# SUPPLEMENTARY APPENDIX

## INTEGRATION OF NEWBORN SCREENING FOR SICKLE CELL DISEASE INTO PRIMARY HEALTH CARE IN BUKAVU, DEMOCRATIC REPUBLIC OF THE CONGO: A PILOT STUDY

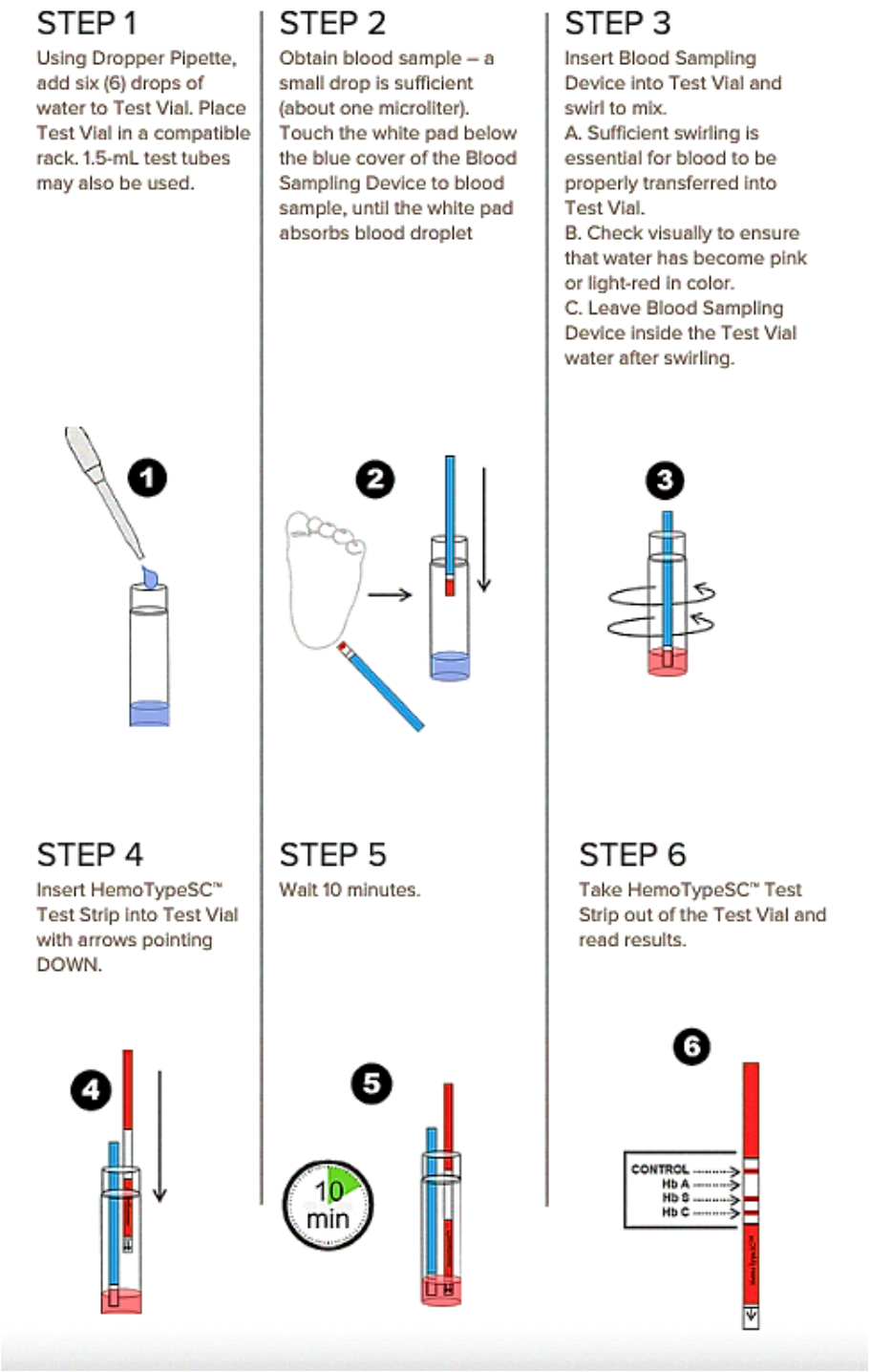


**Figure S1. Step-by-step approach for using the HemoTypeSC test and Protocol for using the HemoTypeSCTM point-of-care testing, based on manufacturer's instructions.**

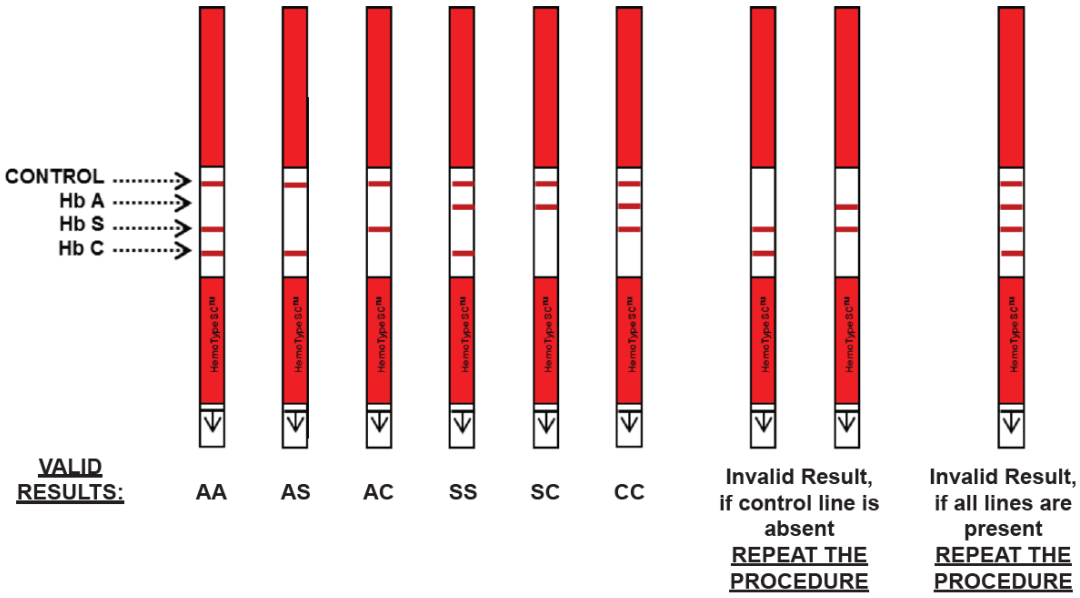


**Figure S2: Interpreting the results of the HemoTypeSC test**

**Expected results and their interpretation for the HemoTypeSC test:**

**Valid Results:**

- AA (Hb A only): The HbA line is absent, while all other lines, including the control line, are present.
- AS (Heterozygous Hb A and Hb S): The HbA and HbS lines are absent, while all other lines, including the control line, are present.
- AC (Heterozygous Hb A and Hb C): The HbA and HbC lines are absent, while all other lines, including the control line, are present.
- SS (Homozygous sickle cell Hb S): The HbS line is absent, while all other lines, including the control line, are present.
- SC (Compound heterozygous Hb S and Hb C): The HbS and HbC lines are absent, while all other lines, including the control line, are present.
- CC (Homozygous Hb C): The HbC line is absent, while all other lines, including the control line, are present.

Invalid Results:

- Absence of the control line: The test is invalid and must be repeated.
- Presence of all lines (Hb A, Hb S, and Hb C): The result is uninterpretable, and the test must be repeated.

If an invalid result is obtained, it is recommended to repeat the procedure to ensure diagnostic accuracy.


**Figure S3. Image showing Gazelle Hb variant point-of-care diagnostic device**

## SETTING UP NEWBORN SCREENING FOR SICKLE CELL DISEASE IN PRIMARY HEALTHCARE FACILITIES IN BUKAVU

Before initiating the screening activity, we sought authorization and collaboration from the Provincial Ministry of Public Health of South Kivu. This step was crucial for informing the primary healthcare centers about the program and gathering information on existing services, such as antenatal clinics, deliveries, and other services offered at these facilities.

Simultaneously, we consulted key personnel and other healthcare workers from primary healthcare centers across the city of Bukavu to gather their views on integrating newborn screening for sickle cell disease using point-of-care tests into services related to childbirth. This information was essential for mapping the primary healthcare centers within the Council's jurisdiction. Participating centers were selected based on patient volume.

For the selected primary healthcare centers, we assessed the existing resources, infrastructure, and services to determine the feasibility of integrating newborn screening for sickle cell disease into childbirth services using point-of-care tests. It is worth noting that all of the center managers confirmed that newborn screening for sickle cell disease could be conducted when mothers visit the centers for deliveries.

We undertook a prospective cohort pilot study from November 2022 to December 2023 in the Ibanda commune, located in Bukavu, DRC. The study was conducted in 15 healthcare facilities offering primary healthcare services in the commune, including Panzi General Referral Hospital, Chai Hospital Center, Cidasa Health Center, Philadelphia Hospital Center, SkyBorn Hospital, Biopharm Hospital Center, Ruhigita Clinic, Malikiya wa Amani Health Center, Bioglodie Medical Center, CELPA Hospital Center, Saint Luc Hospital Center, Olive Lembe Hospital Center, Mulungulungu Hospital Center, Kabuye Health Center, and Maman Mwilu Health Center. This diversity enabled a better representation of the local population and allowed for the collection of meaningful and representative data for this pilot study.

These preparatory efforts laid the necessary foundation for launching the screening program.

**TABLE S1: DISTRIBUTION OF RECRUITED, SENSITIZED, AND LOST TO FOLLOW-UP PARTICIPANTS BY HEALTHCARE FACILITY**

| Facility | Pregnant women who accepted the test | Total sensitized | Lost to follow-up |
|---|---|---|---|
| Panzi General Referral Hospital | 941 | 1272 | 65 |
| Philadelphia Hospital Center | 367 | 497 | 19 |
| Cidasa Health Center | 694 | 873 | 7 |
| Chai Hospital Center | 623 | 860 | 55 |
| Malikiya wa Amani Health Center | 356 | 452 | 24 |
| CELPA Hospital Center | 261 | 321 | 5 |
| SkyBorn Hospital | 426 | 558 | 48 |
| Bioglodie Medical Center | 356 | 444 | 29 |
| Ruhigita Clinic | 135 | 168 | 13 |
| Biopharm Hospital Center | 44 | 58 | 0 |
| Olive Lembe Hospital Center | 57 | 72 | 8 |
| Saint Luc Hospital Center | 20 | 28 | 0 |
| Mulungulungu Hospital Center | 149 | 189 | 10 |
| Kabuye Health Center | 53 | 68 | 16 |
| Maman Mwilu Health Center | 14 | 18 | 0 |
| **Total** | **4496** | **6353** | **299** |

**FORM FOR MOTHER AND NEWBORN SCREENING**

**NEWBORN SCREENING FOR SICKLE CELL DISEASE IN BUKAVU, DRC**

Name of the healthcare facility: .....

Serial No: .....

Dr. Mwanza's team is organizing a free newborn screening for sickle cell disease in Bukavu, after which a certificate will be provided. If you consent to have your child screened and followed up, please fill out the following form.

Number of the participant: .....Mother's Name: .....

Mother's Age: .....

Baby's Name: .....

Date of Birth: ..... Baby's Age: .....

Baby's Sex: .....

Mother's Phone number: ..... Address: .....

Sample collection date: ..... Sample collected by: .....

Mother's screening: HemoTypeSC™: .....

Gazelle® electrophosis: .....

Baby's screening: Gazelle® electrophosis: .....

I have reviewed the information in this consent form, or someone has read it to me and provided an explanation, and I fully understand its contents. All of my questions have been addressed, and I consent to have my baby's sample collected and for my baby to be enrolled in this program for follow-up if diagnosed with sickle cell disease.

| Parent's/Guardian's Name | Signature | Date |
|---|---|---|
| ..... | ..... | ..... |
| Research Associate's Name | Signature | Date |
| ..... | ..... | ..... |

## THE DRC NATIONAL IMMUNIZATION SCHEDULE FOR CHILDREN UNDER 15 MONTHS

In the Democratic Republic of the Congo (DRC), the national immunization schedule for children under 15 months, as recommended by the Ministry of Health, includes the following vaccines:

**At birth:** During the first vaccination appointment, your child will receive two vaccines to protect against:

- Tuberculosis (BCG)
- Poliomyelitis ($VPO_0$)

**At 1.5 months:** During the second vaccination appointment, your child will receive four vaccines to protect against:

- Pneumococcal infections ($PCV13_1$)
- Poliomyelitis ($VPO_1$)
- Diphtheria, tetanus, whooping cough, hepatitis B, and meningitis (Pentavalent, $DTC_1$, $Hep\text{-}Hib_1$)
- Diarrhea ($Rota_1$)

The same vaccines will be administered during the next appointment.

**At 2.5 months:** During the third vaccination appointment, your child will receive four vaccines to protect against:

- Pneumococcal infections ($PCV13_2$)
- Poliomyelitis ($VPO_2$)
- Diphtheria, tetanus, whooping cough, hepatitis B, and meningitis (Pentavalent, $DTC_2$, $Hep\text{-}Hib_2$)
- Diarrhea ($Rota_2$)

The same vaccines will be administered during the next appointment.

**At 3.5 months:** During the fourth vaccination appointment, your child will receive several vaccines to protect against:

- Pneumococcal infections ($PCV13_3$)
- Poliomyelitis ($VPO_3$, VPi)
- Diphtheria, tetanus, whooping cough, hepatitis B, and meningitis (Pentavalent, $DTC_3$, $Hep\text{-}Hib_3$)
- Diarrhea ($Rota_3$)

**At 6 months:** During the fifth vaccination appointment, your child will receive a vaccine to protect against:

- Malaria ($VAP_1$)
- Vitamin A supplementation

**At 7 months:** During the sixth vaccination appointment, your child will receive a vaccine to protect against:

- Malaria ($VAP_2$)

**At 8 months:** During the seventh vaccination appointment, your child will receive a vaccine to protect against:

- Malaria ($VAP_3$)

**At 9 months:** During the eighth vaccination appointment, your child will receive three vaccines to protect against:

- Poliomyelitis (VPi)
- Measles/rubella (VAR/RR)
- Yellow fever (VAA)

**At 12 months:** Deworming with Mebendazole and vitamin A supplementation, with a booster every 6 months until 59 months.

**At 15 months:** During the ninth and final vaccination appointment, your child will receive two vaccines to protect against:

- Measles/rubella (VAR/RR)
- Malaria (VAP4)

## Vaccines Protective for Sickle Cell Disease (SCD):

For children with sickle cell disease (SCD), the following vaccines are particularly crucial to help protect them from infections that they are more vulnerable to due to their compromised immune system:

1. **Pneumococcal vaccine (Pneumo)**: Helps prevent pneumonia, meningitis, and other serious infections caused by Streptococcus pneumoniae.
2. **Meningococcal vaccine (Meningitis)**: Protects against meningitis, which is a serious risk for children with SCD.
3. **Haemophilus influenzae type B (Hib)**: Helps prevent serious infections like meningitis and pneumonia, which are more dangerous for children with SCD.

4. **Hepatitis B vaccine (HepB)**: Prevents liver disease that can be more severe in children with SCD.
5. **Influenza vaccine** (not listed but recommended in clinical practice): Prevents severe complications from influenza, which children with SCD are at higher risk for.

These vaccines are essential for reducing the risk of severe infections in children with SCD, which can otherwise lead to complications or even death. Given the high risk of infections in children with SCD, additional vaccines such as the annual influenza vaccine and a booster dose of pneumococcal vaccine (PPSV23) after the age of two are often recommended in clinical practice.

FONDATION PIERRE FABRE

**UNE COMMUNAUTE SENSIBLISEE SUR LA DREPANOCYTOSE, EVITE LE MARIAGE A RISQUE**

**ENSEMBLE CONTRE LA DREPANOCYTOSE**

## EXISTE-T-IL UN CENTRE DE PRISE EN CHARGE DES DREPANOCYTAIRES AU SUD-KIVU ?

Oui, La Petite Cliniques Ami des Enfants d'Umbrella foundation asbl, située au n°76 Avenue Hypodrome, quartier Nyalukemba, commune d'Ibanda, ville de Bukavu, Province du Sud-Kivu, RDC.

Tél. +243994137748 Email : pcamidesenfants@gmail.com

## LISTE DE STRUCTURES DE SANTÉ

Oui, il existe 14 structures sanitaires de dépistage et de prise en charge de l'anémie SS ou Drepanocytose dans la zone de santé d'Ibanda/Bukavu.

- Hôpital General de Reference de PANZI
- Petite Clinique Ami des Enfants
- Centre Hospitalier de CHAI
- Centre Hospitalier de PHILADELPHIA
- Centre de Santé CIDASA
- Centre Hospitalier OLIVE LEMBE
- Centre Hospitalier BIOGLODIE
- Clinique RUHIGITA
- Centre Hospitalier BIOPHARM
- Centre Hospitalier SAINT LUC
- Centre médical IMPACT MEDICAL
- Hôpital SKY BORN
- Centre de Santé MALKIA
- Centre Hospitalier CELPA

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**LA PETITE CLINIQUE AMI DES ENFANTS**


N° av. d'Ibanda, ville de Bukavu, Province du Sud-Kivu, RDC.

+243994137748

pcamidesenfants@gmail.com

umbrella foundation

foundationumbrella_sud_kivu


République Démocratique du Congo

**PROVINCE DU SUD-KIVU**

Ministère Provincial de la Santé, Genre, famille, des Affaires sociales et humanitaires

# DIVISION PROVINCIALE DE LA SANTE

UMBRELLA FOUNDATION

Point focal du programme national de lutte contre la drépanocytose Sud-Kivu

**CE QUE NOUS DEVONS SAVOIR DE LA DREPANOCYTOSE OU ANEMIE SS**

PROGRAMME NATIONAL DE LUTTE CONTRE LA DREPANOCYTOSE

PNLCD

SICKLE CELL SCREENING LEAFLET FOR PREGNANT WOMEN

## LA DREPANOCYTOSE OU ANEMIE FALCIFORME OU ANEMIE SS

### I. DE QUOI S'AGIT – IL ?

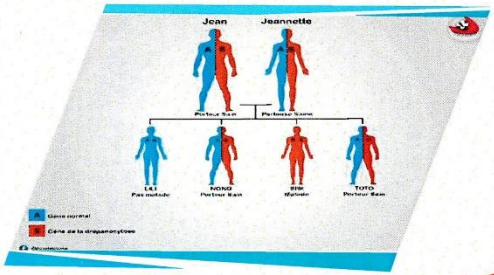
Maladie génétique héréditaire de l'hémoglobine, la plus répandue au monde. L'hémoglobine : une protéine qui se trouve dans le globule rouge qui fixe l'oxygène et permet sa distribution dans l'organisme entier. L'Hémoglobine normale est représentée par la lettre A et hémoglobine hémoglobine anormale par la lettre S.

Une Personne normale : hémoglobine AA. Une Personne porteuse saine : hémoglobine AS. Une Personne malade (drépanocytaire) : hémoglobine SS. Cette maladie touche plus les noirs, les arabes et les indiens. Maladie mal connue par la communauté et entourée de beaucoup de mythes.

### II. COMMENT SE TRANSMET – ELLE ?

C'est une maladie héréditaire que les parents porteurs sains (AS x AS) ou deux parents malades (SS x SS) ou encore un parent porteur sain et un parent malade ((AS x SS) transmettent à leurs enfants

TRANSMISSION




### III. QUELLE EST LA SITUATION DE LA DREPANOCYTOSE EN RDC et SUD-KIVU.

- RDC, troisième pays au monde après le Nigeria et l'Inde qui regorgent le plus grand nombre de cas de drépanocytose. 30% de la population congolaise sont de porteurs sains(AS), 1 à 2% de naissances sont drépanocytaires (SS).

- Au Sud-Kivu on rapporte environ 1300 cas de crises drépanocytaires (anémie aigue, crise de douleur). Les ZS de Fizi, Nundu, Uvira, Lemera, Ruzizi, Ibanda et Shabunda sont les plus concernées.

### IV. COMMENT SE MANIFESTE-T-ELLE ?

- Anémie chronique et aigue (manque de sang) entrainant de multiples transfusions sanguine (hospitalisations)

- Crise de douleurs atroces de localisations diverses (crise vaso-occlusive) obligeant le recours à des médicaments antidouleur majeurs en hospitalisation

- Episodes d'Infection à répétition avec plusieurs hospitalisations

- Plusieurs complications aigues et chroniques dont : les infections respiratoires atteintes ostéo-articulaires, rénales, cardio-vasculaires, cérébrales (AVC), oculaires, cutanées et retard de croissance

- Conséquences socio-économiques et psychiques énormes : appauvrissement des familles dû au coût aux soins et absence des parents au travail, stigmatisation des enfants et surtout de leurs mères (porte-malheur, sorcier, démon), divorce, abandon des enfants aux grands parents, retard scolaire, dépression mentale (maladie de la douleur et de la souffrance), bref l'exclusion sociale.

UMBRELLA FOUNDATION

Journée mondiale de la
# DRÉPANOCYTOSE

«Drépanocytose (anémie SS) et Covid-19 : Sensibiliser c'est bien, Dépister c'est mieux»

PNLCD

unicef

### V. QUEL EST LE TRAITEMENT DE LA DREPANOCYTOSE ?

a. TRAITEMENT CURATIF : jusqu'à ce jour, il n'existe pas de traitement curatif définitif pour les malades SS. Mais il existe des mesures efficaces permettant d'améliorer la qualité de vie de drépanocytaires, de mener une vie quasi normale et leur permettre d'atteindre une espérance de vie au-delà de 60 ans, de se marier et faire les enfants, d'étudier et travailler pour gagner la vie.

**Quelles sont ces mesures :**

**-Dépistage (à la naissance) et prise en charge précoces de la maladie :**

respect du calendrier vaccinal, traitement précoces et prophylaxie (Péni V) des infections, prise de l'acide folique à vie, prophylaxie contre le paludisme (dormir sous Moustiquaire), boire suffisamment, éviter les efforts physiques soutenus, éviter le froid, déparasitage, bonne hygiène de vie, bonne nutrition et un suivi médical régulier.

b. La prévention : est- il possible d'éviter d'avoir les enfants drépanocytaires ?, OUI, par connaissance du statut de son hémoglobine bien avant le mariage voire à la naissance par un diagnostic systématique. Ceci permettre d'éviter : le mariage entre deux personnes porteuses saines (AS x AS), ou une personne porteuse saine (AS) et une personne malade(SS) (AS x SS), ou entre deux personnes malades (SS) (SS x SS).