

# Sickle Cell *Rapper*

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THE CHILDREN'S REGIONAL SICKLE CELL CENTER  
UTHSCSA DEPT OF PEDIATRICS  
DIVISION OF HEMATOLOGY/ONCOLOGY  
CHRISTUS SANTA ROSA HEALTHCARE

## 3 TOP REASONS THAT BRING A SICKLE CELL PATIENT TO THE ER



*This article was presented to ER Staff and was included in the Children's ER Newsletter*

Let's go back to basics; Sickle Cell Disease (SCD) is due to a change in the morphology of the red blood cell. The shape of the cell is critical in its ability to carry oxygen to the tiniest vessels of the body; constant sickling and unsickling damages the cell leading to sticky, sickle shapes.

Sticky cells lead to traffic jams → decreased oxygenation → necrosis → pain crisis

Sickle Cell red cell life is 10-18 days; normal cell life is 120 days. Two major issues to remember regarding SCD: Chronic anemia and pain crisis. Pain crisis is the hallmark of SCD – the complication of the traffic jams on the organs leads to failure of their ability to do their jobs in the body and can affect every organ.

***Let us review the 3 top reasons you or your child will see the Emergency Room***

(Continued on page 2)

***Camp Sky – August 5 -10 More information to come!***

## 3 TOP REASONS THAT BRING A SICKLE CELL PATIENT TO THE ER

### #3 Pneumonia/Acute Chest Syndrome

***Sickling in the capillaries of the lungs leading to alveola collapse:***

- a) Difficult to diagnosis Acute Chest Syndrome (ACS) vs. pneumonia
- b) Progresses very quickly and can be life threatening
- c) Symptoms – Pre: fever, chest/back pain, coughing, shortness of breath

### #2 Infection/Fever

***Major complication of SCD***

Prevention: All children with SCD are on prophylactic antibiotics until age 5 years old and then are reassessed. It prevents 80 percent of life threatening episodes of childhood infections in children with SCD.

Any baby with a temperature of 100 A is required to come to ER. Children with temperature of 101A, the parents are taught to call the Sickle Cell Nurse for telephone triaging. If after hours, they call the Hematologist/Oncologist on Call. They then are instructed to go to ER for blood cultures, blood counts, antibiotics, closer monitoring of potential problems and likely admission

### #1 Pain Crisis

***Leading cause of trips to the ER and/or hospitalizations***

First sign is dactylitis (swelling of hands/feet) in infants about 6 months of age or a bit later when mom's protective hemoglobin F is decreasing in the baby's system. Its presence can be an indicator of the severity of the disease. It can affect: brain, spleen/gallbladder, lungs, bones, penis, kidneys, eyes, heart.

Pain is unpredictable. It can be caused by a number of factors such as weather changes, stress, dehydration, insufficient sleep/rest, anxiety. What causes parents most distress is that one year their child can have had one admission to hospital for pain crisis and the next year it is up to 3 or 4. It is a roller-coaster ride. There is no laboratory marker for presence and/or severity of pain therefore the "gold standard" is what the patient says it is

#### **Types of pain seen in SCD:**

*Somatic Pain:* experienced most frequently – deep structures i.e. muscles, tendons, ligaments, bone marrow, joints and arteries.

*Visceral Pain:* spleen, liver, lungs – vague, poorly localized, dull aching.

*Chronic Pain:* lasts 3-6 months. It is usually somatic and associated with bony changes i.e. avascular necrosis (humerol/femoral heads), vertebral collapse, and chronic/recurrent leg ulcers. You can see an acute episode superimposed on chronic pain.



*Acute Pain:* most common type of pain –abrupt onset. It can vary from mild aches to debilitating pain. Uncomplicated acute pain can last from hours to days and it can move to different sites.

Pain is what causes our families the most distress...seeing your child in agony drives parents mad!! When patients show up in ER they have exhausted all home management remedies: i.e. non-steroidal anti-inflammatory (Motrin/Tylenol), Tylenol and Codeine, with warm packs, distractions, rest and fluids. For those that have Morphine or Roxanol at home, that is their last resort. Can you imagine yourself as an 12 year old growing up and have had to deal with pain all your life ... reality is you will again experience pain unlike anyone else.

The fear of pain encompasses memory of past pain, pain experienced at present and future pain.

In a perfect world when a patient presents to ER the best dose of opioids for severe sickle cell pain is what provided relief the previous time which would be readily available on a user-friendly computer. That is when you will see patients coming in to ER and informing the staff what medication and dose they want. This is often seen as drug seeking or addiction but in reality it is relief seeking. Only 3% of patients actually demonstrate addictive behavior. A patient on long term Morphine/opioids may have developed a tolerance to the dose he/she is receiving and an evaluation needs to be made to either increase the dose of the medication, change the frequency of the medication or use another medication.

There are changes coming! We are working closely with our patients to empower them to take control of their disease.

There is so much to talk about with regard to Sickle Cell Disease. I appreciate that there are many concerns and challenges with regard to this population, please call.

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We will be providing a Portable Medical Card for all patients. This card will fit in a wallet and/or purse and will contain the patient's name, date of birth, diagnosis, allergies, emergency contact person & pertinent phone numbers for the hospital. We will let you know once our first supply is ready for distribution.

**Camp Sky – August 5 -10 More information to come!**

## SICKLE CELL CENTER UPDATES

For our teenagers we will be offering the last 5 minutes of the appointment time with any part of the team to be “alone time”. The purpose of this is to allow the teenager the time to ask any questions that he/she may find difficult in front of their parents – it also promotes steps in taking control of their disease and independence. We will gradually start taking more and more time alone until they reach a level of maturity and independence in their ability to “speak” for themselves. We will also be teaching our younger patients to know more about their disease, the medications that they are on and will be quizzing them at their appointments. This is to help our patients become independent and promote self-confidence with their disease. This will also assist in preparing for their transition to an adult medical setting.

Sickle Cell Disease is no longer considered a childhood disease it is considered a chronic disease. On Feb. 21-23, 07 I attended the first Sickle Cell Research and Educational Symposium – Transition from Pediatric to Adult Care it generated much discussion. There was a panel of adults at a question/answer session and they introduced themselves individually by saying “I am living well with Sickle Cell Disease”. Sickle Cell Disease is a part of you it is not the whole of you. Here is a sample of a research question during one of the educational sessions:

“Factors associated with Resilience (for youth with disabilities): (Hacket & Antonelle)

*What would you think a group of “successful” adults with disabilities would say is the most important factor that assisted them in being successful? Weiner 1992.*

- Self-perception as not handicapped.
- Involvement with household chores.
- Having a network of friends
- Having non-disabled & disabled friends
- Family & peer support
- Parental support without over protectiveness

***In order to protect your privacy and make the most of your visit with the doctor, we are requesting that during your appointment time the cell phone is turned off or put on “silent mode”.***



Answer: #2...So get busy with those chores!

## Do you suffer from Anxiety or Depression?

### Anxiety Checklist

- Pounding heart or palpitations
- Sweating
- Shaking or trembling
- Shortness of breath
- Feeling of choking
- Chest pains
- Nausea
- Dizziness or lightheaded feeling
- Feeling of being detached from oneself
- Fear of losing control
- Fear of dying
- Tingling or numbness
- Chills or hot flashes

### Depression Checklist

- Depressed most of the day
- Loss of interest or pleasure in most activities
- Significant change in weight or appetite
- Inability to sleep or sleeping too much
- Feelings of restlessness or slowed movements
- Feeling of worthlessness or guilt
- Fatigue or loss of energy
- Inability to think or concentrate
- Thoughts of death or suicide

***Consult your doctor immediately if you have thoughts of suicide or death***

If you feel that you suffer from anxiety or depression, there is help for you. Notify your healthcare provider immediately. Below are resources you may find useful:

Anxiety Disorders Association of America

[www.adaa.org](http://www.adaa.org)

Families for Depression Awareness

[www.familyaware.org](http://www.familyaware.org)

National Mental Health Association

[www.nmna.org](http://www.nmna.org)

National Information Center for Children  
And Youth with Disabilities

[www.nichcy.org](http://www.nichcy.org)  
(800) 695-0285

Would you like more information on Sickle Cell Disease?  
These are great links to get you started:

[www.dshs.state.tx.us/newborn/sickle](http://www.dshs.state.tx.us/newborn/sickle)  
[www.sicklecell-info.org](http://www.sicklecell-info.org)  
[www.nhbj.nih.gov/healthy](http://www.nhbj.nih.gov/healthy)  
[www.SCInfo.org](http://www.SCInfo.org)





333 North Santa Rosa, 8th Floor  
San Antonio, TX 78207  
Phone: 210-704-2187  
800-227-3618  
After hours call: 210-704-2011 and ask  
for Pediatric Hematologist on call.

E-mail: someone@example.com

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