PATIENT'S MANUAL - DO YOU HAVE A STUDENT WITH SICKLE CELL DISEASE IN YOUR CLASS GROUP? - REVISED EDITION 05/2010

Do you have a student with **SICKLE CELL DISEASE** in your class group?

A guide of Sickle Cell Disease
for TEACHERS and EDUCATORS

HEMORIO
BLOOD CENTER OF RIO DE JANEIRO

Introduction

This booklet represents the desire of Department of Hematology of HEMORIO, reflecting our quest to always offer the best to patients.

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. Moreover, the daily life of Services and Health teams do not always allow all the information be provided, regardless of the treatment is.

In this sense, we realize the immense value of this print, in which we seek to move in a clear, objective and accessible, the main doubts about the diagnosis and treatment of their disease.

We also intend to clarify issues that may arise during the treatment when the aid outside the hospital is valuable. We refer to the actions of relatives and friends at the time of illness.

Love, understanding and constant presence represent irreplaceable role in the perspective of the treatment and comfort only obtained in this way.

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.

MESSAGE TO THE TEACHER:

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. But there are many things you can do to help encourage your student with Sickle Cell Disease.

You too can minimize the problems caused by sickle cell disease, and with your help, the child or adolescent will be useful to an adult and a valuable member to society.

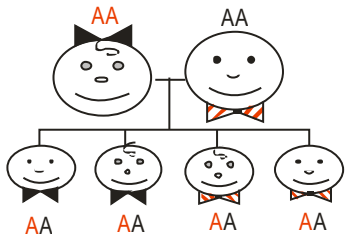
HEMORIO is always available to answer your questions.

What is Sickle Cell Disease?

Sickle Cell Disease is a hereditary blood disorder that affects 40,000 Brazilian citizens, in which red blood cells, before certain conditions alter their shape and become like a sickle.

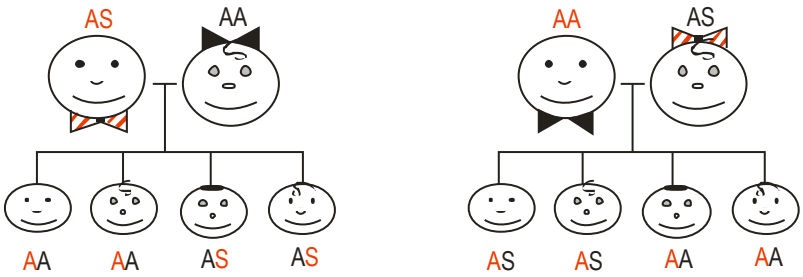
It is also known as "hemoglobinopathy S", that corresponds to a change in hemoglobin, which is a protein in red blood cells and is essential for the health of all organs of the body. It works carrying oxygen to the body.

Usual hemoglobin is called the A and AA is considered normal individuals, receiving a portion of the hemoglobin from the father and another from the mother. (See diagram below).

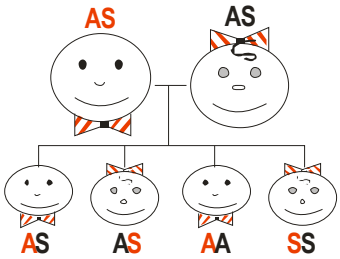


In sickle cell disease, the hemoglobin produced is abnormal and is called S. When a person receives hemoglobin A from a parent, and hemoglobin S from the other one, is called "sickle cell trait," and is referred as AS. People with sickle cell trait are not sick. They are usually asymptomatic and discover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

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.



Sickle cell trait does not mean Sickle Cell Disease?

People with sickle cell trait inherit a gene for usual hemoglobin (Hb A) from one parent, and a sickle hemoglobin gene (Hb S) from the other one. It is a condition in which the person, although not present anemia at a routine examination (CBC) carries a kind of modified hemoglobin gene, called the “S". it affects 3 in 100 Brazilian individuals. He does not cause sickle cell disease, can not become Sickle Cell Disease and needs no treatment.

Sickle cell trait is a genetic disorder inherited from one parent who is not strong enough to manifest as disease.

The importance of sickle cell trait is that if both parents have sickle cell trait, the child has 25% chance of being born with Sickle Cell Disease.

The sickle cell trait can be detected by a blood test specific: Electrophoresis of Hemoglobin.

There are several different types of sickle cell disease ranging in severity, some causing many problems, and others almost none.

What teacher should know about disease?

Complications of Sickle Cell Disease can affect a person's routine at school, making it then necessary that you are aware about the concepts of some of the problems that may occur with your student.

Watch debauchery

People with sickle cell disease may suffer ridicule because they had yellow eyes, short stature and leg wounds. Teacher should explain about the disease to other students to demystify the stigma surrounding the disease.

Regarding sports and physical education

It is important to consider that Sickle Cell Disease school-aged children can often, but not always, take part in physical education or sports. However, they should be encouraged to take part in all physical activities, but should be allowed to stop feeling pain or shortness of breath.

About swimming

Pain related to sickle cell disease may be triggered by exposure to cold and moisture. Children should not play in the water for a long time and when they leave, they must soon be dry and wear dry clothes.

The practice of swimming is not contraindicated, however, must be evaluated case by case basis.

Permission to go to the bathroom

Allow the child to have free access to the bathroom, because children with Sickle Cell Disease produce more urine than normal ones and may need to go to the bathroom more frequently.

Additionally, make sure the child has water readily available at all times in class. However you should ask if he is feeling a burning sensation when urinating, because it may have an infection and, in this case should go to the doctor.

What to eat

The school aged children with sickle cell anemia have high iron stores in the body. Therefore, they should be avoided foods rich in iron such as liver, tongue, heart and kidneys, marine animals nuts, eggs, molasses, whole wheat bread and fortified, enriched and whole grains. That does not mean that these foods should be prohibited. It means they must be consumed in moderation. It is important to drink plenty of fluids. Black tea reduces iron absorption and, therefore, should be used freely.

The stature of the child

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.

About missing school

People with Sickle Cell Disease are more likely to miss school because of the occurrence of bone pain, increased risk of infections, medical visits or hospitalizations. To compensate for these absences they should receive encouragement and extra help, if possible.

What about medical problems?

You can contribute to the health of your student observing some of the symptoms of the disease. If the child is tired, apathetic and looks very pale, he should go to hospital for an evaluation. In case of joint pain or back pain, he should be allowed to lie down to drink and painkillers such as Tylenol or painkillers. If there is high fever, he should be taken to the hospital urgently. Signs and symptoms are as follows. Stay tuned!

Painful crises

Pain is the most common symptom of sickle cell disease, caused by obstruction of small vessels by sickle red blood cells. It may be located in the bones or joints, chest, abdomen, and can reach anywhere on the body.

These crises have variable duration and can occur several times a year. They are usually associated with cold weather, infections, premenstrual, emotional problems, pregnancy or dehydration.

Pain should be seen as a symptom, which although frequent, is not normal. The deal will be effective if the management follow these rules:

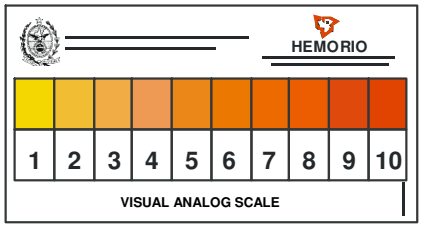
1 - When the pain occurs, it is necessary to maintain the use of analgesics, for a minimum of 24 hours after the relief of pain;

2 - A single dose of an analgesic is generally not sufficient for the rapid relief of pain;

3 -The use of analgesics should be continuous;

4 - For each level of pain, treatment must begin differently;

5 - The level of pain should be measured by a Visual Analog Pain Scale (see illustration)



How to Use a Visual Analog Pain Scale

The chart below summarizes the steps to be followed, according to the degree of pain. Your doctor will give appropriate guidelines.

PAIN INTENSITY	START OF TREATMENT	AFTER 24 H WITHOUT PAIN	AFTER MORE THAN 24 H WITHOUT PAIN	RECURRENCE OF PAIN
1 2 3	DIPYRONA PO 8/8H STOP AFTER 24H WITHOUT PAIN	REMOVE DIPYRONE		
4 5 6	DIPYRONA PO 8/8H INTERIM DICLOFENAC PO 8/8 H - UNTIL 24H WITHOUT PAIN	REMOVE DICLOFENAC AND KEEP DIPYRONE PO 6/6 H FOR 24 H		GO TO EMERGENCY ROOM
7 8 9 10	DIPYRONA PO 8/8H INTERIM DICLOFENAC PO 8/8 H INTERIM CODEINE PO 4/4 H UNTIL 24H WITHOUT PAIN	REMOVE DIPYRONA AND KEEP CODEÍNE PO 8/8 H INTERIM DICLOFENAC PO 8/8 H	REMOVE CODEINE AND KEEP DICLOFENAC FOR 24 H	

Yellow eyes (jaundice)

Is the most common sign of the disease. When red cell is destroyed appears a yellow pigment in the blood called bilirubin. 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.

Leg ulcers

Children and adolescents may be not gentle with other children who are different. One of the most common problems in adolescents with Sickle Cell Disease is the appearance of injured ankles that may take long to heal. These lesions are not contagious, but the child may feel so embarrassed that moves away from school.

People with these wounds should be encouraged to attend school because with the injuries protected by bandages, they will not submit odor and will not cause other problems.

Infections

Infections may occur frequently located in the throat, lungs and bones. These infections should be evaluated by a doctor as soon as appear. All patients must be current on vaccinations. At the first sign of fever should seek medical attention in an emergency room.

Aseptic Necrosis of the Femoral Head

Some patients have obstruction of the small veins of the hip joint, then the head of the femur (thigh bone), without movement, and wears leg shortens. The pain in the legs or hips suggests that complication.

Priapism

Priapism is unwanted, prolonged and painful erection of the penis, caused by obstruction of the vessels. Usually it occurs spontaneously at night.

When it occurs, it is necessary to go to an emergency room immediately. Early treatment can prevent future complications.

Although embarrassing to the student, this complication can be suspected by teachers, particularly if they are aware of the possibility of its occurrence.

These episodes correspond to situations of emergency, and the student should be immediately referred to a doctor. Early treatment can prevent future complications.

Collaborators

General Management
Clarisse Lobo

Coaching Staff
Interdisciplinary Group for Treatment of Patients with Hereditary hemolytic disease
Vera Marra

Publishing
Marcos Monteiro

Further information and help:

SOCIAL SERVICES OF HEMORIO

#8 Frei Caneca Street, room 811 - Downtown
Rio de Janeiro - Phone: 2332-8611 ext 2253

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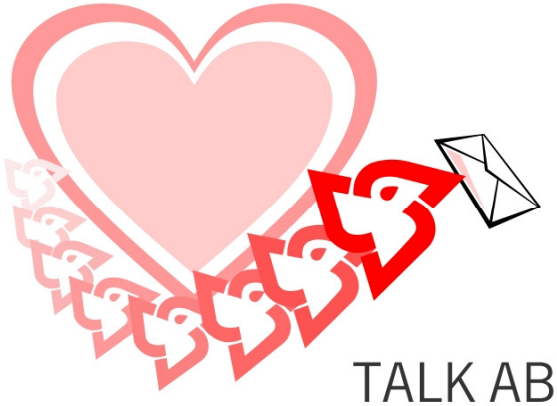

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DISQUE SANGUE 0800 2820708

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