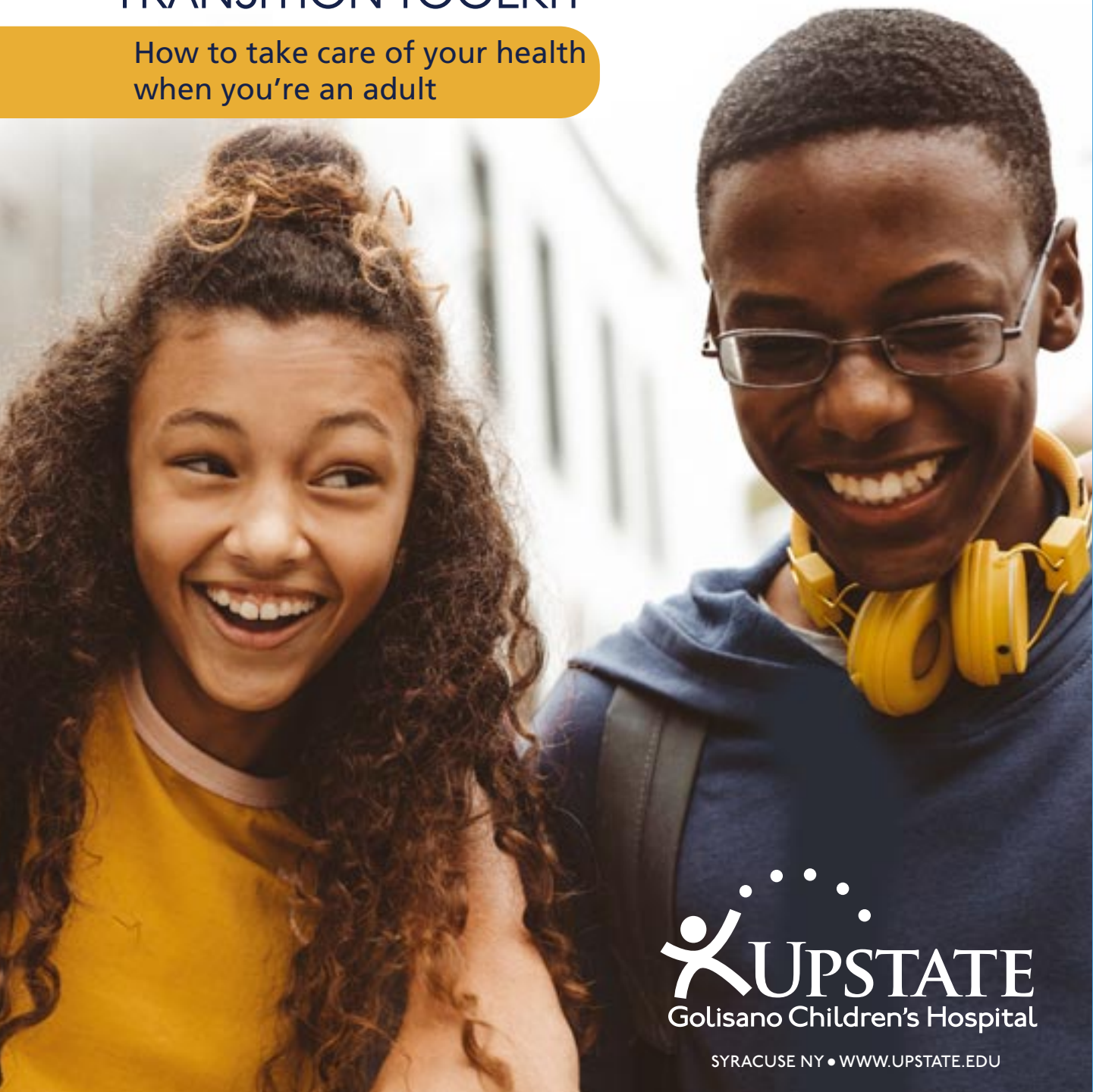


# THRIVING ONWARD

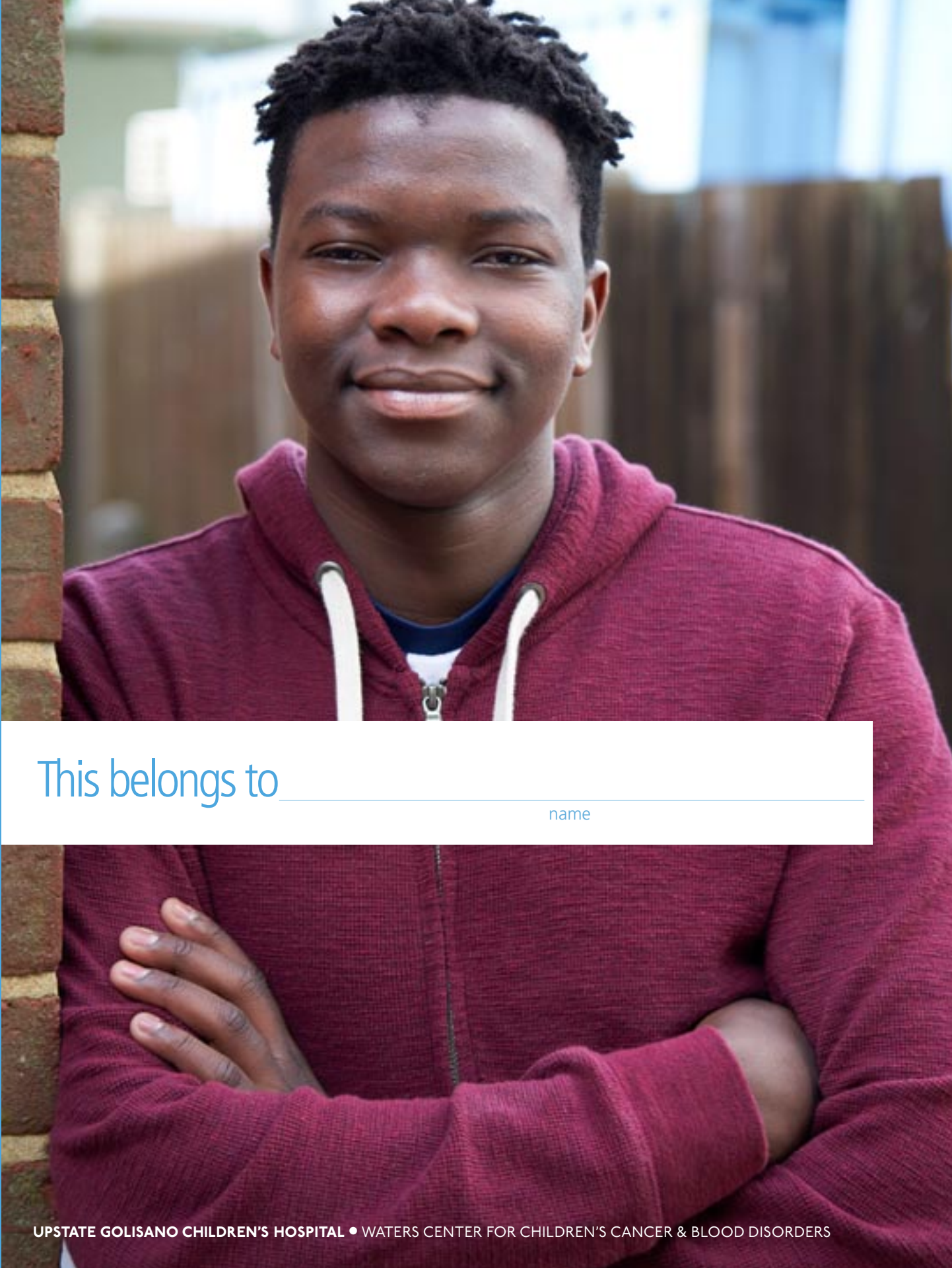
## SICKLE CELL DISEASE TRANSITION TOOLKIT

How to take care of your health  
when you're an adult



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# THRIVING ONWARD

## SICKLE CELL DISEASE TRANSITION TOOLKIT

How to take care of your health when you're an adult

By  
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## Pre-test: Sick Cell Disease Transition Readiness Assessment

Please fill out this form to show what you know about your health care and to identify areas that you want to learn more about. If you need help completing this form, please ask your parent/caregiver or health care provider.

Date	Name	Date of Birth									
On a scale of 0 to 10, please circle the number that best describes how you feel now. (0=not, 10=very)											
How important is it to you to manage your own health care?											
0 (not)	1	2	3	4	5	6	7	8	9	10 (very)	
How confident do you feel about your ability to manage your own health care?											
0 (not)	1	2	3	4	5	6	7	8	9	10 (very)	
How confident do you feel about preparing for/changing to an adult doctor before age 22?											
0 (not)	1	2	3	4	5	6	7	Does not apply:	8	9	10 (very)

 Please check the box that applies to you now.	No, I do not know	No, but I am learning to do this	Yes, I have started doing this	Yes, I always do this when I need to
<b>Disease Knowledge</b>				
I know what type of sickle cell disease I have.				
I know my medical needs and can explain them to someone.				
I know what a hematologist is and why I go to one.				
I know what to do in case of a medical emergency.				
I understand what causes a pain episode.				
I understand how drugs, alcohol and tobacco affect sickle cell disease.				
I have friends that I can talk to about sickle cell disease.				
I know about necessary screening exams (echo annually, kidney function annually, retinal exams, etc.).				
I know how to get blood work and x-rays.				
<b>Medication Management</b>				
I know what my medications are for.				
I know the names and doses of my medications.				
I remember to take my medications without a parent/guardian reminding me.				
I fill prescriptions before I run out of medications.				
I am aware of what hydroxyurea is and how it prevents sickling of my red blood cells.				
I know how to prevent a pain episode and what to do if I have pain.				
<b>Appointments</b>				
I make my own doctors' appointments.				
I know how to get medical care when the doctor's office is closed.				
I fill out my own medical history form				
I keep track of my own medical information.				
I keep track of my doctors' and other appointments.				
I make a list of questions before my visit with my doctors.				
I answer questions on my own during medical visits.				
I arrange my own transportation to medical appointments.				
<b>Insurance</b>				
I carry my own insurance card.				
I understand my insurance plan.				
<b>Privacy Information</b>				
I understand how health care privacy changes at age 18, when I am legally an adult.				



# PART ONE

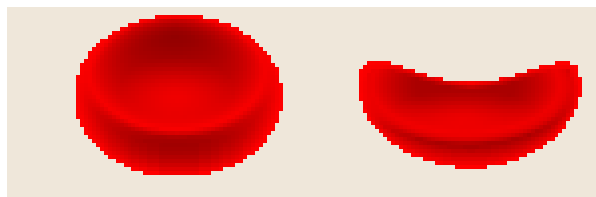
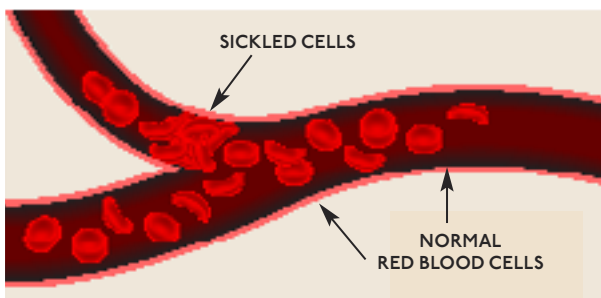
## Understanding Sickle Cell Disease

### What is Sickle Cell Disease?

Sickle Cell Disease (SCD) is an inherited condition of red blood cells passed from one generation to another through your genes. The chart at right shows how the disease can be passed from parents to children.

In people with sickle cell, there has been a change in the gene that makes hemoglobin. Hemoglobin is a protein inside your red blood cells that carries oxygen. In individuals with sickle cell, the hemoglobin gene has a change in the code that changes the shape of the hemoglobin protein inside red blood cells.

Red blood cells usually look like doughnuts. They are bendable and slippery, and easily pass through arteries and veins to bring oxygen and nutrients to the whole body. In people with sickle cell, the changed hemoglobin genes can cause red blood cells to become shaped like sickles or crescent moons. These red blood cells become stiff and sticky and have a hard time traveling through the arteries and veins. Sometimes they clump together and prevent blood from flowing easily.



### SICKLE CELL DISEASE, TYPE SS: AN INHERITED CONDITION

Sickle cell disease and sickle cell trait are inherited.

This chart shows the likelihood of any child being affected by the disease or the trait.

#### KEY

- has sickle cell disease
- has sickle cell trait
- is unaffected (has neither disease nor trait)

**When both parents have sickle cell disease**



**Each child** will have sickle cell disease  
**Zero chance** any child will have sickle cell trait  
**Zero chance** any child will be unaffected

**When one parent has sickle cell disease and one parent has sickle cell trait**



**50% chance** any child will have sickle cell disease  
**50% chance** any child will have sickle cell trait  
**Zero chance** any child will be unaffected

**When one parent has sickle cell disease and one parent is unaffected**



**Zero chance** any child will have sickle cell disease  
**Each child** will have sickle cell trait  
**Zero chance** any child will be unaffected

**When both parents have sickle cell trait**



**25% chance** any child will have sickle cell disease  
**50% chance** any child will have sickle cell trait  
**25% chance** any child will be unaffected

**When one parent has sickle cell trait and one parent is unaffected**



**Zero chance** any child will have sickle cell disease  
**50% chance** any child will have sickle cell trait  
**50% chance** any child will be unaffected

SOURCE: SUNY UPSTATE MEDICAL UNIVERSITY

## Types of Sickle Cell Disease

### Hemoglobin SS (HbSS)

People with HbSS have two copies of the HbS gene, one inherited from each parent. HbSS is the most common and most severe form of sickle cell disease. There are several symptoms that may occur, including chronic (long term) anemia. Anemia develops when your blood lacks enough healthy red blood cells or hemoglobin.

### Hemoglobin SC (HbSC)

People with HbSC have one HbS gene and one abnormal HbC gene. These people may have a mild to moderate anemia. All complications of sickle cell disease may occur but tend to be milder.

### Hemoglobin S-Beta Thalassemia

In this condition, the person has one HbS gene and one Bthalassemia gene. Anemia and other sickle cell symptoms can occur.

There are two types:

**Hemoglobin S Beta Zero (HbSB0) thalassemia** – This is very similar to HbSS.

**Hemoglobin S Beta Plus (HbB+) thalassemia** – The severity of symptoms varies but usually are mild.

### Other forms of sickle cell disease (compound heterozygotes), including HbSE, HbSO and HbSD

These types occur when a person has the HbS gene and another abnormal hemoglobin gene. Symptoms can be different for different people.

## Why do you see a hematologist?

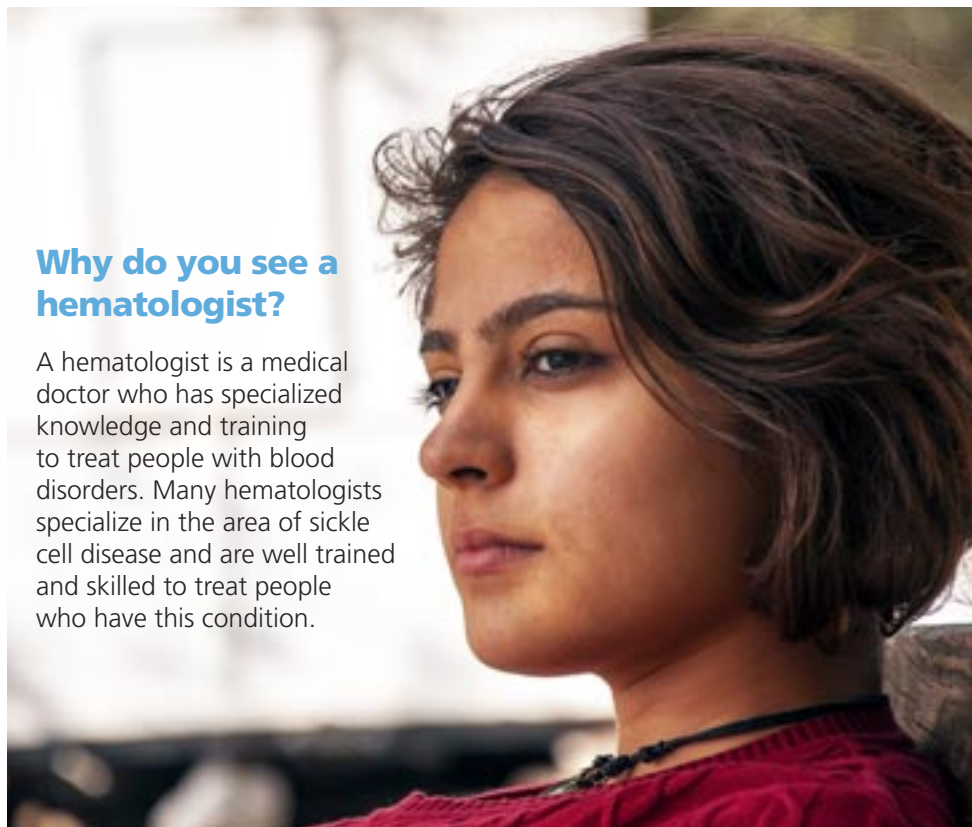
A hematologist is a medical doctor who has specialized knowledge and training to treat people with blood disorders. Many hematologists specialize in the area of sickle cell disease and are well trained and skilled to treat people who have this condition.

## What can you do if you have a medical emergency?

It is important to have a pain and fever plan with your doctor.

This plan will help you know exactly what to do when you feel that a pain episode, fever, or other sign of infection is happening. When possible, you will call your doctor's office to talk about your concerns. If calling the doctor's office is not possible, you may need to go to the emergency room. If you are going to the emergency room, it is important to let the on call physician know so she/he can call ahead and help the emergency room take good care of you.

If there is a true medical emergency, it is important to immediately go directly to an emergency room or call 911. Take a copy of your fever and pain management plan with you to the emergency room. Keep a picture of your updated pain and fever plan on your phone.





## Sickle Cell Disease Complications

### Anemia

This is a drop in the hemoglobin levels in the blood caused by sickling and destruction of red blood cells.

### Pain/vaso-occlusive episode

This is caused from a block in blood flow by sickled red blood cells. It can lead to swelling or redness of the affected area and damage to nearby tissues.

### Acute Chest Syndrome

This is when sickling happens in the lungs and causes pneumonia. ACS is when you have a fever, respiratory symptoms/chest pain and an abnormality on chest xray. You may need oxygen to help you breathe and IV fluids and IV antibiotics.

### Splenic Sequestration

This is caused from a blockage in blood flow by sickled red blood cells in the spleen. It can lead to abdominal pain and swelling as well as anemia and low platelets. You may need a blood transfusion or surgery to take out the spleen if this happens.

### Sepsis

This is caused from a serious bacterial infection in the blood stream. It can be life-threatening and requires hospitalization and IV antibiotics. The first sign of sepsis can be fever. That is why it is very important to come to clinic or the emergency room immediately for a blood culture and IV antibiotics as soon as you have a fever.

### Stroke

This is a blockage of blood flow in the brain caused by sickled red blood cells. It can cause neurologic symptoms and be life threatening. It requires a special kind of blood transfusion called an exchange transfusion to help lower the amount of sickled hemoglobin in the body quickly. You may also require blood thinners.

### Retinopathy

This is abnormal blood vessel growth on the retina of the eye due to sickling in the blood vessels of the eye. It can lead to retinal detachment and blindness if not diagnosed and treated early. Annual eye doctor appointments are important to watch for retinopathy after the age of 10yo.

### Avascular Necrosis

This is damage to the joints from sickling which causes joint pain and limited mobility. You may need to see an Orthopedic doctor to help treat this condition.

## Prevention and Treatments for Sickle Cell Disease

### Hydroxyurea

This is medication taken once a day by mouth starting at 9 months old. It helps the body to produce fetal hemoglobin (Hgb F), a type of hemoglobin normally found in unborn and very young children. This will help decrease risk for all of the above sickle cell complications.

### Penicillin

This is an antibiotic medication taken twice a day by mouth for the first 5 years of life. Only some types of sickle cell disease require this antibiotic. Penicillin decreases the risk for infection and sepsis.

### Routine Vaccinations

It is critically important to stay up-to-date with all routine childhood shots including all booster doses as recommended by your doctor. Vaccines can help prevent sepsis and death from infection. Part of decreasing risk for infection also includes washing hands frequently and seeing the dentist every 6 months.

### Good hydration

Staying well hydrated with water intake will decrease the sickling of red blood cells and decrease the risk for pain episodes and other sickle cell complications.

### Ibuprofen (also called Motrin or Advil) and other narcotic pain medications

These help decrease inflammation in the body during a sickling event and decrease the feeling of pain.

### Transcranial Doppler Ultrasound

This is a special ultrasound of the head that measures the speed of blood flow in the brain. High blood flow speeds mean that the risk for stroke is high. Some sickle cell patients require this ultrasound once a year to watch for stroke risk.

### Transfusion

Blood transfusion may be given in serious situations to treat sickle cell complications in the hospital. It can also be given on a planned monthly schedule if the patient has a severe form of sickle cell disease to help decrease the risk for all of the above complications.

### Iron Chelators

This is a medication taken once daily to help the body get rid of extra iron. Iron chelators are usually only used in sickle cell patients on a chronic transfusion regimen who have too much iron in their body, called iron overload.

## PART ONE – UNDERSTANDING SICKLE CELL DISEASE

# Quiz

1. What kind of cells are affected by sickle cell disease?

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2. What type of sickle cell disease do you have?

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3. Name two medical complications that you may experience due to sickle cell disease.

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4. Name two treatments for sickle cell disease.

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# PART TWO

## Understanding Pain and Sickle Cell Disease

### Acute Pain

This is the typical sickle cell pain, also called a vaso-occlusive crisis or episode. This is treated with rest, heat, hydration, ibuprofen and sometimes narcotic pain medication.

### Chronic Pain of Unknown Etiology (unknown cause)

This type of pain is chronic, or long-term. It may be due to having several pain episodes over time. **It is important to treat your sickle cell pain as soon as it becomes noticeable to try to prevent pain from becoming chronic.** You may have chronic pain in a specific area of your body, such as a specific organ or tissue. When people have chronic pain from sickle cell disease, they might describe it as constant, deep, or achy. It can happen in several parts of your body, including your chest, back, arms and legs, neck, head, or abdomen.

### Neuropathic Pain

Another type of pain you may experience is called neuropathic pain. People with sickle cell disease describe this as burning, numbness, tingling, shooting, or a sensation of pins and needles. Some of the possible causes of this kind of pain may be due to tissue damage from blockage of blood vessels that go to nerves. It can also be from chronic pain, which may be from chronic inflammation (irritation and/or swelling).



**More on pain in  
Part Five, page 15**

## Harmful substances that increase pain and damage your health

### How can alcohol, drugs and tobacco affect you?

#### Alcohol

Alcohol is a depressant, which means that it slows your brain. It can also cause you to urinate more and you may lose too much fluid in your body. This can lead to dehydration and a pain episode. In addition to avoiding alcohol, it is important for people with sickle cell to stay hydrated (drink plenty of water) to avoid blood cells from sticking together and blocking blood vessels.



#### Tobacco

Tobacco – cigarettes, chew, vaping, and cigars – decreases the amount of oxygen you receive in your lungs. Decreased oxygen can cause infections and lead to acute chest syndrome and damage to your lungs. People with sickle cell disease are especially prone to problems associated with tobacco. Complications from using tobacco products include chronic obstructive pulmonary disease (a disorder that causes increased shortness of breath and gets worse as time goes on), emphysema (damage to the air sacs of the lungs), pulmonary hypertension (increased pressure in the blood vessels that carry blood from the heart to the lungs), stroke (blockage of blood flow to the brain), ulcers (open sores caused by breaks in the skin that do not heal), and more frequent pain episodes.

#### Street Drugs

All street drugs can be dangerous and cause health problems. Many cause more severe side effects and worsen symptoms of sickle cell. For example, smoking marijuana can increase heart rate and cause coughing and bronchitis. Stimulant drugs such as cocaine or crack cocaine can increase heart rate, breathing rate, and blood pressure. It can also cause stroke and increases the risk for organ damage.

#### Prescription Drugs

Using prescription drugs in any way other than what your doctor tells you or any prescription drugs that your doctor did not prescribe for you can lead to addiction, harm to your body, and even death.



## PART TWO – UNDERSTANDING PAIN AND SICKLE CELL DISEASE

## Quiz

1. Name three types of pain that people with sickle cell disease can have.

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2. What is one problem that can be caused by using alcohol?

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3. What is one problem that can be caused by using marijuana?

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4. What is one problem that can be caused by using cocaine or crack cocaine?

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# PART THREE

## Finding Support and Getting Medical Tests

### Types of Support

#### Social Support

This can include several people. You might consider family, friends, faith groups, community groups, school, or others to be social support. This means you can talk with them openly about challenges and they are willing to listen and help in any way they can.

#### Patient education

Nurses, doctors, and other providers at your medical facility will be able to answer questions and teach you many things you need to know to help you manage your sickle cell disease.

#### Behavioral Health

There are several providers who can be supportive with behavioral health concerns. This might include coping with thoughts, feelings, behaviors, and pain, or adjusting to a new diagnosis or hospitalization.

#### Social Work

Social workers provide emotional support and assistance with financial questions and challenges, such as working with health insurance companies, getting transportation, or other expenses.

#### Caregiver Support

Caregivers such as parents, family members, and other loved ones also experience challenges when a child, adolescent, or young adult they love has sickle cell disease. Because of this, there are support groups and behavioral health providers that can support caregivers as well.



### How can I get blood work and imaging?

In order to get blood work, imaging, or other medical tests done, your doctor will request them. It is important for you to choose a primary medical facility and a backup in case your first choice has no openings or does not provide the specific test. You may be able to get these tests done where you see your hematologist, such as at Upstate University Hospital.

### What screenings do I need?

**When you have sickle cell disease, the disease can affect several parts of your body.**

It is important to know about all of the screenings or tests that you need to have to keep yourself well. Talk with your doctor about the types of tests you need and where to have them done. Write this down so you have the information when you have questions or need to schedule a screening test. **These are some of the screening tests that people with sickle cell disease often get:**

- Electrocardiogram (measures the electrical activity of your heart)
- Echocardiogram (provides a video of your heart function)
- Pulmonary function testing (determines how well your lungs are functioning)
- Renal function testing (urine or blood tests that measure kidney function)
- Serial eye/retinal examination (examines the blood vessels behind the eye)
- Brain imaging (assesses the structures and blood flow in your brain)
- Transcranial Doppler (another test that measures blood flow in your brain)

**PART THREE – FINDING SUPPORT AND GETTING MEDICAL TESTS**

# Quiz

1. Name two types of support you may use for your sickle cell disease.

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2. Name two types of yearly screening tests you should have if you have sickle cell disease.

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3. Where would you go for your bloodwork or imaging test?

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# PART FOUR

## Understanding and Managing Medications

### What medications do you take to manage your sickle cell disease?

There are several types of medication you may need to use if you have sickle cell disease. For example, you may take Hydroxyurea, non-steroidal anti-inflammatory drugs (ibuprofen, Motrin or Advil) or other pain medicines, asthma medicines, or antibiotics.



### What are the names and doses of all of your medications?

It is important for you to know the types of medication you take, the reason you take them, the doses that you take, and how often you take your medicine. This will help you achieve your health care goals and prevent future problems like pain crises. It will also protect you from using your medicine incorrectly, which could cause harm.



### Overcoming obstacles to taking your medication

It can be challenging to take all of your medications the way you are supposed to take them. It may be hard to take your medicine at the same time each day. You may worry about side effects. You may have several medications you need to take. You may wonder if all of your medication is necessary, or you may feel like the medication isn't helping you get better.

Because these challenges may affect your ability to take your medicine the way you need to, it is helpful to consider using some of these tips:

- **Talk with your doctor to learn more about your medication.** For example, ask why you take it, how it works, share your concerns about side effects, and learn why it is important to take it the way your doctor tells you to take it.
- **Ask your support system to help you take your medicine the way you should.** For example, ask a friend, significant other, or family member to help you take all of your medications as prescribed.
- **Talk with a behavioral health provider about behaviors and ideas that interfere with you taking your medicine.** These may include understanding, attitudes, beliefs, a desire to have what is normal for others, motivation, intention to change, rewards and benefits, financial concerns, and challenges with other resources.

**PART FOUR – UNDERSTANDING AND MANAGING MEDICATIONS**

# Quiz

1. Name all of the medicines you take, the reason you take them, the dose that you take, and how often you take them.

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2. Name two ways that you can get help taking your medicine the way it is prescribed by your doctor.

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## PART FIVE

### More on Medication and Pain Management

#### Filling prescriptions before they run out

To prevent running out of your medication, it is important to have a pharmacy that is easy for you to get to and accepts your insurance. Search online for a pharmacy near your home by typing “pharmacy” and your home’s zip code. You should also call our sickle cell clinic if you are having trouble with your medications.”

#### What is hydroxyurea and how does it work?

Hydroxyurea is a medicine that can help adults and children with sickle cell disease. Studies have shown

that hydroxyurea can decrease the number of pain events, pneumonia, acute chest syndrome, and admissions to the hospital. It must be prescribed by a doctor, and is often covered by health insurance, which makes it affordable to many people. It can come in a capsule or a liquid.

Hydroxyurea works by allowing red blood cells to have more fetal hemoglobin. This allows the cells to become larger and less sticky, and to move more easily throughout the blood vessels in the body. Hydroxyurea can also reduce inflammation and can be used by a wide age range of people with sickle cell disease.



#### How can you prevent pain episodes?

- Drink plenty of water
- Try not to get too hot or too cold
- Avoid places and situations with high altitudes (flying, mountain climbing, or cities with high altitudes).
- Avoid activities and places with low oxygen levels, such as mountain climbing or intense exercise.
- When you need to use the bathroom – do so. Don’t try to “hold it.”
- Avoid constipation. If having hard stools, tell your doctor and you may be prescribed laxatives for relief of constipation.”
- Take your medicine as prescribed by your doctor.
- Stay up-to-date on your routine health care needs (Appointments with your primary doctor, routine visits with your hematologist, immunizations, and care for other medical conditions that you may have)

#### What do I do if I have pain?

When you feel pain coming on, it is important to remember the pain plan that you have created with your doctor.

- If you have mild pain, your doctor may recommend you take a specific dose of a non-steroidal anti-inflammatory medication and use a warm pack on the location where you have pain. Consider using heat before ice for sickle cell pain.
- If you experience severe pain, your doctor may tell you to contact his or her office if it is open (often Mon.-Fri., daytime), or go to the nearest emergency room if the doctor’s office is closed so you can be evaluated and treated.
- Be sure that you have spoken with your doctor to create a pain plan and follow through with the recommendations.



## PART FIVE – MORE ON MEDICATION AND PAIN MANAGEMENT

## Quiz

1. How can you get a prescription refilled?

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2. Why do some people take Hydroxyurea?

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3. Name four ways that you can work to prevent a sickle cell pain episode.

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4. What is the pain plan that you have created with your doctor?

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## PART SIX

# Scheduling and Attending Appointments

It is important to know all your doctors and other health care providers so you can make medical appointments. Write down the name, title (ex.: primary care, hematologist, social worker, eye doctor), phone number, and address of each of your medical providers so you are prepared to make your own medical appointments.

It can be challenging to make your own medical appointments if you have not done this before. It is OK to be nervous. It is useful to write down notes to refer to while making a phone call. For example, before you call, you could write:

“This is Jay. I am calling to make an appointment to see Dr. Scott for a follow-up visit.”

The person on the phone will likely ask for your date of birth, then tell you some days and times that you can see Dr. Scott. Be sure to have a calendar (such as phone calendar) nearby so you can know when you are available to see the doctor, and can record the date, time and location of the appointment on your calendar.

If you cannot make it to an appointment for any reason, it is very important to call the doctor’s office to tell them. You can reschedule your appointment when you call to tell them you can’t make this visit. By cancelling an appointment ahead of time, you help the office so they may fill the time slot with another patient who needs to receive medical care. You may also avoid “no show” fees which many offices charge unless they hear about a cancellation at least 1-2 days before the appointment.



## PART SIX – SCHEDULING AND ATTENDING APPOINTMENTS

## Quiz

1. What is the first thing you need to know to make an appointment to see a medical provider?

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2. How can you make the phone call easier when you are calling to make a medical appointment?

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3. What can you do to save the information you talk about when you are making an appointment over the phone?

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4. What should you do if you are late or have to cancel an appointment?

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# PART SEVEN

## After Hours Care & the Value of Medical History



### Getting medical care when the doctor's office is closed

Sometimes you need to speak with a doctor or see a doctor outside of normal business hours, which are usually Monday-Friday 8am-4pm. You can look at your pain and fever card and/or call the on-call number to speak to the on-call physician. She or he might give you instructions on how to feel better at home or direct you to the emergency room for more concerning problems. If you are coming to the emergency room, the on-call sickle cell doctor will call the emergency room to tell them that you are coming and give them advice on how best to care for you.

### Filling out my own medical history form

When you go to the doctor's office, you may need to fill out a medical history form. There are a few ways that you can provide them with your medical

history information. First, you may have your **sickle cell disease card** with you, which provides the doctor with your specific diagnosis, medications, treatment plan, and pain and fever plan. This will help them understand your sickle cell disease.

In addition to your sickle cell disease and management, there is other information that doctors may ask you to provide. This might include your immunization history, childhood illnesses, and family medical history. Don't panic! It is likely too much information for you to have memorized. Good news. There are places that you can enter your medical history and retrieve it when you need it.

**MyChart** is an excellent resource. If you activate MyChart through Upstate University Hospital, you are able to review your medical history, test results, medications, and much more. You may also choose to upload your information into another form of electronic medical record or have a copy of your medical history saved and available to you.

## PART SEVEN – AFTER HOURS CARE AND THE VALUE OF MEDICAL HISTORY

# Quiz

1. What should you do if you need to see a doctor and the doctor's office is closed?

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2. Name two things you can use to help provide your medical information when you need to complete a medical history form.

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# PART EIGHT

## Preparing for Medical Appointments



### Keeping track of your medical information

An excellent way to keep track of your medical information is by using MyChart or having a copy of your medical records available to you.

### Keeping track of your appointments

When you have a chronic medical condition like sickle cell disease, you may be required to have several doctors' appointments and medical tests. It is important to organize all of your appointments and find a way to remind yourself about them.

One of the easiest and most effective ways of remembering your appointments is to **use the calendar app in your phone**. By doing this, you can enter an appointment with details such as date, time, place and who your appointment is with. Some phone calendar apps have a reminder option. Appointment information will pop up to remind you hours or days before your appointment so that you can remember and plan for it. **Or, you can use a planner notebook** that you carry with you whenever you leave home.

### List of questions for doctor's visits

When you visit your doctor, **ask** her or him any **questions** you have and describe any concerns you have about your health. This will help you get all the medical care that you need.

**Write your questions and concerns** on paper or in your phone's notepad. Bring the questions to the appointment so you remember to talk with the doctor about them.

### Answering questions on your own during medical visits

As you are becoming an adult and preparing to transition into adult health care, it is important to learn how to communicate with health care providers and staff when scheduling and attending your appointments.

You may be accustomed to a parent or other loved one answering questions or providing information for you during your medical visits. But, this is the time for you to start working toward doing this on your own.



For example, when a nurse checks you in and asks how you have been since your last visit, tell him or her if you have had any pain, any concerns with mood or worries, or any other physical symptoms. If you haven't had any problems, share that as well. You know yourself the best, so you are best able to answer the questions.

If during your transition you find it difficult to answer some questions, that is OK. If you have done your best to provide information and you need help to provide all the information to your doctors or nurses, that is OK. This is a process. Practice will help you to become independent.

### Planning transportation to and from medical appointments

In planning to attend your medical appointments, transportation is very important. You may drive yourself, have a loved one drive you, use public transportation, or use a cab service.

**Plan ahead.** If you need a loved one to drive you, be sure to make plans with them in advance. If you need to take a bus or a cab, check the schedule or call ahead to have a cab pick you up. If finances for traveling are a concern, you may request to speak with a social worker at the doctor's office to ask if they can provide financial assistance for transportation.

## PART EIGHT – PREPARING FOR APPOINTMENTS

## Quiz

1. Name two ways you can remind yourself about your scheduled appointments.

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2. Why is it important to write down your questions for your doctor before your appointment?

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3. Why is it important for you to be able to answer your doctor's questions on your own?

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4. Name two options you have for receiving transportation to your medical appointment.

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# PART NINE

## Understanding Health Insurance

### What is health insurance?

Health insurance covers much of the cost of health care (seeing your doctor, spending time in the hospital, buying medication, and receiving medical tests). When you have health insurance, you pay a set amount of money each month. With health insurance, you receive your health care at a significantly lower cost, or at no cost.

### These are ways to get health insurance.

- You may qualify through your **workplace**. Ask the human resources department about this. They should provide you with written information about health insurance you are eligible for.
- You may look into other health insurance plans at [www.HealthCare.gov](http://www.HealthCare.gov).
- You may qualify for a government assistance plan such as **Medicaid**, which provides health insurance to people depending on several factors, such as income. You can look into options at [www.HealthCare.gov](http://www.HealthCare.gov).

### Why should I have individual health insurance?

1. **Always be prepared for health care needs.** You never know when you will need to see a doctor or have tests or procedures.
2. **Preventing health problems.** With many health insurance plans, you can have screenings and routine medical care that can keep you healthy and prevent you from becoming sick.
3. **It's the law.** Under the Affordable Care Act, there may be a penalty if you don't have health care coverage.

### Carrying your insurance card

Your medical insurance card is the key to seeing your medical providers. It provides them with information about who provides your insurance and your personal information so they may communicate with your insurance company to ensure that you do not receive bills that your health insurance should cover. Be sure to carry this with you in your wallet or electronically. You never know when you may need it.

### Understanding your health insurance plan

You may have a lot of questions about your health insurance plan. There is a lot of information that goes into these plans. If you have questions, there are a few simple solutions to having your questions answered.

- First, you may call the phone number that is listed on your insurance card to ask a representative from the company about the questions you have.
- Second, you may have a packet or a document you received from your insurance company that provides you with a great deal of information that may help to clarify your questions.

### Expiration of Health Insurance Plan

It is very important that you know when your health insurance plan will expire so you can enroll in a new plan. There are several reasons why your health insurance may expire or be discontinued. Some of the ways you can know that your health insurance needs to be renewed include:

- You received a letter in the mail that tells you that you need to renew your insurance plan or that it has been discontinued
- You are turning 26 years old
- You got married
- You got divorced
- You are moving to a new home, especially out of state
- You are leaving your current job that offers your health insurance
- You are being told your current insurance plan is being discontinued

If your health insurance expires or discontinues, there are ways to become insured again. It is important to get insurance as soon as possible to avoid large medical bills or other consequences. To become insured again, you can:

- Contact the New York State of Health Official Health Plan Marketplace: 855-355-5777, <https://nystateofhealth.ny.gov/>
- Call the phone number on the back of your insurance card to discuss options
- Call the human resources department at your job that provided your insurance plan



## PART NINE – UNDERSTANDING INSURANCE

## Quiz

1. What is health insurance? Why should you have it?

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2. What are two benefits of having health insurance?

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3. Why is it important to always have access to your insurance card?

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4. What are two ways that you can have your questions about your health insurance answered?

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5. What are two ways you can know that your health insurance might expire?

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6. If your health insurance expires or is discontinued, what are two ways you can reapply for a health insurance plan?

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# PART TEN

## Understanding Privacy Rights

**Health care privacy changes when you become a legal adult at age 18.**

Until your 18th birthday, your parents or guardians have access to your health care information and have the ability to make medical decisions for you. When you turn 18, you will likely be asked to fill out and sign a Health Insurance Portability and Accountability Act (HIPAA) form. The HIPAA Privacy Rule provides national standards to protect individuals' medical records and other personal health information and applies to health plans, health care clearinghouses, and health care providers who conduct certain health care transactions electronically.

On the HIPAA form, you will state who, if anybody, can have access to your medical information (documents, chart, or the ability to speak with your providers). This is your choice when you turn 18. You may choose to have anyone you would like to be able to access your information or participate in your health care.

This is a big step. When you are 18, especially if you choose not to have another person have access or participate in your health care, you are now responsible for communicating with all of your providers, making your appointments, managing your health insurance, etc.

This may sound scary, but you probably know much more than you think you do! If you need help or have questions about the process, ask your health care providers. If you feel that you need additional support with this process, there are professionals such as care managers who can help guide you to meet your healthcare needs.

### Health Care Proxy

The New York Health Care Proxy Law allows you to appoint someone you trust — for example, a family member or close friend – to make health care decisions for you if you lose the ability to make decisions yourself. By appointing a health care agent, you can make sure that health care providers follow your wishes. Your agent can also decide how your wishes apply as your medical condition changes. Hospitals, doctors, and other health care providers must follow your agent's decisions as if they were your own. You may give the person you select as your health care agent as little or as much authority as you want. You may allow your agent to make all health care decisions or only certain ones. You may also give your agent instructions that he or she has to follow. This form can also be used to document your wishes or instructions with regard to organ and/or tissue donation.

You can find more information and see the Health Care Proxy form at: <https://www.health.ny.gov/publications/1430.pdf>



## PART TEN – UNDERSTANDING PRIVACY RIGHTS

## Quiz

1. At what age will you have a major change in your privacy information?

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2. What is the name of the law that protects your health information and maintains privacy with your healthcare information?

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3. Are you able to allow anybody else to access your healthcare information after your transition to adulthood?

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4. Will you have access to support if you have questions about your healthcare privacy and communication about your healthcare? If so, who may be able to help?

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5. What is a health care proxy/agent? Why is it important?

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# Sickle Cell Disease Transition Readiness Assessment

Please fill out this form to show what you know about your health care and to identify areas that you want to learn more about. If you need help completing this form, please ask your parent/caregiver or health care provider.

Date	Name	Date of Birth
<b>On a scale of 0 to 10, please circle the number that best describes how you feel now.</b> (0=not, 10=very)		
How important is it to you to manage your own health care? 0 (not)    1    2    3    4    5    6    7    8    9    10 (very)		
How confident do you feel about your ability to manage your own health care? 0 (not)    1    2    3    4    5    6    7    8    9    10 (very)		
How confident do you feel about preparing for/changing to an adult doctor before age 22? 0 (not)    1    2    3    4    5    6    7    8    9    10 (very)		
Does not apply:		

 <b>Please check the box that applies to you now.</b>	No, I do not know	No, but I am learning to do this	Yes, I have started doing this	Yes, I always do this when I need to
<b>Disease Knowledge</b>				
I know what type of sickle cell disease I have.				
I know my medical needs and can explain them to someone.				
I know what a hematologist is and why I go to one.				
I know what to do in case of a medical emergency.				
I understand what causes a pain episode.				
I understand how drugs, alcohol and tobacco affect sickle cell disease.				
I have friends that I can talk to about sickle cell disease.				
I know about necessary screening exams (echo annually, kidney function annually, retinal exams, etc.).				
I know how to get blood work and x-rays.				
<b>Medication Management</b>				
I know what my medications are for.				
I know the names and doses of my medications.				
I remember to take my medications without a parent/guardian reminding me.				
I fill prescriptions before I run out of medications.				
I am aware of what hydroxyurea is and how it prevents sickling of my red blood cells.				
I know how to prevent a pain episode and what to do if I have pain.				
<b>Appointments</b>				
I make my own doctors' appointments.				
I know how to get medical care when the doctor's office is closed.				
I fill out my own medical history form				
I keep track of my own medical information.				
I keep track of my doctors' and other appointments.				
I make a list of questions before my visit with my doctors.				
I answer questions on my own during medical visits.				
I arrange my own transportation to medical appointments.				
<b>Insurance</b>				
I carry my own insurance card.				
I understand my insurance plan.				
<b>Privacy Information</b>				
I understand how health care privacy changes at age 18, when I am legally an adult.				







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