) in group 4 (Table I). None of the children had orbital disease, trauma, surgery, endocrine disease, myopia of more than 5 diopters, buphthalmos, anophthalmos, craniofacial anomalies or any obvious external ocular disease.

We performed the same measurements on 98 healthy children of similar ages. The results have been previously published (2).

MEASUREMENTS

The measurements were performed (1,2) with the Hertel exophtalmometer for the proptosis and the outer orbital distance, with the pupillometer model PD-2 meter for the interpupillary distance and with a ruler for the inner canthal distance. All measurements were made by one of us (KWK.D). For each subject, the mean value of three consecutive measurements was considered as the representative measurement in this study.

STATISTICAL ANALYSIS

Values are given in mm as mean ± standard deviation. Data were tested for significance by means of the *t*-test for unpaired data for continuous variables. $P > 0.05$ was considered significant.

Tab. II: *The outer orbital distance in 66 Congolese children with homozygous sickle cell disease*

Age groupe (years)	Boys			Girls			Both		
	N	Mean	SD	N	Mean	SD	N	Mean	SD
2-6	16	100.1	3.1	14	97.0	2.9	30	98.7	3.2
7-10	5	103.2	1.9	6	102.8	2.2	11	103.0	2.0
11-14	9	105.7	3.3	8	105.5	2.5	17	105.6	2.9
15-18	4	114.2	4.2	4	110.0	2.3	8	113.0	3.0

Tab. III: *The inner canthal distance in 66 Congolese children with homozygous sickle cell disease*

Age groupe (years)	Boys			Girls			Both		
	N	Mean	SD	N	Mean	SD	N	Mean	SD
2-6	16	25.8	1.6	14	26.5	1.6	30	26.1	1.6
7-10	5	28.0	3.2	6	29.5	2.5	11	28.8	2.7
11-14	9	31.2	2.6	8	30.1	2.6	17	30.7	2.7
15-18	4	32.0	2.5	4	31.5	1.2	8	32.1	1.4

Tab. IV: *The interpupillary distance in 66 Congolese children with homozygous sickle cell disease*

Age groupe (years)	Boys			Girls			Both		
	N	Mean	SD	N	Mean	SD	N	Mean	SD
2-6	16	51.4	4.8	14	51.4	3.6	30	51.4	4.2
7-10	5	57.0	2.0	6	51.2	6.8	11	53.8	5.5
11-14	9	56.3	4.4	8	55.8	5.1	17	56.1	4.8
15-18	4	63.0	3.0	4	61.8	1.8	8	63.9	2.2

Tab. V: *The proptosis in 66 Congolese children with homozygous sickle cell disease*

Age group (years)	Right eye								
	Boys			Girls			Both		
	N	Mean	Sd	N	Mean	SD	N	Mean	SD
2-6	16	13.7	2.3	14	12.8	1.6	30	13.3	2.1
7-10	5	13.2	1.3	6	15.0	1.0	11	14.2	1.1
11-14	9	13.8	1.8	8	15.4	1.8	17	14.5	1.8
15-18	4	16.2	0.8	4	18.3	1.3	8	17.2	1.3

Age group (years)	Left eye								
	Boys			Girls			Both		
	N	Mean	SD	N	Mean	SD	N	Mean	SD
2-6	16	13.6	1.9	14	12.9	1.8	30	13.3	1.9
7-10	5	12.8	2.0	6	15.0	1.3	11	14.0	1.4
11-14	9	12.9	1.9	8	15.1	1.7	17	13.9	2.1
15-18	4	12.2	1.2	4	18.3	1.4	8	17.4	1.4

Tab. VI: *The outer orbital, inner canthal and interpupillary distances in healthy and sickle cell Congolese children*

Age group (years)	The outer orbital distance						The inner canthal distance					
	Sickle cell disease			Healthy children			Sickle cell disease			Healthy children		
	N	Mean	SD	N	Mean	SD	N	Mean	SD	N	Mean	SD
2-6	30	98.7	3.2	25	100.0	4.4	30	26.1	1.6	25	27.4	2.7
7-10	11	103.0	2.0	37	106.5	4.6	11	28.8	2.7	37	29.7	3.1
11-14	17	105.6	2.9	22	111.7	6.8	17	30.7	1.7	22	30.0	2.4
15-18	8	113.0	3.0	11	118.5	6.4	8	32.1	1.4	11	32.2	3.1

Age group (years)	The interpupillary distance						The proptosis (Right eye)					
	Sickle cell disease			Healthy children			Sickle cell disease			Healthy children		
	N	Mean	SD	N	Mean	SD	N	Mean	SD	N	Mean	SD
2-6	30	51.4	4.2	25	56.1	3.8	30	13.3	2.1	25	14.4	2.2
7-10	11	53.8	5.5	37	61.2	3.6	11	14.2	1.1	37	15.4	3.2
11-14	17	56.1	4.8	22	65.5	2.9	17	14.5	4.8	22	16.6	1.8
15-18	8	63.9	2.2	11	69.2	3.9	8	17.2	1.3	11	17.6	2.2

RESULTS

Tables II, III, IV and V give the mean ± SD values of outer orbital distance, the interpupillary distance, the inner canthal distance and the proptosis. All measurements were aged related ($P < 0.05$). No significant difference was found between the two sexes for all measurements. There was no significant difference between right and left eye for the proptosis ($P < 0.05$) (Table V).

THE OUTER ORBITAL DISTANCE

The mean ± SD outer orbital distance varied from 98.67 mm for the first age group to 113.00 mm for the fourth age group. There was no significant difference ($P > 0.05$) between boys and girls in all but the youngest (group 1) age categories.

THE INTERPUPILLARY DISTANCE

The mean interpupillary distance varied from 51.4 mm for the first age group to 63.88 mm for the fourth age group. Although average values for boys were higher, there was no significant difference ($P > 0.05$).

THE INNER CANTHAL DISTANCE

The mean inner canthal distance ranged from 26.13 mm for the first age group to 32.125 mm for the fourth age group. No statistically significant difference was found between boys and girls for the average inner canthal distance in all age groups ($P > 0.05$).

THE PROPTOSIS

The mean proptosis varied from 13.27 mm (group 1) to 17.25 mm (group 4) for the right eye and from 13.27 mm (group 1) to 17.38 mm (group 4) for the left eye.

DISCUSSION

We studied the outer orbital distance, the interpupillary distance, the inner canthal distance and the proptosis in children with homozygous sickle cell disease and evaluated the relative effects of age and gender.

The results of this study were compared with those of healthy Congolese children of the same age, examined with identical methods by the same investigators. Their results have been published previously (2). In Table VI a comparison between results in healthy and diseased children is shown.

Values of the inner canthal distance in children with homozygous sickle cell are similar to those of healthy children in every age group. For children with homozygous sickle cell disease the values of the interpupillary and outer orbital distance, and the proptosis are smaller than those of healthy children in all ages groups. Differences are more pronounced in the age groups 7-10 years and 11-14 years.

Results in children with homozygous sickle cell disease suggest a slow growth of orbital contents giving the paediatric clinical impression of hypertelorism.

Other studies comparing biometric orbital data with those of the rest of the body (body mass index, cranial measurements) in these children have been started in Kinshasa to support or refute this hypothesis.

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