



14th

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& WMDA Meetings

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Sickle Cell Anemia, a Burden in Africa



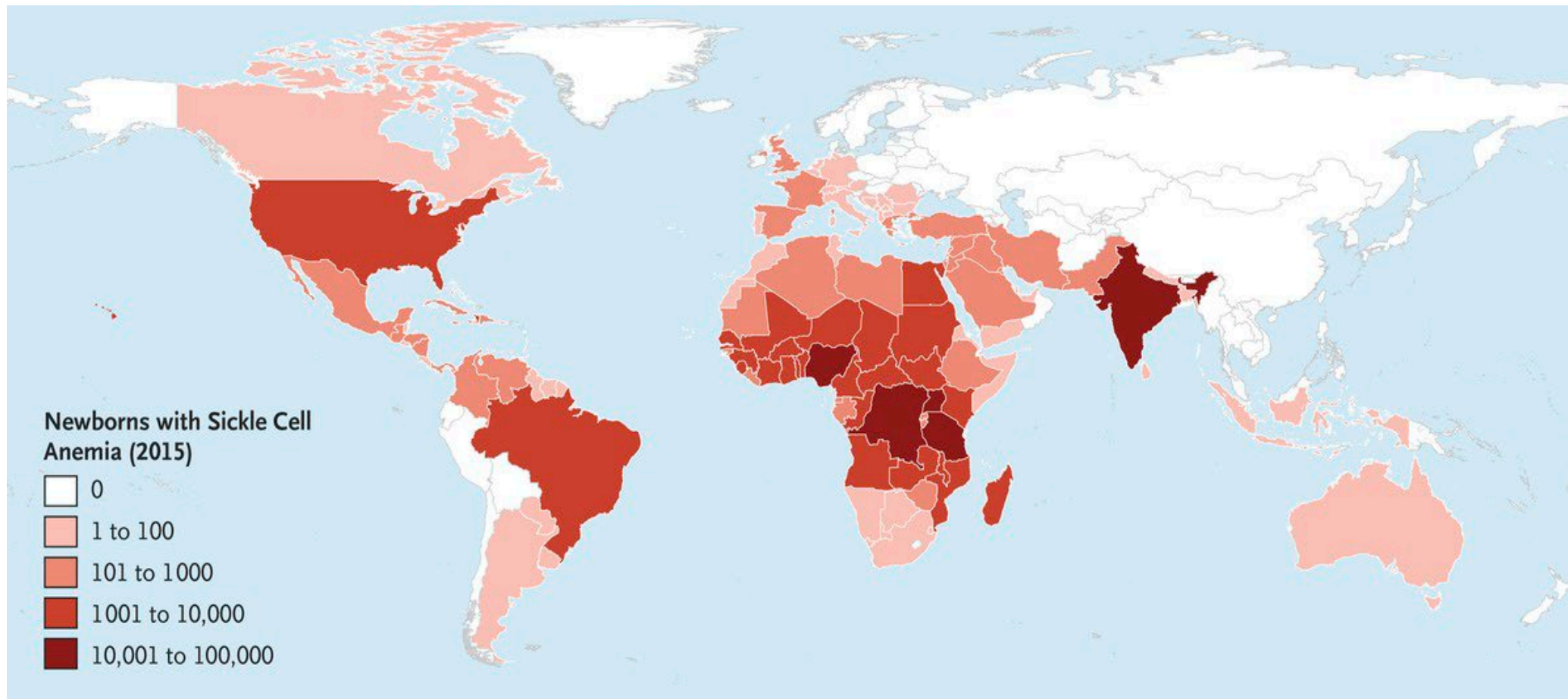
Challenges Associated with Sickle Cell Disease in Low-Middle Income and Low-Income Countries

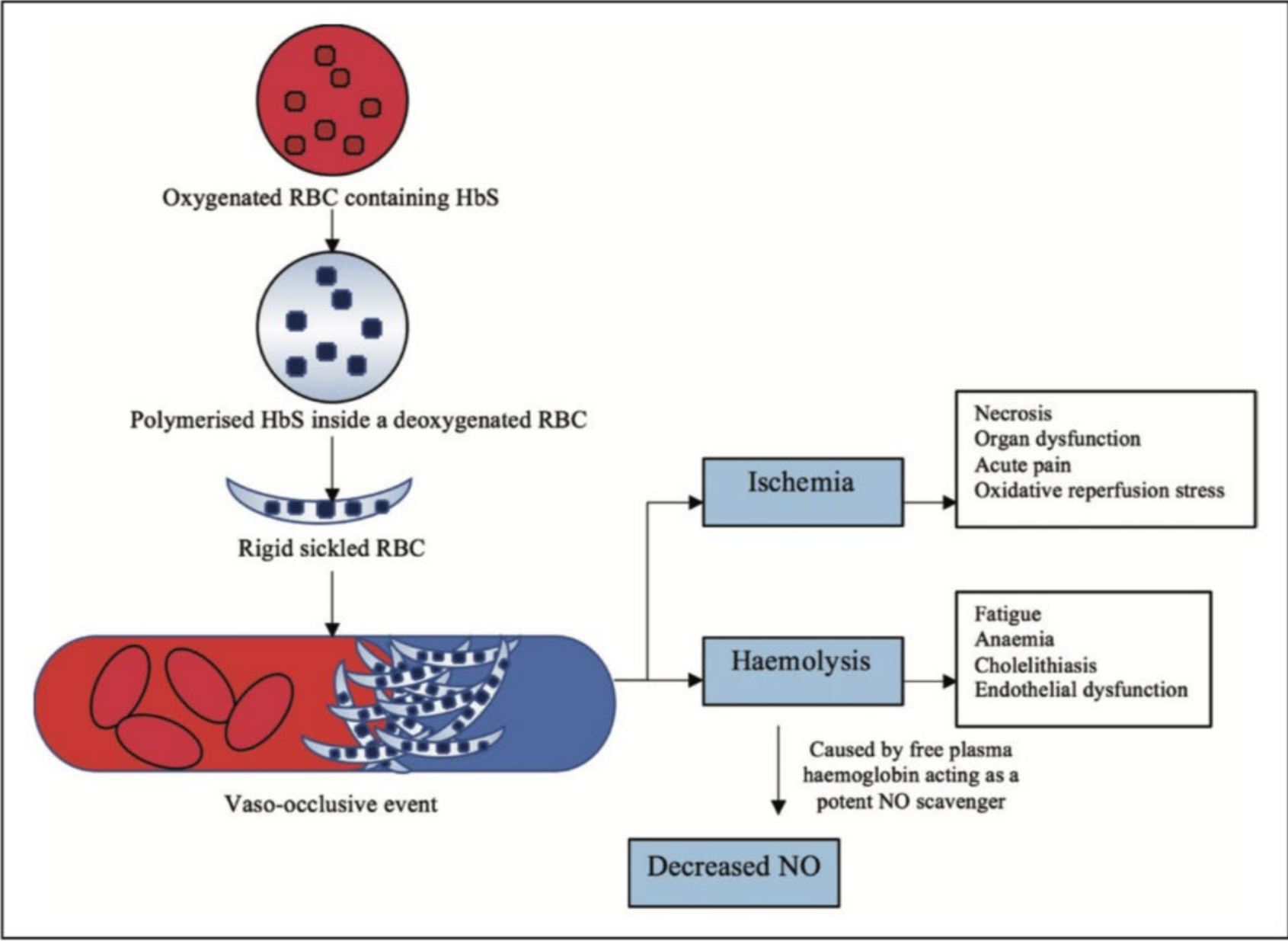


Introduction

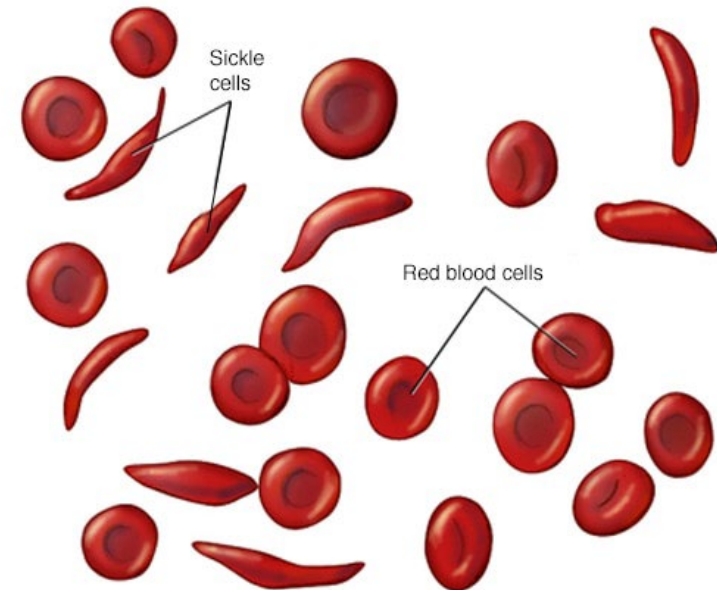
- SCD is a hereditary red blood cells disorder.
- Most of the world's SCD burden is in Africa where it is a major contributor to child morbidity and mortality. Most of the cases occur in Nigeria & Democratic Republic of Congo.
- SCD is one of the main causes of childhood anemia in Africa. Around 1000 children in Africa are born with SCD every day and more than half will die before they are 5 years old.
- Around 40% of people in some African countries have SC Trait.
- *Subsequently, SCD is a major burden on Blood Establishments in most of African countries.*

Sickle Cell Disease Burden in Africa





Sickle cell shape



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Survey among African countries

- 1- Is Sickle Cell Disease (SCD) a burden on blood banks in your country?
- 2- What is the percentage of carrier rate in your country?
- 3- What is the percentage of the affected persons in your country?
- 4- What is the classical treatment of SCD in your country?
- 5- What is the percentage of your RBCs stocks that are spared for SCD patients?
- 6- what is the percentage of TTIs infected SCD patients due to transfusion in your country?
- 7- Do you have any national awareness program to reduce the SCD newborns in your country?
- 8- Is there a national SCD patients society or NGO to care for these cases in your country?
- 9- Who pays for the blood needed to transfuse SCD patients in your country?
- 10- Is chelation of excess iron deposition as part of treating SCD patients available in your country?

Responding countries to the survey

Response to email survey

- Malawi
- South Africa
- Nigeria
- Botswana
- Burkina Faso
- Uganda
- Madagascar
- Egypt
- Tunisia

Personal contact

- Morocco
- Algeria
- Libya

1- Is Sickle Cell Disease (SCD) a burden on blood banks in your country?

Country	Response
Nigeria	SCD is a big burden on the blood transfusion Service
Malawi	Yes. It is a very big problem.
South Africa	Yes. Specifically, when red cell exchange is required
Madagascar	Yes
Botswana	SCD is not a burden
Burkina Faso	Yes, because many cases of SCD use many blood bags for treatment.
Uganda	It is the 2 nd leading medical condition utilizing blood in SSA – at 18%.
Egypt	Only in west desert (Siwa & Dakhla owasis)
Tunisia	No.

2- What is the percentage of carrier rate in your country?

Country	Response
Nigeria	Carrier rate, About 25%.
Malawi	Carrier rate; It is 9.1%.
South Africa	Carrier rate is Unknown.
Madagascar	10%.
Botswana	Extremely rare disease, 1 seen and was foreigner.
Burkina Faso	Not known.
Uganda	Carrier rate 13.3% .
Egypt	9% only in western desert oases.
Tunisia	1.9%.

3- What is the percentage of the affected persons in your country?

Country	Response
Nigeria	About 1 to 2 % homozygote SS or SC Hb phenotype out of a country population of over 200 million.
Malawi	It is 0.1% (SCD).
South Africa	Unknown, but prevalence is <1%.
Madagascar	1.2%
Botswana	Almost 0.
Burkina Faso	The prevalence rate of sickle cell disease was 4.63% including 54.12% of homozygous SS form in the country
Uganda	0.7% affected persons
Egypt	Few hundreds are registered in Cairo University hospital.
Tunisia	500 registered.



4- What is the classical treatment of SCD in your country?

Country	Response
Nigeria	Routine Folic acid and Antimalarials, RBC transfusion for haemolytic crisis, Analgesics for pain, and Antibiotics for infection, A few HSCT have been carried out successfully.
Malawi	Monthly prophylaxis of: SP (Sulfadoxine-Pyrimethamine) tablets, folic acid, Iron, Benzathine injections.
South Africa	Treatment is Hydroxyurea.
Madagascar	Folic acid, Symptomatic treatment : analgesic, antibiotics, oxygen, rehydration, Hydroxycarbamide (when affordable), RBC transfusion : simple transfusion or exchange transfusion.
Botswana	Medications and blood transfusion.
Burkina Faso	Blood treatment : simple transfusion, transfusion exchange, Therapeutic blood letting, Symptomatic treatment, infections treatment, Pain treatment.
Uganda	Folic acid - daily, Anti-malarial, chemoprophylaxis with SP- Monthly, Antibacterial chemoprophylaxis for U5 children with Pen-v - daily, Hydroxyurea, Pneumococcal Vaccine – booster, NSAIDs – for mild painful crises, while oral morphine for severe pain management.
Egypt	Blood transfusion, hydroxyurea,
Tunisia	Blood transfusion, exchange transfusion, hydra, BM transplant.

5- What is the percentage of your RBCs stocks that are spared for SCD patients?

Country	Response
Nigeria	Most transfusion data is not entered into data collection systems
Malawi	Most transfusion data is not entered into data collection systems
South Africa	No RBC stock are spared specifically for SCD patients in the SA setting
Madagascar	20%
Botswana	No RBCs stocks assigned for SCD
Burkina Faso	not available
Uganda	About 7% of RBCs stocks at the national referral hospitals go to SCD
Egypt	No RBCs stocks assigned for SCD
Tunisia	10% to all hemoglobinopathies.

6- what is the percentage of TTIs infected SCD patients due to transfusion in your country?

Country	Response
Nigeria	No figures available but very infrequent
Malawi	Not known. We do not have a good hemovigilance program.
South Africa	We do not have this specific information. We have had 1 proven HIV and 2 proven HBV TTIs since the implementation of NAT on 03 October 2005 though.
Madagascar	1%
Botswana	No TTIs prevalence rate
Burkina Faso	not available
Uganda	is Not extensively studied, except for HCV. 3% of transfused SCD patient may be with HCV.
Egypt	Not available.
Tunisia	Residual risk is 1/3500 for HBV, 1/3600 for HCV.

7- Do you have any national awareness program to reduce the SCD newborns in your country?

Country	Response
Nigeria	There is no national Newborn Screening Program in the country
Malawi	Only activity on the international day recognizing sickle cell disease, on the 19 th June every year.
South Africa	due to low prevalence rate, done only to extended families. However, there were 2 sessions with Q&A on national TV. For healthcare workers, SA national guidelines for management of SCD.
Madagascar	Essentially during SCD day and through SC patient organization
Botswana	NOT A PROBLEM IN BOTSWANA
Burkina Faso	No national awareness program
Uganda	A national sickle cell screening program was started in 2014 for awareness
Egypt	Through the national SCD NGO.
Tunisia	No.

8- Is there a national SCD patients society or NGO to care for these cases in your country?

Country	Response
Nigeria	Yes, One is called Sickle Cell Foundation of Nigeria. Another is called Sickle Cell Support Society of Nigeria (SCSSN) and another is the Association for the Prevention of Sickle Cell Anemia in Nigeria (APOSCAN)
Malawi	Yes, NGO/society
South Africa	No SCD NGO or Society
Madagascar	Yes
Botswana	No NGO or Society for SCD
Burkina Faso	There is an association which campaigns for therapeutic care of SCD patients: its name is Initiative committee against sickle cell disease (Comité d'initiative contre la drépanocytose)
Uganda	Yes , NGO/Society
Egypt	NGO/Society.
Tunisia	No .

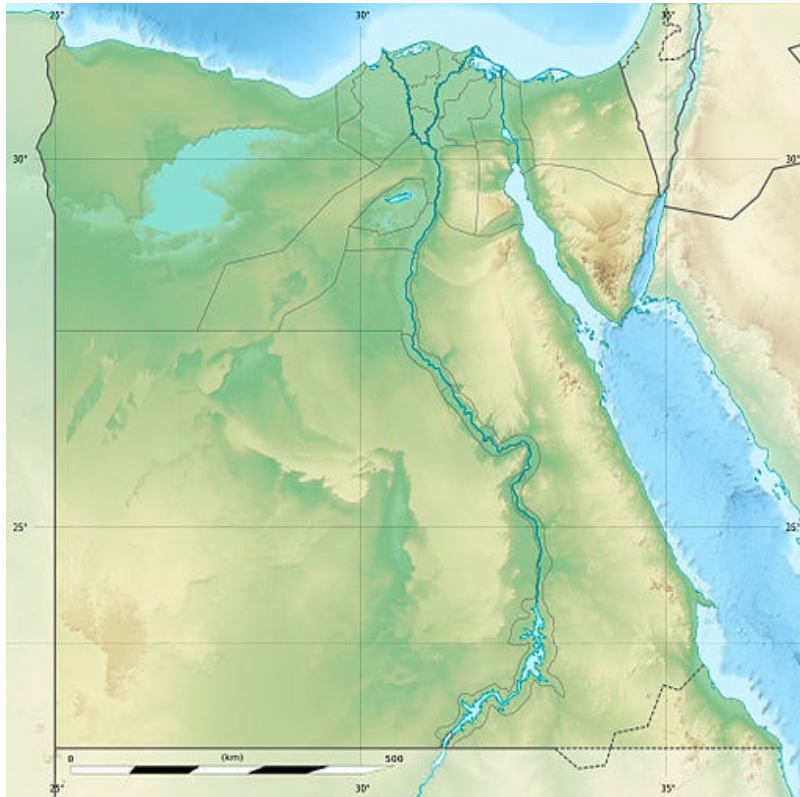
9- Who pays for the blood needed to transfuse SCD patients in your country?

Country	Response
Nigeria	Patients pay out of pocket. Coverage by the official National Health Insurance System is very limited.
Malawi	In Malawi Government hospitals blood is freely available to the patient. The Government then pays the Blood Transfusion centres a Cost recovery Fee on behalf of the patient.
South Africa	South Africa has two healthcare sectors: Private healthcare sector, funded by formal healthcare funders / medical aids, or the government-funded Public healthcare sector. A minority of patients in SA would be private patients, without healthcare funding / medical aids, and would pay out-of-pocket.
Madagascar	Blood transfusion is free, but blood donation is very low, 20% regular blood donation
Botswana	Government pays for all transfusions
Burkina Faso	The state of Burkina Faso pays for all patients. blood is free for all patients
Uganda	Government (Public) pays for transfusion
Egypt	Government, few cases out of the pocket in case of shortage they seek private BBs.
Tunisia	National Health insurance.

10- Is chelation of excess iron deposition as part of treating SCD patients available in your country?

Country	Response
Nigeria	Chelation therapy is available only in a few private hospitals at great cost.
Malawi	No, it is not available.
South Africa	Yes, for chelation.
Madagascar	Yes, when affordable.
Botswana	Chelation not available.
Burkina Faso	Chelation is not available in our country.
Uganda	Chelation not universally available, Also, screening for iron overload is not standard-of-care.
Egypt	Yes.
Tunisia	Yes.

Hb As 9% carrier western desert, Egyptian Oases (El_Kharga and El-Dakhla)



Hbs in Egyptian Siwa Oasis. (Western Desert)

- Hb S 22.17%
- Hbc 0.49%
- Siwa Oasis dates back at least to the time of Ramsis III (Circa 1175 B.C) When it is populated by the Ancient Egyptians (Simpson 1929).
- By the coming of Islam in Egypt (640 A.D) Arab tribes settled in Siwa. (Simpson 1929).

General information from NA countries

Blood Banks

- Blood Banks are not burdened by this disease.
- Only transfusion is offered, no apheresis.
- Patients are referred for transfusion from hospitals upon crisis.

Health Authorities

- No national registry for SCD.
- Governments do not consider it a priority.
- No specific national guidelines for treatment.
- General impression that the real numbers of cases are bigger than known.

Reflections

Challenges Associated with SCD in LMICs

- Lack of support by Health authorities.
- Lack of resources for diagnosis, treatment, follow up, etc.
- Size of burden is not quantified. Lack of national registries.
- No clear standards for management.

Recommendations

- Support by organizations like WHO, AfSBT, ISBT, etc.
- Health Authorities to consider this group of patients as priority; provide national awareness program, standardized management, provide blood and medications, etc.
- African countries need to cooperate and exchange experiences.

Thank you.

