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Incidence of Sickle Cell Disease and Other Hemoglobinopathies in Burkina Faso: Results of a Five-Year Systematic Neonatal Screening (2015-2019) in Four Urban Hospitals

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Abstract

Hemoglobinopathies, mainly Sickle cell disease (SCD), are the most common monogenic disorders in Africa. In Burkina Faso, data on these diseases are scarce, mainly hospital-based in Ouagadougou and its surroundings. In order to assess the incidence and allelic frequencies of the main hemoglobinopathies in newborns in Burkina Faso, we conducted a cross-sectional study from 2015 to 2019 in four hospitals. The study included babies of both sexes, regardless of ethnic group and parents' hemoglobin status. It was a newborn screening and hemoglobin variants were detected using isoelectric focusing on cord blood samples and confirmed using hemoglobin electrophoresis by high-performance liquid chromatography. The proportions and cumulative incidences of the different hemoglobinopathies were computed. Hardy-Weinberg equilibrium law was applied to calculate genotypic and allelic frequencies. The significant level was $p < 0.05$. Out of 11,337 newborns included, 47.8% were males and 60.2% were from Bobo-Dioulasso. Abnormal hemoglobin was found in 27.1%, representing a cumulative incidence of 1:4 newborns. The incidence of SCD was 1.9% (1:53 newborns) with 27.9% of homozygous SS. Homozygous CC and compound heterozygous Cβ-Thalassemia accounted for 1.1%. SCD cases

were 1.51 times higher in Bobo-Dioulasso (OR = 1.51; 95% CI [1.09 - 2.10]: p = 0.013). The observed genotype frequencies were significantly different from the expected ones (p < 0.001). The βS and βC alleles represented 5.1 and 9.9%, respectively. This study showed a high incidence of hemoglobinopathies. Such results raise the question of control strategies for these hemoglobinopathies in our country.

Keywords

Abnormal Hemoglobin, β-Globin Gene, Newborn Screening, Isoelectric Focusing

1. Introduction

The inherited disorders of hemoglobin (Hb), mainly sickle cell disease (SCD) and thalassemia, are the most common and worldwide distributed monogenic disorders. Around 7% of the world's populations were assumed to be carriers of a significant hemoglobinopathy mutation [1]. In Sub-Saharan Africa, SCD affects up to 2% of newborns and the prevalence of sickle cell trait (SCT) reaches 40% in some areas [2].

SCD constitutes a group of autosomal recessive β-globin abnormalities with the hemoglobin S-variant (HbS) as a common pathognomonic feature. The anomaly can occur homozygously (βS/βS) leading to the homozygote phenotype HbSS or as a compound heterozygous state (βS/βX) combining the basic mutation βS on one allele with the other allele, one of a number of another abnormal β-globin allele (βX) giving the composite heterozygous phenotypes HbSC, Sβ0- or Sβ+-Thalassemia, SD-Punjab, SO-Arab, etc.) [3] [4].

In SSA, deaths due to SCD mostly occur in children under five years old [3] [5]. Therefore, the WHO declared SCD a global health priority and urged each country to set up a national comprehensive control program [2]. Many studies indicate that effective strategies that contribute significantly to the reduction of morbidity and mortality include, among others, the early diagnosis and treatment of affected subjects through neonatal screening (prevention of 70% of deaths). However, few countries in SSA have such SCD programs. Sometimes, reliable data to support the development of a relevant program is lacking.

In Burkina Faso, the hospital prevalence of all hemoglobinopathies and SCD varied between 44.8% - 41.2% and 8.4% - 12.1% respectively [6] [7], accounting for more than 60% of non-malignant hematological pathology [8]. Among children over two years of age and adolescents in a community study, their prevalence was found to be at 30% and 1.4% respectively [6]. These data are quite partial and concentrated on the city of Ouagadougou and its surroundings. The first study on neonatal screening was conducted sequentially between 2000 and 2004 in the peripheral maternities of Ouagadougou. It is the only one that has made available the most complete and reliable data on hemoglobinopathies that

the country has ever known. Abnormal hemoglobin was found in 27.4% of newborns and SCD occurred in one out of 57 newborns [9].

In this work, we report data from a somewhat larger study on SCD and other significant hemoglobinopathies that may be useful for policy decisions.

2. Material and Methods

2.1. Study Setting and Design

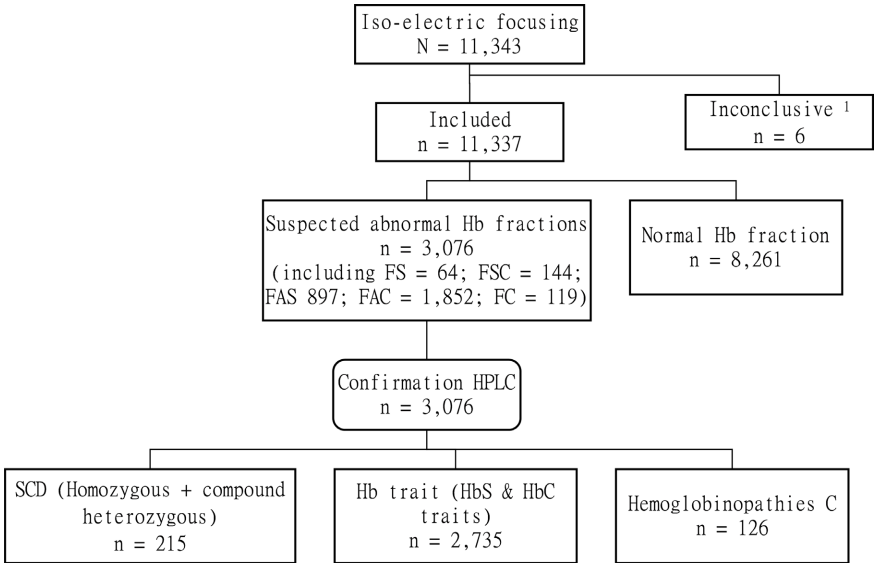
We conducted a cross-sectional study from 2015 to 2019 in Ouagadougou and Bobo Dioulasso, the administrative and economic capitals of Burkina Faso, respectively. The maternities of the Yalgado Ouedraogo Teaching Hospital, Saint-Camille Hospital and Schiphra Protestant hospital in Ouagadougou and the maternity of the Sanou Sourô Teaching Hospital were included in the systematic newborn screening (NBS) program of SCD. Babies of both sexes and of all ethnic groups, born during the study period in the selected maternities and whose mothers and/or fathers consented to screen were included, regardless of the parents' hemoglobin status.

2.2. Blood Specimen Collection and Hemoglobinopathy Screening Tests

A sample of cord blood was taken, either on blotting paper (Dried blood spot - DBS) or in an EDTA tube (whole blood), prior to delivery, suitably labelled, treated and kept in temperature-controlled refrigerators. Samples from maternities in Ouagadougou were sent to the laboratory for analysis twice a week, and samples from maternities in Bobo-Dioulasso every two weeks for DBS and as soon as possible for EDTA tubes.

Iso-electric focusing (IEF) on agar gels (Isolab Inc., Akron, OH, USA) was carried out as the first-line screening test. The principle of the hemoglobin study is based on the migration of Hb variants under an electric field in a pH gradient. Thus, the migration of each variant in a hemolysate preparation stopped at the pH corresponding to its isoelectric point. For each DBS, a punch of 2 - 3 millimeters was removed, placed into a microtube for hemoglobin elution with an appropriate solution. For whole blood, red blood cells (RBC) were lysed to release hemoglobin using demineralized water. Samples of these hemolysates were carefully applied on gel paper alongside Hb A, Hb F, HbS, and HbC home-made control samples. The results were then read according to the migration pattern in comparison with the control hemoglobin samples to identify the hemoglobin variants as AA, AS, AC, SS, or SC by trained laboratory technicians and validated by biologists.

Cases with an ambiguous migratory profile (e.g., contamination by maternal blood) were considered indeterminate and excluded (**Figure 1**). The result of the suspicious abnormal Hb fraction was disclosed to the parents between 1 and 3 months later by the pediatrics during the routine healthy babies' consultation. A capillary blood sample with EDTA microtubes was taken on this occasion for



1: Inconclusive: Ambiguous migration profile; Abbreviations: HPLC: high performance liquid chromatography; SCD: Sickle cell disease; Hb: Hemoglobin; HbC: Hemoglobinopathy C.

Figure 1. Flowchart of screening for hemoglobinopathies.

confirmatory tests using a Bio-Rad® high-performance liquid chromatography (HPLC) instrument (BioRad, Hercules, CA, USA). The confirmation of the exact Hb fractions (HbA, HbS, or HbC) was based on their retention time. Beta-thalassemia syndrome was suspected if the HbA2 fraction was greater than 3.5% on HPLC. No further testing was performed for samples with no anomalous fraction. No testing has been done for the newborn's mother and her family members. According to the interpretation of the results, newborns were classified into four major groups (**Figure 1**): normal hemoglobin (absence of HbS or HbC), sickle cell disease (homozygous SS, compound heterozygous SC, Sβ0- and Sβ+-Thalassemia), Hb trait (HbAS and HbAC), and hemoglobinopathy C (CC and Cβ-Thalassemia).

Figure 1 above shows the flowchart of screening for hemoglobinopathy.

2.3. Quality Management

Before the NBS started, obstetricians and midwives were trained in cord blood sample collection and processing. Pediatrics and nurses were trained in the disclosure of results. All samples were tested according to the manufacturers' recommendations. A homemade control sample (composed of a mixture of AA cord blood, AS and AC adult blood) was inserted every 24 samples on the IEF agar gels. The results were double interpreted by the technician and biologists before validation.

2.4. Ethical Considerations

Ethical and administrative approvals were obtained before the start of the study. Mothers were informed by the obstetricians or midwives staff of the maternity units during antenatal consultations and sometimes at admission for childbirth.

All parents of babies gave informed consent for the NBS. Parents of newborns with severe hemoglobinopathy received result disclosure and appropriate advice from pediatrics, obstetricians or midwives. Babies with SCD were registered for follow-up. The screening tests were free of charge for all the babies included.

2.5. Statistical Analysis

Data were entered into an MS Access database, then exported and analyzed with Stata 15 software. The proportions and incidences of the different hemoglobinopathies were computed. The comparison between groups was made using an odd ratio. Hardy-Weinberg equilibrium (HWE) theorem was applied to calculate genotypic and allelic frequencies. For this purpose, β-thalassemia variant alleles were considered as β alleles.

Assuming AA_o, AS_o, AC_o, SS_o, SC_o, and CC_o are the observed genotype frequencies and N total study population; the following equations were used to calculate the allelic frequencies p of the β allele (Equation (1)), q of the β^S allele (Equation (2)) and r of the β^C allele (Equation (3)):

$$p = \frac{2\mathrm{AA}_o + \mathrm{AS}_o + \mathrm{AC}_o}{2N} \tag{1}$$

$$q = \frac{2\mathrm{SS}_o + \mathrm{AS}_o + \mathrm{SC}_o}{2N} \tag{2}$$

$$r = \frac{2\mathrm{CC}_o + \mathrm{AC}_o + \mathrm{SC}_o}{2N} \tag{3}$$

The expected genotype frequencies were calculated using the following formulas:

$$\mathrm{AA}_e = Np^2; \mathrm{AS}_e = 2Npq; \mathrm{AC}_e = 2Npr; \mathrm{SC}_e = 2Nqr; \mathrm{SS}_e = Nq^2; \mathrm{CC}_e = Nr^2 \tag{4}$$

We tested the HWE using the chi-square test as follows:

$$\chi^2 = \sum \left[(\text{Observed} - \text{Expected})^2 / \text{Expected} \right] \tag{5}$$

To assess the evolution of hemoglobinopathy and Hb variants over time in our country, we compared the confidence intervals of the incidence of SCD, abnormal Hb variants, and allele frequencies of β-globin in the current study with those observed in other studies in Burkina Faso. There was a difference in proportions when the confidence intervals did not overlap. The significant level was $p < 0.05$.

3. Results

Over the 5-years study period, a total of 11,343 newborn samples were tested. Six (0.05%) were excluded for the inconclusive profile (**Figure 1**). Males accounted for 47.8%. The majority of samples (60.2%) were from Bobo-Dioulasso. In the first year (2015), all samples were from Ouagadougou. 2/3 of the samples were tested during the first three years (**Table 1**). As shown in **Table 1**, at least an abnormal Hb fraction was found in 3076 samples; *i.e.*, 27.1% representing a cumulative incidence of 1:4 newborns, with a relative increase in frequency from

Table 1. Baseline distribution of the different haemoglobinopathies among screened newborns.

	2015	2016	2017	2018	2019	Total
Newborns screened (n; %)	1070	3405	2716	1685	2461	11,337
Ouagadougou	1070 (100)	1182 (34.7)	498 (18.3)	814 (38.3)	945 (38.4)	4509 (39.8)
Bobo-Dioulasso	0 (0)	2223 (65.3)	2218 (81.7)	871 (51.7)	1516 (61.6)	6828 (60.2)
Hb fractions (n; %)						
Abnormal	271 (25.3)	870 (25.6)	754 (27.8)	473 (28.1)	708 (28.8)	3076 (27.1)
Normal	799 (74.7)	2535 (74.4)	1962 (72.2)	1212 (71.9)	1753 (71.2)	8261 (72.9)
Type hemoglobinopathies						
All SCD (n; %)	24 (2.2)	30 (0.9)	41 (1.5)	40 (2.4)	80 (3.2)	215 (1.9)
Homozygous SS	5 (0.5)	11 (0.3)	11 (0.4)	14 (0.8)	19 (0.8)	60 (0.5)
Heterozygous SC	19 (1.8)	19 (0.6)	30 (1.1)	26 (1.5)	61 (2.5)	155 (1.4)
Total Hb traits (n; %)	234 (21.9)	801 (23.5)	686 (25.3)	412 (24.4)	602 (24.5)	2735 (24.1)
Hb AS	73 (6.8)	267 (7.8)	207 (7.6)	137 (8.1)	206 (8.4)	890 (7.8)
Hb AC	161 (15.0)	534 (15.7)	479 (17.6)	275 (16.3)	396 (16.1)	1845 (16.3)
Hemoglobinopathies C (n; %)	13 (1.2)	39 (1.1)	27 (1.0)	21 (1.2)	36 (1.5)	126 (1.1)
Hb CC	12 (1.1)	38 (1.1)	27 (1.0)	21 (1.2)	21 (0.8)	119 (1.0)
Hb Cβ-Thal	1 (0.1)	1 (0.03)	0 (0.0)	0 (0.0)	5 (0.2)	7 (0.1)

n, number; %, percentage; Hb, Hemoglobin; SCD, sickle cell disease; Cβ-Thal, Cβ-Thalassemia.

year to year (Kh^2 = 29.96; p< 0.0001). Among them, SCD was found in 1.9% of all samples (cumulative incidence of 1:53) with homozygotes SS cases which accounted for 27.9% (Table 1). Hemoglobinopathies C (HbCC and Hb Cβ-Thalassemia) accounted for 1.1% (Table 1), representing and cumulative incidence of 1:90 newborns.

The incidence of SCD was 1.51 times higher in Bobo-Dioulasso than in Ouagadougou with a cumulative incidence of 1:49 versus 1:59, OR = 1.51; 95% CI [1.09 - 2.10]: p = 0.013, while hemoglobinopathies C were almost 50% lower in Bobo-Dioulasso than in Ouagadougou (1:188 versus 1:66, OR= 0.56; 95% CI [0.39 - 0.79]: p = 0.001).

The observed genotype frequencies were significantly different (χ^2 = 50.09, p < 0.001) from the expected ones (Table 2).

The deviation is induced by the difference of 107% in the number of homozygote cases (β^S/β^S) observed compared to the expected cases (p = 0.001). For the other classes of genotypes, the observed and expected frequencies were not significantly different (p > 0.05). The allelic frequencies of β^S and β^C were 5.1 and 9.9%, meaning that the HbC variant occurred twice as often as the HbS variant (1:5 versus 1:10 newborns) (Table 3).

As shown in Table 3, when compared our results to those found in children and adolescents a fifteen year earlier, the cumulative incidence of total SCD and

Table 2. β-globin gene alleles frequency and observed and expected genotypes absolute frequencies of hemoglobin variants in newborns.

Genotype	Observed number	Expected number
AA (β/β)	8261	8191
AS (β/β^S)	901	983
SS (β^S/β^S)	60	29
AC (β/β^C)	1852	1908
SC (β^S/β^C)	144	114
CC (β^C/β^C)	119	111

$\chi^2 = 50.09$, 6df, $p < 0.001$.

Table 3. Comparison of the incidences of hemoglobinopathies, haemoglobin variants and allelic frequencies of β-globin gene with two previous studies on newborns and children/adolescents populations in Burkina Faso.

Parameters	Current study (2015-2019) N = 11,377	Kafando *et al.* [9], (2000-2004) N = 2341	Simporé *et al.* [6], (1997-2000) N = 28,226
Incidence of hemoglobinopathies: Incidence; [95% CI]; (cumulative incidence)			
Total SCD	1.90; [1.65 - 2.16]; (1:53)	1.75; [1.26 - 2.37]; (1:57)	1.36; [1.23 - 1.50]; (1:73)[a]
SS	0.53; [0.40 - 0.68]; (1:189)	0.60; [0.33 - 1.00]; (1:167)	0.20; [0.15 - 0.26]; (1:487)[a]
SC	1.37; [1.16 - 1.60]; (1:73)	1.15; [0.76 - 1.67]; (1:87)	1.16; [1.04 - 1.29]; (1:86)
AS	7.95; [7.45 - 8.46]; (1:12)	7.30; [6.28 - 8.43]; (1:14)	8.09; [7.77 - 8.41]; (1:12)
AC	16.33; [15.65 - 17.02]; (1:6)	16.87; [15.38 - 18.45]; (1:6)	19.17; [18.71 - 19.63]; (1:5)[a]
CC	1.05; [0.87 - 1.25]; (1:95)	1.37; [0.94 - 1.92]; (1:73)	1.43; [1.29 - 1.58]; (1:70)[a]
Incidence of abnormal Hb S and C: Incidence; [95% CI]; (cumulative incidence)			
HbS	9.75; [9.20 - 10.31]; (1:10)	9.05; [7.92 - 10.29]; (1:11)	9.45; [9.11 - 9.79]; (1:10)
HbC	18.65; [17.93 - 19.38]; (1:5)	19.35; [17.77 - 21.01]; (1:5)	21.76; [21.28 - 22.25]; (1:4)[a]
Allelic frequencies of β-globin gene alleles: Relative frequency; [95% CI]			
β	0.850; [0.843 - 0.856]	0.848; [0.833 - 0.862]	0.836; [0.832 - 0.840][a]
β^S	0.051; [0.047 - 0.055]	0.048; [0.039 - 0.057]	0.048; [0.045 - 0.050]
β^C	0.099; [0.093 - 0.105]	0.104; [0.092 - 0.117]	0.116; [0.112 - 0.120][a]

[a]: Significant difference compared to current study; N, number; 95% CI, confidence interval; SCD, sickle cell disease.

homozygous SS was significantly higher in newborns (1:53 in the current study 1:57 in 2000-2004 versus 1:73 in children and adolescents). This means a decrease in the number of cases of SCD disease as age increases. However, HbC trait, homozygous CC and βC allele were most frequent in children and teenagers compared to newborns.

4. Discussion

The purpose of this study was to assess the incidence of SCD and other hemog-

lobinopathies in Burkina Faso through systematic NBS. An abnormal Hb variant was found in 27.1% with a cumulative incidence of SCD of 1:53 newborns. The β^S and β^C alleles represented, respectively, about 5% and 10% of β-globin alleles in the studied population.

This study is the first to focus on NBS in another geographic and ethnic area of Burkina Faso in addition to Ouagadougou and its surroundings. It provides data to better appreciate the extent of hemoglobinopathies in the country. Indeed, in the context of poor care services, at ages beyond infancy, individuals with homozygous deleterious mutations (HbSS) are more likely to die. Moreover, those that were heterozygous trait, HbAS, and HbC due to a selective advantage for malaria, will have better survival than homozygous AA [10] [11] [12]. Therefore, NBS allows us to measure the true genotype frequencies.

However, our study has some limitations. Although Ouagadougou and Bobo-Dioulasso where the study took place, are the cities with the greatest ethnic mix (due to rural exodus), the results cannot be extrapolated to the entire country. These two major cities of the country are likely to combine several malaria spread factors (overcrowding, poor sanitation, and waste management…) that can alienate the random transmission of β-globin gene alleles across generations, due to the malaria implications discussed above. We also failed to recruit all newborns during the study period. In addition, we had not performed parental testing or molecular analysis to confirm certain findings, including thalassemia.

This study reported an incidence hemoglobinopathies of 27.1% (cumulative incidence of 1:4), confirming their importance in Burkina Faso, as previously noted [6] [7] [9]. The HbAS, and AC traits were the most prevalent and accounted for about 7.8% and 16.7%, in agreement with previous findings in the country. These results differ from those reported in Senegal and Mali, where, although the prevalence of the sickle cell trait HbAS was in the same proportions as in the current study, those of the heterozygote HbAC were much lower with 1.9% [13] and 5.6% [14] respectively. In Central Africa (Gabon, The Democratic Republic of Congo) and in Nigeria, HbAC is much rarer (1.1%, 0%, and 1.1% respectively) compared to HbAS which was 1.5 to 2 times higher than in the current study [15] [16] [17] [18]. These results confirm that the β^C allele is more concentrated in West Africa, especially around the Voltaic plateau, covered by Burkina Faso and Ghana [9] [12].

SCD was found in 1.9% of newborns, corresponding to a cumulative incidence of 1:53 newborns with a predominance of compound heterozygous SC or Sβ-thalassemia (1:73 versus 1:189 for homozygous SS). Compared with children and adolescents aged 2-25 years [6], the incidences in newborns were one-quarter (1:53 versus 1:73), two-thirds (1:189 versus 1:487) and one-sixth (1:73 versus 1:86) higher respectively for all SCD cases, homozygous SS and compound heterozygous (**Table 3**). These differences were not significant for compounds heterozygous. This trend confirms previous hypotheses that, in the absence of adequate health care, more than half of SCD cases are expected to die of malaria

or bacterial infectious diseases by age 5 years [3]. This was particularly true for homozygous SS (61% more in newborns). For compound heterozygotes, the difference was much modest, presumably due to the natural history of this clinical form of SCD, which is more characterized by chronic degenerative complications than by acute complications [19].

The observed frequencies of Hb genotypes compared with those expected differed statistically ($\chi^2 = 50.09$; $p < 0.001$) indicating that the distribution of these genotypes did not follow Hardy-Weinberg equilibrium. The frequency of the β^S (5.1%) and β^C (9.9%) alleles were five and ten times the theoretical equilibrium threshold of 1%. The deviation from Hardy-Weinberg equilibrium is due to the observed homozygotes (β^S/β^S) and heterozygotes (β^S/β^C) frequencies that were, respectively, 50% and 20% higher than the expected frequencies (60 cases versus 29 and 144 versus 114). Such findings have been reported previously [12] [20].

Compared to the results of the first NBS study in Burkina Faso [9], the incidences of overall homozygous HbSS, and compound heterozygotes were slightly, but not significantly ($p > 0.05$) higher in the current study (**Table 3**). This should indicate that the pool of abnormal β^S genes is maintained stable over the 15-year interval (2000-2015) between the two studies. The incidence of the HbS and the frequency of β^S allele in the two populations in **Table 3** confirmed this stability. This raises the question of the lack of a comprehensive sickle cell disease program in the country, as previously mentioned by Kafando *et al.* [9]. However, it is relevant to note that a period of 15 years seems very short to achieve involution in a complex health problem of genetic origin such as SCD, without radical measures such as banning marriage between people at risk or terminating pregnancies with affected fetuses. This is far from being the case in Burkina Faso, given the political, ethical, and moral controversies surrounding these issues. Currently, premarital screening is not mandatory and prenatal diagnosis of SCD is neither legal nor technically feasible. Therefore, the strategy remains to focus on public awareness and newborn screening. To date, this strategy is hampered in resource-poor countries by many of the logistical and organizational barriers already discussed [9] [21]. To overcome these barriers, it is suggested that newborn screening be linked to an existing accepted program. In Benin, a strategy targeting at-risk pregnant women during prenatal consultations to sensitize them to the need to test their children at birth has shown that about 80% of women adhere [22]. In Nigeria, authors recommended that newborns screening for SCD be linked to the vaccination program [16] [23].

5. Conclusion

SCD and other haemoglobinopathies appear to be frequent in Ouagadougou and Bobo-Dioulasso. Such results call for policy decisions in favor of newborn screening in our country. Integration of such a program with other public health interventions into a large multisectoral strategic plan will facilitate its implementation and increase its potential effectiveness. A centralized approach, based

on a few reference laboratories could be adopted for biological analyses, mainly confirmatory tests. Rapid diagnostic tests could be used in the first intention.

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Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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