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Néonatal screening the drepanocytosis and Glucose 6 Phosphate Deshdrogenase deficiency in Kinshasa

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Abstract
Objectives: The objective of this work was to carry out the neonatal tracking of the drépanocytose and the deficit in G6PD in order to establish the prevalence of these pathologies in the town of Kinshasa.
Patients and Methods: We worked with 4 maternities chosen by the national program of fight against the drépanocytose (PNLD).). The samples were taken on blotting paper. The tracking of the drépanocytose realized by Iso-focusing related to 6540 blood samples of the new-born babies.
1350 samples drawn dice samples having been used for tracking from the drépanocytose were used for the tracking of the deficit in G6PD with the méhtode of fluorescence of Beutler.
Results: The tracking of the drépanocytose revealed that 1.7% of the new-born babies had the homozygous form of the disease and 15.3% had the heterozygous form. The tracking of the deficit in G6PD carried out showed a prevalence of 6.3% including 1.1% for the girls and 5.2% for the boys.
Conclusion: The prevalence of the drépanocytose and the deficit in G6PD in this work is high. We suggest the installation of a systematic tracking through the town of Kinshasa accompanied by a sensitizing of the population. Also, we ask to proscribe the regulation of certain drugs at the overdrawn ones in G6PD. The Congolese government should require manufacturers of the drugs before any marketing of the data on the activity of the G6PD of all new active ingredients.

Keywords: drepanocytose, deficit in G6PD, neonatal, tracking, prevalence

Introduction

Several hereditary pathologies can affect the integrity of the red globule, like the hémoglobinopathies, the membrane enzymopathies and anomalies [1, 2, 3]. the neonatal tracking of some of these pathologies proves to be crucial in order to take preventive measures in order to reduce the clinical complications which are dependent there.
The drépanocytose and the deficit in Glucose 6 Phosphate Déshydrogénase (G8PD, EC1.1.1.49) dyd were selected to be part of this study and this taking into account their frequency raised throughout the world and that they are detectable by reliable tests, inexpensive and applicable on a large scale [2, 4].
The drépanocytose, which is one of the hémoglobinopathies more rependue, constitutes public health problems in certain areas of the world.
As a Democratic republic of Congo, the frequency of the allele S can reach 30% to 40% in certain areas [2, 3]. the neonatal tracking of the drépanocytose will make it possible to set up strategies of early assumption of responsibility having for goal, at the patients drépanocytaire, a significant reduction of their mortality and their morbidity [5, 6, 7]. the neonatal tracking of the deficit in G6PD, the most frequent enzymopathy érythrocytaire in certain ethnos groups, make it possible to take precautions as for the use of certain oxidizing medications responsible for haemolytic crises at the overdrawn individuals in this enzyme [8, 9].
A systematic neonatal tracking of the drépanocytose realized in certain maternities of the province city of Kinshasa was set up by the laboratory of Biochemistry-Hematologic of the pharmaceutical Faculty of Science of the university of Kinshasa with an aim of establishing the incidence of this pathology at the new-born babies. With that, we associated the neonatal tracking of the deficit in G6PD.

Equipment and methods

It is within the framework of an inter-University Cooperation program that a project of fight against the drépanocytose was set up. This program enabled us to make a systematic tracking of the drépanocytose in some maternities of the town of Kinshasa. This tracking was carried out in maternities of the province city of Kinshasa located in various communes.
The annual number of birth on average in these maternities was of 400 in Bondeko, of 1300 in Kintambo, 850 in Ngaba and of 1800 in Bomoi. In Kinshasa, we have 210,000 births in general a year and in DRC, a figure of at least two million birth was advanced by the national plan of the health of the reproduction.
The choice of maternities was established according to the national program of fight against the drépanocytose and this in order to avoid the overlapping with other structures working for the same objective. The blotting papers one provided to maternities by the public health clinic, the collection of the taken samples was carried out to the maximum 1 days after the taking away. After analysis, the results were returned to maternities the next day
The results returned to maternities were accompanied by two

cards, a general card gathering the results of all the detected children and the individual record sheets to give to the moms of each detected child. A Committee of wise consisted each hospital, was in charge of the handing-over of the results. The assumption of responsibility of the diagnosed patients drépanocytaires intervened three months after birth by pediatrists also trained by the Project.

On the whole 6540 capillary blood samples (heel) of the new-born babies collected on blotting paper was used for the tracking of the drépanocytose. Among the 6540 taking away, 1350 samples were used for the neonatal tracking of deficit in G6PD. The quantity of blood on the blotter had not made it possible to work on all the samples. The blotting papers were preserved at the laboratory between 2 - 8 °C.

The Tracking of the drépanocytose was made by an isoelectric technique of focusing [10]. A test of confirmation per electrophoresis with acid pH (SEBIA System electrophoresis on acetate) was carried out on the same sample for all the new-born babies detected like drépanocytaire (homozygous for HbS) [11].

The Tracking of the deficit in G6PD was carried out by the test of fluorescence. This qualitative method is based on the observation of the fluorescence of the cofactor reduces NADPH [12]. In short, a blotting paper spot is introduced into a tube containing 50 µl of the solution of 1 ml Glucose 6P 0.1 M, 1 ml NADP 0,0075M, 2 ml Saponin 1%; 3 ml Sorting HCl buffer pH 7.8 0,75M; 1ml GSSG 0.008 M; 2 ml H20) and to mix during 1 minute. To deposit 10 µl on a blotting paper and To make the spot after 5 minutes, 10 minutes, 15 minutes of incubation.

The evaluation of the fluorescence of the reduced cofactor is carried out under lamp UV to 254 Nm. The normal samples give a green fluorescence whose intensity of colouring is in keeping with the time of incubation whereas the overdrawn samples in G6PD do not give any fluorescence to all times of incubations [12].

Results

Neonatal tracking of the drépanocytose

The number of children detected of 6540 was distributed as follows between maternities: 1485 in the center mother and children of Ngaba, 2396 with the maternity from KintamBo, 1860 with Bomoi maternity and 799 with the maternity of Bondeko.

The assumption of responsibility should concern all the detected children drépanocytaires, however only the children drépanocytaires pertaining to stripped families were dealt with. It sied to announce that only one case of death had been recorded. It is only case is allotted to nonthe adherence of the parents of the patient to the assumption of responsibility. The results are presented in table I.

Table 1: Results of the tracking of the drépanocytose by Maternity

Maternités	AA(%)	AS(%)	SS(%)	Total
CME- NGABA	1198 (18,3)	255(3,9)	32(O, 5)	1485
KINTAMBO	1933(29,6)	422(6,5)	41(0, 6)	2396
BOMOI	1618(24,7)	211(3,2)	30(0, 5)	1860
BONDEKO	681(10,4)	109(1,7)	9(0, 1)	799
Total	5430(83)	997(15,3)	112'(1,7)	6540

Neonatal tracking of the deficit in G6PD

The neonatal tracking of the deficit in G6PD was carried out on 1350 samples obtained starting from the samples used for the neonatal tracking of the drépanocytose. Among the 1350 new-born babies having profited from a qualitative analysis of the activity of the G6PD, 6,3% were shown like defective in this enzyme. In this population tested, 1.1% (15/1350) and 5.2% (70/1350) of the female and male new-born babies respectively of sex were defective in G6PD

Discussion

With an incidence of new-born babies drépanocytaires of 1.7%, our data collected on a population of 6540 new-born babies are similar to those found in the literature (Burkina Faso-Faso 1.6%) (13) and with those (1.4%) established by Tshilolo *et al*. as a Democratic republic of Congo on a population of 31,204 new-born babies (14). In this study, tracking was carried out on the blood of the cord while the present study used capillary blood. The blood of cord does not pose any problem especially when the taking away are carried out by wise women trained for this kind of activity. What makes it possible to avoid the contamination of the blood of the baby by that of the mother. On this precise point, the capillary blood taken on the heel does not require a particular formation. Although the mode of taking away is not identical, the results are concordant. A cost reduction D ` analyzes was carried out in our study by using reagents prepared locally (solutions and freezing for IEF) making it possible to more reduce the cost of the analysis to less than 2.5$.

The results were transmitted one day after analysis by the person in charge selected to the level of the hospital. For new the, born ones detected like drépanocytaires, the hospitals were contacted by telephone as of obtaining the résulatt in order to or not obtain a second making it possible to confirm the results. In the study of Tshilolo and al. the confirmation was done at least three months after the birth. The strategy installation by our tracing routine made it possible to reduce the number of lost of sight, The confirmed children drépanocytaires were dealt with by pediatrists trained within the framework of the project and referents in each center concerned with the study. This assumption of responsibility consisted of free consultations in the event of the crises, exemption from payment of the drugs prescribed thus that antibiotic disease prevention (penicillin) and of the antimalarial ones.

We analyzed 6540 samples for the neonatal tracking of the drépanocytose, these results show that 15.3% of the new-born babies are heterozygous for HbS and 1.7% of the new-born babies are homozygous for HbS. The prevalence of the homozygous form equivalent to that of 1.4% obtained in a previous study carried out as a Democratic republic of

Congo On a sample of 31,204 new-born babies [14].

A systematic tracking, carried out well under our work conditions with some hospitals, supported by an adequate program of assumption of responsibility is able to decrease morbidity and mortality related to this maldie but for that we do not have a figure. A training of professionals proves to be essential in this assumption of responsibility.

An estimate of the incidence of the deficit in G6PD was given starting from a troop made up of 1350 new-born babies. She is of 6.3% is 1.1% from the girls and 5.2% among boys. They are the first recent data available for the area of Kinshasa. These results are higher those observed than Rwanda (3.8%) [15], east of Guinea (5.7%) [16]. The knowledge of this incidence in our population, enables us to take precautions in the use of certain drugs whose consumption supports the installation of an oxydative stress de facto involving a state of anaemia. That justifies the founded good of this study which confirms the thesis according to which these anomalies érythrocytaires (drépanocytose, deficit in G6PD) developed in paludous zones of which the DRC.

The deficit in G6PD generally appears as an acute haemolytic anaemia with all the symptoms of anaemia and often of the abdominal pains. The deficit in G6PD must always be taken into account with regard to the etiologies of anaemias as a Democratic republic of Congo. The oxidizing drugs must be proscribed with all the defective children because of their harmful effect on the red globule [8]. Among these drugs, we can quote certain antibiotics of the acid type nalidixic and unquestionable antalgic of noramidopyrine type: metamizole sodic and sulfasalazine [17].

In-outside haemorrhages, the assumption of responsibility of acute anaemias in DRC is limited primarily to paludism. Nevertheless other pathologies, like the drépanocytose and the deficit in G6PD, can be the cause of those. In addition, a crisis of paludism passes by certain treatments which can worsen anaemia if the patient presents a deficit in G6PD. Our study shows once again that the drépanocytose is a frequent pathology in DRC as well as the deficit in G6PD.

Acute anaemias are regarded in general as a crisis of paludism and are treated by blood transfusions but the transfusional security is not yet optimal, in particular in DRC.

Conclusion

In conclusion, in order to avoid any useless or inadequate treatment but also to found an adapted assumption of responsibility of the patients drépanocytaires, a generalization of the neonatal tracking of the drépanocytose should be founded. Moreover the diagnosis of this affection as well as deficit in G6PD, at least at the time of an acute anaemia on a phenomenon of hémolyse, should be available in the laboratories of clinical biology in DRC. For the G6PD, sight the prevalence (6.3%), the Congolese government should require manufacturers of the drugs before any marketing of the data on the activity of the G6PD of all new active ingredients.

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