



Research Article

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Prevalence of sickle cell disease in newborns in the Yaounde Central Hospital

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Abstract

Introduction: Sickle cell disease (SCD) in Africa is associated with very high incidence and morbidity and mortality rates, especially for children aged 0-5, yet reliable up-to-date information is lacking. Significant reduction in mortality rates has moreover been shown to be achieved with early neonatal diagnosis, followed by early management of affected children, as recommended by the United Nations. This is yet to be implemented in our context, thus underscoring the importance of well researched and published data on sickle cell disease neonatal prevalence. **Method:** We conducted a cross-sectional descriptive study at the Yaoundé Central Hospital maternity from January 2016 to July 2016. Blood samples were collected from the heel of new-born babies and analysed by high liquid performance chromatography. Results were made available to parents and affected cases were orientated for follow-up by a specialist. **Results:** A total of 703 new-borns were included out of which 9 babies had SCD making a prevalence of 1.27; 5 (0.75%) were homozygous SS and 4 (0.52%) were compound heterozygotes for HbS/ β^+ Thalassemia. One-hundred and eighteen new-borns (16.8%) were AS and 576 (81.9%) AA. Prevalence of the S gene was 18.2% (127). Sex distribution was 366 males and 337 females. No correlation was found between anthropometric parameters and abnormal haemoglobin genotype except for the head circumference. **Conclusion:** Prevalence of SCD in new-borns is high. Neonatal screening should be extended to the rest of the territory with methods adapted to the local population.

Keywords: Sickle cell disease, New-borns, Prevalence, Yaoundé Central Hospital.

INTRODUCTION

Sickle cell disease (SCD) is a group of inherited red blood cell disorders which are characterised by the presence of an abnormal haemoglobin called sickle haemoglobin (HbS) [1]. Sickle cell disorders are associated with severe clinical manifestations exception made for the haemoglobin A and haemoglobin S heterozygous coexistence, also called sickle cell trait (SCT), which generally has mild clinical expressions. The most common form of SCD in Africans is the homozygous HbS disease (HbSS), an autosomal recessive disorder first described by James Herrick in 1910 [2]. Besides this are the heterozygous composite forms namely; HbS/HbC, HbS/ β^0 thalassemia, HbS/ β^+ thalassemia HbS/HbD and HbS/HbO for the most frequent.

United Nations (UN) in 2008 estimated over 20 to 25 million people affected with SCD in the world, 60% of these lived in Sub-Saharan Africa. Recent estimates suggest that 300.000 children are born with SCD worldwide every year and that 3/4 of these births occur in Sub-Saharan Africa [3]. Besides the very high prevalence rates, Africa is equally faced with high morbidity and mortality rates as over 50 to 70% of deaths before 5 years of age [4]. SCD is thought to target the black population, from where it is thought to have emerged as an adaptation to endemic malaria spreading worldwide after migration [5]. Prevalence of the SCT in Cameroon is 21.6% [6]. Similar prevalence rates are found in other West African countries namely; Benin, Ghana, Burkina Faso and Nigeria with 20-30% prevalence rates; 40% in certain regions of Uganda and 30% in the Democratic Republic of Congo (DRC) [3]. The alarming situation, added to the high socio-economic burden of SCD led to its recognition as a public health issue by UN in 2008 [7]. A number of policies were there after stated, targeting secondary prevention by identification of affected babies at birth and subsequent early management [8].

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Neonatal screening for SCD alongside early follow-up of patients is associated with a marked reduction of morbidity and mortality rates [9, 10]. Implementation of this approach which is attributed a major role in the fight against SCD has been done in several countries worldwide. Nigeria, Benin, Burkina Faso and the Democratic Republic of Congo, have researched and published neonatal prevalence rates ranging from 1.32 to 3% [11, 12, 13, 14]. Such data are inexistent in our Cameroun context hence emphasising the importance of our study which will make it possible to carry out an early detection of sickle-cell anemia in newborns born at the Yaoundé Central Hospital.

METHODOLOGY

We conducted a cross-sectional descriptive study at the Yaoundé Central Hospital (YCH) maternity from January 2016 to July 2016. All babies of the YCH maternity who were born during our study period were included in the study upon mother's approval. Sampling was consecutive.

We included new-borns aged 30 weeks to 43 weeks gestational age before delivery. Mothers were approached a day after delivery for consent. After complete physical examination of the new-borns including the weight, height, head and mid-upper arm circumference we proceeded with filling of our data questionnaire. Blood samples were hereafter collected from the heel of new-borns using a lancet needle and placed as blots on a whatmann paper, appropriately labelled for each participant.

After collection, samples were placed in polythene envelopes before transfer to the laboratory of the Yaoundé Central Hospital where they could be stored at 2-8°C, if necessary, for a maximum of 3 days. Analysis were done by High Liquid Performance Chromatography. Babies with an FA(AA) pattern result were considered normal, FAS(AS) patterns were considered carriers, and FS (SS) or FSA patterns were considered to have SCD: FS were homozygous sicklers (SS) and FSA were heterozygous S/β+thalassemia. SPSS version 20.0 was used for data entry and analysis.

RESULTS

We approached 1030 mothers for participation in the study. Out of these, 676 mothers approved participation leading to the inclusion of 703 babies. Acceptability rate was 65.63%. Of the babies included, 52% (365) were males and 48% (337) were females giving a sex-ratio of 1.08. We obtained different genotypes of haemoglobin by high performance liquid chromatography (Table I).

Table 1: Distribution of haemoglobin genotype of new-borns as obtained with high performance liquid chromatography

Chromatography Results	Frequency (N)	Percentage (%)
FA	576	81.9
FAS	118	16.8
FS	5	0.75
FSA ₂	4	0.52
Total	703	100

Key: FA = Haemoglobin F/ haemoglobin A
 FAS= Haemoglobin F/ haemoglobin A/ haemoglobin S
 FS = Haemoglobin F/ haemoglobin S
 FSA₂= Haemoglobin F/ haemoglobin S/ haemoglobin A₂

Out of the 703 new-borns we included in our study, 9 were found to have SCD, amongst which 5 (0.7 %) were homozygous for sickle cell anaemia and 4 (0.6 %) had an association of HbS and β+ thalassaemia mutations. 118 (16.8%) of babies were AS and 576 (81.9%) were AA. Prevalence of SCD in our study population was hence estimated to be 1.27%.

We recorded the anthropometric data of the newborns (Table 2) and we analyzed them to see their relevance according to the type of hemoglobin

Table 2: Distribution of the anthropometric profile of new-borns

Anthropometric parameters	Mean	Minimum	Maximum
Weight (g)	3202.62±588.47	1690	5000
Height (cm)	49.82±1.47	46	55
Head circumference (cm)	33.88±1.23	31	37
Brachial circumference (cm)	11.32±0.85	7	13

Analysis of the anthropometric profile of babies showed that majority of babies included in our sample weighed 3202.62±588.47g, ranging from 1690g to 5000g. The tallest baby measured 55cm while the smallest measured 46cm and the mean height was 49.82±1.47 cm. The head circumference of new-borns ranged from 31cm to 37 cm and brachial circumference 7 cm to 13 cm, with means 33.88±1.23 cm and 11.32±0.85 cm respectively. It should be noted that we included in our study sample, babies of 30 to 42 weeks gestational age before delivery (Table 3).

Table 3: Distribution of anthropometric parameters with respect to new-born haemoglobin electrophoresis

Anthropometric parameters	Chromatography result				p-value
	FA	FAS	FS	FSA	
Mean weight (g)	3207.24	3192.54	2672.00	3410.00	0.29
	±611.54	±453.26	±708.99	±220.00	
Mean height (cm)	49.80	49.98	48.80	50.25	0.39
	±1.60	±1.52	±1.78	±0.50	
Mean head circumference (cm)	33.93	33.66	32.80	33.75	0.04
	±1.26	±1.07	±1.30	±1.50	
Mean brachial circumference (cm)	11.30±	11.41±	10.60	11.75	0.56
	0.85	0.72	±2.07	±0.50	

We went further to search for a correlation between anthropometric profile of babies and their chromatography results, so as to determine a possible influence of an abnormal haemoglobin genotype on the weight, height as well as the head and brachial circumferences of new-borns. The table above shows that we found no statistically significant difference between anthropometric parameters and chromatography results, except for the head circumference (p-value=0.04) as babies with abnormal haemoglobin profiles on chromatography tend to have smaller heads than those with FA patterns.

DISCUSSION

Out of the 703 babies we screened during our study, 9 were suspected to have SCD making a prevalence of 1.27%; 118 (16.8%) babies were AS and 576 (81.9%) were AA. Five (0.75%) of the new-borns with SCD were homozygous SS and 4 (0.52%) were heterozygous composite for HbS/β+ Thalassaemia. This prevalence is similar to that obtained by Tshilolo *et al* in Congo (1.4%), Nga Motaze *et al* in Centre Hospitalier d'Essos Cameroon (0.8%) and Kafando *et al*, Burkina Faso (1.75%), who have equally invested on neonatal screening for SCD [11, 13, 14]. Our sample was larger than that of Nga Motaze *et al* and so could account for the higher prevalence we got in our study. In addition to this, we came to realise that educational level was a huge promoting factor to the widespread of SCD as the major route of information on the

disease in our context is via lessons, which are only taught in high school. Contrary to Centre Hospitalier d'Essos which is attended by patients of the medium to high social class, Yaoundé Central Hospital is attended by patients of low economic background who are furthermore often illiterates and as such have never heard of SCD [13]. Ignorance about the existence of this pathology can explain that many parents are often unaware of their haemoglobin genotype hence increasing the probability of having a sick child or carrier and as such could account for the relatively higher prevalence [13].

Results are expressive and reflect that new-borns are still delivered with SCD despite the seemingly tough efforts of health authorities. With regards to the recommendation of The World Health Organisation which recommends the development of disease-specific programs when birth rate of affected infants is above 0.5 per 1.000 births, we can conclude that our prevalence rate is high and requires appropriate control measures [1]. Looking at our country Cameroon in particular, we notice that comparisons between this prevalence rate and those obtained several years back, points out a non-negligible reduction in SCD incidence rate, as we have literally observed a 4.8% decrease in the SCT prevalence rate, relative to the 21.6% rate in the past [5]. This could be as a result of the systematic request of a haemoglobin electrophoresis exam during antenatal visits and increased awareness of the existence of SCD by the general population over the years, even though the latter remains unsatisfactory and as such requires to be improved and upgraded, as pointed out by Bazuaye *et al* [15]. SCD prevalence rates in neighbouring Democratic republic of Congo and Burkina Faso averages 30% and corresponds to the relatively higher result they obtained during neonatal screening, added to the endogamy marriage system practiced in Congo [11, 14].

Anthropometric parameters of weight, height, head and mid-upper arm circumference of new-borns delivered during our study period were normal for gestational age. Mean weight was 3202.62±588.47g (1690-5000g), mean height 49.82±1.47 cm (46-55cm), mean head circumference was 33.88±1.23cm (31-37), mean mid-upper arm circumference was 11.32±0.85cm (7-13cm). Male sex was preponderant over females with a sex ratio of 1.08, akin to results found by Nga Motaze *et al*, in 2013 [13]. Similarities were observed with results on anthropometric parameters published by Munyanganizi *et al*. in Rwanda [16]. On comparing the anthropometric parameters of babies with the SCT gene to those with normal electrophoresis results, we found no statistically significant difference but for head circumference (p=0.04) which tend to be smaller in babies with the S gene. The anthropometric measurements of SCD affected new-borns may nevertheless be affected as they grow. More emphasis is brought by Sadarangani *et al* who demonstrated that children with SCD generally have low anthropometric parameter measurements compared to non-SCD children. Significant differences are found with the weight-to-age and mid-upper arm to age measurements of children (aged 0.8 to 13.7years) compared to non SCD children of the same age, often due to infections, anaemia and nutritional deficiencies [17].

CONCLUSION

At the end of the study we note that: Prevalence of sickle cell disease is high.

There is no relation between new-borns' anthropometric parameter measurements and abnormal haemoglobin genotype results except for the head circumference.

Mothers are mostly uneducated and awareness of sickle cell disease is low.

Early detection of sickle-cell anemia enables rapid and adequate management and thus a better quality of life.

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