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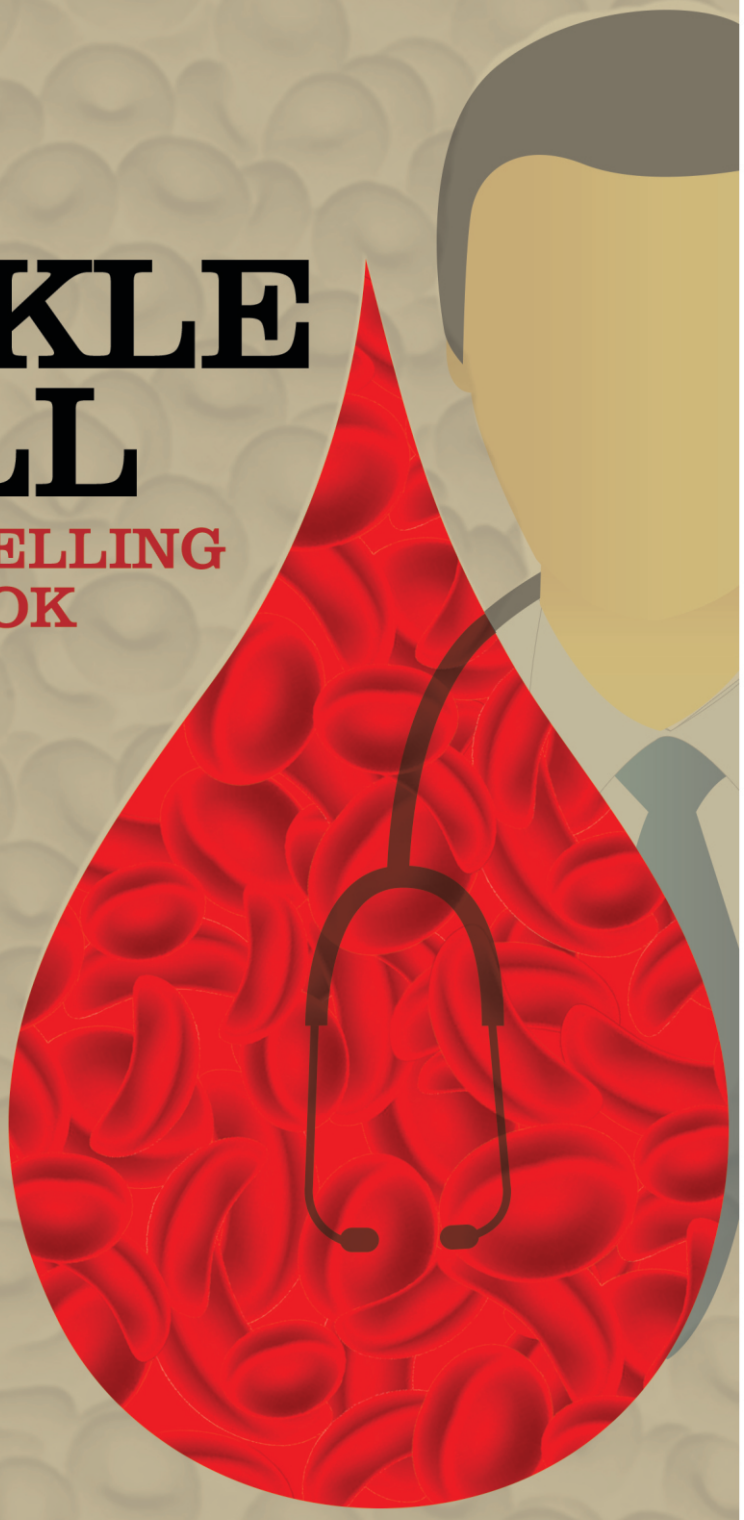
SICKLE CELL

A COUNSELLING HANDBOOK



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SICKLE CELL INSTITUTE CHHATTISGARH, RAIPUR

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INDEX

Symptoms of

Sickle Cell Anaemia

Red = In severe anemia

Central
- Fatigue
- Dizziness
- Fainting

Eyes
- Yellowing

Skin
- Paleness
- Coldness
- Yellowing

Blood vessels
- Low blood pressure

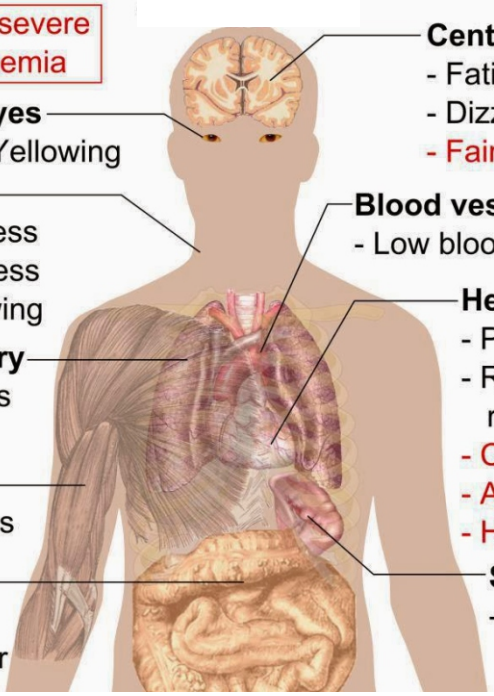
Heart
- Palpitations
- Rapid heart rate
- Chest pain
- Angina
- Heart attack

Respiratory
- Shortness of breath

Muscular
- Weakness

Intestinal
- Changed stool color

Spleen
- Enlarge-ment

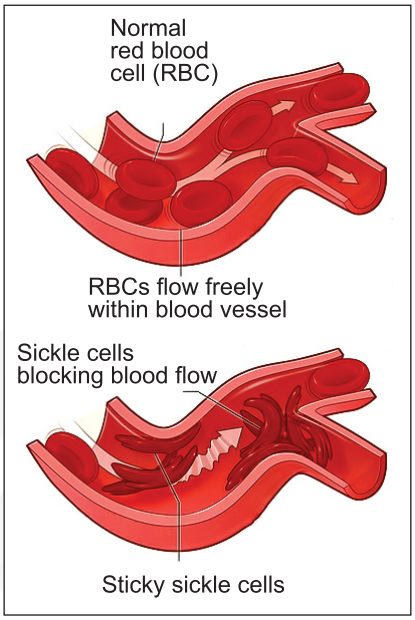


I
INTRODUCTION

What is Sickle Cell?

Sickle cell is a disease that affects haemoglobin inside our Red Blood Cells (RBCs). The haemoglobin in the RBCs is important as it carries oxygen to every part of the body. Normal life depends on the regular availability of oxygen in the cells and tissues.

A person with this disorder has a different kind of haemoglobin, which causes the RBCs to change their shape. Instead of being smooth and round, the cells become rigid and sticky. They have the shape of a sickle, which gives the disease its name.



The rigid, sticky sickled RBCs have trouble moving through the small blood vessels. Many times they clog up the blood vessels, thus preventing blood carrying oxygen from reaching the tissues. This causes pain or damage to these areas.

Sickle Cell Anaemia (SCA) is the result of both Trait and Disease. The persons with the Trait are the carriers, while those with Sickle Cell Disease (SCD) suffer from this disorder. Traits are generally asymptomatic.

It is genetic, ie., this disorder is hereditary.

What is its prevalence?

Sickle cell affects about 12% of the population of the state of Chhattisgarh. It is found mostly in the semi-tribal areas of the state, although with migration of people, even urban areas now have affected population. Certain castes in Chhattisgarh have been observed to have a very high incidence of SCA/SCD.

In India, it is mainly found in central regions.

What are the types, and how does it affect human body?

There are many different types of sickle cell disorder: **Sickle Cell Trait** or carrier (AS phenotype) & **Sickle Cell Disease** (SCD with SS phenotype). Other varieties include Sickle Beta Thalassemia Disease (S beta thal disease) which is less common in Chhattisgarh, sickle-haemoglobin C disease (HbSC), sickle beta-plus-thalassaemia (HbS/β^{+}) and sickle beta-zero-thalassaemia (HbS/β^{0}). Some types of sickling cause fewer problems than others. For example, Trait is generally less serious than SCD. However, it varies widely from patient to patient.

Sickle cell disease is a chronic disease. It can't be cured, but it can be treated. People with SCD can live well into middle and late adulthood. With penicillin to help stop infections, today children with SCD are also living longer than ever before.

SCD can cause many kinds of problems, including anaemia, infections, inflammations, severe pain, and organ damage. It also reduces life expectancy. However, **not everyone who has sickle cell disease will have all of these problems.**

Sickle cell disease affects the body, ***not*** the mind. SCD will not affect how smart a person is.

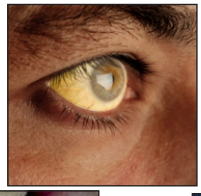
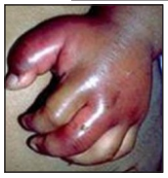
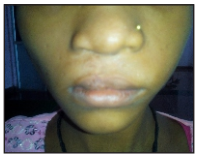
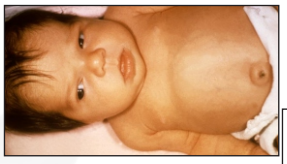
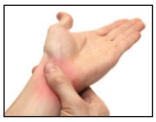
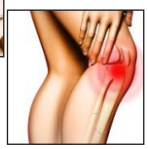
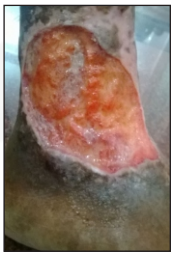
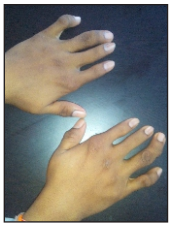
Pain & its severity:

Pain is an inevitable part of SCD. Its severity and frequency are unpredictable.

The frequency, severity and duration of pain vary widely among affected persons – from rare to several times a year. Approximately 30% of patients have few pain episodes, 50% have occasional pain episodes, and 20% have weekly to monthly episodes. Frequency of pain episodes generally increases in the second decade of life and decreases in the fourth decade. Patients with more than 3-4 episodes a year should be priority for round the clock treatment. ***Emotional stress, depression, changes in temperature, wind, dehydration, infection, fatigue and over-exertion often precipitate pain and crisis events.***

Symptoms:

- Delayed growth, late puberty, sub-normal weight & height
- General complains of weakness & fatigue
- Highly anemic appearance
- Pale skin and nails
- Yellow discoloration of skin & eyes (jaundice)
- Flattening of nasal bones
- Continuous mild fever over long period
- Shortness of breath / breathlessness
- Loss of appetite for extended periods
- Frequent urination, dark urine
- Frequent viral & bacterial infections
- Pneumonia (long term)
- Enlarged spleen
- Pain in bones & ribs
- Swelling in hands & legs (hand-foot syndrome)
- Leg ulcers
- Priapism
- Infertility, or delayed fertility
- Irritability & prominent mood swings

Who should be screened for Sickle Cell Disorder?

- All new-borns.
- If any child is detected with it, then parents too; and vice-versa.
- Any adult who displays symptoms of severe anaemia, long term fatigue, delayed growth pattern, hyphema, haematuria, pregnant mothers, couple planning for pregnancy, and those having a family history of SCA/SCD should test for it.
- Selection of spouse (for blood compatibility)

Screening tests include solubility tests, Electrophoresis, HPLC (high performance liquid chromatography), isoelectric focussing, & DNA-based techniques, for definitive diagnosis of different variants.

II
COUNSELLING

The patient realizes that he/she has to live with the disorder. Only the painful conditions can be ameliorated and the crisis episodes minimized. In trying to reduce them, the afflicted person can try to live life as normally as possible and attempt to increase his/her own life expectancy.

Counselling is necessary because the patient knows that the disorder is life-long.

Counselling focuses on **three things**:

- ***General pain management,***
- ***Handling of crisis episodes, &***
- ***Planning for living a life as normal as possible***.

SCD affects different persons in different ways. No one can know how serious the disease will be for themselves or for their children, but we do know that **four things** can make a difference:

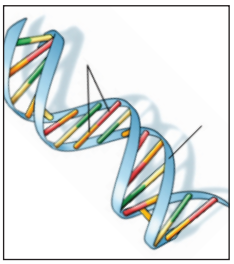
- **The type of Sickle Cell Disorder (Trait/ Disease)**
- **The kind of care a person gets**
- **How the person and the people around him deal with/react to the disease**
- **Awareness about the disease/disorder and the limitations it imposes (the more they know, the better care they will be able to give to themselves/their children)**

Therapy must be individualised.

What Patients should know at various stages:

Pre-Marriage Counselling in Young Adults:

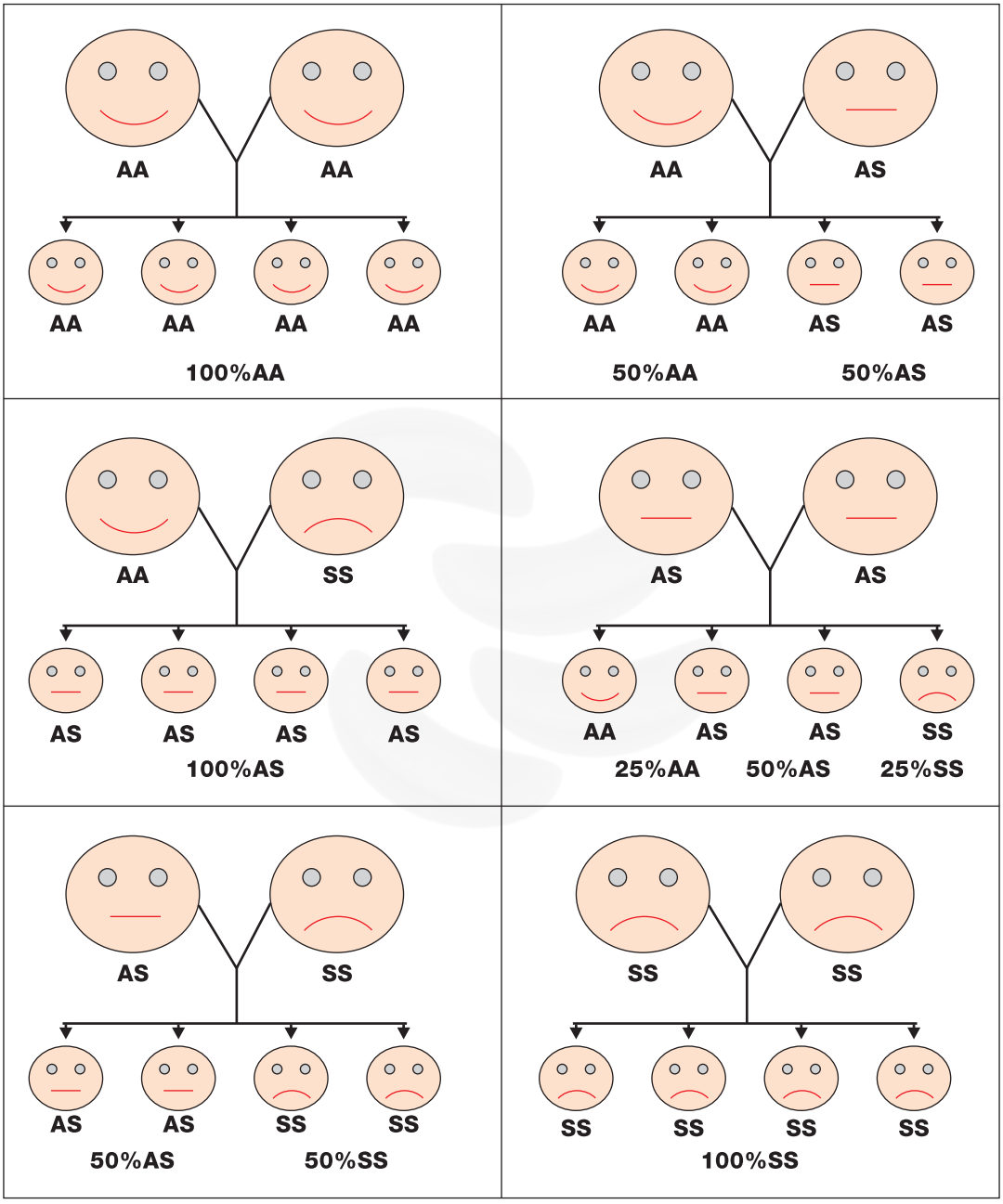
Young adults should go in for pre-marriage screening and genetic counselling, to know the risks involved for a couple affected by SCA/SCD, and the chances of a normal offspring. While planning a family, 3 important things are to be kept in mind:



- **Genetic Counselling:** This information can help patient figure out the chances of having future children with sickle cell disease, and lets them know options for family planning and pre-natal testing.

- **Testing your baby before it is born:** Amniocentesis and DNA testing is possible at the pre-natal stage.
- **Chances of passing on the sickle cell genes:** The chances of passing on genes and making a child a carrier or an affected individual is determined by the combination of parental genes. This is given in the **Inheritance Chart.**

SCA/SCD INHERITANCE PATTERNS



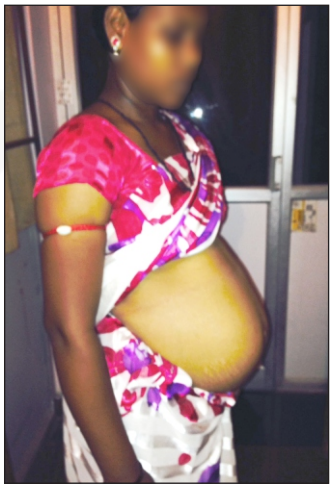
AA=Normal AS=Sickle Trait SS=Sickle Disease

Remember: These are *indicative* chances of births, and the chances are the *same for each child and in each pregnancy with the same partner*, independent of any earlier birth.

Young Couples/ Just Married:

- Genetic counselor will **not** tell the patient what to do. He/she will only provide to the parents knowledge of how sickle gene affects the newborn baby.
- Partners are free to choose the path which is most comfortable to them.
- They have to decide *whether* to go in for a child – based on inheritance pattern, and given the factor of random chance.

Pregnancy phase / Pre-natal:



- Planning & early pre-natal care are the key to survival of the new-born. In early phase of pregnancy, parents should be aware of how Sickle Cell gene affects a new-born baby.
- Pre-natal care should be done by an obstetrician (OB), who is an expert in **high-risk** pregnancy.
- Parents, especially the mother-to-be, to avoid infections of all types – viral, bacterial, including UTI.
- Hydroxyurea **not** advised for pregnant mothers and women planning for pregnancy.
- Take healthy, high protein diet, rich in fibre and roughage. Avoid dehydration.
- Pre-natal vitamins, folic acid supplements may be necessary.
- Amniocentesis and DNA testing of the foetus is possible at the pre-natal stage.
- High possibility of premature/pre-term baby.
- Patient and her family should know that frequency and severity of pain

episodes may increase during pregnancy.

- During labour, IV fluids may be required to prevent dehydration.
- Oxygen (through mask) may be given during labour (depending on patient condition).

Neo-natal / Early Infancy:

- There is a high possibility of a premature / pre-term baby, hence sickling parents should be forewarned and prepare themselves accordingly.
- Newborn screening for SCA/SCD: HPLC test. (Solubility test **not** conclusive for infants).
- Mothers should breast-feed the infant.
- Test for sepsis.
- High risk of infections.
- All standard Immunizations – against Hepatitis B, polio, diphtheria, influenza, MMR, BCG, etc.
- Vaccinations – anti-tetanus, pneumococcal vaccination (PCV) till age 5.
- Penicillin prophylaxis.
- Children may also need vitamin, zinc and iron supplements.
- However **additional iron/folic acid should not be given *unless*** the infant is proved iron-deficient (as SCD accumulates iron faster than normal, leading to complications). Hence, it should be taken under doctor's advice.

Childhood and schooling, and Role of Parents & Teachers:

A little extra care needs to be given to SCD children when they step out of home for extended durations, especially when they start going to school. Both parents and teachers need to be sensitized

- Parents should give affection, and provide a safe and understanding environment at home.
- Parents should know that pneumococcal vaccination and penicillin prophylaxis have reduced the risk of morbidity in SCD children.
- Teachers should be informed/made aware enough to provide the little extra attention that such children require. They have to provide a comfortable and understanding environment at school.

- Give moderate work load in schools.
- Give extra time to complete classwork, tests, etc.
- Children should be allowed to carry extra water bottles, medications, umbrella, etc.
- Motivate to drink water frequently in school.
- Allow them to go to the bathroom as many times as they want.
- Give 2 sets of books/copies as far as possible, one at home, one at school, so that they do not have to carry load daily.
- Avoid strenuous exercise, long distance movement or running.

- Exempt from outdoor activities when temperatures are very high or low, instruct for alternate indoor activity.
- At the hint of fever/ morbidity/ pain/ change in complexion or pallor/ breathing difficulty/ dizziness/ differential palpitation/ swelling/ muscular weakness, they should be immediately allowed to go home or referred to a doctor.
- ***Teachers and student both should know who to contact in emergency.***
- ***Child should have on his/her possession a card containing contact numbers and necessary medical details.***

Adolescents and Young Adults, and Role of Parents:

Role of parents is very important in this stage of a child's life.

Parents should

- discuss nature of the disease/trait & its impact, with the growing child.
- discuss & review ways to manage pain/crisis (what to do & how to do).
- counsel them about adverse effects of tobacco, smoking, alcohol & drugs.
- review school & work-place performance, watch out for drops in performance as they indicate problems and crisis episodes.
- watch what words they say & the way they interact with children, as such children will be sensitive.
- encourage their adolescent child to try different activities to see what he/she enjoys. This will divert their attention from the problem or crisis.
- praise the child frequently, hug often, and make the child feel wanted.
- create a safe & loving home environment and a positive atmosphere at home.

Friends also play an important role in making the patient at this age feel wanted.

Friends should

- treat the patient as just another normal person, and should help as & when required.
- help catch up with school work when missed, do

homework together, etc.

- encourage and promote healthy habits through peer choice & pressure.
- know the warning signs and get help as/when required.

Despite others help or support, adolescent patients have to learn to deal with the disease themselves and cope.

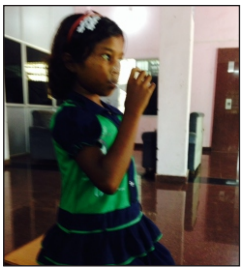
They should

- learn to identify onset of pain/crisis by self.
- know who to contact, when to contact – doctor's name, hospital, etc.
- know how to make appointments.
- have a personal health summary at all times, and should carry own health insurance, cards, etc.
- understand what complications can arise, & under what conditions.

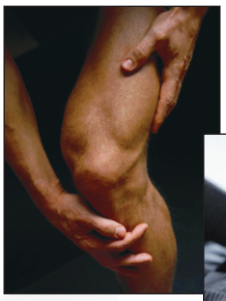
- Affected adolescent/young adult should discuss plans (with parents, peers, and professionals) for independent living, vocational & economic self-reliance.
- After puberty, the patient should be educated about bodily changes, safe sexual practices.
- Adolescent girls should be aware of the risks of pregnancy with SCD, safe birth control practices, and of the merit of pregnancy at the right age.
- Parents wanting patient's marriage & unmarried patient himself/herself should consider the inheritance patterns, before deciding future action.

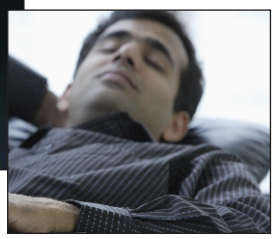
III
MANAGEMENT OF EPISODES

Pain Management:



The degree and duration of pain cannot be predicted in advance. In general, the following can be adopted for keeping pain levels low and bearable:





- Take plenty of water.
- Take warm bath.
- Place heating pad/warm moist towel on sore part.
- Massage the part that hurts, in direction of blood flow.
- Avoid infections.
- Take rest and/or play quietly, relax.
- Avoid exposure to high heat or cold air, wind or water.
- Use painkillers and analgesics, if & when required.

Education/awareness & reassurance greatly improve ability to cope with pain.

Excessive pain precipitates a crisis.

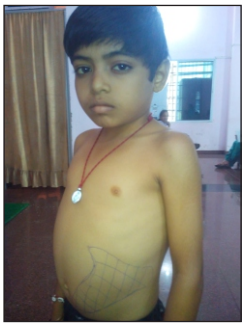
Crisis Management:

Remember, crisis can occur at any time. Its frequency and severity are unpredictable.

To handle crisis, patient/parent/spouse should know how severe the pain is, what kind of pain it is, & where it is located. One should know when to visit the doctor.

The following kinds of specific crises should be known by a SCA/SCD patient:

SPLENIC SEQUESTRATION (SPLEEN CRISIS):

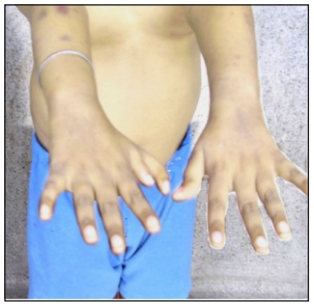


- Occurs when spleen enlarges rapidly.
- Know what a rapidly enlarging spleen is.
- Family members should be educated to assess splenic palpitation & emergency of a rapidly enlarging spleen.
- Abdominal pain.
- Pallor (esp. of lips, gums, nails).
- Education and awareness are very important.
- Early diagnosis helps treatment & relief. Seek a doctor immediately.

ACUTE CHEST SYNDROME (VASO-OCCLUSIVE CRISIS):

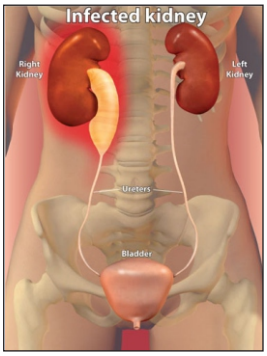
- Occurs when sickle cell blocks the flow of oxygen.
- Generally occurs in children under 16, and in people with chronic haemolytic anaemia.
- High fever, cough, rapid breathing, grunting, hypoxia, asthma.
- Rapid heartbeat.
- Chest pain.
- Can be life-threatening.
- Seek a doctor immediately.

APLASTIC CRISIS:



- Caused when retic count reduces & worsens the patient's baseline anaemia, ie, it prevents RBC production for durations upto 10 days. Availability of RBCs drops suddenly, resulting in an abrupt life-threatening situation.
- Headache, irritability, poor appetite, unusual tiredness.
- Sudden change in complexion, resulting in pale colour.
- Symptoms of jaundice.
- Take rest. Seek a doctor immediately.

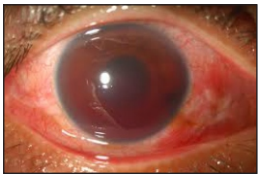
KIDNEY & BLADDER INFECTION/DYSFUNCTION:



- Caused by dehydration (as they drink less water), and due to loss of fluid through diarrhea, vomiting, etc.
- Severe dehydration.
- High fever, UTI.
- More frequent urination than normal (inability to control urination).
- Painful urination.
- Hematuria (blood in urine).
- Take more fluids. Seek doctor immediately.

OPHTHALMIC DYSFUNCTION:

- Caused due to retinal artery occlusion affecting the vascular bed in the eye
- May result in bleeding in eyes, retinal detachment, etc.
- Early stages of eye disease usually do not result in visual symptoms, hence goes undetected in most cases
- May result in permanent loss of vision
- Regular annual or six monthly complete eye check up is advised for all SCD patients
- In case of pain or bleeding, to immediately seek a doctor.

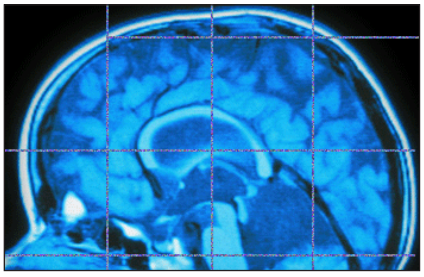


PRIAPISM:

- Occurs due to painful swelling of penis caused by trapped RBCs blocking drainage of the corpora cavernosa of penis, without relation to sexual excitement.
- Can occur both in young children & adults.
- It can occur anytime without sexual drive.
- Painful erection.
- Immediate remedial measures: Warm bath (never use ice packs or cold water); mild massage; mild physical exercise; take more fluids; urinate more.

STROKE & BRAIN:

- Stroke is associated with low baseline haemoglobin, high leukocyte count, frequency of chest syndrome, high BP, and stroke in siblings. Risk mainly dependent upon patient genotype. Affects the large arterial vessels.
- Difficulty with memory/speaking/understanding.
- Fainting, dizziness.
- Persistent headaches.
- Sudden loss of vision or blurred vision.
- Muscular weakness in parts of the body.
- Children are also susceptible to strokes.
- Take rest. Have plenty of fluids. Seek a doctor.



When to call for a doctor right away...

Fever	101 degree F or higher. Persistent mild fever.
Pallor	Noticeable changes in complexion, skin & nails.
Head	Fainting spells. Dizziness. Difficulty in remembering. Stroke symptoms.
Chest	Pain. Coughing. Rapid breathing. Pneumonia.
Stomach	Vomiting more than once in short succession. Sudden gastric upsets. Acute/sharp pain in stomach. Spleen enlarged.
Kidney	Excessive or minimal urination. Blood in urine. UTI.
Bones	Infection. Breakdown of the bones. Severe pain in bones.
Liver	Enlarged liver. Gallstones. Jaundice (yellowing of eyes & skin).
Eye	Sudden blurred vision. Hyphema (blood in the front chamber of the eye).
Leg ulcers	A cut or wound that doesn't heal. A patch of dry & itchy skin.
Hands & Legs Syndrome	Abnormal swelling in extremes of limbs. Painfulness.

Prescription medication:

If a doctor prescribes a pain medication, follow the directions exactly. Do not increase or decrease the dose of your medication without checking with your doctor first — no matter how you're feeling.

Here are some of the other ways to protect yourself:

- Make a note of the effects the drug has on your body and emotions, especially in the first few days as your body gets used to it. Tell your doctor about these.
- Never use someone else's prescription and don't allow a friend or another patient to use yours. Not only are you putting him/her at risk, but you could suffer, too.

Right to Health Benefits:

A patient of SCD should be aware of the various government schemes that carry potential benefit, including but not limited to health care insurance, smart cards, free treatment for all BPL persons, etc. These help reduce personal and financial burdens on poor families for treatment.

Having health benefit insurance is important for patients with special needs because it affects their ability to get medical care, stay well, and prevent future problems. Treatment or insurance coverage can be government or private.

Living Well With Sickle Cell – Social Support:

Living well with SCD requires social support.

Social support is the physical and emotional comfort given to SCD patients by their families, friends, co-workers and others. It involves knowing that you are part of a community of people who value you and think well of you. Social support also involves guidance, social reinforcement and help with the tasks of daily life, as well as help during emergencies.

There are many care-providers in the society, other than parents and family members. A patient may enlist the help of any one or more of these

persons/institutions on a regular basis or when the need arises. Choice depends on emotional bonding, proximity, confidence, knowledge, skill-levels, and privacy issues.

These include:

- Immediate family: parents, spouse, children
- Near family: siblings, uncles/aunts, cousins
- Distant relatives & friends
- Society in general, including teachers, colleagues, casual acquaintances
- Professional persons: doctors, nurses, MPWs/ANMs, para-medical personnel
- Providers: hospitals, health-services units
- Groups: institutions, activists, NGOs, organized society.

The role of these care givers, including family members, friends and outsiders, is directly related to their understanding of the causes and impact of SCA/SCD, and their perception of contributing to a person in need.

Friends, Family, and Relationships:

The qualities of a healthy relationship include:

- Trust
- Honesty
- Respect, and mutual appreciation and respect
- Encouragement
- Support
- Give-and-Take
- Safety
- Shared decision making.

Personal Relationships:

As SCD patients enter adolescence and young adulthood, their personal relationships will change and develop. While these relationships can be exciting, they may be more complicated than when they were younger.

Here are a few things to consider as a patient, when thinking about relationships:

- Friends respect each other and share ideas, feelings, and experiences.
- In a good relationship, you have room for your own interests and friends.
- Good friends will never pressure you into a decision, or an activity that makes you uncomfortable.
- Trust yourself if you begin to feel uncomfortable with someone you have recently met.

Family support:

Friends are important, but one's own family is still essential, too. Things will get stressful and challenging from time to time, and a patient will need his/her family to provide love and affection and support. However a child or adolescent patient will have to gradually learn to become independent as he/she grows up.

Peer support:

If there are other sickling affected adolescents or young adults in the neighbourhood, it would be a good idea of having or creating a peer group, and taking part in a peer support group or attending meetings with others in the transition process. These can provide a young patient with the opportunity to meet and learn from the experiences and coping of others.

As an adolescent or young adult, it is required, even necessary, to take more steps towards personal independence in tasks and financial autonomy, to become competent and responsible for one's own needs, feelings and behaviours. A teenager will have to gradually emerge as a separate, independent person with his/her own identity and values.

Treatment:

Treatment	Category	Benefits	Side-effects
Hydroxyurea	Standard drug prescribed for primary treatment	• Has chemotherapeutic effect which increases "F" Fetal Haemoglobin • Reduces incidence of acute painful events & acute chest syndrome • Mortality reduced by 40% • Less haemolysis (less anaemia) • Fewer hospitalization • Reduced medical costs • Improved physical capacity	• Potential for carcinogenicity/ cytotoxicity • Not recommended for patients trying to conceive & for pregnant women
Blood Transfusions (BT)	During Aplastic & other crisis	• Increases normal haemoglobin levels immediately • Can prevent strokes in high risk patients	• Repeated transfusions leads to serious complications of iron overload • High risk of alloy-immunization • Risk of transmission infections increases
Bone Marrow Transplant (BMT)	Destroys original bone marrow, replaces with new bone marrow	• New bone marrow starts manufacturing sickle-free RBCs • In most cases, a permanent cure	• Technique is toxic & risky • Other complications • New technique, research driven
Stem Cell Treatment		• New RBCs are manufactured sickle-free • Possible permanent cure	• Technique in infancy in India
Gene Therapy	Changes the hemoglobin gene	• Possible permanent cure	• Toxic nature • Technique in infancy

IV
HEALTHY HABITS

Nutrition:



- Take a high protein diet.
- Take a high fibre diet, plenty of roughage (as pain medication causes constipation).
- Take plenty of fluids (water).
- Have a diet rich in anti-oxidants, and in vitamins C & E (Antioxidants, Vitamin C & E may help prevent formation of the dense cells that trigger a sickle cell pain episode) .
- Avoid oily and fatty foods (masala).

Avoid Smoking:

- Smoking decreases lung capacity and carries a high risk for cancer.
- Smoking has been linked to a higher incidence of chest syndrome, a leading cause of hospitalization and mortality in people with SCD at any age.
- Even second-hand smoke (passive smoking, i.e. being in the room or car where someone else is smoking) can cause lung problems in people with SCD at any age.

Avoid Drinking Alcohol/Taking Tobacco:


- Too much alcohol may cause dehydration, with adverse impact for SCD patients.
- Alcohol can damage liver.
- Tobacco puts pressure on heart, can cause cancer.

Have daily routines:

- Normal regular walking or simple exercise for short durations helps.
- Exposure to early morning sunshine is recommended.
- Work, home, and daily routines should avoid large fluctuations in temperatures.

Vocation and career choice:

Children and young adults with SCD should set their educational and career goals in accordance with their ambitions and innate abilities. **Jobs requiring heavy manual labour and strenuous activity, military service, long work hours, exposure to hypoxia, and working in extreme temperatures are not options for SCD persons.** Role models are important as a person with SCD who has been vocationally successful can provide useful tips for youngsters with the same problem.

Body Image and Self-Esteem:

For a patient, half the battle is for one's own confidence. Whatever one does, or steps one takes, it is important to know that there are risks associated with each of these behaviours.

Living with a long-lasting health condition, like sickle cell, diabetes or asthma presents a person with new challenges. Learning how to meet those challenges is a process — it doesn't happen right away. But understanding more about one's own condition, and doing one's own part to manage it, can help a patient take health challenges in his/her stride. Many people find that taking an active part in the care of a chronic health condition can help them feel stronger and better equipped to deal with most of life's trials and tribulations.

It takes time to adjust to and accept the realities of a long-term illness, but teens and young adults who are willing to learn, seek support from others, and participate actively in the care of their bodies usually get through the coping process much simpler and much easier.

V
COPING WITH THE DISEASE

The Coping Process:

Most people go through 3 stages in learning to cope.

Some people feel vulnerable, confused, and worried about their health and the future. Others feel sad or disappointed, while for some, the situation seems unfair, causing them to feel angry at themselves and the people they love. These feelings are spontaneous, and form at the start of the coping process. Everyone's reaction is different, yet they are all completely normal.

The next stage in the coping process is learning. The fear of the unknown needs to be removed. Most people living with sickle cell find that knowledge is power: The more they find out about their condition, the more they feel in control and the less frightening it is.

The third stage in coping is all about taking it in stride. At this stage, patients feel comfortable with their treatments and with the tools (like inhalers or medications) they need to use to live a normal life. A good coping strategy effectively ameliorates chronic pain, reduces psychological distress, and boosts confidence in SCD patients.

Remember – ***there is no definite time frame or limit as regards a person's coping process.*** Everybody's process of coming to terms with and accepting a chronic illness is different. In fact, most people will find that emotions surface at all stages in the process. Even if treatments go well, it's natural to feel sad or worried from time to time. Recognizing and being aware of these emotions as they surface is all part of the coping process.

Sad Too Long? Signs and Symptoms of Depression:

Living with a chronic illness such as sickle cell disease increases the chances of developing depression. It is because the afflicted person keeps thinking about his predicament all the time, without any optimism for the future.

If a sad feeling goes on for too long…hurts too deeply….and makes it hard to enjoy the good things about your life, it may be **depression. Talk to a trusted person or call a doctor or counsellor**.

Some signs and symptoms of depression are:

- Feeling hopeless (like there is nothing to look forward to)
- Feeling guilty or worthless
- Feeling lonely or unloved
- Feeling irritable and annoyed at small things (every little thing gets on the nerves)
- Feeling like things are not fun anymore
- Having trouble focussing attention on schoolwork/homework, or getting bad grades
- Having trouble keeping the mind on things like reading or watching TV, or not remembering what a book or a TV show was about
- Feeling less energetic, and feeling tired all the time
- Sleeping too much or not enough
- Not eating enough and losing weight, or eating too much and having weight gain
- Thinking about death or thinking about suicide
- Spending less time with friends and more time alone
- Crying a lot, often for no reason
- Feeling restless (being unable to sit still or relax)
- Having certain body feelings, like lot of stomach aches, headaches, or chest pain.

Remember – many of the above symptoms may be more perceived or imagined than real!

How Is Depression Different From Regular Sadness?

No one's life is perfect. Sadness is a natural emotion because life has its ups and downs, and not everything desired can be achieved. The normal stresses of life can lead anyone to feel sad every once in a while. An argument with a loved one or a friend, a breakup, doing poorly in an exam, not being selected for a job – any or all of these can lead to feelings of sadness, hurt, disappointment, or grief. These reactions are usually brief and go away with a little time and care, or with the mind getting diverted.

Depression is more than occasionally feeling low, sad, or down in the dumps. It is a strong mood involving sadness, discouragement, despair, or hopelessness, and it lasts for weeks, months, sometimes longer. It interferes with a person's ability to be normal or participate in normal activities.

Depression affects a person's mood, thoughts, outlook, and behaviour. In addition to a depressed mood, a person with depression can also feel tired, irritable, and have major changes in appetite.

When someone has depression, it can cloud everything. The world looks bleak and the person's thoughts reflect that hopelessness and helplessness. People with depression tend to have negative and self-critical thoughts. Sometimes, despite their true value, people with depression can feel dispirited, lethargic, worthless and unloved.

Because of feelings of sadness and low energy, people with depression may pull away from those around them or from activities they once enjoyed. This usually makes them feel lonelier and isolated, making the depression and negative thinking worse.

Depression can be mild or severe. In its mild form, it can lead to withdrawal or aloofness. At its worst, depression can create such feelings of despair that a person thinks about committing suicide.

Depression can cause certain physical symptoms, too, including but not limited to stomach upsets, loss of appetite, weight gain or loss, headaches, and sleeping problems.

Getting Help:

Depression is one of the most common emotional problems among SCD patients in Chhattisgarh and in India. The good news is that it is also one of the most treatable conditions. Counsellors and therapists can help. In fact, about 80% of people who get help for their depression have a better quality of life — they feel better and enjoy themselves in a way that they were not able to before.

Treatment for depression can include talk therapy, medication, or a combination of both.

Talk therapy with a psychologist or psychiatrist or health professional is very effective in treating depression. Therapy sessions can help people understand more about why they feel depressed, and ways to combat it.

Sometimes, doctors prescribe medicine for a person who has depression. When prescribing medicine, a doctor will carefully monitor patients to make sure they get the right dose. The doctor will adjust the dose as necessary. It can take a few weeks before the person feels the medicine working. As every person's brain is different, what works well for one person may not be work for another.

Everyone can benefit from mood-boosting activities like exercise, yoga, dance, journaling, or art. It can also help to keep busy no matter how tired or dispirited one feels.

Living Well With Sickle Cell – Travel tips:

Live with optimism – it doesn't hurt. Do what brings joy and satisfaction, but know the limits and always be prepared!

Travelling can ease pessimism, and broaden horizons. However a patient should be prepared.

Taking a Flight:

It is a fact that high altitudes can contribute to the breakdown of red blood cells. SCD patients are particularly susceptible. Although modern aeroplanes fly at a height of about 30,000 feet, they have pressurized cabins, which are adequate for sickling patients. For added safety, SCD patients planning long flights may play safe, and call the airline ahead of time to find out what altitude the flight will reach and what procedures are in place for the use of supplemental oxygen.

Altitude of a destination:

If traveling to a high altitude destination, such as Shimla or Ladakh, SCD patients should take time to get used to the new high altitude surroundings. Never try to do too much too fast. Drinking plenty of water is advised.

High altitudes and the cold air, wind and water there may trigger a painful event, as sickle cell hampers the smooth movement of RBCs when the body requires more blood in an effort to keep warm.

Dehydration:

The air in Chhattisgarh and in many places in India is dry, so one needs to drink more water than usual to avoid becoming dehydrated. While travelling, especially in a bus or taxi or even in an aeroplane, or moving in the open, drinking plenty of water is a must.

Dressing Appropriately:

Dress should be appropriate so that a patient does not get too cold or too warm while traveling. Taking along a light blanket or wrap is advisable in an AC train or car journey, or at a cool destination. Similarly for a hot destination, care is to be taken that the body is not exposed to extremes of temperature.

Keep Moving:

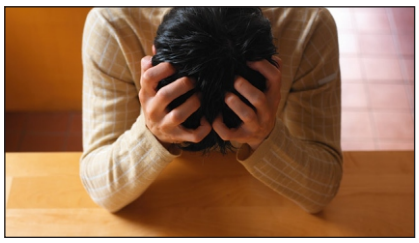
Regardless of how one is traveling, if the trip lasts more than two hours, it is strongly advised to get out of the seat/berth and move about the cabin, or take frequent rest stops. This keeps blood from pooling in the legs and feet, and enhances circulation, thereby reducing cramps and painful episodes.

Keeping A Portable Medical History or Summary:

Whenever travelling, a patient should always carry some documentation of his/her medical history in the event of requiring emergency care. It would also be wise to have the contact numbers for the doctor/hospital who normally caters to him/her.

Stress and fatigue:

Although indirectly related, stress and fatigue often are part of any travel experience. However these can trigger a painful episode. Hence, after arriving at a new destination, one should spread out schedules, relax and enjoy the holiday!



VI
Self-Assessment

Each SCA/SCD patient must know enough about the disease/disorder in order to cater to any arising eventuality.

Do you have the personal confidence and the social supports you need? Are you confident about your medical condition and what it takes to combat it?

Take 2 minutes to identify your areas of strength, and highlight where there is room to grow!

Patient Self-Assessment Form

1. Personal Skill Set	Yes/Fully	Partly	No	I am not sure /I don't understand the issue
I know & understand what SCA & SCD is.				
I understand what limitations SCA/SCD places on me and my activities.				
I know what foods to take and what to avoid.				
I know what kind of regular activities to undertake.				
I have hobbies or personal activities that I enjoy.				
I have no hesitation in asking for help whenever required.				

2. Social Skill Set	Yes/Fully	Partly	No	I am not sure /I don't understand the issue
I understand what self-esteem is.				
I understand the difference between being sad and being depressed.				
I understand what "Guardianship" means.				
I understand what healthy relationships are and what they mean to me.				
I participate in activities at school and/or outside the home.				
I participate in small informal gatherings like picnics, etc.				
I participate in social gatherings like festivals, marriages, community events, etc.				
I have a person(s) that I can talk to without hesitation about SCD.				
I have a vision about my future, and a plan for what I would like to do in the years to come.				

3. Health Skill Set	Yes/Fully	Partly	No	I am not sure /I don't understand the issue
I understand how my age is a factor in assessing impact of SCA/SCD.				
I think I can handle general pain episodes by myself.				
I understand how drugs, smoking, alcohol and tobacco usage affect sickle cell disease.				
I know the various types of health benefits available to me.				
I understand how my age can affect my medical benefits.				

3. Health Skill Set (Continued)	Yes/Fully	Partly	No	I am not sure /I don't understand the issue
I always carry my health insurance & other benefit cards with me.				
I always carry a brief record of my medical history when I leave home for more than a day.				
I always carry drinking water and/or money with me for emergency requirements.				
I try to keep myself at a steady pace, warm, and avoid extreme temperatures and strenuous activities.				

Wherever you are unsure of yourself, do find out details, take appropriate measures and re-assess yourself to a higher confidence and better coping level!

Last but not the least, it is up to an informed individual to know about the disease and prepare himself/herself accordingly.

The decisions he/she makes on a daily basis result in real consequences that shape his/her future.

We at the ***Sickle Cell Institute of Chhattisgarh*** trust that you will ask for the support you need as you make your own decisions. As your healthcare team, it is our job to provide you with reliable information affecting your health. We advise you NOT to smoke, drink, or take drugs or other medication irresponsibly. We want you to make informed decisions, and act responsibly.

We believe that your health and well-being should be a priority and that true friends will respect and support your choices.

Life with Sickle Cell is a challenge. If you wish to know more about the problems at hand, potential mitigating factors, or have any queries or require any kind of guidance or counselling, we, at the SCIC Raipur, would be glad to handhold you at the time of your need. You may visit us or call us at the Toll Free number.

We wish you a comfortable and a confident life ahead!

2014

Dr. (Mrs.) Radharani Sahoo
Counsellor

HOLDING YOUR HAND - THROUGH LIFE

Sickle Cell Institute of Chhattisgarh

For further details and for counselling, contact:

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