Sickle-Cell Anemia

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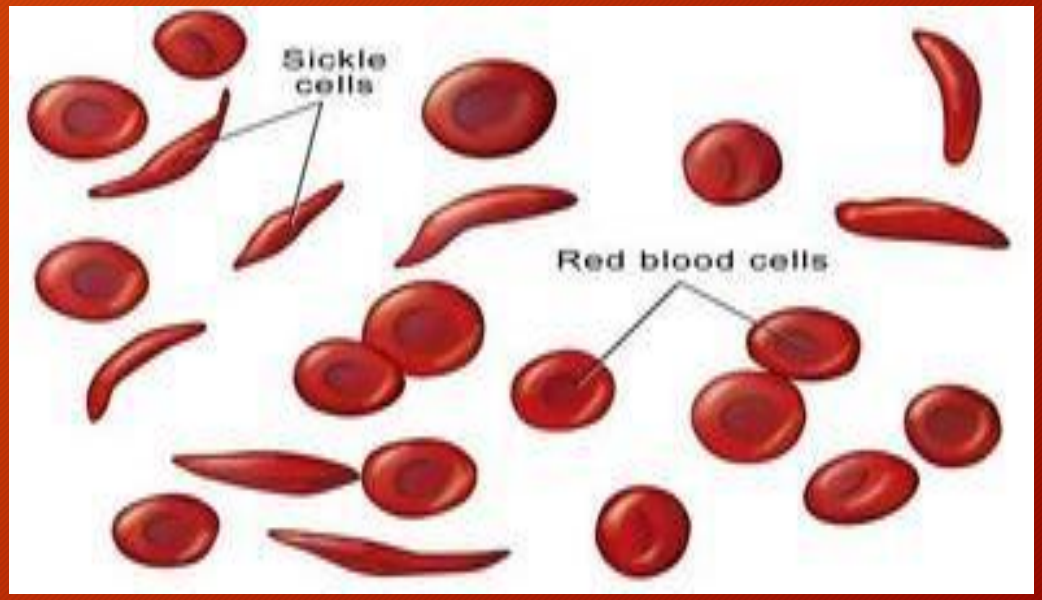
SICKLE STRONG INITIATIVE

What is Sickle-cell Anemia?

- An inherited disease that results in the production of abnormal hemoglobin in red blood cells (Hemoglobin SS)
- Hemoglobin is a protein that binds to and helps transport oxygen to the body's tissues
- When hemoglobin is abnormal, it forms rods in the red blood cell and causes it to become crescent- or sickle-shaped

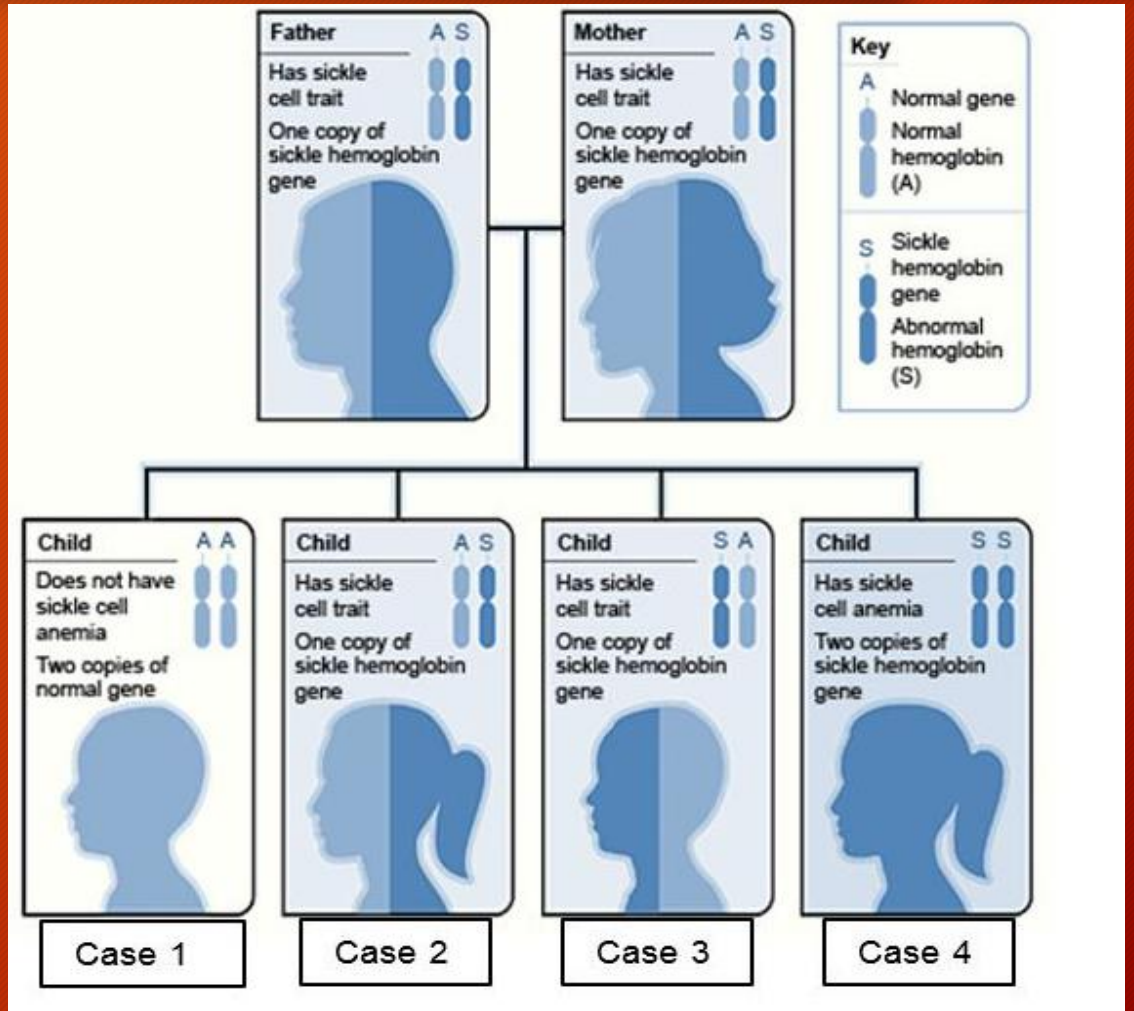


What does this mean in the body?

- Severe pain and swelling: The sickle-shaped cells will often get stuck in blood vessels and will be unable to transfer its oxygen to the tissues
- Anemia: Inefficient red blood cells are destroyed by the body and the rate of destruction for sickle red blood cells is often faster than the rate of production, meaning the overall amount of red blood cells in the body is less than it should be
- Organ damage: the inefficiency in oxygen delivery can also affect the spleen, brain, eyes, lungs, liver, heart, kidneys, penis, joints, bones, or skin
- Acute chest syndrome: sickling in the blood vessels of the lungs that results in chest pain, hypoxia (low oxygen), cough, fever and lung infiltrates

How do you get it?

- It is inherited meaning that you can only get it if both of your parents carry mutated hemoglobin genes, Hemoglobin S
  - Genes are segments of your DNA that code for a particular protein in the body
- If both of your parents are carriers, meaning they have one normal gene and one HbS gene, their children have a 1 in 4 or 25% chance of inheriting 2 copies of HbS gene. They will have sickle cell anemia
- Even though a carrier has one mutant gene, they do not experience symptoms of sickle cell anemia
  - Carriers have been shown to have a tolerance of or protection to malaria



How do you know if you have sickle-cell anemia?

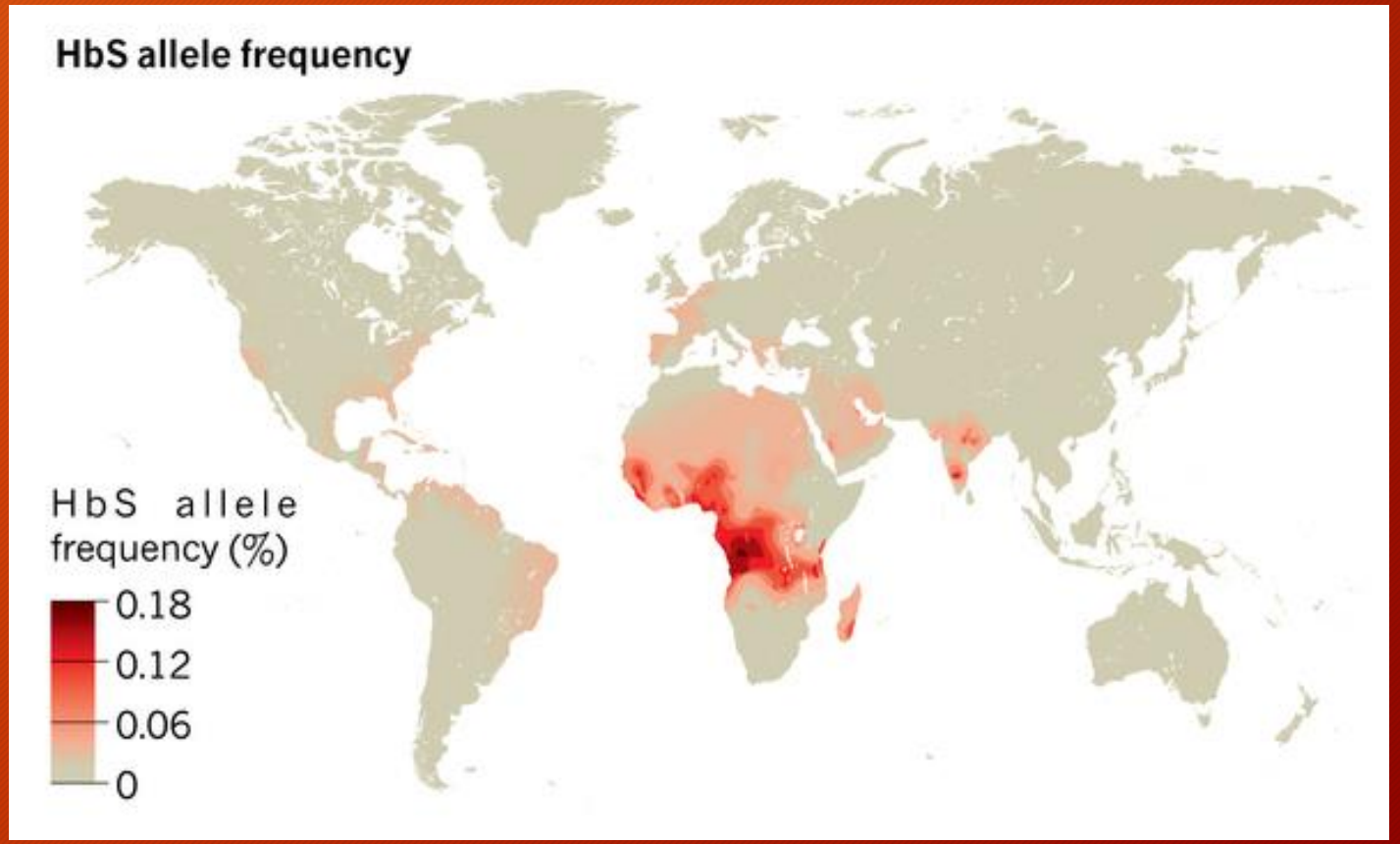
- Adults and newborns can undergo a blood test to see if they carry sickle hemoglobin
- Prenatal screening is also possible 8-10 weeks into a pregnancy to see if the abnormal hemoglobin gene is present in the amniotic sac or placenta
- EARLY DETECTION MEANS EARLY INTERVENTION to provide the best quality of life for the child

What are the treatments for sickle cell anemia?

- Early intervention includes: penicillin, vaccinations and folic acid supplementation
- Treatment depends on the severity and specific symptoms felt by the person suffering from sickle cell anemia
- Antibiotics and Vaccinations: due to damage to the spleen, people with SSA are more susceptible to infections
- Oral or IV Fluids: to decrease swelling and pain due to clogged blood vessels
- Blood transfusions: If suffering from severe anemia
- Hydroxyurea: found to decrease the number of pain crises, need for transfusions and frequency of acute chest syndrome

Who is at risk of obtaining SSA?

- People with sickle cell anemia exist all over the world
- It is most common in West and Central Africa
  - 25% of the people have sickle cell trait and 1-2% of all babies are born with a form of the disease
- Adults and children suffer from sickle cell anemia, but in places without access to early intervention or proper treatment, **most children do not make it to adulthood**
- There are reports that suggest 50-90% of children born with SSA die before their 5th birthday



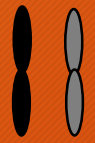
Links to resources used

- http://www.nature.com/nature/journal/v515/n7526_supp/full/515S2a.html
- http://www.sicklecelldisease.org/index.cfm?page=scd-global
- https://www.nhlbi.nih.gov/health/health-topics/topics/sca/treatment

Testing your knowledge…

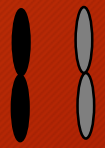
- Which protein is abnormal in sickle cell anemia?
- What is a gene?
- If someone coughs or sneezes on you and they have sickle cell anemia, can you get it?
- Name one important early intervention treatment.
- Name one symptom and its treatment for SSA.
- Who is most affected by sickle cell anemia?
- What do red blood cells do?

Your father has one normal hemoglobin gene and one abnormal hemoglobin gene, HbS



Hb HbS

Your mother has one normal hemoglobin gene and one abnormal hemoglobin gene, HbS



Hb HbS

List the 4 possible genetic outcomes for you:

1. _1_ normal hemoglobin gene, _1_ abnormal hemoglobin gene = _C_
2.
3.
4.

C= carrier, SSA = sickle cell anemia, ND = no disease