Bahrain success story in controlling sickle cell disease 1984-2012

STUDENT SCREENING

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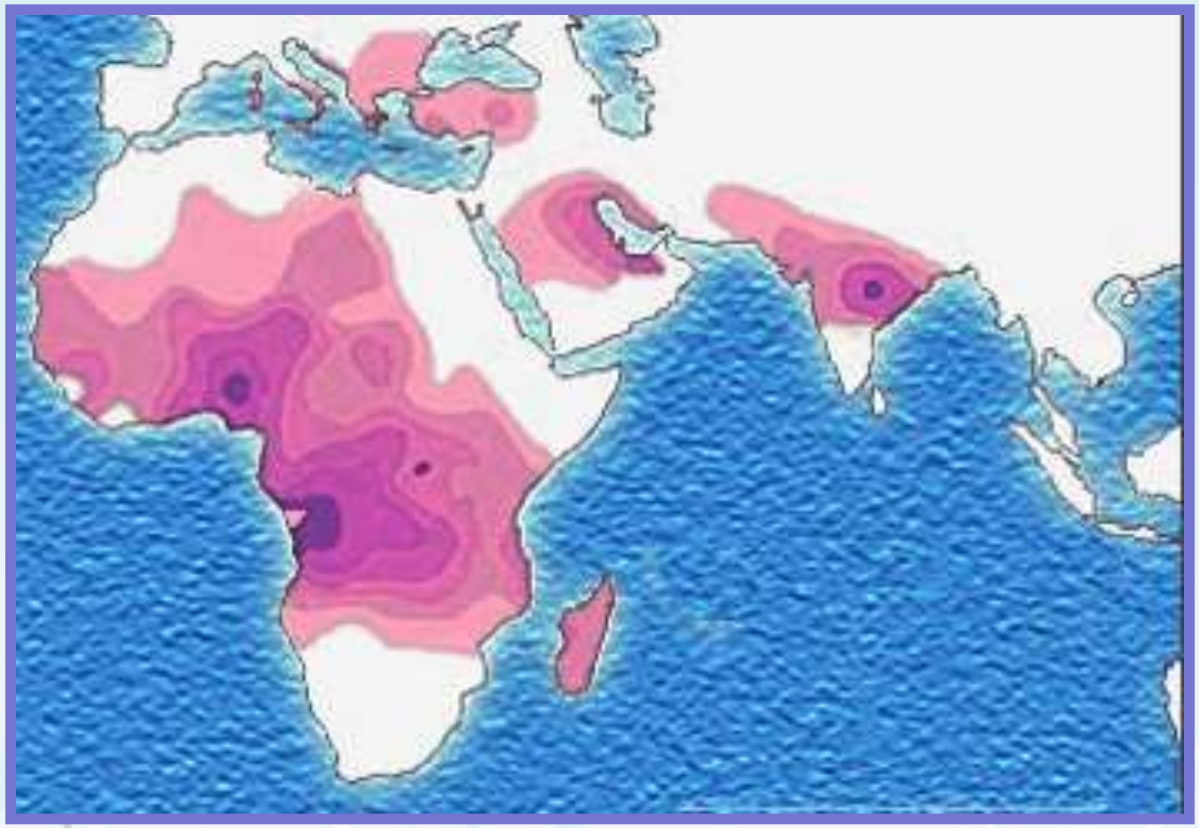
Global SCD distribution

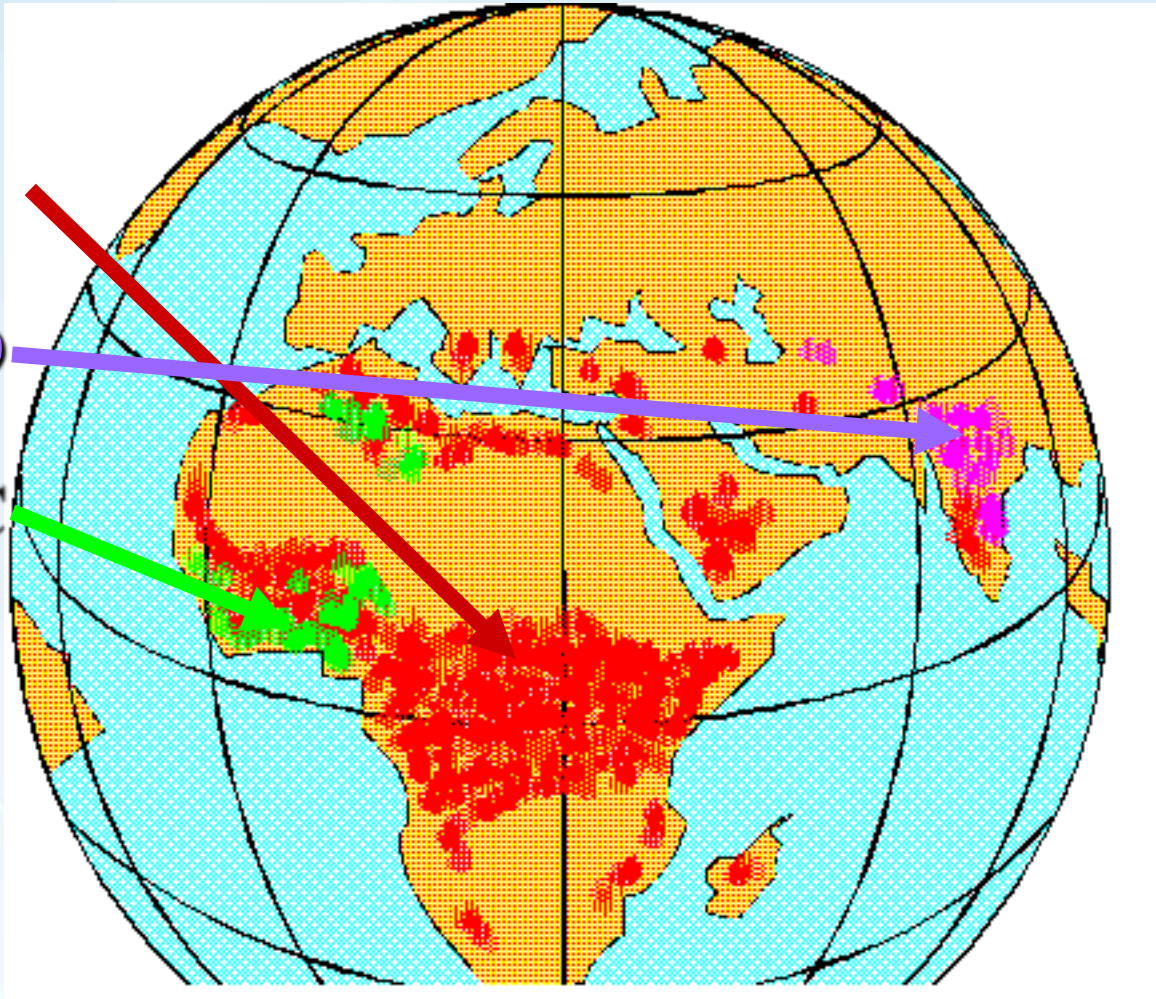


Worldwide Distribution

Hemoglobin S

Hemoglobin D

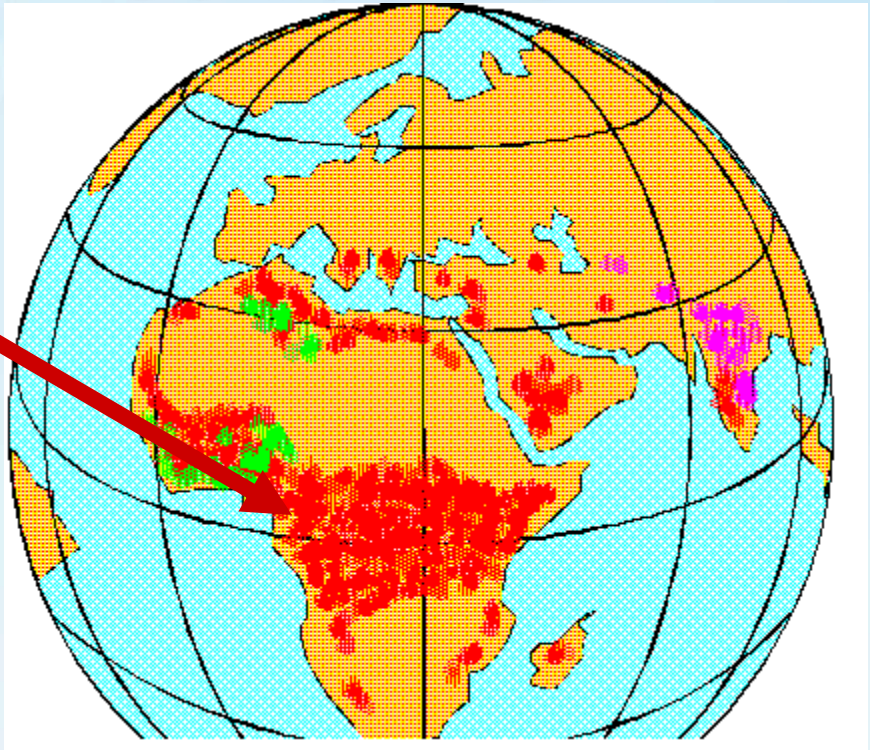
Hemoglobin C



Worldwide Distribution

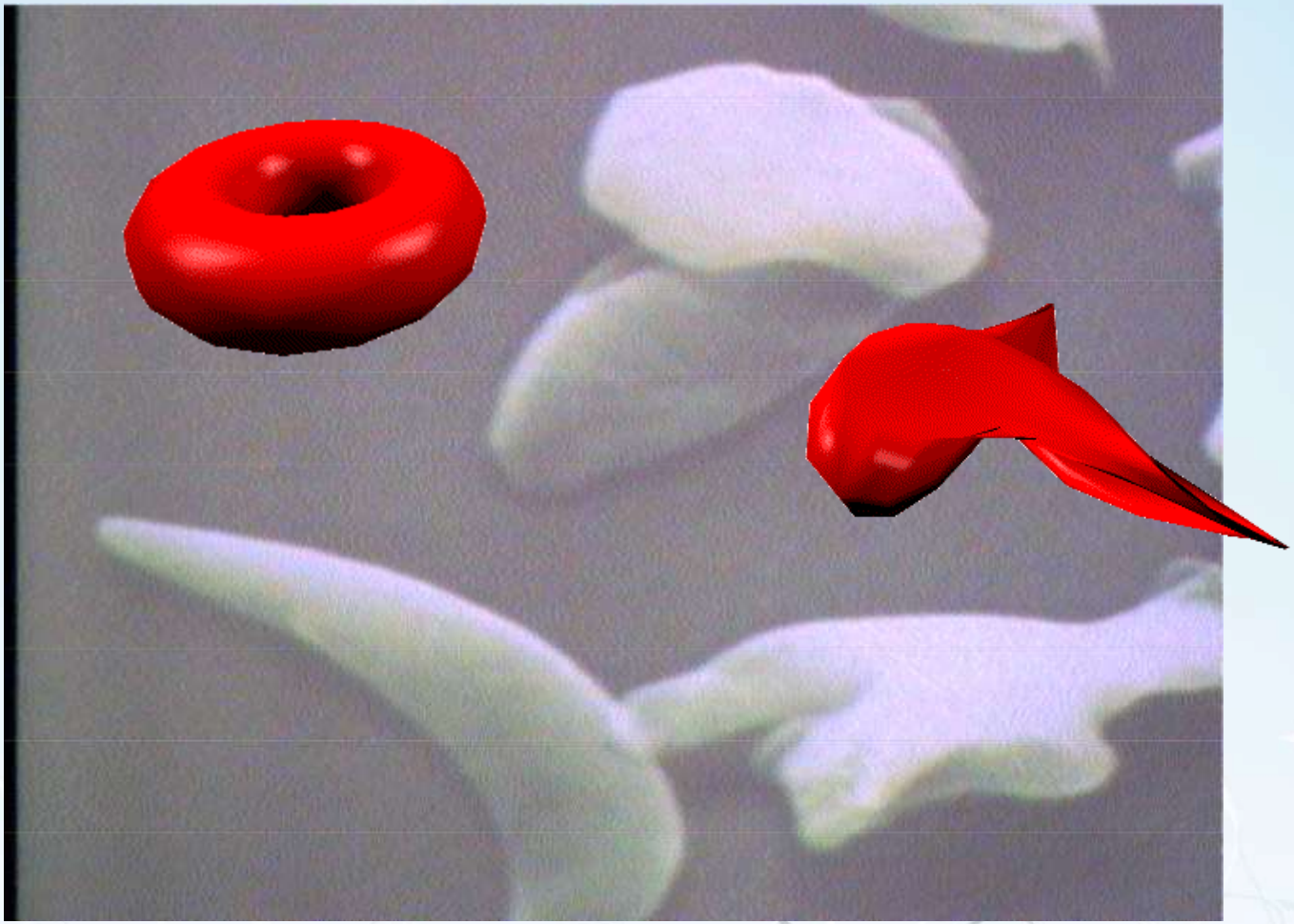
Hemoglobin S

Sickle Cell Disease is found in Africans, Turks, Greeks, Saudi Arabians, Egyptians, Iranians, Italians, Latin Americans and Asiatic Indians.

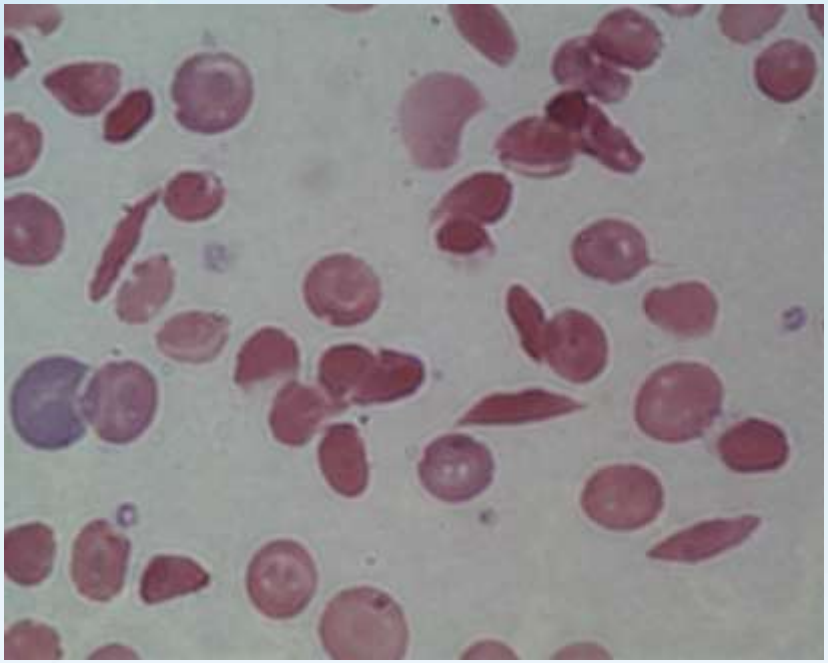


Sickle cell disease is present in one out of four hundred African Americans in the United States. It is the most common genetic disease in this country. All new born babies should be tested at birth for sickle cell disease, so prevention can be started right away. A simple blood test can be done from the baby's blood.

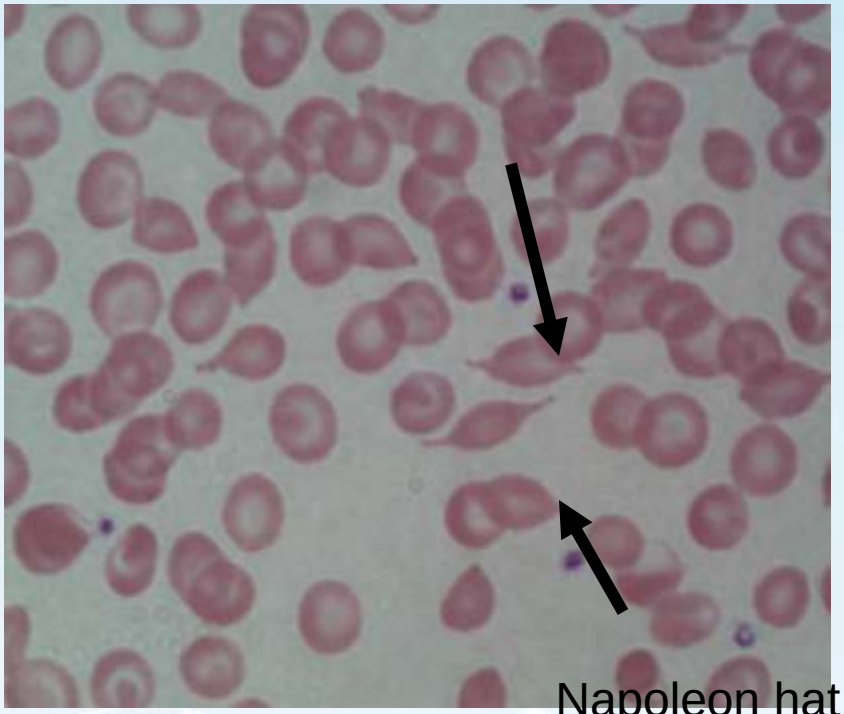
What is Sickle Cell Disease?



Hb S : sickle cell disorder



S/S



S Oman trait

Normal vs. Sickle Hemoglobin

Normal

- disc-Shaped
- soft(like a bag of jelly)
- easily flow through small blood vessels
- lives for 120 days

Sickle

- sickle-Shaped
- hard (like a piece of wood)
- often get stuck in small blood vessels
- lives for 20 days or less

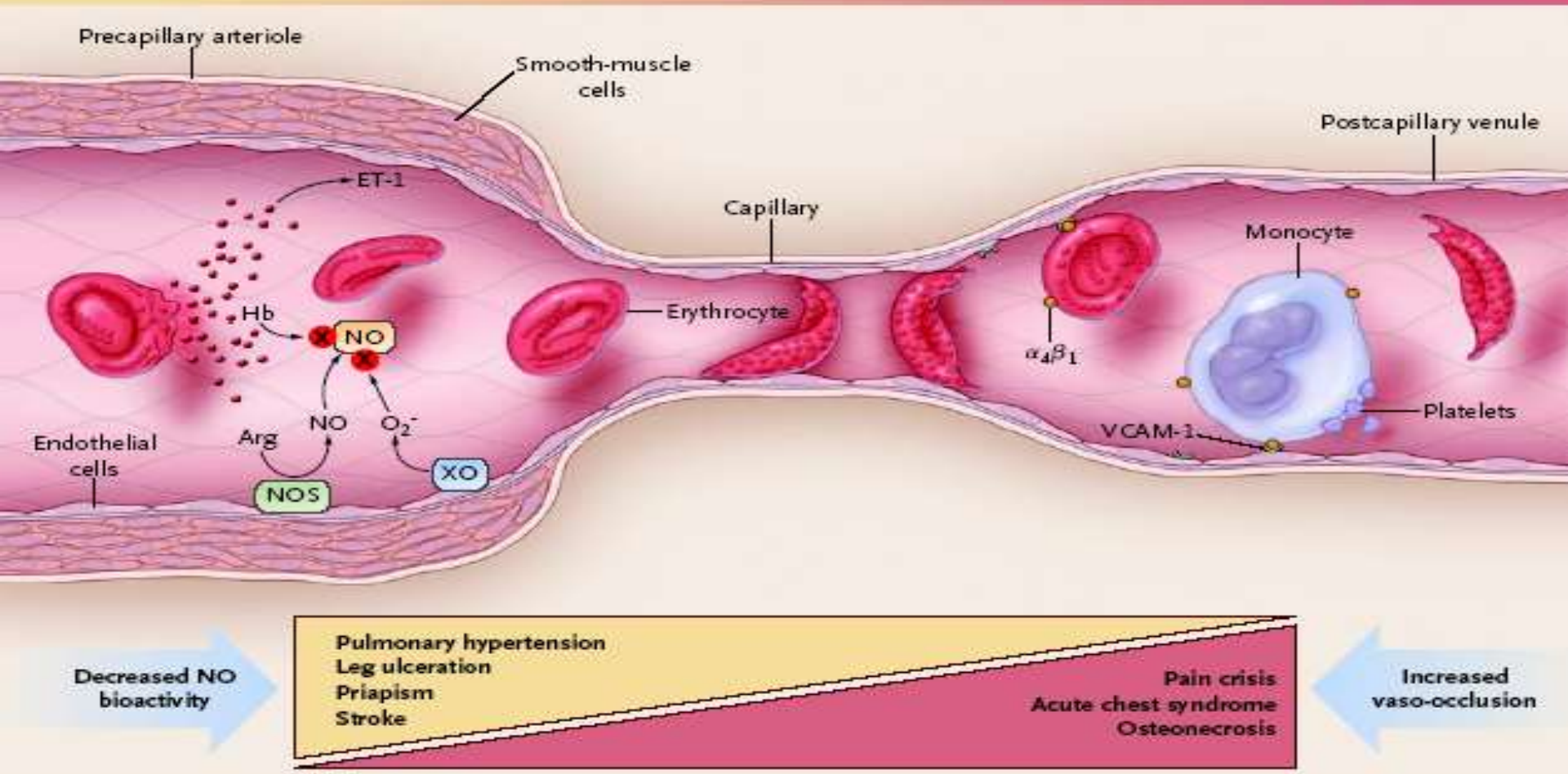


Figure 1. Hypothetical Mechanisms of Clinical Subphenotypes of Sickle Cell Disease.

It is hypothesized that many of the complications of sickle cell disease can be divided into two overlapping subtypes, each driven by distinct mechanisms. Cutaneous leg ulceration, priapism, pulmonary hypertension, sudden death, and stroke are associated with low steady-state hemoglobin (Hb) levels and an increased rate of intravascular hemolysis, shown on the left side of the figure. These vasculopathic complications probably result from endothelial dysfunction, mediated by both inactivation of nitric oxide (NO) by free-plasma hemoglobin and vascular reactive oxygen species as well as arginine (Arg) catabolism by plasma arginase. This process of hemolysis-associated endothelial dysfunction may also cause hemostatic activation and intimal and smooth-muscle proliferation. Such clinical complications as vaso-occlusive pain crisis, the acute chest syndrome, avascular necrosis of bones, and retinal vasculopathy are associated with high steady-state leukocyte counts and high hemoglobin levels. These complications are likely to result from obstruction of capillaries and postcapillary venules by erythrocytes containing polymerized hemoglobin S and by leukocytes (a monocyte is shown), as shown on the right side of the figure. ET-1 denotes endothelin 1, NOS nitric oxide synthase, O_2^- superoxide, VCAM-1 vascular-cell adhesion molecule 1, and XO xanthine oxidase.

Painful crisis

Precipitating factors

Skin cooling		90%
Seasonal	74%	
Rain	50%	
Swimming	40%	
Exercise		66%
Infection		24%
Psychological stress		22%

Avascular necrosis of lumbar vertebrae

Photos courtesy of the late Prof. Lemuel Diggs

Complications From Sickle Cell Disease

- Sickle cells become trapped and destroyed in the spleen causing Splenic Sequestion
- Shortage of red blood cells or anemia
- Pain episodes
- Stroke or Brain Damage
- Kidney failure
- Pneumonia or Chest Syndrome
- Increased Infections

Therapies for SCD

- There are 3 therapies that are of proven value
  - Blood transfusions
  - Hydroxyurea
  - Bone marrow transplant

Healthy Living

- **FARMS**
- F - Fluids, Fever, and Food
- A - Air, and Oxygen
- R - Rest
- M - Medications and Medical care
- S - Situations and Support

in Bahrain

- 1984 The hereditary disease program started in Bahrain;

- 1991: a National Committee for prevention of Genetic disease was formed to plan the control program of these diseases in the country.

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The strategic goals were:

-to reduce the incidence of hereditary diseases in Bahrain,

-and to improve the standard of management and treatment of patients suffering from these diseases.

The Campaign to control SCD included:

1- education and public awareness campaign,

Information booklets were prepared and distributed widely in schools and to the public

2- screening of different categories:

Antenatal, carriers, students, premarital newborns, prenatal screening, newborn screening followed by genetic counseling.

1991, the Bahrain Hereditary Anemia Society was established which strengthen these efforts .

1993, a premarital counseling (PMC) service was started,

1998, a student-screening project began.

2000: A molecular laboratory was established to study the genotype of the difficult cases

Health Education Programme

A comprehensive health education programme Has been launched , to increase public awareness of the diseases and methods to avoid them.

This programme used mass media and target key opinion leaders in society. In schools and other public places

The programme included:

- General publicity campaign through media , periodic education programmes. presentations , leaflets and booklets ,etc

screening for Genetic Blood diseases

To raise awareness about genetic blood diseases, and to identify carriers

1- Antenatal screening(Pregnant women)

2-Premarital counseling

3- Screen school leavers (15-16 graders)

4- prenatal testing.

5- Cascade Screen

6- New born screening

Result:

These efforts continued for the past 25 years.

It had tremendous effect in reducing the prevalence of Genetic Blood Diseases (GBD) among the newborns,

from 20 /1000 to 4/1000,

with 75% decline .

Consanguinity rate also declined gradually due to increase awareness about genetic diseases.

The total consanguinity rate and the first cousin marriages rate.

in 1990 were 39% and 24%. Respectively, while it became 20% ,and 11% in 2007 with 70% decline.

During the screening campaign the ELSI measures were taken care of, such as:
equity,

informed consent,

privacy,

confidentiality

and to prevent stigmatization and discrimination

المشروع الوطني لمكافحة أمراض الدم الوراثية

برنامج
فحص طلاب المدارس

إذا كنت مريضاً أو حاملاً للمرض فتأكد من ارتباطك بشخص سليم

مع تحيات
اللجنة الوطنية لمكافحة الأمراض الوراثية في البحرين

تحت رعاية

سعادة الدكتور عبد العزيز يوسف حمزة

وكيل وزارة الصحة

تقيم وزارة الصحة حفل تدشين المرحلة الحادية عشر من

برنامج فحص الطلاب عن أمراض الدم الوراثية

يوم الثلاثاء الموافق 24 - 2 - 2009م

مدرسة المعرفة الثانوية للبنات

1- student screening for GBD

1999-The project started ,as collaborative project between MOH, MOE. It was initially funded by ROTARY CLUB OF MANAMA.

Later it was funded by BNHAS (cost 240,000 BD)

- The target group was all students at high school(11 grade students).
- It continue for fourteen year.
- More than 88,000 students were screened for genetic blood diseases.
-A card with the results was given to each student.
-The project has been very popular among students and families,
- it resulted in tremendous raise in the awareness among the community. the awareness among the community.

-The aims included:

1- Raising awareness among the youth in Bahrain about hereditary anemia.
-Aiming at empowering them to take the right decision to prevent these diseases in their future families.
2-Determining prevalence of the common genetic blood disorders among Bahraini secondary school students.

The student screening is a collaborative project between

1-The Ministry of Health
2-Ministry of Education
3- Bahrain Hereditary Anemia Society
and initially funded by Rotary club of Manama

-Permission for screening was taken from parents.
-positive response found in 80-85% of families every year.

The students were fully informed about these diseases through educational sessions.

-Technicians visited the school to withdraw the blood samples and these were tested on the same day.
-More than 30000 informational booklets were distributed annually

-High pressure liquid chromatography (HPLC machine) was used to perform the hemoglobin electrophoresis.

-The blood samples were also tested for blood grouping, and G6PD deficiency testing.

-A coded form was designed with the demographic information of each student .it included the results of the tests.

-At the end of the campaign each student received a card with the lab results.

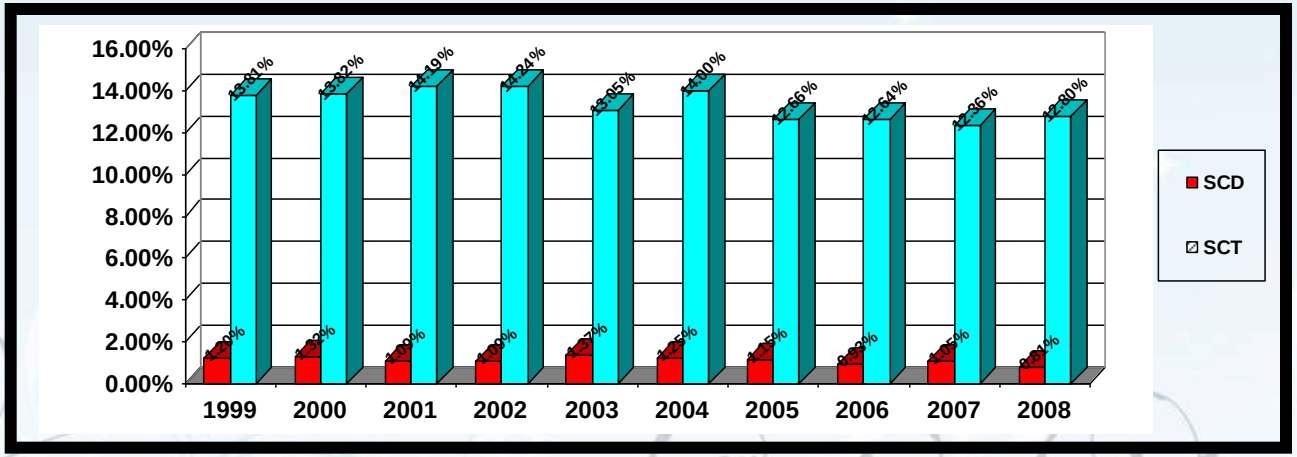
-Reports were also sent to schools M.O.H and M.O.E officials.

Privacy was provided as cards with test results were given to students in a sealed envelope, and were delivered via their school, to be opened at home. In order to protect privacy, confidentiality, and prevent stigmatization.

Screening of Student for Genetic Blood Disorders (1999 – 2008): Prevalence of Abnormal Hemoglobin

Year	1999		2000		2001		2002		2003	
Disease	No	%	No	%	No	%	No	%	No	%
SCD	68	1.20%	75	1.32%	68	1.09%	64	1.09%	74	1.37%
SCT	785	13.81%	787	13.82%	886	14.19%	838	14.24%	706	13.05%
No. of Students	5685		5694		6244		5894		5418	

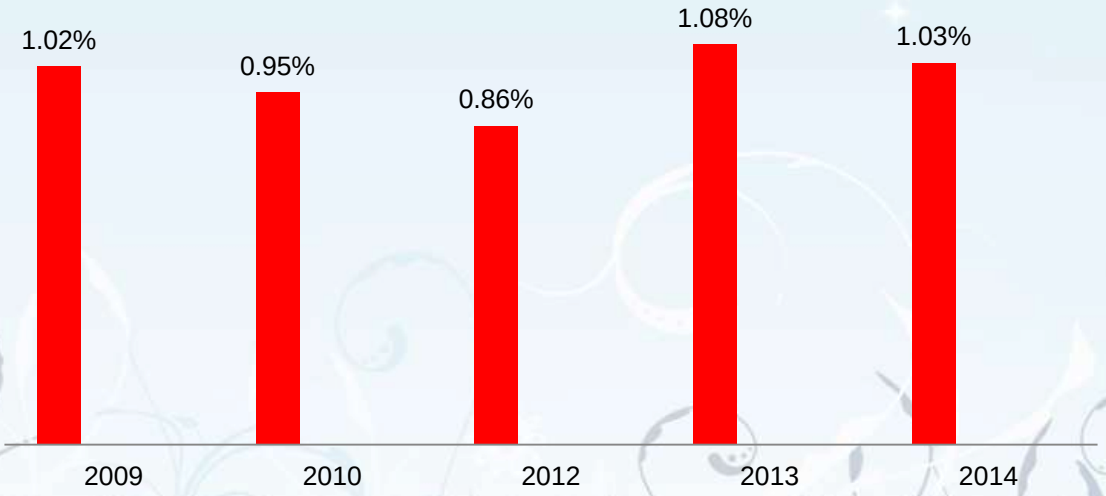
Year	2004		2005		2006		2007		2008	
Disease	No	%	No	%	No	%	No	%	No	%
SCD	78	1.25%	73	1.15%	59	0.93%	67	1.05%	50	0.81%
SCT	873	14.00%	805	12.66%	803	12.64%	788	12.36%	789	12.80%
No. of Students	6273		6358		6352		6376		6166	



Screening of Student for Genetic Blood Disorders (2009– 2014): Prevalence of Abnormal Hemoglobin

Year	Total no.	No. Of patient	Percentage
2009	5877	60	1.02%
2010	5920	56	0.95%
2012	9565	82	0.86%
2013	4554	48	1.08%
2014	5029	52	1.03%

Percentage Of Sick patient Student Screening Program



أهداف

المشروع الوطني لمكافحة أمراض الدم الوراثية

- رفع وعي المجتمع عن أمراض الدم الوراثية .
- التأكيد على أهمية الفحص قبل الزواج .
- إعطاء كل طالب بمرحلة الثاني ثانوي بطاقة تعرفهم عن وضعهم الوراثي لأمراض الدم الوراثية .

إذا كنت مصابا أو حاملا للعامل الوراثي فتأكد من ارتباطك بشخص سليم

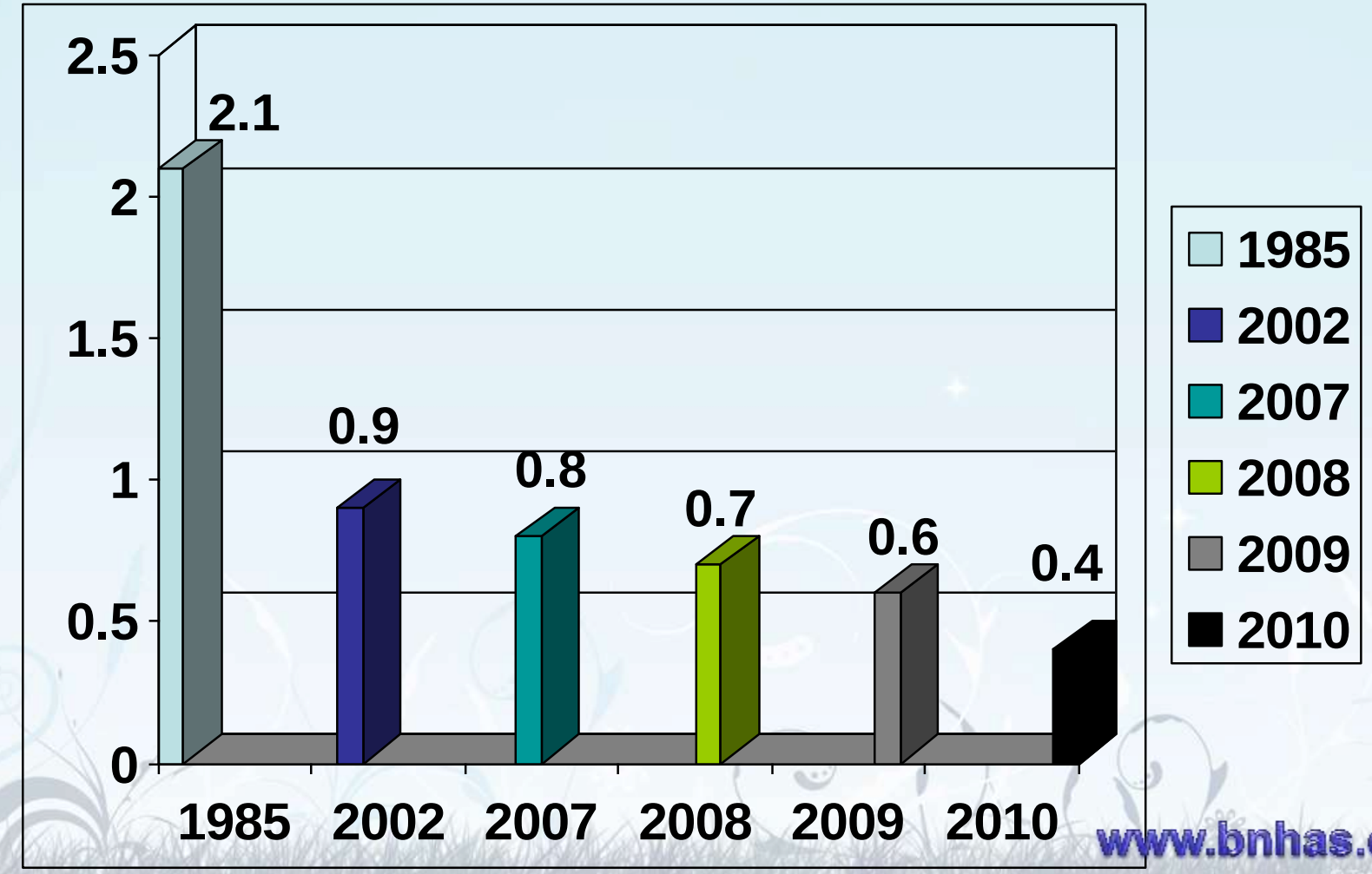
المشروع الوطني لطلبة المدارس لمكافحة أمراض الدم الوراثية

قسم التثقيف الصحي

www.bnhas.org

PERCENTAGE OF SCD AMONG NEWBORN

YEAR	1985	2002	2007	2008	2009	2010
PERCENTAGE OF SCD	2.1%	0.9%	0.8%	0.7%	0.6%	0.4%



نادي روتاري المنامة يتصدر قائمة الأندية الدولية عن مشروع مكافحة أمراض الدم الوراثية>1999

- وحصل نادي روتاري المنامة على كأس ((كمال بدوي)) لأحسن مشروع قدم لخدمة المجتمع وهو المشروع الوطني الذي تبناه نادي روتاري المنامة لمكافحة أمراض الدم الوراثية المتمثلة في الأنيميا المنجلية الثلاسيميا وأنيميا نقص الخميرة، والتي تعتبر من أهم المشاكل الصحية التي تواجه العائلات في البحرين ومنطقة الخليج العربي والتي تنعكس سلبياً على النواحي الاجتماعية والاقتصادية.
- وقد تسلم أحمد ميرزا علي جواهري رئيس نادي روتاري المنامة الكأس من دوروس جوايولوس محافظ المنطقة.

Conclusion:

The ethical legal social issues have been implemented during the campaign to control GDB in Bahrain. The campaign gets support of the community and the policy makers, and was successful in reducing the rate GBD in the country.

we need to continue this program for few years to get the maximum benefits .

More efforts are needed to reduce the rate of other genetic diseases, and to improve the treatment to the patients .

THANK YOU

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