Cayman Islands Health Services

PRESS RELEASE

What is Sickle Cell Disease?

Sickle cell disease is an inherited chronic disorder that affects red blood cells. It has a worldwide distribution, and is one of the most common genetic disorders.

All persons have two genes that make haemoglobin. Normal red blood cells contain haemoglobin A, a protein that helps red blood cells carry oxygen around the body. With sickle cell there is a different form of protein, haemoglobin S. With sickle cell disease, both genes are affected, causing severe symptoms.

Normal red blood cells are round, flat and very flexible. However, when the oxygen comes out of the red blood cells of sickle cell disease, the cell becomes stiff and takes on the shape of a sickle – hence, the name. The sickle cells clump together, are not able to squeeze through the small blood vessels, and so the sickle cells get destroyed more quickly. A normal red blood cell lives approximately 120 days but a sickle cell may only live 11 or 12 days.

What is Sickle Cell Trait

When only one gene is affected, it is called sickle cell trait, or persons are called sickle cell carriers. Having sickle cell trait means that the person stays healthy under normal circumstances, and the main significance is that it can be passed down to one’s children. However, under certain extreme circumstances, a person with the trait may experience complications as if having sickle cell disease. Persons with just the trait cannot later develop the disease.

Persons with the trait can, under extreme conditions, have some symptoms, so it is recommended that persons with sickle cell trait:

* Avoid high altitudes
* Avoid scuba diving and free diving
* Drink adequate fluids to avoid dehydration
* Pace themselves when exercising/playing sports
* Rest as needed when exercising/playing sports
* Seek medical care right away if feeling ill when playing sports

How to Find out if you have sickle cell trait

A blood test can be done to determine if a person has the trait.

How is Sickle Cell Disorder Inherited?

If a man and a woman are both sickle cell carriers, with each pregnancy, there is a:

* 25 per cent chance of the child having the disease;
* 25 per cent chance of the child being completely free from sickle cell;
* and a 50 per cent chance of the child having the sickle cell trait, i.e., being a carrier

If only one parent is a carrier, then there is a

* 50 per cent chance of the child having the trait;
* 50 per cent chance of the child being completely free of sickle; and
* No chance of the child having the disease.

What Symptoms Are Associated with Sickle Cell Disease?

* Anaemia, jaundice and gallstones due to rapid breakdown of the red blood cells
* Painful swelling of fingers and toes in babies
* Painful attacks of joints, back and abdomen as there may be damage to the bone marrow
* Infections may develop, such as pneumonia
* Leg ulcers may develop due to less oxygen to the lower legs

Management of Sickle Cell Disease

* Full recommended immunisations plus pneumococcal vaccines
* Penicillin, starting from about two months of age to five years of age to help prevent serious infection
* Management of symptoms by using medications as needed
* Folic acid daily to help make new red cells

These are some of the management strategies. For further details, please consult your doctor.

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