## Sickle Cell Anemia

November 9th, 5-6pm By: Destiny E., Cora H., Grant H., Tanaya J., Sophie K., Shyann M., Jaida M., Hailey M., Chidozie O., & Richie W.



#### What is Sickle Cell Anemia?

- Inherited disease of Anemia
  - Not enough healthy, red blood cells (erythrocytes) to carry oxygen throughout the body
- Affects a protein (hemoglobin) inside the red blood cells
- Red blood cells become rigid, sticky, are shaped like sickles or crescent moons
  - These can get stuck in blood vessels



#### History of Sickle Cell Anemia

- In 1910, Dr. James B. Herrick and Dr. Ernest Irons discovered sickle shaped red blood cells within a patient in the United States.
  In 1927, Hahn and Gillespie found that red blood cells gained the sickle shape from the lack of oxygen.
- In 1951, Dr. Linus Pauling and Dr. Harvey
   Itano discovered hemoglobin which helps
   to identify numerous blood diseases.



#### Normal Hemoglobin Vs. Sickle Cell Hemoglobin

- Disc-shaped
- Soft
- Easily flows through blood vessels
- Lives for 120 days

- Sickle-shaped
- Hard
- Often gets stuck in blood vessels
- Lives for 20 days or less





#### Genetics of Sickle Cell Anemia

#### Trait

- Offspring can inherit the sickle cell trait from one of their parents.
- This happens to people who inherit 1 sickle cell gene and 1 normal gene.
- A person who has the trait will be able to pass it onto their offspring.
- Usually, no symptoms are present.

#### Disease

- Offspring can inherit the sickle cell disease from both parents.
- This happens to people who inherit 2 sickle cell genes (1 from each parent).
   A person who has the disease can pass this and the sickle cell trait to their offspring.
  - Symptoms are present.

#### Who is Affected

Sickle Cell Anemia affects around 100,000 Americans alone:

- 1 out of every 365 African American
  - 1 out of 13 African American babies are born with the sickle cell trait
- 1 out of every 16,300 Hispanic American



#### Mechanism of Sickle Cell Anemia

Caused from a mutation that produces iron rich hemoglobin to the bloodstream.

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- The irregular shaped hemoglobin leads to the red blood cells to deform their original shape.
- Sickle cell anemia can lead to strokes, pulmonary hypertension, and even blindness in patients.



### Symptoms of Sickle Cell Anemia

- Joint pains (sickle cell crisis)
- Swelling in hands and feet
- Shortness of breath
- Low haemoglobin (Anaemia)
  - Weak immune system
  - Jaundice
  - Fatigue



#### Treatments

Management of sickle cell anemia is usually aimed at avoiding pain episodes, relieving symptoms and preventing complications:

- Hydroxyurea-reduces the frequency of painful crisis and might reduce the need for blood transfusions and hospitalizations.
- L-glutamine oral powder- helps in reducing the frequency of pain crises.
- Crizanlizumab-helps reduce pain.
- Voxelotor-improves anemia in people with sickle cell disease.



#### Case Study

Patient: Male, 27

Final Diagnosis: <mark>Myonecrosis of sickle cell anaemia</mark>

Symptoms: Pain • redness to feet • swelling foot

Medication: —

**Clinical Procedure: MRI** 

**Specialty: Podiatry** 

#### Background

In myonecrosis, red cells containing sickle hemoglobin become rigid, resulting in reduced blood flow and myonecrosis.

#### Case Report

A patient in sickle cell crisis with an episode of acute pain and swelling to the intrinsic muscles of the foot as a prominent feature of the crises.

Vascular insufficiency to the muscles was found in an MRI scan of the patient's foot, resulting in diminished warmth to the area.

This swelling, often along with a fever, is caused by the sickle cells getting stuck in the blood vessels and blocking the flow of blood in and out of the hands and feet.



#### Patient



Turaga, L. P., Boddu, P., Kipferl, S., Basu, A., and Yorath, M. Myonecrosis in Sickle Cell Anemia: Case Study. *PMC* Vol. 18, 100-103 (2017)

#### What is my Hemoglobin Type?

-A hemoglobin test requires a medical professional to draw blood from the arm to send to the lab or they may choose to do a finger stick test.

-A hemoglobin test measures how much hemoglobin is in the blood. Abnormal levels could indicate anemia or another blood disorder.

-These tests are prescribed by your doctor.



# Now Kahoot!

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