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Sickle Cell Anemia

Sickle cell anemia is a genetic red blood cell disorder that affects people the around the world every day. This particular blood disorder has a specific shape, is classified as a hemoglobin abnormality that is incurable and often times can be fatal. It is not contagious but is transmitted by one's genes from parents to their children. To better understand sickle cell anemia, one must understand the genetic aspect of this blood disorder as well as what a blood disorder entails.

The overall definition for anemia is defined as being a condition where you do not carry enough healthy red blood cells to carry enough oxygen to a person's body tissues. When a person has anemia or any type of classified anemias, they have a low hemoglobin. What that means is you will often feel tired and weak because of the lack of this protein in your red blood cells that is meant to carry oxygen and carbon dioxide to integral parts of the body. A person with sickle cell anemia has abnormal hemoglobin that forms stiff rod that affect the way oxygen is passed on to the red blood cells. When a person has sickle cell anemia, their red blood cells are sickle shaped or have an appearance of a crescent moon. Compared to normal red blood cells

that are disc shaped and flexible so that they can move freely through the blood vessels; a person with sickle cell anemia has cells that are not flexible and do not bend or move easily making it difficult for blood to flow properly to the rest of the body. When inherited, a person will two abnormal hemoglobin genes from each parent. The gene is called hemoglobin S or HbS. Subsequently, the lack of proper oxygen flow or blockage to flow through the blood vessels effectively other organs are then also affected, such as the liver the kidneys, the gallbladder, the eyes, the heart, the bones and the joints. This can then result in pain and organ damage.

With sickle cell anemia you can also carry the trait and thus pass it down to your child if the other parent also carries the trait. This normally would characterize the person that has the sickle cell trait as being a carrier of the hemoglobin S gene. “A 25%, or 1 in 4, chance of inheriting two normal hemoglobin A genes. This child does not have sickle cell trait or disease. A 50%, or 1 in 2, chance of inheriting one normal hemoglobin A gene and one hemoglobin S gene. This child has sickle cell trait. A 25%, or 1 in 4, chance of inheriting two hemoglobin S genes. This child has sickle cell disease.” (National Heart, Lung, and Blood Institute).

For sickle cell anemia disease, the only current treatment and cure for some people would be to get a blood and bone marrow transplant. There are varies medications that are used for early prevention, to reduce pain once diagnosed with the disease, medications to further reduce other complications, as well as blood transfusions. Voxelotor is one of the medications used to prevent the sickling or forming of a crescent shape of the red blood cells. It is an oral medication used to treat adults and children of 4 years of age or older. With this medication it reduces the damage of the red blood cells which in return lowers the changes of anemia which then improves blood flow. Crizanlizumab-tmca is another medication used to prevent the sickling or forming of a crescent shape of the red blood cells. This medication is given intravenously and has only been

approved for adults and children of 16 years of age and older. Various other medications include Hydroxyurea, an oral medication that aids with the complications of the disease. Penicillin can also be given to lower the risk of infection. When it comes to the blood and bone marrow transplant, it is currently only performed on children who have had severe complications. The transplants are known to have an 85% success rate when the donor is related. There are also some genetic therapy treatments that are in the workings to better treat sickle cell anemia.

Although sickle cell anemia is and inherited disease there are efforts being made to correct the abnormality in the gene created. Along with be aware of your family history and what treatments and tests that are constantly being done we have a far way to go but we are making progress. When trying to decide what disease I would choose I knew I wanted it to be one of the blood diseases/disorders. I have always been fascinated with the science of it when it comes to genetics and DNA. Another reason for me was the fact that I have a friend with sickle cell anemia who while growing up I always wondered why she was always sick and was always in and out of the hospital. Later in life I also came to the information that my dad had the sickle cell trait as well and passed it on to my younger sister. So just wanting to better understand and educate myself on this disorder has always been one of my goals.

References

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