# Sickle Cell Disease in short

## What is Sickle Cell Disease?

SCD is a blood disorder. Instead of being round and supple, the red blood cells in SCD tend to become half moon-shaped, brittle and sticky. Because of this, they shred fast and tend to block small blood vessels, thus preventing the flow of blood, oxygen and nutrients to the tissue. When this happens, it causes severe, incapacitating pain and/ or organ damage. Patients are also more susceptible to severe infections than the average population.

## How to you get SCD?

SCD is inherited. It is not catching. People who carry the sickle trait (HbAS or HbAC), can pass on the sickle gene to their children. People with the sickle trait do not know that they carry one sickle gene, unless they get tested, because this carrier status does not cause any symptoms. If two partners both have the trait, each of their children has a 25% risk of being born with Sickle Cell Disease (HbSS or HbSC).

## Some symptoms of Sickle Cell Disease

* Anemia : patients may be dizzy at times, easily exhausted or have headaches. They may look pale or have a yellow tinge to their eyes.
* Pain crises due to blockage of tiny blood vessels
* swollen fingers or toes in toddlers
* severe infections. Vaccines against some infections are available.
*Stroke and silent minimal strokes: Patients at risk can be identified through transcranial doppler sonography and placed on prophylactic treatment.
*Eye problems: Sickling can cause damage to the retina and eventually lead to blindness. Regular eye exams and laser treatment when needed, can prevent handicap.
* Joint damage: may develop over time and require hip replacement in some cases.
* Splenic sequestration: blood pools in the spleen, causing hard, tender abdomen and life-threatening, acute anemia. Immediate transfusion and iv fluids are necessary.

## Treatment options

SCD is caused by an error in the patient's genome. At present, the only permanent cure is bone marrow transplantation, which is still risky, unsatisfactory in a number of cases, very expensive and therefore not an option for us in Antigua. We are placing our hopes on further advances in gene therapy.

However, there is a lot that we can do to help patients with SCD:
* daily folic acid (needed to produce red blood cells)
* Penicillin prophylaxis for children
* non-routine vaccines (in particular Prevnar 13) for both children and adults
* prompt and sufficient pain management of vaso-occlusive episodes
* Hydroxyurea (an oral medication for patients with frequent pain crises or severe complications).
* life style: plenty of fluids, enough rest, no alcohol, no smoking, avoid exertion, regular check-ups.

## Know your sickle status – get tested!