



19/06/2020

sickle cell disease

World Sickle Cell Day 19th JUNE

- The 19th june is World Sickle Cell Day and this year the Sickle Cell Society is celebrating 40years of working in the sickle cell community.
- World Sickle Cell Day is a United Nation's recognised day to raise awareness of sickle cell at a national and international level.

SPORTS AND SICKLE CELL DISEASES

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Sickle Cell Disease

- Terminology
- Epidemiology
- Complications
- Screening/Diagnosis
- Management
- Return to Play

Sickle Cell Disease

- Normal adult hemoglobin

- ✓ $2 \alpha + 2 \beta = \text{Hb A tetramer}$

- Hémoglobine S

- ✓ Mutation in the beta-globin chain

Poorly soluble hemoglobin, which forms long, inflexible chains when deoxygenated

- ✓ Inflexible hemoglobin chains

Stiff & sticky (sickle) red blood cells

- ✓ Sickle cells logjam in small vessels

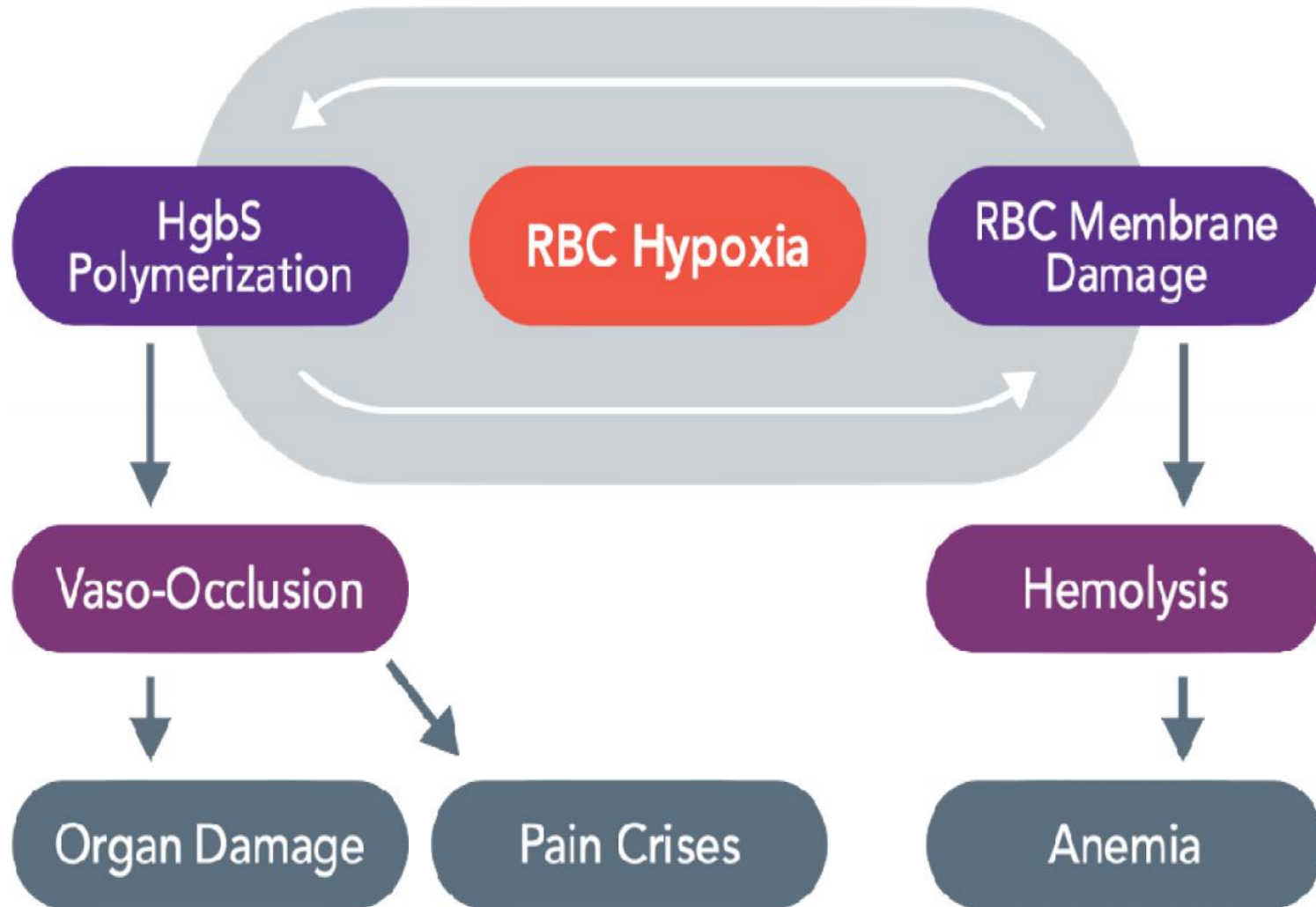
About sickle cell disease

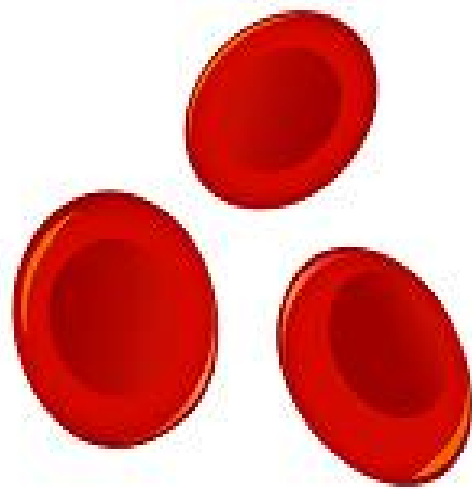
- ▶ Sickle cell Anaemia / Drepanocytosis.
- ▶ Various cell genotypes or variants of the sickle cell syndrome:
 - Hb AS (SCT) Hb SD- Punjab
 - Hb SS etc
 - Hb SC
- ▶ Genetic or hereditary blood disorder
- ▶ Inheritance -Autosomal recessive
- ▶ Perpetuation of the sickle cell gene can be controlled.

Genotype

- Sickle cell disease
 - ✓ All conditions associated with sickling
- Sickle cell anemia (Hb SS)
 - ✓ Homozygous for hemoglobin S
- Hemoglobin SC disease (Hb SC)
 - ✓ Hemoglobin S and hemoglobin C (typically 50:50)
- Sickle cell trait (Hb AS)
 - ✓ Hemoglobin A and hemoglobin S (typically 60:40)

PATHOPHYSIOLOGY SSA

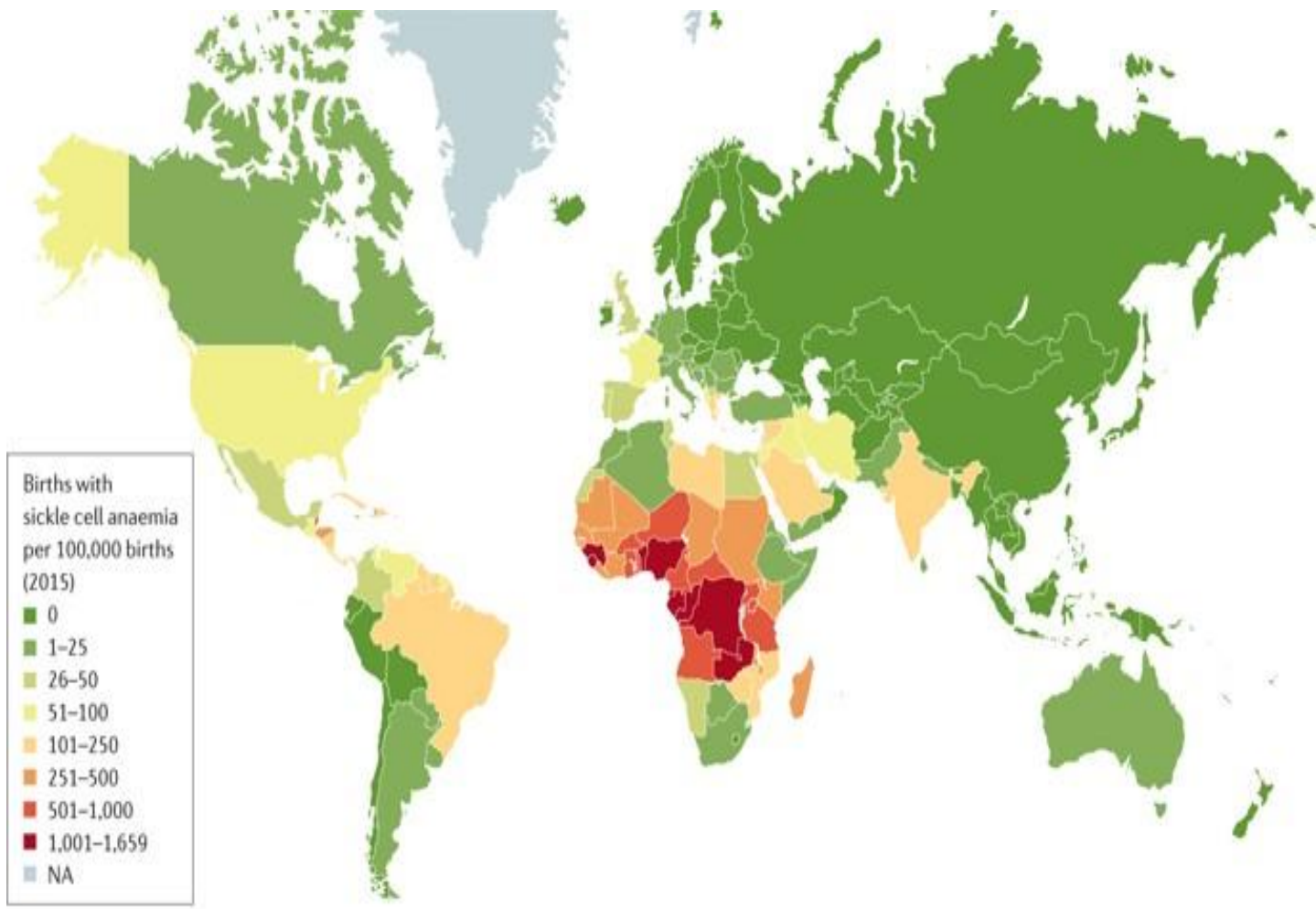




Normal
Red Blood Cell



Sickled
Red Blood Cell



Sickle Cell Disease: A Global Public Health Issue

COUNTRY	SICKLE CELL BIRTHS/YEAR
Nigeria	91,011
Democratic Republic of the Congo	39,743
Tanzania	11,877
Uganda	10,877
Angola	9,017
Cameroon	7,172
Zambia	6,039
Ghana	5,815
Guinea	5,402
Niger	5,310
Sub-Saharan African Total	242,187
Worldwide Total	305,773

www.MyThreeSicklers.org

Phenotype Sickle cell anemia (SCA)

- ✓ Numerous complications; rarely compete in sports
- Most common complication is pain crisis
- Anemia, infection, stroke, CVD, renal disease, leg ulcer, priapism
- Life expectancy: 45 years
- ✓ No reported cases in NCAA athletes

✓ Hemoglobin SC disease

- ✓ Less severe than SCA; more severe than SCT
 - Most common complication is pain crisis
 - At risk for same complications above—much less common
 - Life expectancy: 64 years sometimes more
- ✓ Four reported cases in NCAA athletes
 - All eventually retired due to complications
 - **Sickle cell trait**
 - “Not a disease;” athletes compete in sports with few complications
 - Roughly 2,000 current NCAA athletes

Question

- ✓ What can go wrong with these athletes?
=Complications of SCT
- ✓ **Royce Moore, MD** UK Sports Medecine :
- ✓ Gross hematuria
- ✓ Venous thromboembolism
- ✓ Splenic infarction
- ✓ Exertional collapse

- **Gross hematuria**
- Results from sickling deep in the renal medulla
Initial therapy is relative rest and hydration
Typically resolves within two weeks

Venous thromboembolism

Studies conflict on the relative risk of VTE in athletes with SCT compared to those without

- Range from 1.5-4 times relative risk
- Monitor athletes with immobilizing injuries for deep venous thrombosis especially those taking oral contraceptives

Splenic infarction

- Hypoxic splenic sickling infarct, typically at altitude
- Most cases occur with mild to moderate activity
- ✓ Consider **oxygen** for long flights/road trips to altitude
- ✓ Think of SCT splenic infarction in **anyone** who develops left lower chest pain while at altitude
- ✓ Diagnosed early, it responds to Conservative therapy, including descent

Exertional collapse

- NCAA Division-1 football
 - ✓ 16 deaths from 2000-2010
- All from conditioning; zero from practice/play
- 10 (63%) attributed to exertional sickling
 - ✓ SCT independent risk factor
- Black athletes with SCT 37 times more likely to experience exertional death than those without SCT

Exertional collapse

- Perfect storm of undue exercise intensity, sustained for at least a few minutes, and a heroic effort beyond the physical limits of an athlete on any given day.

- ✓ Intense exertion → Hypoxemia → Sickling

- ✓ Sickle cells → Muscular ischemia → Rhabdomyolysis

- Deaths are largely due to rhabdomyolysis

- ✓ Acute renal failure

- ✓ Hyperkalemia

- ✓ Fatal arrhythmias

Prevention

- Athletes with SCT should train *consciously*...

Set their own pace

- ✓ Engage in a slow and gradual preseason conditioning regimen
- ✓ Build up slowly while training, allowing adequate rest and recovery between repetitions
- ✓ Be excused from performance tests such as serial sprints or timed mile runs, avoiding all-out exertion beyond 2-3 minutes without a break
- ✓ Access supplemental oxygen at altitude as needed

Stay well hydrated at all times, especially in hot and humid conditions

- ✓ Maintain proper asthma management
- ✓ Refrain from extreme exercise during acute illness
- ✓ Stop activity immediately upon struggling or experiencing symptoms such as muscle pain, weakness, undue fatigue, or breathlessness

Management

- In the event of a sickling collapse...
 - ✓ Be prepared with an Emergency Action Plan
 - ✓ Check vital signs
 - ✓ Administer oxygen
 - ✓ Cool the athlete if necessary
 - ✓ Conditioning the patient and transport the athlete to the Emergency Unit and check for rhabdomyolysis

Return to play

Limited evidence supports recommendations

- ✓ Once the athlete is asymptomatic at rest and has normal end-organ function, re-visit precautions for safe participation.
- ✓ If the athlete desires to resume activity, allow a gradual supervised return to activity as tolerated

Recommendations ACC TF4

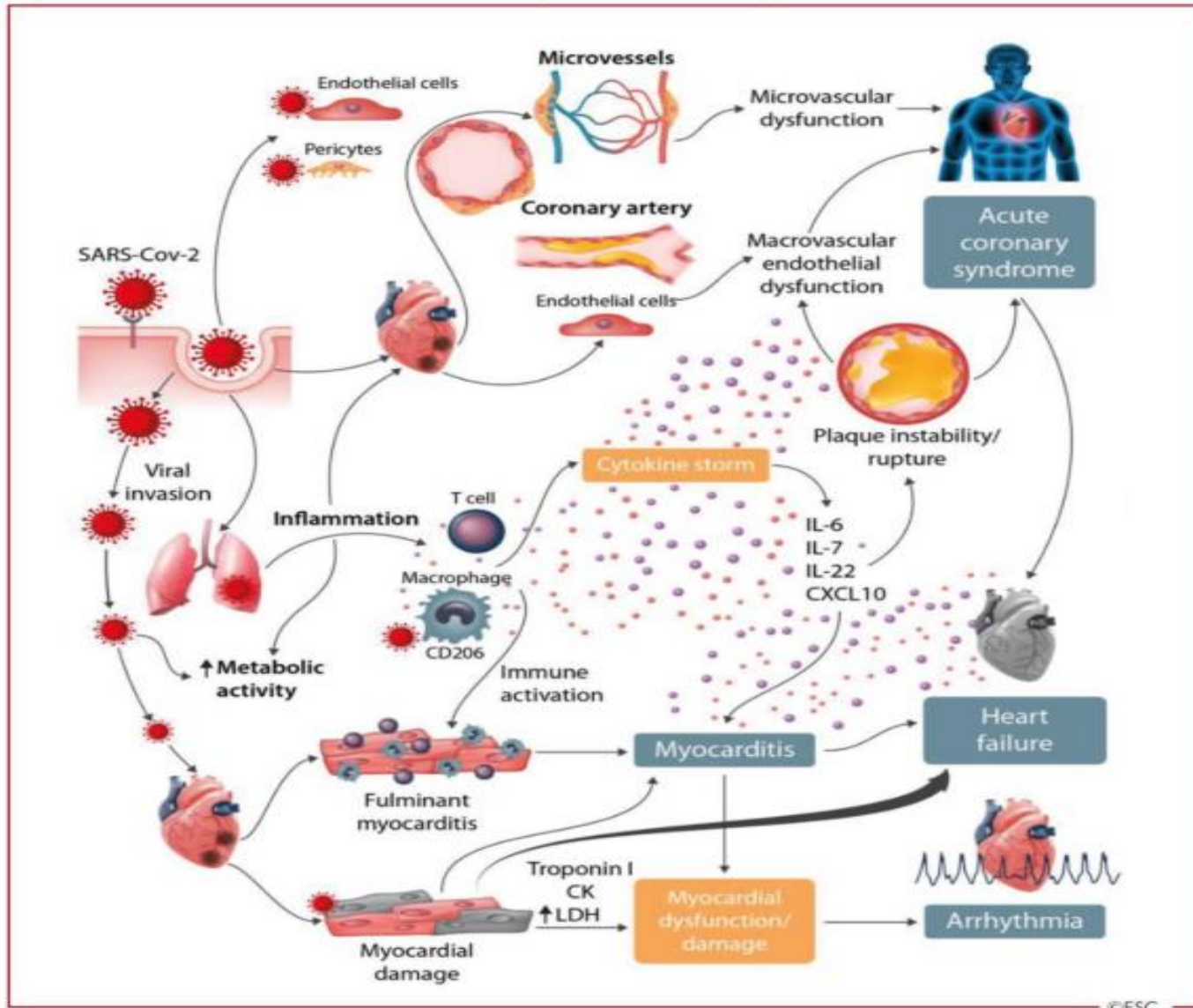
- 1) Recognition of SCT status is not itself a justification for disqualification from competitive sports (Class I; Level of Evidence C).
- 2) Recommended preventive strategies (including adequate rest and hydration) should be performed to minimize the likelihood of an event occurring on the athletic field in a person known to have SCT (Class I; Level of Evidence B).
- 3) It is critical to be prospectively aware of acute emergency medical strategies should suspicion of an emerging event arise in an athlete known to have SCT (Class I; Level of Evidence C).

4) Particular caution should be exercised for athletes known to have SCT who are competing or training in high environmental temperatures or at extreme altitude (Class I; Level of Evidence C).

SPORTS, SICKLE CELL DISEASES AND INFECTIONS

Most common infections in Africa :

- ✓ Malaria
- ✓ RHD due to streptococcal infection Other bacterial infections
- ✓ Viral infections: HIV
- ✓ Corona virus infection with endothelitis+++
- ✓ Most of these infections occur with anemia in African context.



Sports practice after Covid19

Resumption of physical activity must be progressive in duration and intensity in order to readjust the body to the effort (heart, muscles, tendons) without forgetting the usual hydration during the effort.

IN ALL CASES, YOU MUST ALWAYS STOP ALL PHYSICAL ACTIVITY AND QUICKLY CONSULT A PHYSICIAN BEFORE THE APPEARANCE OF THE FOLLOWING WARNING SIGNS:

chest pain (in the chest);

dyspnea: abnormal shortness of breath;

palpitations: feeling that your heart is beating too fast or irregularly; abnormal change in heart rate at rest or during exercise;

✓ sudden loss of taste and / or smell;

✓ abnormal fatigue;

✓ temperature greater than or equal to 38 ° at rest at a distance from the activity;

✓ resumption or appearance of a dry cough.

Carefully respect the conditions and modalities of resumption of aps fixed by the government, in particular by applying barrier measures and rules of physical distancing:

- ✓ Only individual sports activities practiced outdoors can be authorized in the current state of the pandemic of corona virus 19.
- ✓ Sports activities involving or promoting contact between people are not allowed.

The rules for physical distance are as follows for sporting practice:

- ✓ All sports activities involving or promoting contact between athletes are not allowed
- ✓ it is advisable to provide between two people a contactless space beyond 1 meter :

- ✓ 10 m for cycling and running;
- ✓ 5 m for fast walking;
- ✓ 1.50 m laterally between two people;
- ✓ for other activities, provide a space of 4 m² for each participant.

Barrier measures must be maintained:

- ✓ Frequent hand washing with soap or hydro alcoholic gel;
- ✓ Snacks and hydration must be managed individually (personalized bottles, etc.);
- ✓ The exchange or sharing of personal effects (towel ...) must be prohibited;
- ✓ The use of personal sports equipment is preferred, otherwise, the common sports equipment is cleaned and disinfected before and after each use;

✓ Wearing a mask makes it difficult to practice many sports. It is however justified in certain situations where the distancing measures could not be strictly respected.

follow the following tips:

- Respect the 10 golden rules of sports cardiologists;
- Do not take paracetamol as a preventive measure (risk of masking the fever);

Do not take anti-inflammatory drugs including aspirin and ibuprofen without medical advice;
do not self-medicate with hydroxychloroquin;
Do not practice alone in isolated areas and / or difficult to reach by the emergency services;
Monitor your temperature regularly at rest, away from exercise.



Thank you very much