# My baby has got
# SICKLE CELL DISEASE
## What to do?

Guide for orientation on
SICKLE CELL TRAIT

HEMORIO
BLOOD CENTER OF RIO DE JANEIRO

# Introduction

The objective of this manual is to provide information to patients and their relatives about Sickle Cell Trait.

We know that medical information is composed, in most cases of technical terms, difficult and incomprehensible to most people. Thus, we hope it can answer your questions.

Any comment is welcome, either about the clarity or omission of some information considered important. It can be sent via a suggestion to HEMORIO or by e-mail ouvidoria@hemorio.rj.gov.br.

# What is Sickle Cell Disease?

Sickle Cell Disease is a hereditary blood disorder that affects one in every 1,000 Brazilian citizens.

There are several different types of Sickle Cell Disease, which vary in severity, causing some problems and many others, almost none.

Some complications of sickle cell disease can affect your baby, making it then necessary that you are aware about the concepts of some problems that may occur with your child, and how to prevent them.

## Why my baby was born with sickle cell disease?

To answer this question, you need to understand a bit about the production of red blood cells and hemoglobin.

**Function:** The red blood cell operates carrying oxygen to tissues, constituting thus the main fuel for organs.

**Composition:** The red blood cells contain hemoglobin which is a protein rich in iron. The oxygen is picked up by hemoglobin, when it passes through the lungs. Oxygen is carried by red blood cells and distributed to various organs and tissues in the body. When the amount of hemoglobin is low, anemia occurs.

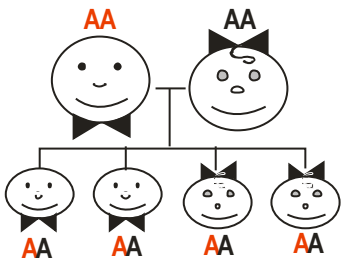
**Evolution:** The development of blood cells is a process called Hematopoiesis. Red blood cells enter the bloodstream after having already gone through the process of growth that occurs in bone marrow. Bone marrow is a soft tissue, like a sponge and is found in the cavities of many bones. Adult red blood cells must be in adequate numbers to develop their functions properly. The red cell membrane should be extremely tough yet highly flexible and deformable to withstand the aggressions in circulation and move through tiny capillaries and "pores" of the spleen.

By losing their ability to function properly, they are destroyed by spleen and replaced with newer erythrocytes that go through the same process, and so on.
In the case of sickle cell disease, red blood cells are destroyed (hemolysis), because of its sickle-shaped. They aggregate and cause blocked blood flow in small vessels of the body. With decreased circulation in the affected organs, injury occurs causing pain, destruction of blood cells, jaundice (yellow eyes) and anemia.

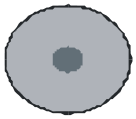
## How do people get Sickle Cell Disease?

Hemoglobin, the pigment that gives color to red blood cells, is essential for the health of every organ in the body. It works carrying oxygen to the body.

The normal hemoglobin is also referred Hb A, and normal people are called AA, because each portion is inherited from both parents.



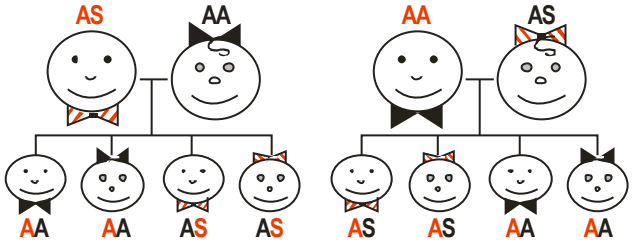
In sickle cell disease, the hemoglobin produced is abnormal and is called S. When a person receives hemoglobin A from a parent, and hemoglobin S from the other one, it is called "sickle cell trait," and it is referred as AS. People with sickle cell trait are not sick. They are usually asymptomatic and discovered to have this condition only when a family study is performed.



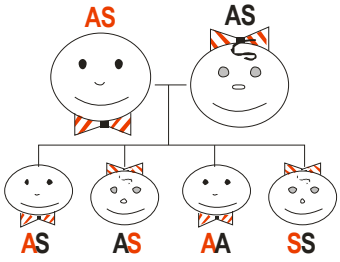
ERITROCYTE WITH USUAL HEMOGLOBIN (AA)

ERITROCYTE WITH SICKLE CELL HEMOGLOBIN (SS)

For example, if one parent has sickle cell trait (AS), there is a 50% chance with each pregnancy of having a baby with sickle trait (AS).




But if both parents have sickle cell trait (AS), there is 50% chance with each pregnancy of having a baby with sickle cell trait (AS) and 25% chance of having a baby with sickle cell disease (SS). Different from sickle cell trait, sickle cell disease can cause pain and lead to serious health problems.




There are other forms of the Sickle Cell Disease. When one parent is a carrier of sickle cell trait and the other is the bearer of some hemoglobin variant. The most common are hemoglobin C trait, hemoglobin D trait and thalassemia trait.

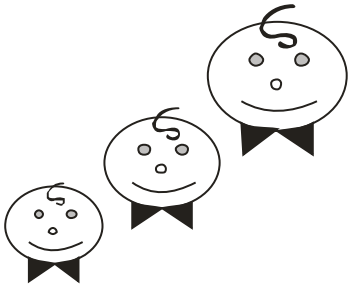
## What are the most common symptoms of Sickle Cell Disease?

Sickle Cell Disease (including SS, SC, SD, S-thalassemia) can manifest itself differently in each individual. Some have only a few mild symptoms while others have one or more signs or symptoms that we will describe:

Symptoms related to the disease usually appear after six months of age. The table below summarizes those warning signs of the disease that indicate that you should take your baby to the emergency, even without an appointment, and how they can be recognized.

| SIGNS / SYMPTOMS | WHAT CORRESPONDS |
|---|---|
| IRRITATION CONTINUOUS CRYING LACK OF APPETITE | PAIN: It is one of the most frequent symptoms of Sickle Cell Disease caused by obstruction of small vessels by red blood cells sickle. Pain may be located in bone or joints, chest, abdomen, reaching anywhere in the body. These painful crises have variable duration and may occur several times a year. They are usually associated with time cold, infection or dehydration. |
| FATIGUE DISCOURAGE-MENT SLEEP EXCESSIVELY | ANEMIA: The sickle cell is normally produced, however as it is abnormal, it is destroyed rapidly, which leads to the development of anemia. This anemia is not is corrected with vitamins and is part of the disease that will accompany the baby for life. |
| YELLOW EYES DARK URINE | JAUNDICE: Is a condition that results from the high rate of red blood cell breakdown. It is one of the most common sign of the disease. The urine becomes the color of Coca-Cola and the whites of the eyes become yellow. It is not contagious and should not be confused with hepatitis. |
| BLOATED BELLY ABDOMINAL PAIN | ENLARGED SPLEEN: Sometimes, you may experience what is called "splenic sequestration" which is a complication of the disease, caused by obstruction of the vessels of the spleen (an organ located in the upper belly, left). Leads to pain in this region and swollen belly. This complication is an emergency situation and can lead to severe anemia and should be treated in hospital. Your doctor will teach you how to palpate the spleen of your baby, for you to control its size. It's the best way to identify this complication. |
| FEVER ABOVE 38ºC COUGH DIARRHEA | INFECTION: In children up to five years, the infection, which usually presents as flu, cold or fever, corresponds to a factor that can cause pain, and worsening anemia. So the baby with Sickle Cell Disease should make use of antibiotics, with the goal of prevent infectious episodes from the first months of age, up to 5 years, plus they also should receive special vaccines to prevent some infections. |
| SWELLING AND REDNESS OF HANDS AND FEET | "HAND – FOOT SYNDROME": In young children the pain crises can occur in small vessels of the hands and feet, causing swelling, pain and redness at the site. This is a situation that requires immediate medical action. Thus, it is important to recognize it immediately and send the child to the hospital emergency room. |

## Will my son grow normally?



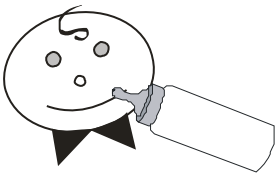
Children with Sickle Cell Disease can grow and develop more slowly than normal children. They are thinner and often enter puberty later than normal.

Generally, in adulthood, his stature is normal, although in most cases, remain lean.

Sexual development and puberty will eventually be normal. It occurs around age 17.

## Can I breastfeed my baby?

Your baby should be breastfed usually up to six months, breast milk, as this represents the main source of nutrients for proper growth.



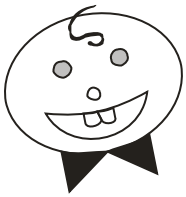
After six months, feeding should be similar to any child, which should be rich in fruits, vegetables and animal protein.

## What about the ability to learn in school?

Children with sickle cell disease often face a school career in which their disease process hinders their ability to concentrate and may interfere with learning. Additionally you may need an increase in school programs, to retrieve possible failures during hospitalizations.

## And about my baby's teeth?

Dental development is normal in sickle cell disease children. Special precautions are indicated for the prevention of dental caries. Everyone should brush their teeth after eating any food, especially those high in sugar.



Importantly, the continued use of antibiotics in the first five years of life does not cause any harm to your baby's teeth.

The first dental visit should be within 3 months of life. From 2 years old, the dentist will make the application of fluoride, at intervals of three months.

## Should I follow the normal vaccination schedule?

,0 you must follow the vaccination schedule achieved by health centers and participate in all public vaccination campaigns. Furthermore, your baby should receive some special vaccines, as demonstrated below.

You must bring the vaccination card for each query of your baby.

## Vaccination Schedule for Children with Sickle Cell Anemia:

| | |
|---|---|
| **BIRTH** | - BCG<br>- Hepatitis B |
| **1 MONTH** | - Hepatitis B |
| **2 MONTHS** | - Triple<br>- Anti-pneumococcal<br>- Anti-meningococcal<br>- Polio<br>- Haemophyllus anti-influenza type B |
| **4 MONTHS** | - Triple<br>- Anti-pneumococcal<br>- Anti-meningococcal<br>- Polio<br>- Haemophyllus anti-influenza type B |
| **6 MONTHS** | - Triple<br>- Anti-pneumococcal<br>- Anti-meningococcal<br>- Polio<br>- Haemophyllus anti-influenza type B<br>- Hepatitis |
| **9 MONTHS** | - Anti-measles |
| **15 MONTHS** | - Triple reinforcement<br>- Polio<br>- MMR<br>- Haemophyllus strengthening anti-influenza type B |
| **2 YEARS OLD** | - Anti-pneumococcal |
| **4 TO 6 YEARS OLD** | - Triple reinforcement and oral polio |
| **5 YEARS OLD** | - Strengthening of anti-pneumococcal |
| **> 6 YEARS OLD** | - Adult-type double every 10 years |

## Will my son have problems with antibiotic use for so long?

No. The use of antibiotic prophylaxis (prevention of infections) is ALWAYS indicated for sickle cell disease patients until the age of 5. This practice is recognized as a major advance in treating this disease, avoiding serious complications.

It should be noted that the daily use of antibiotics, does not "spoil the teeth" does not cause diarrhea, stomach pain or vomiting and does not "decrease the appetite" as well. If there is any doubt, talk to your doctor about it.

## What can I expect to treat my baby in HEMORIO?

HEMORIO has not only doctors and nurses to care for your baby. A team of several health professionals with extensive experience in the treatment of Sickle Cell Disease is available to help.

## These are professionals who are part of the Comprehensive Group for Treatment of patients with hereditary hemolytic anemia:

**Doctors:** Your baby will have a physician who will accompany you in all queries. Besides him, you can count on to other specialists such as nephrologists, dermatologists, neurologists, cardiologists, ophthalmologists, orthopedists, whenever requested by your doctor.

**Nurses:** The nursing staff is available for the treatment of babies with sickle cell disease, especially in monitoring the vaccine schedule.

**Dentists:** HEMORIO has Dentistry Service, which you must bring your baby within the first three months of life, to receive the first orientations. These additional consultations will help him grow stronger.

**Social Workers:** Social workers will tell you about the features of HEMORIO and their rights about Social Security and / or labor, resources and community services.

**Nutritionists:** Our nutritionist will make nutritional counseling, and monitoring of body weight gain of your baby.

**Physiotherapists:** Our physiotherapists will evaluate your baby, whenever requested by the attending physician, in order to improve possible postural problems, respiratory or neurological advising physical exercise training specific to each case.

**Psychologists:** The counseling is intended to help to understand the difficulties of the individual in various stages of life, helping to living with the disease. The family can also receive counseling, to assist in understanding the child's behavior as well as living with them.

## What should I do in case of emergency?

You should take your baby to HEMORIO emergency room, where he will be attended by doctors on duty, who shall take all necessary steps.

If you and your baby are in place well away from HEMORIO, or are being treated at another hospital, you must show the doctor that is making the call, the portfolio of HEMORIO, stating the diagnosis and the baby's blood group.

You should always take this manual with you, because the doctor may refer you. If the doctor still has any doubt about the action to be followed he should call HEMORIO (021-2332-8611), and talk to our duty doctor.

## What support groups or other sources of information are available to me?

There are many other sources of information that you receive over your child's development.

Still others, you can get through AFARJ, "Association of sickle cell disease and thalassemia of Rio de Janeiro", based in HEMORIO, room 524, phone: 2332-8611 ext 2246.

## Ten Commandments of Caring for Your Baby

Pay close attention to the items below. They are intended, to guide the family of the baby with sickle cell disease, the basic care that should be taken with him. It is essential that you follow them strictly.

| |
|---|
| **1 - Strictly follow the medical guidelines.** |
| **2 - Giving your baby, antibiotic prophylaxis, DAILY.** |
| **3 - Giving your baby folic acid.** |
| **4 - Call a doctor, each warning sign.** |
| **5 - Offer plenty of fluids, especially on hot days.** |
| **6 - Keep the baby wrapped up (socks and gloves), on cold days.** |
| **7 - To prevent tooth decay and dental treatment.** |
| **8 - Following the immunization schedule.** |
| **9 - Controlling the spleen size of your baby through the abdominal palpation, guided by your doctor.** |
| **10 - Maintain a good diet.** |

# Collaborators

**General Management**

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**Coaching Staff**

Interdisciplinary Group for Treatment of Patients with Hereditary hemolytic disease

Vera Marra

**Publishing**

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## FURTHER INFORMATION AND HELP:

**SOCIAL SERVICES OF HEMORIO**

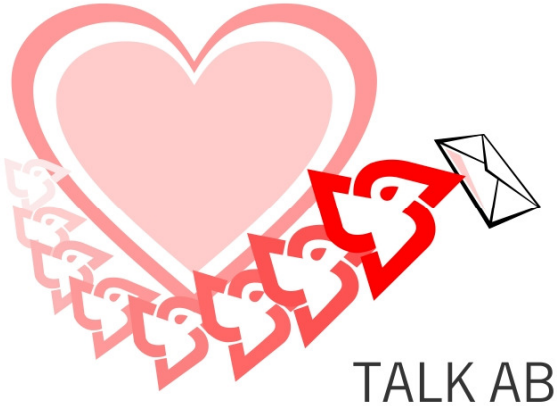
#8, Frei Caneca Street, room 801, Rio de Janeiro
RJ - ZIP CODE:20.211-030, room 811
Downtown - Phone: 2332-8611 ext 2253

**AFARJ, Association of sickle cell and thalassemia Rio de Janeiro**

# 8, Frei Caneca Street, room 801, Rio de Janeiro
RJ- ZIP CODE: 20.211-030
phone: 2332-8611 ext 2246.

**"Here you will find people who care about you."**

WHEREVER YOU
GO TAKE A
MESSAGE OF LOVE.



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GOVERNO DO Rio de Janeiro SECRETARIA DE SAÚDE

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