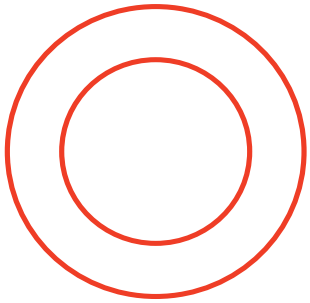
FOR PATIENTS & CAREGIVERS

Learning about sickle cell disease

People want to learn about sickle cell disease (SCD) for different reasons. Your newborn baby or young child may have it. You may have SCD and want to know more. Or you might care for a family member with SCD. Learning all you can about SCD can help you take better care of yourself, your child, or your loved one.



Normal red blood cell

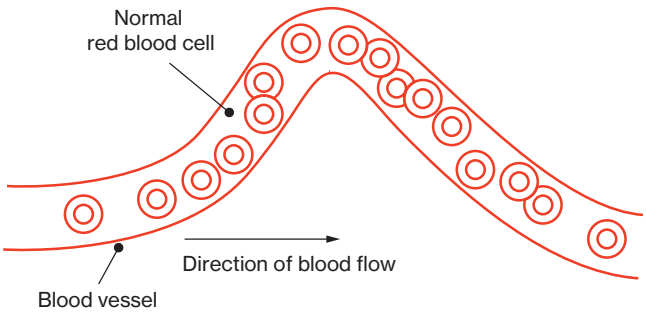
Sickle cell

What is SCD?

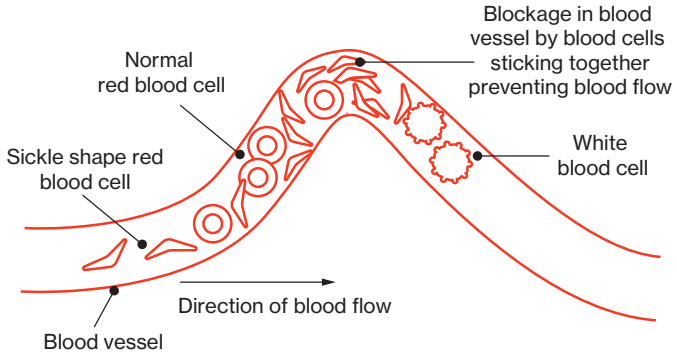
SCD is a disease that affects red blood cells. Normal red blood cells look like round discs and are very flexible. They flow through blood vessels with ease. Red blood cells carry oxygen to all parts of the body.

SCD causes red blood cells to change shape. With SCD, they are shaped like a sickle or a crescent moon. Sickle cells are stiff, not flexible. They break apart easily and stick to each other. They can clog up blood vessels, especially small ones. This slows or stops the flow of blood and oxygen to nearby tissues. A lack of oxygen to tissues can cause severe pain attacks called ***pain crises***.

Healthy individual



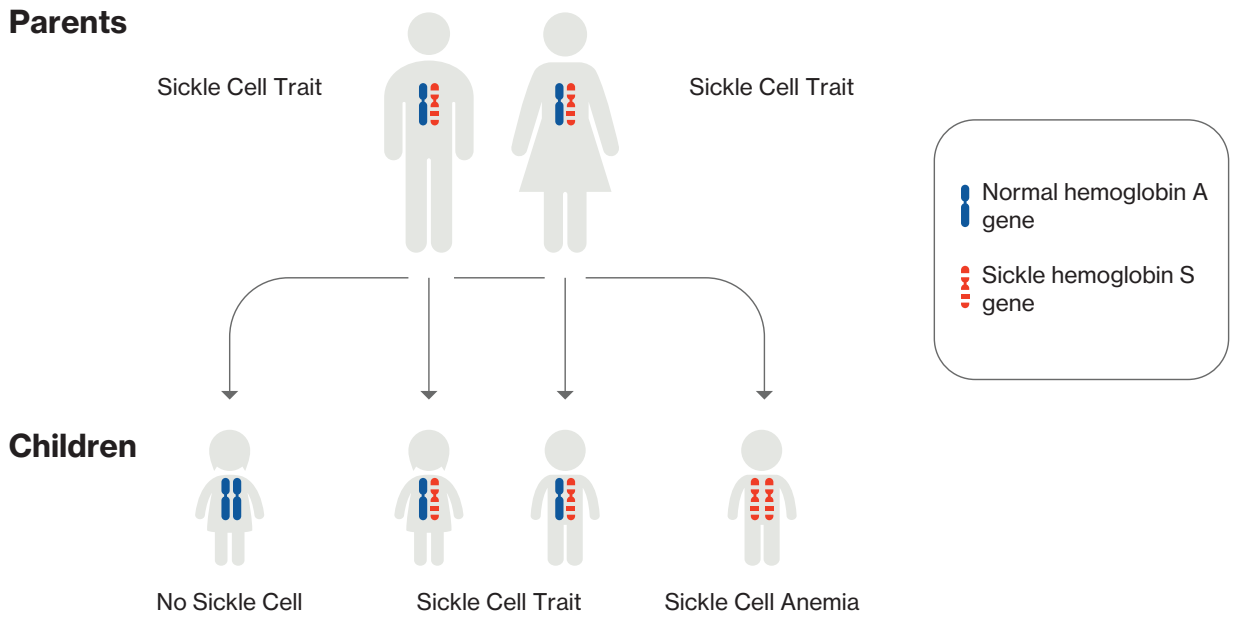
Individual with sickle cell anemia



What causes SCD?

People who have SCD are born with it. This means it is an ***inherited*** disease. You can't catch SCD, like the flu, or get it later in life.

SCD happens when both parents pass on the sickle cell gene to their child. Sometimes, the sickle cell gene is passed on to a child from only 1 parent. This child will be born with ***sickle cell trait.*** When both parents have sickle cell trait, their children have a 1 in 4 chance of having SCD. You can see how all this works in the image below.



Who gets SCD?

Most children born with SCD are of Black or of African ancestry. But people from other racial backgrounds, such as Hispanic, can be born with it, too. Here are some facts about SCD in the United States:

1 of every 365 African-American babies is born with SCD. About 100,000 people have SCD.

News and better news

Here's some news that's not so good: SCD does not go away, and it gets worse over time. But here's better news—SCD used to be thought of as a childhood disease. Today, people with SCD often live well into their 40s.

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Sickle cell disease diagnosis

Receiving a diagnosis of sickle cell disease (SCD) can be difficult

Learning that you, your child, or loved one has SCD can be quite upsetting. You may feel a wide range of normal emotions such as:

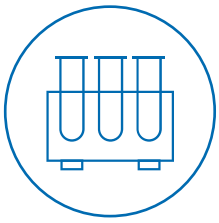
- Shock and disbelief, especially for parents who did not know they had sickle cell trait
- Depression
- Fear
- Anger
- Guilt
- Helplessness
- Confusion
- Anxiety

Tests can show if a person has SCD or sickle cell trait

Here are some tests doctors use to know if someone has SCD:

In the United States and many other places, ***all*** newborn babies must be tested for SCD.

Parents may choose to have their baby tested for SCD ***before*** birth. Babies float in fluid inside a mother's womb. The fluid or tissues from the womb can be tested. Tests look for the sickle cell gene. The test can be done as early as 8 to 10 weeks into pregnancy.



Anyone can have a blood test to learn if they have sickle cell trait. This information helps people know if they might pass on the sickle cell gene to their children.

Early signs of SCD

Most babies born with SCD do not show signs of it right away. Signs of SCD often do not start until 5 or 6 months of age. It is around this age that a baby's red blood cells start to sickle and cause problems.

It is smart to know these early signs of SCD:
Baby's skin or whites of the eyes look yellow. This is called ***jaundice*** (JAWN-dis)
Baby is often very sleepy or fussy. This may be caused by anemia, which is when the body does not have enough red blood cells
Baby's hands and feet are swollen. This happens when sickle cells start to block blood flow in and out of the hands and feet

Early signs of SCD are not the same for every baby. If your baby shows any of these signs, be sure to let your doctor know right away.

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Sickle cell disease may cause health problems

When blood cells clump together and reduce or stop blood flow to parts of the body, health problems can happen. **Sometimes, problems are serious enough that you must go to the doctor or hospital right away for treatment.**

Some samples of serious health problems that need to be treated as soon as possible

Pain crisis – ***A very bad pain that happens suddenly with no warning. This sudden pain can:***

- Happen in any part of the body at any time
- Feel **very** bad, like the worst pain you've ever had

Pain crisis happens when blood flow is blocked by sickle cells and other blood cells sticking together somewhere in the body.
It can occur when a person with sickle cell disease (SCD):

- Gets too cold
- Does not drink enough water
- Is sick or stressed out

Acute chest syndrome (ACS) – ***Happens when sickle cells block blood vessels in the lungs.*** This can cause lung damage and stop oxygen from going to other parts of your body. ACS often starts a few days after a pain crisis begins. **ACS can be life-threatening and must be treated right away in a hospital.**

Symptoms and signs of ACS include:

- Chest pain
- Cough
- Hard time breathing
- Fever

Anemia ***(uh-NEE-me-uh) – When the body has too few healthy red blood cells to carry oxygen to all parts of the body.*** Sickled red blood cells die early.

The body cannot make new red blood cells fast enough to replace them. When this happens, a child or adult might feel:

- Tired all the time
- Cranky
- Dizzy or lightheaded
- Short of breath

Other symptoms of anemia can be:

- A fast heartbeat
- Yellowish color of the skin and whites of the eyes (jaundice)
- Slow growth and late puberty

Severe anemia – is caused by a type of infection. This infection stops the body from making new red blood cells for a while. It can put the lives of newborns and children in danger.

Severe anemia can also be caused by a problem in the ***spleen***, an important organ in the body. In people with SCD, the spleen can suddenly grow much larger than normal. It happens when sickle cells trap red blood cells inside the spleen. Red blood cells can't flow out of the spleen and through the body.

You should know these signs of severe anemia in infants and children:

- They seem too quiet and don't have any energy
- They do not want to eat

Infections – ***People with SCD, especially infants and children, are at high risk of infections.*** The spleen helps protect the body from certain kinds of infections. If the spleen is damaged by SCD, it can't do its job well and infections happen. A lung infection called ***pneumonia (new-MOH-nyah)*** can be deadly in infants and children with SCD.

These are things you can do to decrease infections:

- Wash your hands many times each day with soap and clean water
- Prepare food safely
- Get all recommended vaccines

Many children take medicine, like penicillin, every day to fight infections until age 5 years or older.

Stroke – ***Damage to the brain caused when blood flow to a part of the brain is stopped by sickle cells.*** Strokes damage the brain, and they can be silent or non-silent. A silent stroke damages the brain with no outward obvious sign to others.

- Silent strokes can happen in adults and children with SCD
- Strokes can cause learning problems and lifelong disabilities

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Priapism ***(PRY-uh-piz-im) – A painful erection that lasts a long time***. It happens when blood flow out of an erect penis is blocked. Over time, priapism can damage the penis and a man can no longer get an erection.

If priapism lasts more than 4 hours, it is an emergency. Men or boys should go to the hospital right away.

Other health issues, some of which may develop over time

Organ damage – ***Cells can block the flow of oxygen-rich blood to nerves and organs in the body***. Organs are parts inside your body such as your lungs, heart, kidneys, liver, spleen, and others. When organs are damaged, they don't work correctly and can lead to complications. Ask your doctor what you can do to keep these organs as healthy as possible.

Loss of vision – ***SCD can damage blood vessels in the eyes.*** You or your child may not be able to see well as a result. Unfortunately, if eye damage is very bad, you or your child can have visual impairment or loss. This is why it's so important to have your eyes tested as often as your doctor recommends.

Leg ulcers – ***SCD can cause open sores on legs***. Ulcer is another word for this kind of sore. They often start to appear after 10 years of age. Sores can heal quickly or slowly—this varies among people.

Gallstones – ***Small stones form in the gallbladder.*** The gallbladder is a small organ under the liver. When red blood cells die, a part of them turns into something called ***bilirubin (bill-ee-ROO-bin)***. Bilirubin can form stones that get stuck in the gallbladder. Gallstones are a common problem in SCD.

Problems during pregnancy – ***Pregnant women with SCD are at increased risk of certain problems***. These risks include:

- High blood pressure and blood clots
- Miscarriage, premature birth, and low-birth-weight babies

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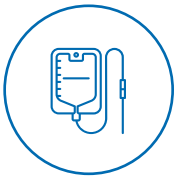
FOR PATIENTS & CAREGIVERS

Treating sickle cell disease

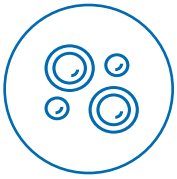
There are many ways to manage sickle cell disease (SCD).

Medicines – Medicines for SCD can help manage problems the disease can cause. In the past few years, new treatments have become available that can help treat symptoms of SCD.



Blood transfusions – In a blood transfusion, extra blood is put into a person's body through IV injection. There are a few reasons why a person with SCD might need a blood transfusion:

- ***Acute transfusion*** – Treats severe anemia, acute stroke, acute chest crises, and organ failure to prevent more problems
- ***Red blood cell transfusion*** – Adds extra normal red blood cells resulting in better oxygen delivery to the body
- ***Repeat transfusions*** – Transfusions may be given often to someone who has had an acute stroke to lower the chance of more strokes. A child who is at risk of having a first stroke may be given transfusions



Stem cell transplant – In a transplant, a doctor puts healthy stem cells into the bloodstream of someone with SCD. These new stem cells replace that person's damaged stem cells.

What are stem cells? Stem cells are special cells that can become many different types of cells. For someone with SCD, the new stem cells they receive become healthy new red blood cells that replace sickle cells.

Talk to your doctor to learn more about what treatments may be right for you.

A key step: when a child with SCD moves from pediatric care to adult care

The move from childhood SCD care to adult care is a big and sometimes scary step for both teenagers and parents. You and your teen's health care team should work together to make a plan that can help your teen make the switch to adult care. **This time in a teen's life is when more health problems and hospital stays often start.**

These are some of the reasons why:
Aging can bring about organ damage and disabilities
The shift to adult care happens just when teens are going through physical, emotional, social, and academic changes. Being more independent and taking more responsibility for themselves can be hard for teens and parents to cope with. Teens may not stick to their treatment plan well
There are often fewer SCD medical and social resources nearby for adults than for children

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Staying with a treatment plan

When you or your child follow your treatment plan:

 SCD problems may be reduced

 Health care costs could go down

Unfortunately, many people with SCD don't stick with their treatment, especially teens and young adults. There are many understandable reasons for this. But you can take steps to help yourself stay on track.

Here are some common reasons why people get off track and some suggested ways to get back on board:

REASON	SUGGESTION
I can't get to the clinic.	Find out if there is a free transportation service in your town to take you to the clinic or hospital. Don't be embarrassed to ask a friend or family member to take you there.
I forget or am too busy to fill or refill my prescriptions.	Have the pharmacy send you refill alerts; ask for a 90-day supply of medicine and have it automatically refilled and delivered by mail.
My child is or I am afraid of needles.	Ask the doctor if there is another treatment option for you or your child.
I can't afford the medicine.	Many drug companies have assistance programs that may help people pay for their medicines. Ask someone at the clinic to look into this for you.

REASON	SUGGESTION
I often forget to take my medicine.	Do you have a cell phone? If so, it can help you remember to take your medicine. You can use your cell phone to: • Get text messages each day that remind you to take medicine and stick to your treatment plan • Set alarms for times to take medicine • Enter dates for appointments, tests, IV infusions, and refills into the calendar • Go online to learn all you can about SCD and find others like you who have SCD
People in my family don't believe in taking medicine.	Ask someone you trust on your health care team to explain why it is so important to stick with all parts of your SCD treatment plan, including taking medicines.
I am scared of medicine side effects.	Please discuss possible side effects associated with your medication and any concerns you may have with your doctor.

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Living with SCD

Life with sickle cell disease (SCD) can be a challenge. But there are things you can do to help yourself, your child, or your loved one live life better:

- Recognize and understand your feelings
- Adapt your lifestyle
- Stay with your treatment plan
- Know the warning signs of trouble

Understanding your feelings

You, your child, or loved one has SCD. You may have a wide range of natural and common feelings. Shock and disbelief may be your first feelings. You might feel upset, angry, or guilty that you passed SCD to your child. When you realize that you can't take away this illness from your child or yourself, you may feel helpless and not in control. Your feelings will no doubt change over time. It's important to recognize your feelings and not feel guilty for having them.

Know the steps you can take to stay as healthy as possible

Make changes to help with sickle cell disease

- Follow your treatment plan every day
- Drink lots of water or other liquids each day
- Try not to get too cold or too warm
- Whenever you can, stay on low ground, not at a high altitude
- Avoid very hard exercise that could lower your oxygen levels

Adopt a healthy lifestyle

- Be physically active, but skip hard exercise
- Get enough sleep
- Drink lots of fluids but limit alcohol
- Follow a heart-healthy diet that's right for you
- Quit smoking

Care for your mental and emotional health

- Get support from others to help lower your stress and anxiety. Share your feelings and be heard
- Seek help from a mental health expert such as a counselor or therapist. Getting this kind of help shows you are strong, not weak

Know the warning signs of serious SCD problems and have a plan

Some problems caused by SCD can be life-threatening. So it's really important for you to learn what they are and have a plan to get medical help right away.

Severe anemia – Here's how you might feel with severe anemia: you are very tired, dizzy, or lightheaded; your heart beats very fast; you are short of breath; and your skin is very pale or looks yellow. Get medical help right away.

Fever – Newborns and very young children should be taken to the doctor or hospital right away if they have any temperature above normal. If your older child or you have a fever over 101.3°F or 38.5°C, go to your doctor or hospital quickly. Treatment must start right away.

Acute chest syndrome – Go to the doctor or the hospital right away if you have chest pain, cough, fever, or have a hard time breathing. You may need to go to the hospital and get antibiotics, oxygen therapy, or a blood transfusion.

Stroke – If you think your child or someone else is having a stroke, act F.A.S.T.

F	A	S	T
Face	**Arms**	**Speech**	**Time**
Ask the person to smile. Does one side of the face droop?	Ask the person to raise both arms. Does one drift down?	Ask the person to repeat a simple phrase. Is their speech slurred or strange?	If you see any of these signs, call 911 right away. Early treatment is crucial.

Priapism – If you or your child has an erection that lasts 4 or more hours, go to the hospital immediately.

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FOR CAREGIVERS

Care for the caregiver

Are you a caregiver for someone with sickle cell disease (SCD)? If you are, this flashcard is all about you. If you care for a baby or young child, you are responsible for his or her physical care and emotional support. Or you may be helping an older child or young adult to navigate life with SCD. In both cases, caring for a person with SCD can be rewarding. But it can also be challenging and stressful.

Self-care is very important

Caring for yourself is one of the most important things you can do, but caregivers often ignore their own needs. You need to understand that self-care is not selfish. It is crucial. When your needs are met, you are helping the person you care for. Here is a quote from a caregiver that sums things up nicely: ***The care you give yourself is the care you give to your loved one.***

Caregiving shows love and commitment but, over time, constant care demands can take their toll on you. Many caregivers find it hard to pay attention to their own health and well-being. But when you ignore these things, you can actually make yourself sick. Here are some things you should be aware of.

Caregivers are at a higher risk than non-caregivers of:
Depression
Alcohol abuse and long-lasting health problems such as high blood pressure and being overweight
Exhaustion from lack of sleep, poor eating habits, lack of exercise, and lack of self-care when they are sick

How to recognize your own needs and get them met

It's not always easy to know what you want and need as a family caregiver. A first step might be to make an I Need list. Do this when you are alone and can take time to think about it before you begin. Here's a good example of an I Need list:

I need help with:

- Grocery shopping and meal prep
- Taking care of the house and garden
- Walking the dog

I need time to:

- Take a nap
- Walk in the park
- Go to girls night out
- See the dentist
- Attend church

If you decide to make an I Need list, you may want to share it with family and friends. That way, they will know how and when to help you.

Take action to reduce stress

Make time to do things that can lower stress. Doing these things may help you feel better and more in control. Think about activities that help relax your body and your mind. Here are some things that are stress busters for some people:

- Spend time with nature – walk in the woods, work in the garden
- Listen to music or read
- Meet a friend for lunch
- Work out at the gym or play a sport
- Soak in a warm bath

You may want to try different types of therapy that help reduce stress and anxiety. Some examples are:

- Massage
- Yoga or tai chi
- Meditation or guided visualization
- Deep-breathing exercises

Get knowledge and feel the power

Knowledge is power. When you learn about SCD, you can better understand what's happening now and what to expect over time. Knowledge can help you communicate better with health care providers. It may help improve your confidence as a caregiver. Knowledge helps you speak smartly on behalf of your child or family member.

You may want to join an SCD caregivers support group if there is one close by. There you can meet people who understand what you are going through. You may also learn caregiving tips and new ways to cope.

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Questions for your care team

You may have found out recently that you or a loved one has sickle cell disease (SCD), or you may have questions about SCD. No matter what, it is important to talk to your care team about the questions that you have. That is why it is a good idea to write down your questions for your care team so that you do not forget them.

Keep a list of your questions in a handy place. Take your list to your next care team visit. Doing so may help you talk with your care team about the things that are important to you. It also shows your care team that you want to take an active role in your SCD care.

Here are examples of questions you might want to ask your care team:

1. Is there a cure for SCD?
2. What treatment option might be right for me?
3. What are the side effects of my SCD treatment?
4. How often should I be assessed or visit my doctor?
5. Are there any clinical trials that I can join?
6. I am having more pain crises. What does this mean? What can I do to help manage the pain?
7. If my baby has the sickle cell trait, will they get SCD later in life?
8. What kinds of physical activities are best for me/my child and what precautions should I take?
9. Is it safe for my child with SCD to go to school/work? Do I have to tell the school or my work that either of us has SCD?
10. What do I need to know before I travel by air?
11. Are there local SCD support groups for teens, adults, or caregivers?
12. I am worried about paying for my SCD treatment. Is there any way I can get help?

Notes

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Sickle cell disease support and assistance

There are many support and assistance resources for adults with sickle cell disease (SCD), parents of a child with SCD, and family caregivers. Here are some of them:

EDUCATION, SERVICES, AND SUPPORT

Sickle Cell Disease Association of America (SCDAA)

www.sicklecelldisease.org

This is a national organization with over 50 local chapters. Come to this site to find free educational materials about living and coping with SCD. SCDAA also offers:

- Screening tests recommended for people living with SCD
- Counseling and social services
- Transportation
- Summer camp
- A sickle cell chat room

American Sickle Cell Anemia Association

www.ascaa.org

This national organization provides many services, including testing, counseling, and support services to people and families who are at risk of or already have SCD.

ONLINE COMMUNITIES

Sick Cells

www.sickcells.org

This is an online community for people with SCD. Its purpose is to make sure the voices of people with SCD are heard. The goals of the organization are to discover what people with SCD need and to stand up for them.

Generation S: Sickle Cell Awareness

www.joingens.com

This online community helps people with SCD learn about SCD and find support. Visitors to the website can read and share stories about living with SCD.

LEARN ALL ABOUT SCD

National Heart, Lung, and Blood Institute (NHLBI)

www.nhlbi.nih.gov/health-topics/sickle-cell-disease

NHLBI is part of the National Institutes of Health (NIH). Here you will find much information about SCD, research, and ways to manage SCD.

Centers for Disease Control and Prevention

www.cdc.gov/ncbddd/sicklecell

Learn more about SCD, who has it, and how SCD health problems can be prevented.

American Society of Hematology (ASH)

www.hematology.org/advocacy/sickle-cell-disease-initiative

The national organization for hematologists (blood experts) offers education about blood diseases and raises awareness of the importance of healthy blood.

KidsHealth

www.kidshealth.org/en/kids/sickle-cell.html

This excellent website is very user-friendly. It has separate sections for kids, teens, and parents to learn about SCD in simple, easy-to-understand language. You can also learn about many other kinds of diseases and health problems, from cancer to warts. The website includes articles, slideshows, and videos.

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