Sickle cell disease tools for patients and non-medical audiences

95 paper and digital tools

95 Total

Sickle Cell Tools

Tools Analyzed

95 Total
Sickle
Cell
Tools

42/95
Acute
Pain
Tools

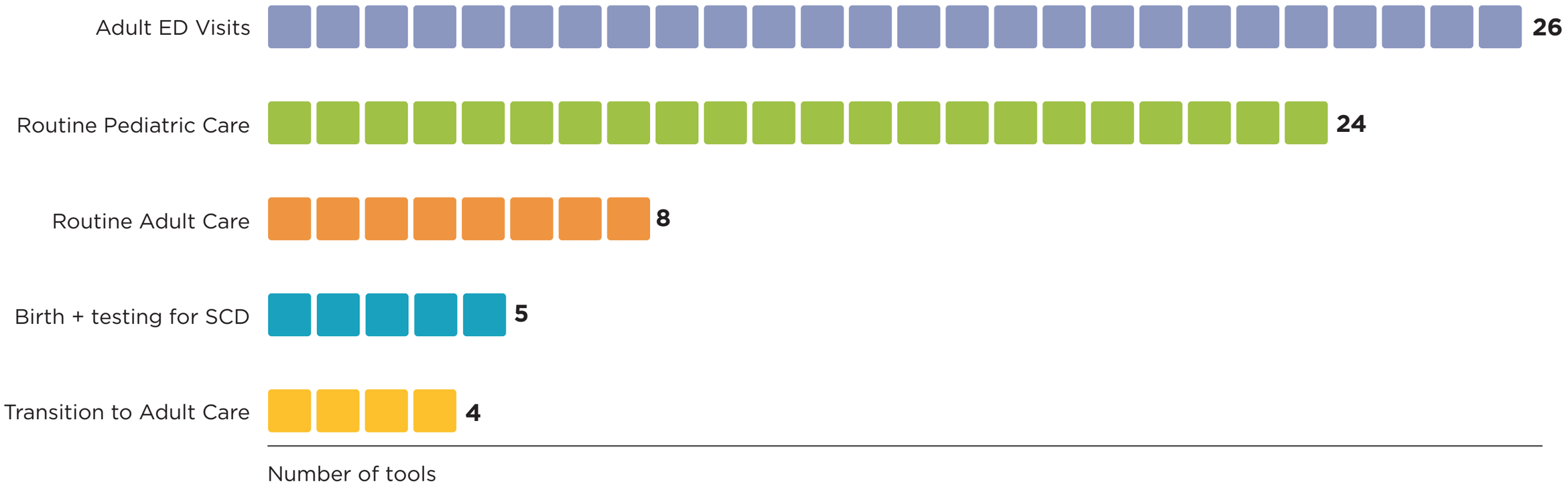
For the 42 tools related to acute pain, we looked at each tool's:

- additional content areas
- intended audience

Other content areas of acute pain tools

42

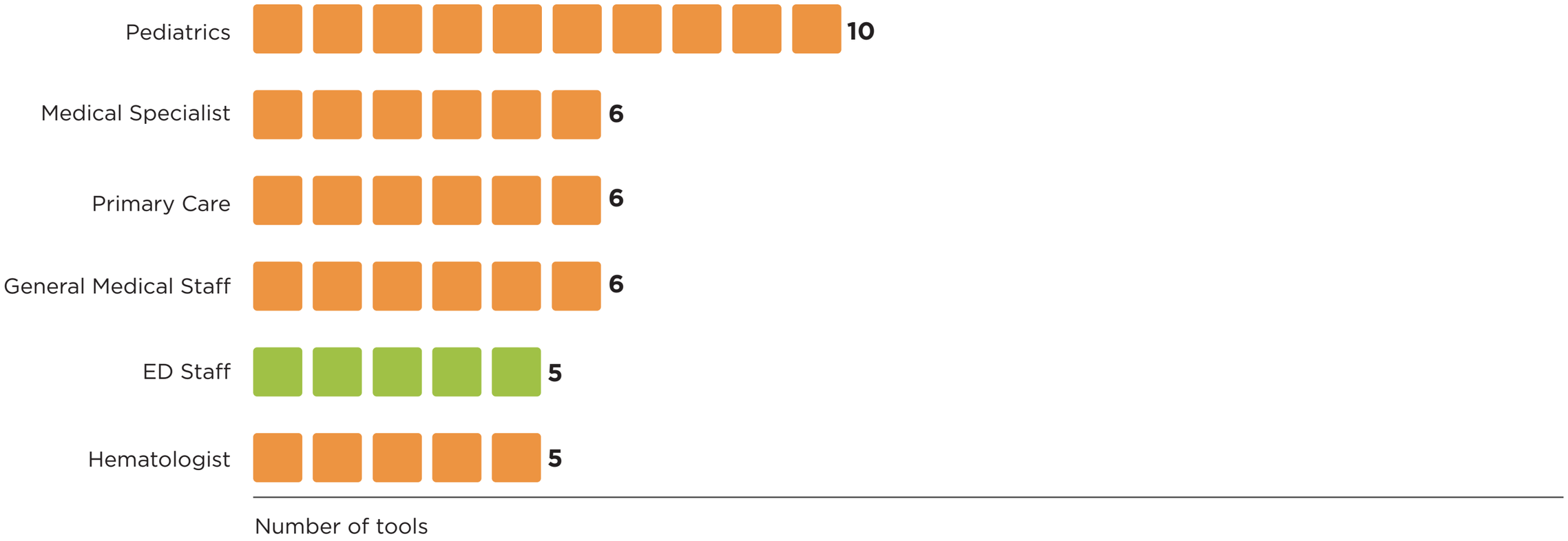
tools that contain content about acute pain also cover the following subject areas:



Intended audience of acute pain tools

16

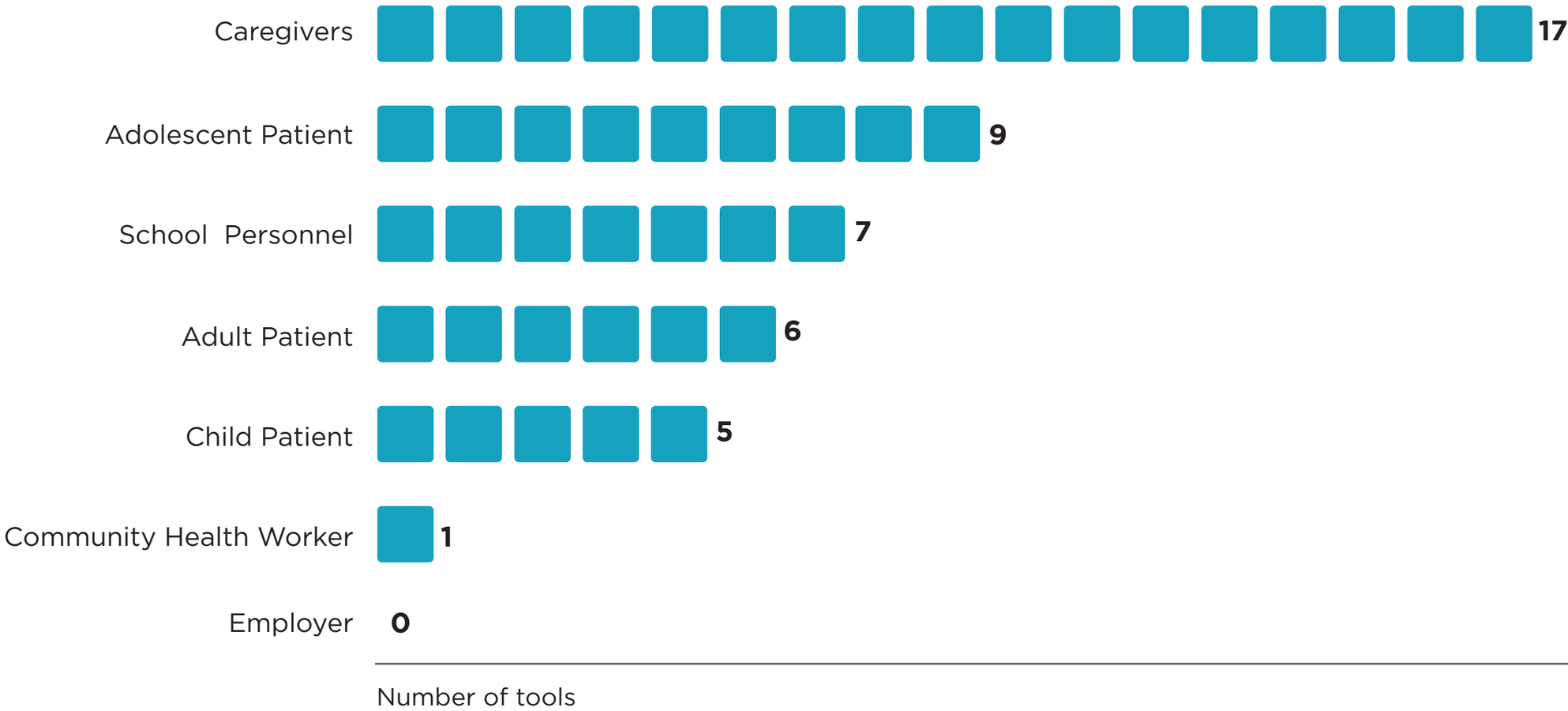
tools collectively serve 6 different medical audiences



Intended audience of acute pain tools

26

tools collectively serve 6 different non-medical audiences



We analyzed each tool through the lenses of:

1. Usability
2. Accessibility
3. Actionability

1. Usability refers to a tool's "human factors" elements: *how well can a person engage with the tool?*

We evaluated known information design principles that include:

- visual hierarchy
- typography
- use of color to highlight and differentiate content

Usability criteria: A good example

Unit 2: Complications of Sickle Cell Disease

Because sickle-shaped red blood cells can slow blood flow to many parts of the body, a number of complications* (health problems) can occur. You need to be aware of these problems and how to treat them. Each child is different. Not every child with sickle cell disease will have all of these health problems. However, knowing about these complications in advance can help prevent them and may even save your child's life.

Blood Infections

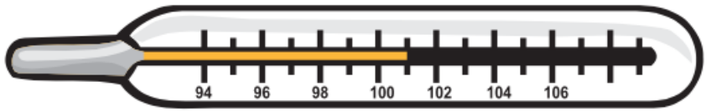
One of the most serious problems facing young children with sickle cell disease is a blood stream infection* (in-FEK-shun). The infection risk is greatest during your child's first 3 years of life. Blood infections can be deadly, and they require treatment right away.

What is the main symptom of a blood stream infection?

Fever is one of the first symptoms and sometimes the **only** symptom of a blood stream infection*.

If your child has a fever of 101 degrees F (38.4 degrees C) or higher, seek medical treatment right away. If you delay treatment for even a few hours, you will put your child in danger.

Do not forget to tell the health care team that your child has sickle cell disease.
Fever **must not** be ignored in a child with sickle cell disease.



Color differentiates information

27 of 42 acute pain tools use color

Typographic hierarchy signals priority

Typesetting creates readability

Serif font, 10-12pt font size
Optimal line length 50-60 characters

4 tools are within the optimal range
Averaging: 72
Ranging between: 24-103

Action items are highlighted

Images support content

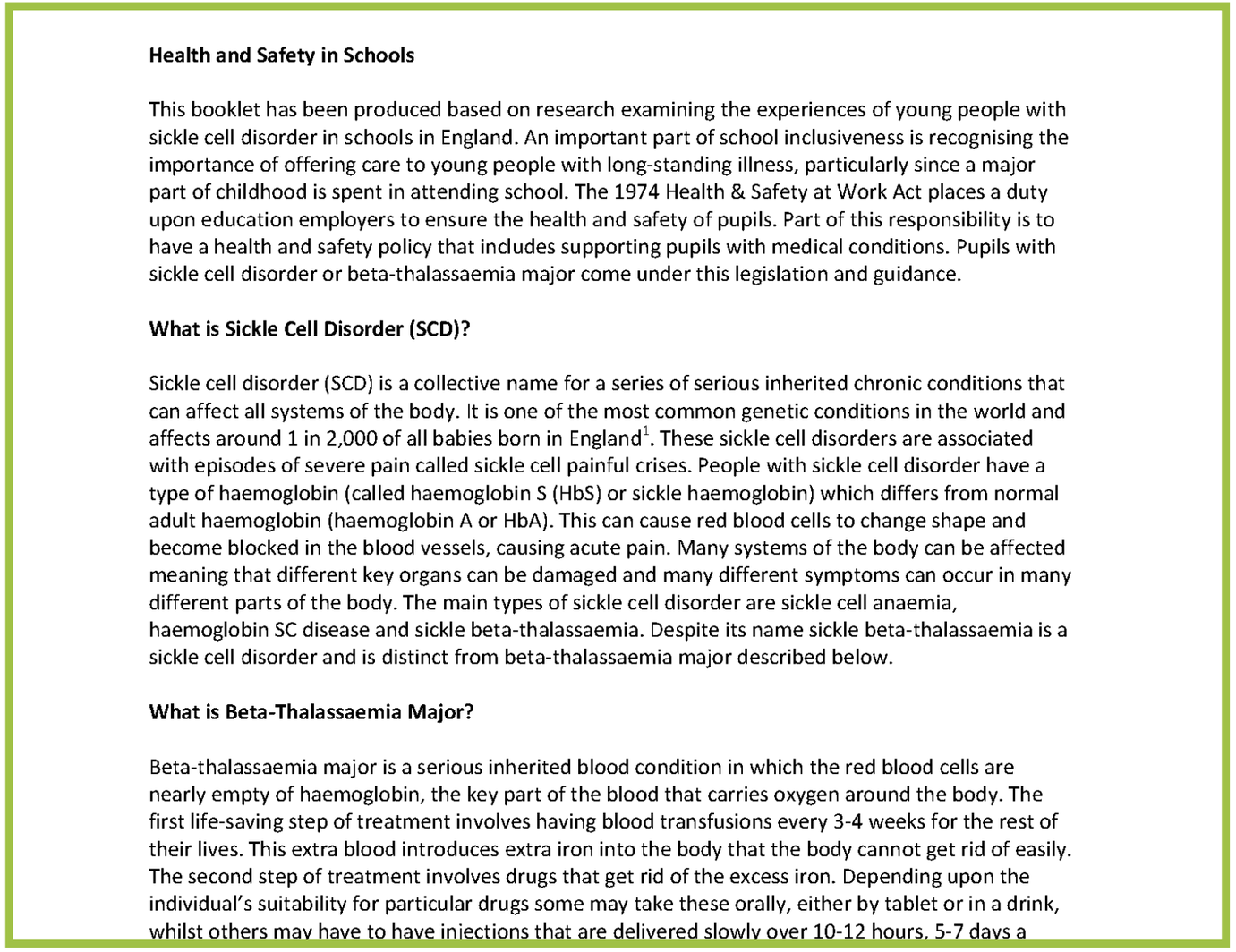
Good example from *Your Child and Sickle Cell Disease*

2. Accessibility refers to how well a tool's content can be understood.

We evaluated:

- reading grade level (Flesch-Kincaid)
- definition of medical terms

Accessibility criteria: Understanding reading levels



Health and Safety in Schools

This booklet has been produced based on research examining the experiences of young people with sickle cell disorder in schools in England. An important part of school inclusiveness is recognising the importance of offering care to young people with long-standing illness, particularly since a major part of childhood is spent in attending school. The 1974 Health & Safety at Work Act places a duty upon education employers to ensure the health and safety of pupils. Part of this responsibility is to have a health and safety policy that includes supporting pupils with medical conditions. Pupils with sickle cell disorder or beta-thalassaemia major come under this legislation and guidance.

What is Sickle Cell Disorder (SCD)?

Sickle cell disorder (SCD) is a collective name for a series of serious inherited chronic conditions that can affect all systems of the body. It is one of the most common genetic conditions in the world and affects around 1 in 2,000 of all babies born in England[1]. These sickle cell disorders are associated with episodes of severe pain called sickle cell painful crises. People with sickle cell disorder have a type of haemoglobin (called haemoglobin S (HbS) or sickle haemoglobin) which differs from normal adult haemoglobin (haemoglobin A or HbA). This can cause red blood cells to change shape and become blocked in the blood vessels, causing acute pain. Many systems of the body can be affected meaning that different key organs can be damaged and many different symptoms can occur in many different parts of the body. The main types of sickle cell disorder are sickle cell anaemia, haemoglobin SC disease and sickle beta-thalassaemia. Despite its name sickle beta-thalassaemia is a sickle cell disorder and is distinct from beta-thalassaemia major described below.

What is Beta-Thalassaemia Major?

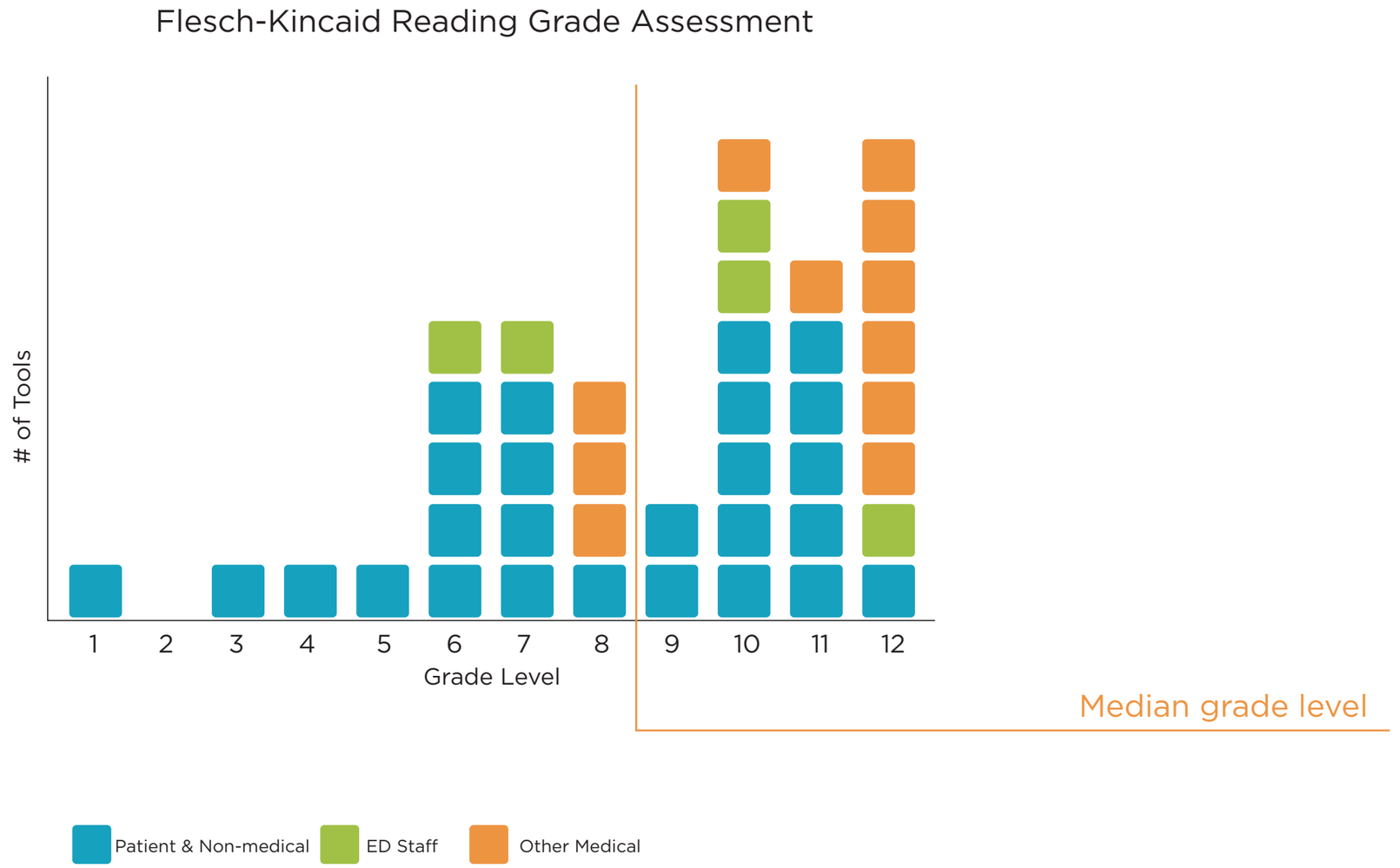
Beta-thalassaemia major is a serious inherited blood condition in which the red blood cells are nearly empty of haemoglobin, the key part of the blood that carries oxygen around the body. The first life-saving step of treatment involves having blood transfusions every 3-4 weeks for the rest of their lives. This extra blood introduces extra iron into the body that the body cannot get rid of easily. The second step of treatment involves drugs that get rid of the excess iron. Depending upon the individual's suitability for particular drugs some may take these orally, either by tablet or in a drink, whilst others may have to have injections that are delivered slowly over 10-12 hours, 5-7 days a

Sickle Cell and Thalassaemia: School Health and Safety

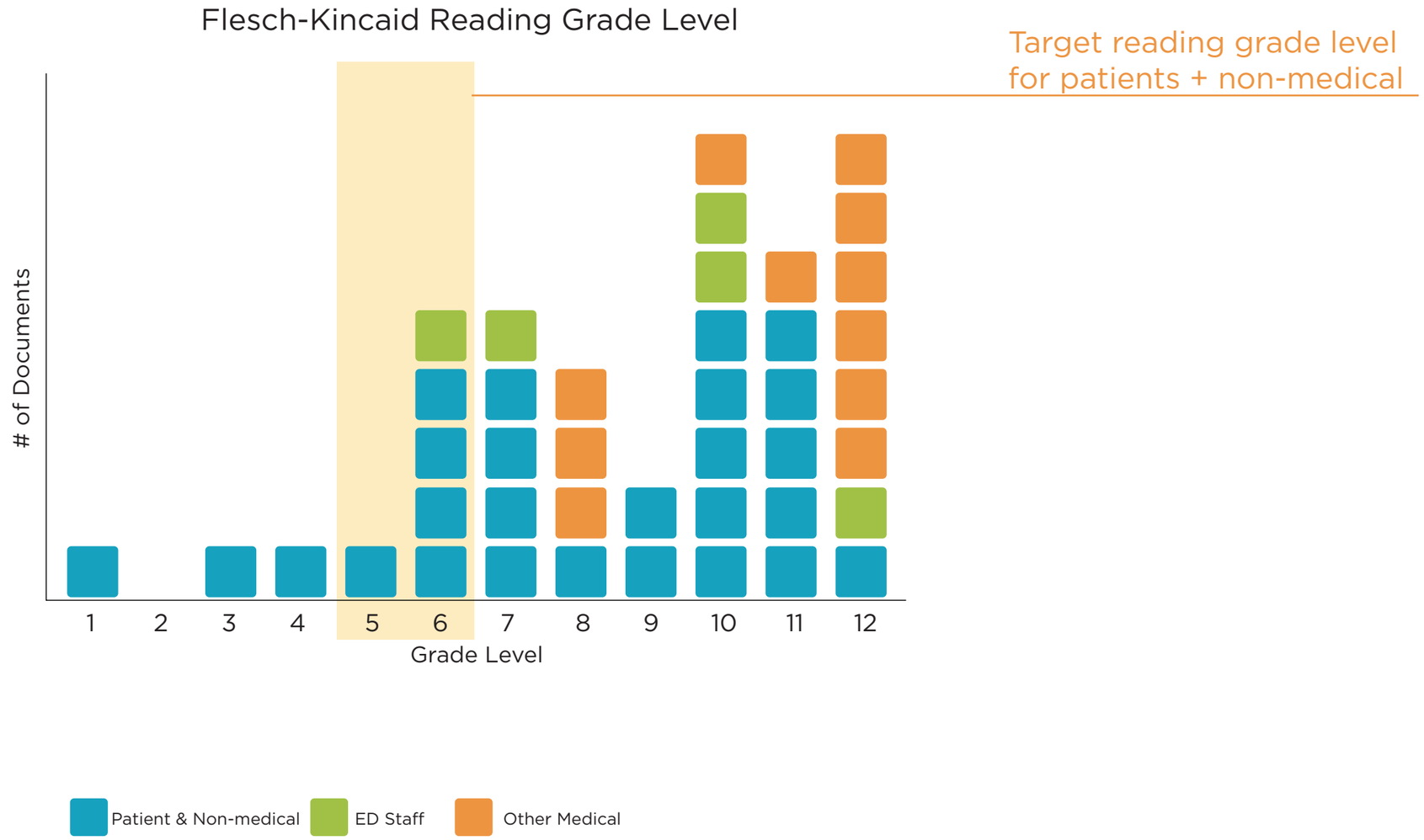
Flesch-Kincaid reading grade assessment

Designed to indicate how difficult a passage written in English is to understand by presenting a U.S. grade level. The core measurement is pulling the word length and sentence length into a formula calculating the Flesch-Kincaid reading grade level.

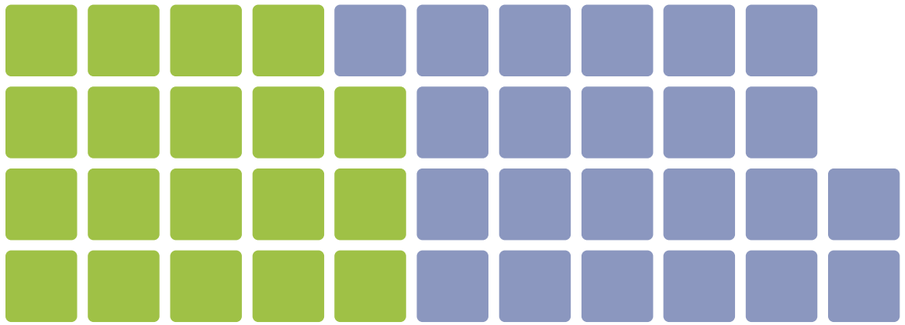
Accessibility criteria: Reading grade level of tools



Accessibility criteria: Reading grade level



Accessibility criteria: definition of medical terms



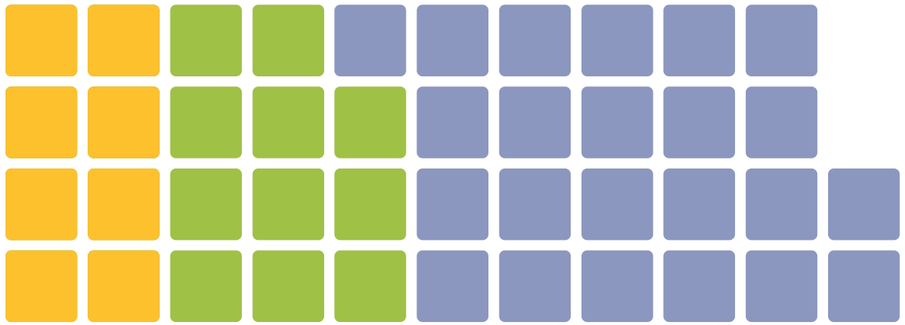
19 of 42 tools for acute pain provide definitions of medical terms

Glossary

- **Anemia** (uh-NEE-mee-uh)**:** Occurs when the blood does not have enough red blood cells. The hemoglobin and hematocrit are laboratory tests used to find out if a person is anemic.
- **Antibodies** (AN-tih-bah-deez)**:** Proteins that fight bacteria and other foreign toxins in the body.
- **Bacteria** (bak-TEER-ee-uh)**:** Germs that are made up of one cell. Certain types of bacteria can cause illness when they get inside the body.
- **Blood stream infection** (in-FEK-shun)**:** When bacteria* get in the blood stream and start spreading throughout the body, making a person ill.
- **Complications** (KOM-plih-kaa-shunz)**:** In people with sickle cell disease, these are health problems caused by the disease.
- **Dactylitis** (DAK-tih-ly-tus)**:** Pain and swelling of hands and feet that is also called Hand-Foot Syndrome.
- **Gene:** A "blueprint" that is passed from parent to child. It carries the instructions for a certain trait, such as hair color, eye color, or skin color.
- **Hemoglobin** (HEE-muh-glow-bin)**:** The main substance of the red blood cell. It carries oxygen from the lungs to all parts of the body. Normal red blood cells contain hemoglobin A. Hemoglobin S and hemoglobin C are abnormal types of hemoglobin.
- **IV:** A needle placed in a vein to deliver fluids and medicines directly into the bloodstream.
- **Spleen:** An organ on the left side of the abdomen. It helps protect against infection by filtering bacteria from the bloodstream. It also produces antibodies*.

Good example from *Your Child and Sickle Cell Disease*

Accessibility criteria: medical terms defined with illustrations



8 tools use illustration to support definitions of medical terms

Your child can control her own pain medicine with a PCA pump.

A TENS unit may help to relieve pain.

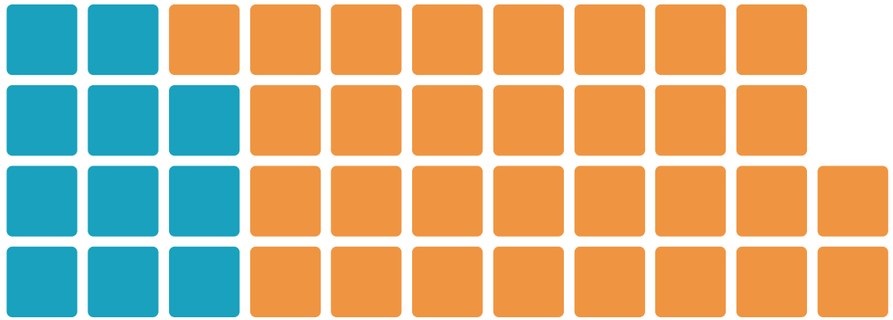
A Parents' Handbook for Sickle Cell Disease includes illustrations of terms and concepts that may not be commonly familiar, such as a patient controlling a PCA pump or using a TENS unit to relieve pain.

3. Actionability refers to how clearly a tool provides instructions for next steps or engages with a user.

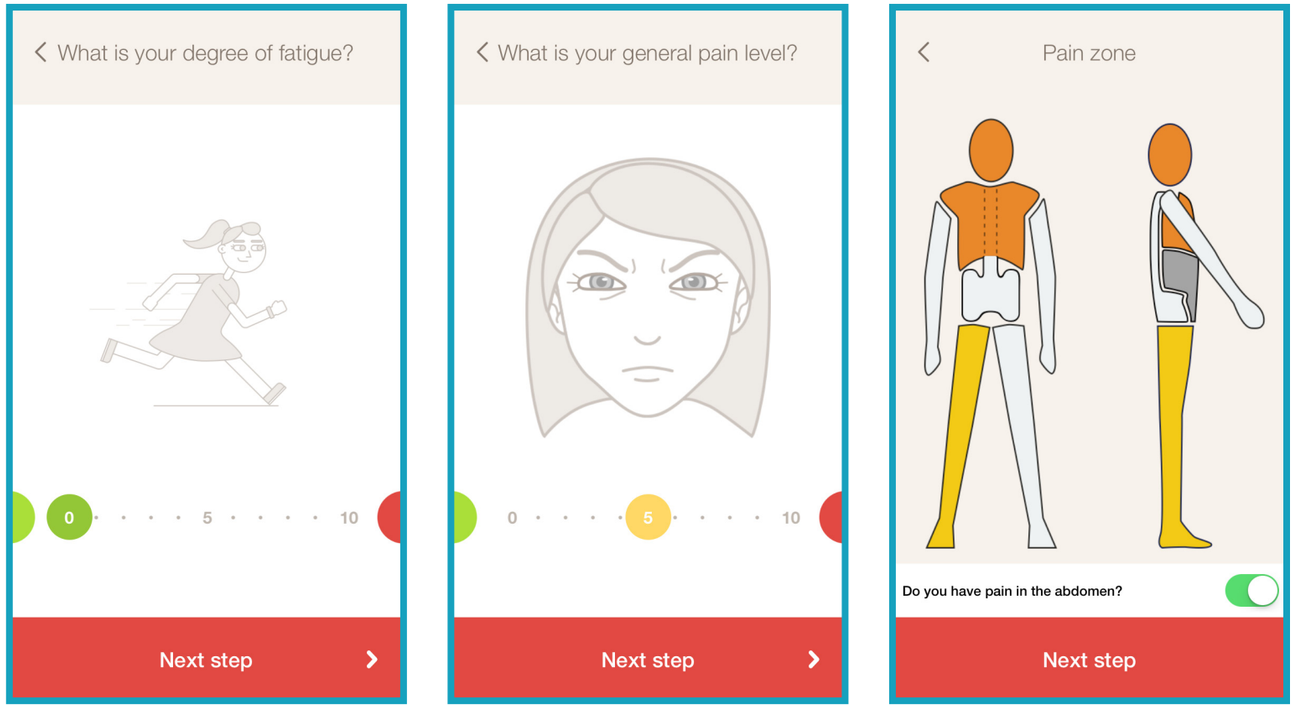
We evaluated:

- interactivity
- number of action items
- type of action items

Actionability criteria: encourage user interaction

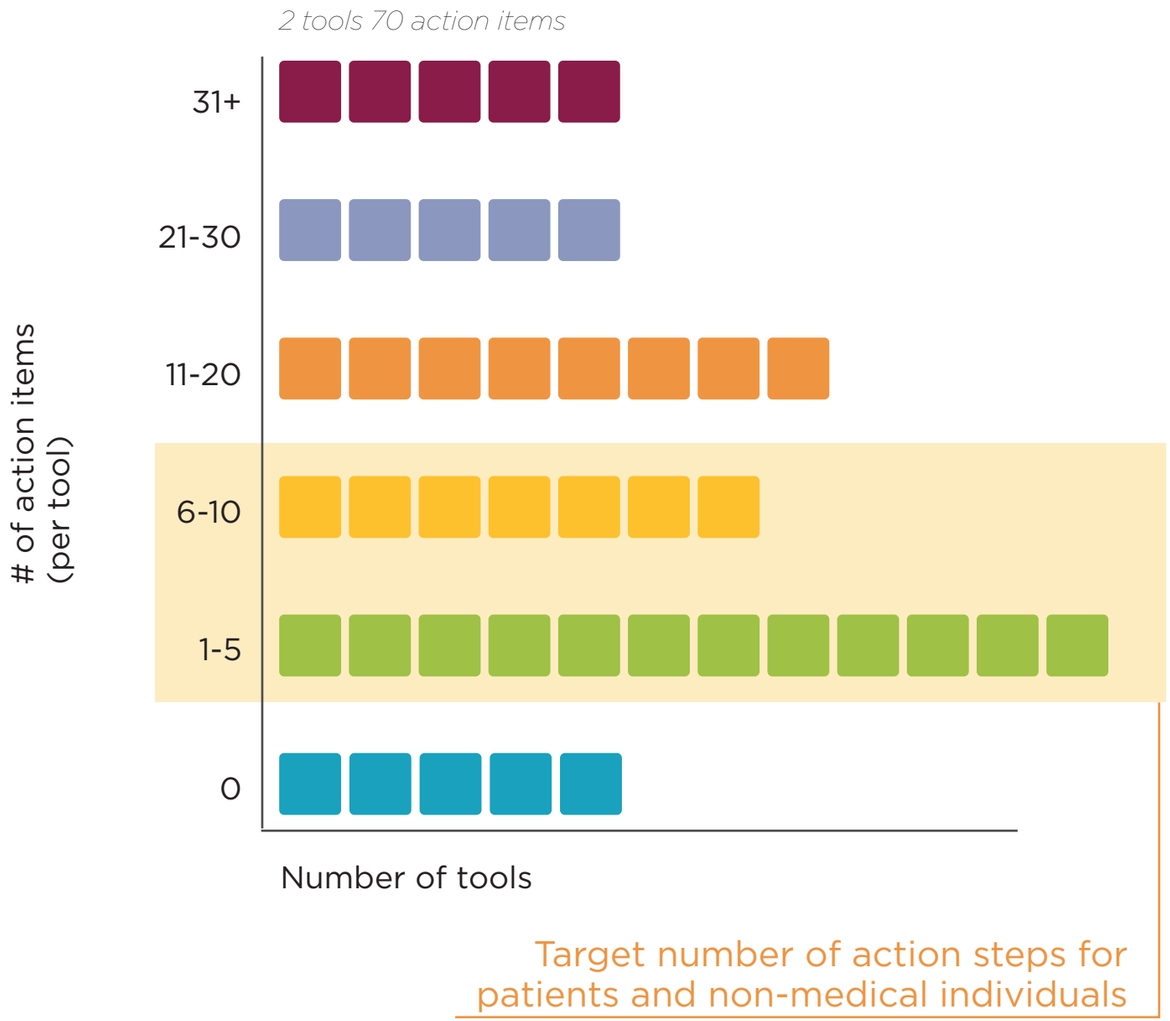


11 of 42 acute pain tools allow for users to interact



Sickle-O-Scope is a mobile application that allows individuals to keep a daily diary of their illness and symptoms.

Actionability criteria: number of action items



Section 2:
Living Well With Sickle Cell Disease

Six Steps to Living Well With Sickle Cell Disease

You can live a full life and enjoy most of the activities that other people do. The following tips will help you stay as healthy as possible:

Find good medical care—Sickle cell disease is a complex disease. Good quality medical care from doctors and nurses who know a lot about the disease can help prevent some serious problems. Often, the best choice is a hematologist (a doctor who specializes in blood diseases) working with a team of specialists.

Get regular checkups—Regular health checkups with a primary care doctor can help prevent some serious problems.

Prevent infections—Common illnesses, like influenza, quickly can become dangerous for a person with SCD. The best defense is to take simple steps like washing your hands frequently to help prevent infections. See "Five Tips to Help Prevent Infection" for more information.

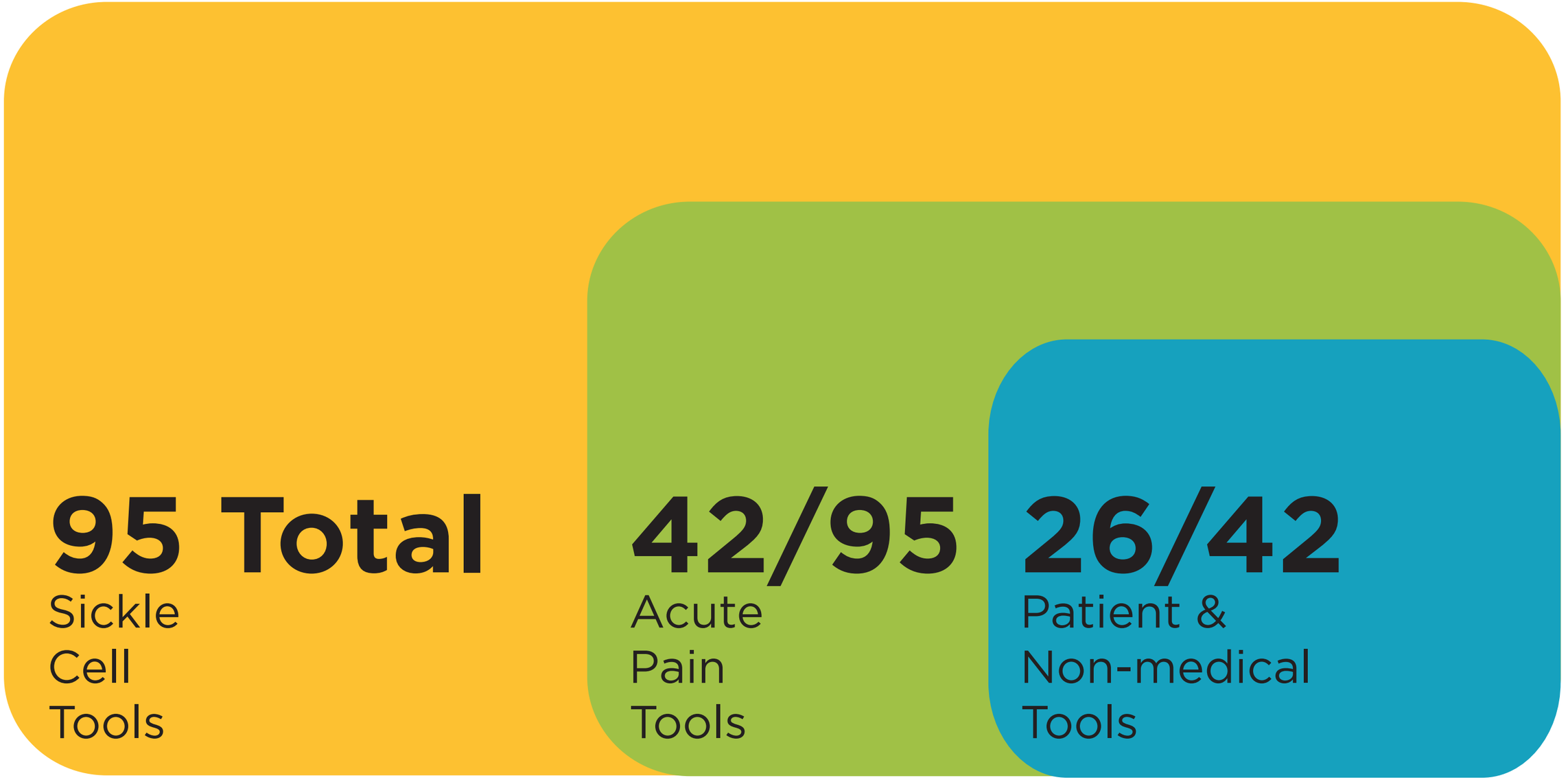
Learn healthy habits—Drinking 8 to 10 glasses of water every day and eating healthy food will help to maintain hydration and proper nutrition. People with SCD should maintain a balanced body temperature, getting neither too hot nor too cold. Participating in physical activity to help stay healthy is very important. However, it's essential that you don't overdo it, rest when tired, and drink plenty of water.

Look for clinical studies—New clinical research studies occur frequently and these studies might give you access to new medicines and treatment options.

Get support—Find a patient support group or community-based organization that can provide information, assistance, and support.

Good example from *Living Well with Sickle Cell Disease Self-Care Toolkit* clearly states 6 steps to Living with Sickle Cell Disease

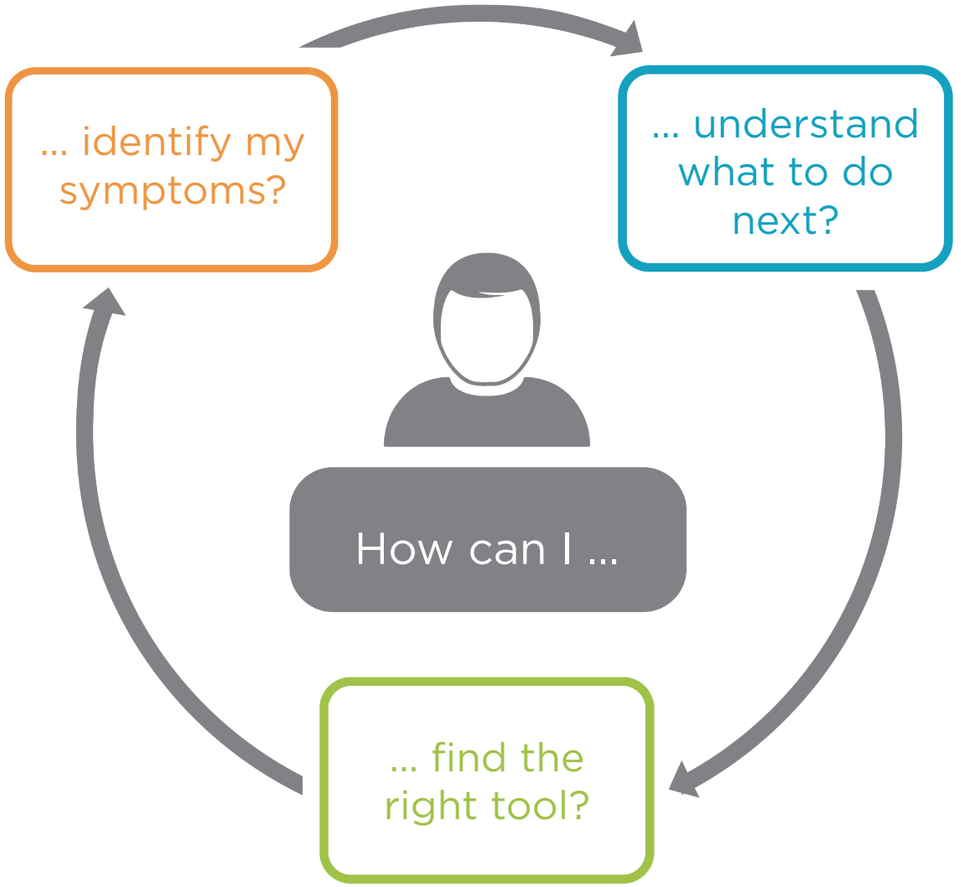
Acute pain tools intended for patient or non-medical audience use

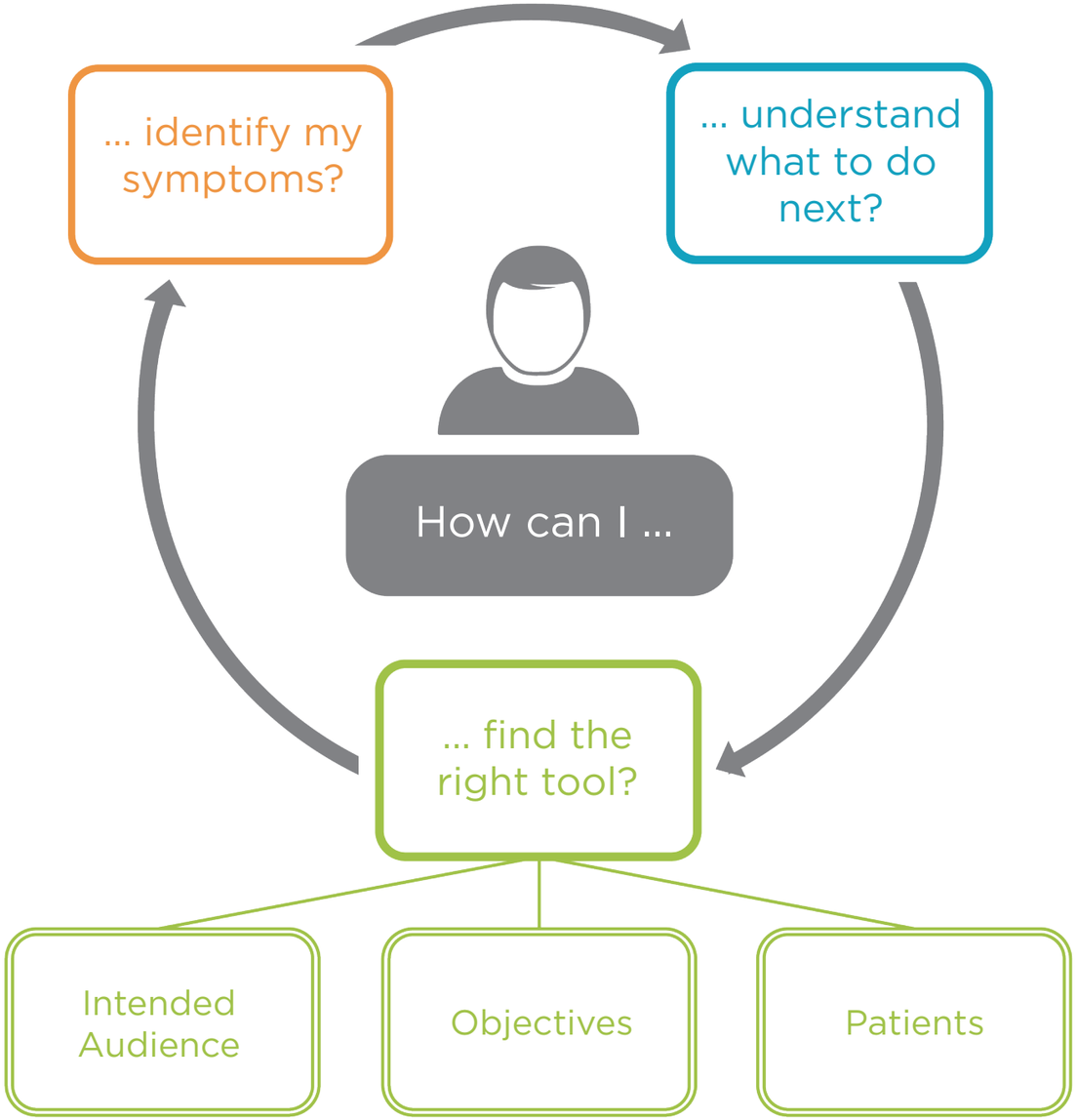


We used our analysis of the 26 patient and non-medical audience-facing tools to develop a model.

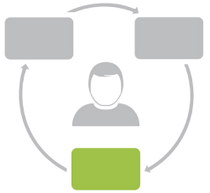
This model focuses on the needs users have of their tools during an acute pain crisis.

How can I find useful information when I have an acute pain crisis?


... identify my symptoms?
... understand what to do next?
How can I ...
... find the right tool?


... identify my symptoms?
... understand what to do next?
How can I ...
... find the right tool?
Intended Audience
Objectives
Patients

Actionability: Intended audience

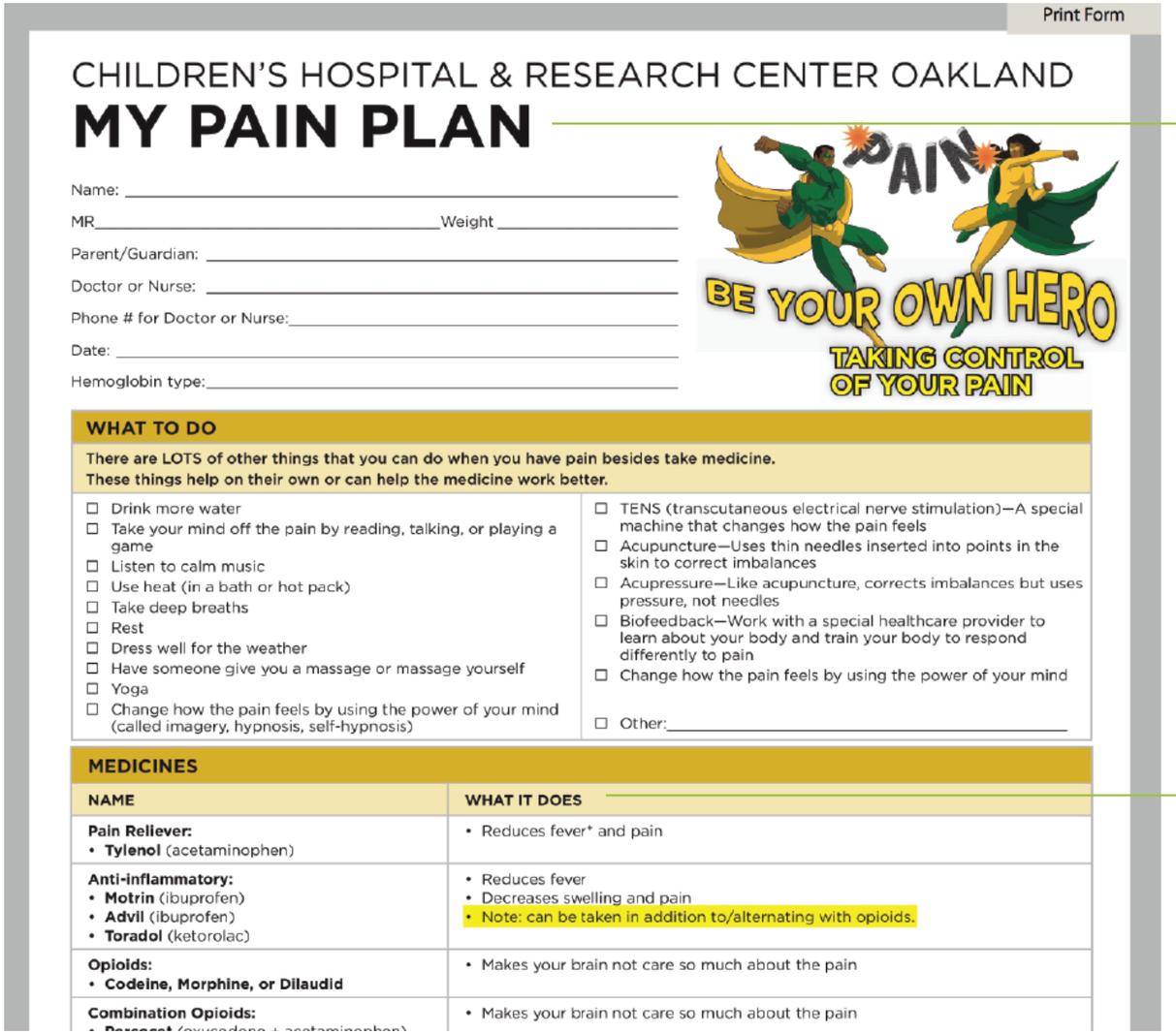


What Can You Do To Help?

Intended audience of each section is clearly identified

Good example from *Educator's Guide to Sickle Cell and School*

Actionability: Objective



Print Form

CHILDREN'S HOSPITAL & RESEARCH CENTER OAKLAND

MY PAIN PLAN

Name: _____

MR_____ Weight _____

Parent/Guardian: _____

Doctor or Nurse: _____

Phone # for Doctor or Nurse:_____

Date: _____

Hemoglobin type:_____

WHAT TO DO

There are LOTS of other things that you can do when you have pain besides take medicine. These things help on their own or can help the medicine work better.

- ☐ Drink more water
- ☐ Take your mind off the pain by reading, talking, or playing a game
- ☐ Listen to calm music
- ☐ Use heat (in a bath or hot pack)
- ☐ Take deep breaths
- ☐ Rest
- ☐ Dress well for the weather
- ☐ Have someone give you a massage or massage yourself
- ☐ Yoga
- ☐ Change how the pain feels by using the power of your mind (called imagery, hypnosis, self-hypnosis)
- ☐ TENS (transcutaneous electrical nerve stimulation)—A special machine that changes how the pain feels
- ☐ Acupuncture—Uses thin needles inserted into points in the skin to correct imbalances
- ☐ Acupressure—Like acupuncture, corrects imbalances but uses pressure, not needles
- ☐ Biofeedback—Work with a special healthcare provider to learn about your body and train your body to respond differently to pain
- ☐ Change how the pain feels by using the power of your mind
- ☐ Other:_____

MEDICINES

NAME	WHAT IT DOES
Pain Reliever: • **Tylenol** (acetaminophen)	• Reduces fever* and pain
Anti-inflammatory: • **Motrin** (ibuprofen) • **Advil** (ibuprofen) • **Toradol** (ketorolac)	• Reduces fever • Decreases swelling and pain • Note: can be taken in addition to/alternating with opioids.
Opioids: • **Codeine, Morphine, or Dilaudid**	• Makes your brain not care so much about the pain
Combination Opioids:	• Makes your brain not care so much about the pain

Objective of tool is clear and prominent

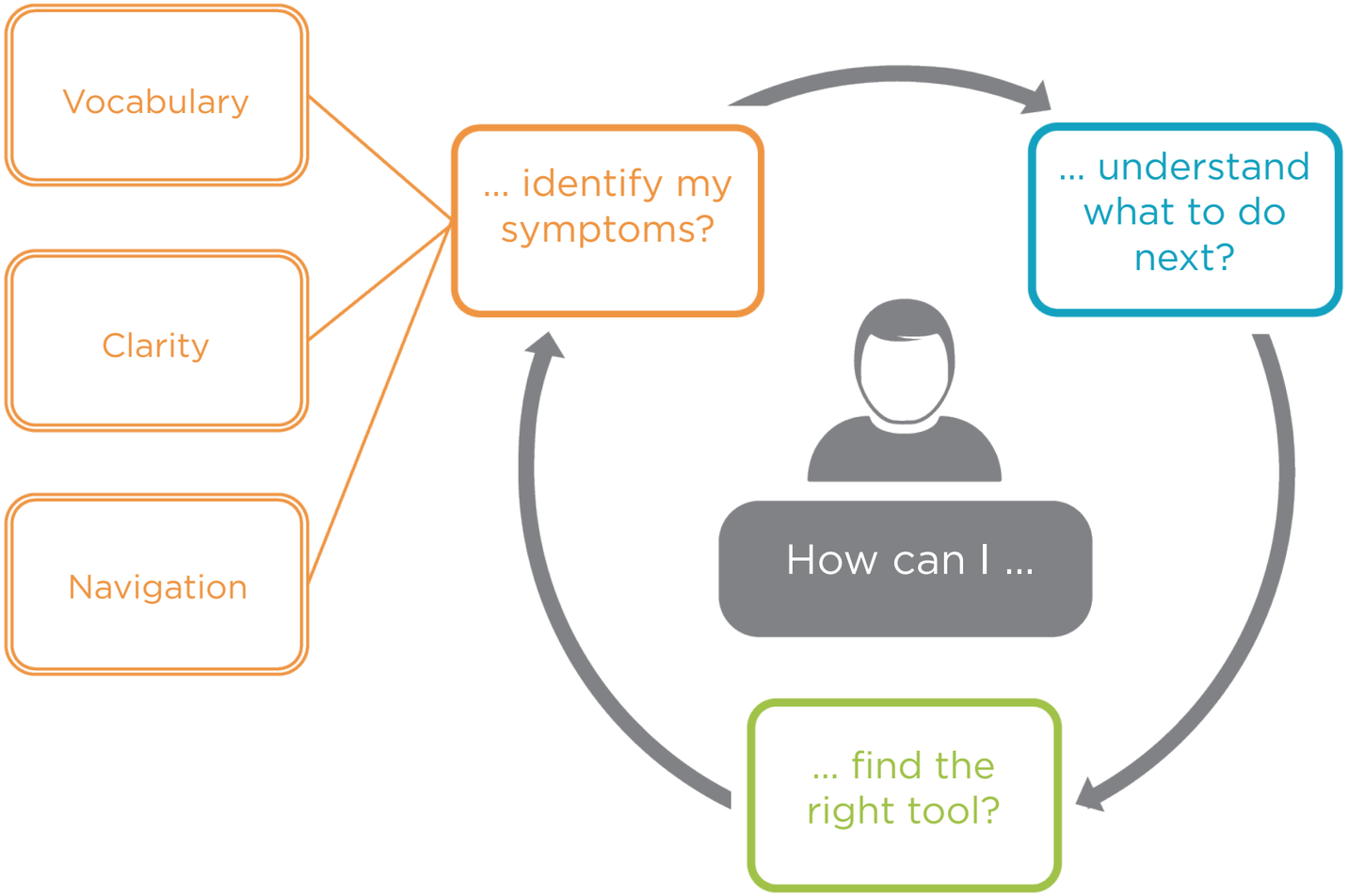
Objective of individual sections of tool are clear

Good example from *My Pain Plan*

Actionability: Patient

Intended patient demographic is clear

An example where the tool clearly states the intended patient age range from *A Parents' Handbook for Sickle Cell Disease*


Vocabulary
Clarity
Navigation
... identify my symptoms?
... understand what to do next?
How can I ...
... find the right tool?

Accessibility: Vocabulary

ORGAN/TISSUE INVOLVED	PROBLEMS CAUSED
KIDNEY	* Enuresis * Hematuria * Nephrotic Syndrome * Unconcentrated urine * Urinary frequency
SPLEEN	* Increased risk for serious infections * Splenic Sequestration * Abdominal pain
LUNGS	* Pneumonia * Acute Chest syndrome
BONES	* Infection * Aseptic Necrosis
BRAIN	* Stroke * Headache
SKIN	* Slow healing ulcers
PENIS	* Priapism
EYES	* Sickle Cell Retinopathy
LIVER	* Hepatomegaly * Cholelithiasis * Jaundice
• Not all these problems happen to everyone with Sickle Cell Disease. You need to know, however, that they **can** happen. **Notify the parents/caregivers immediately if you think their child has any of these problems.**	

Poor example from *Understanding the Child with Sickle Cell Disease, A Handbook for School Personnel.*

Readily understood signs are used to group symptoms

Grouping of problems by affected organ tissue is not intuitive to laypeople

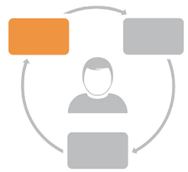
Everyday language is used to describe symptoms

Symptoms are described using unfamiliar medical terms

SIGNS	SYMPTOMS
FEVER	• 101 degrees or higher
PALLOR	• Noticeable change in complexion, lips, fingernails
BREATHING	• Dyspnea (difficulty breathing) • Tachypnea (fast rate of breathing) • Stertorous breathing (labored breathing)
HEADACHE	• Sudden or constant • Dizziness
HEARTBEAT	• Tachycardia (rapid heart beat) • Pounding
PAIN	• Head • Chest • Joints • Abdomen (abdominal distention) • Penis (prolonged erection)
SWELLING	• Hands • Feet • Joints (with redness)
MUSCULAR WEAKNESS	• Either side of the body
• Contact the child's family if you notice any of these signs and symptoms. Any change from what you feel is normal for the child should be reported to the family.	

Better example from *Understanding the Child with Sickle Cell Disease, A Handbook for School Personnel.* The symptoms and signs are frequently conflated in this example.

Accessibility: Vocabulary



Clear and concise descriptions of symptoms and recommended actions eliminate confusion in crisis situations

Poor example

There may be a prolonged, painful erection that does not go away for **more than several hours**. This can last up to **several days or weeks**. This type of priapism **needs attention by a doctor**. (Source: *Priapism-Sickle Cell Information Center*)

Better example

If your child has a painful erection that does not go away **within 30 minutes, call your doctor**. He **may need treatment right away** with a blood transfusion, IV fluids, and pain medication. (Source: *Parents' Handbook for Sickle Cell Disease*)

Accessibility: Navigation - Buried Content

9 of the tools for patients and caregivers contain content about more topics than acute pain. In fact, acute pain may only be a small portion of the tool.

Total document pages

Acute pain pages

1 of 16 pages Community Health Worker Training Manual

6 of 30 pages Sickle Cell Disease A Handbook for School Personnel

2 of 8 pages Tips for Supporting Students with Sickle Cell Disease

2 of 25 pages Educator's Guide to Sickle Cell and School

6 of 34 pages Living Well With Sickle Cell Disease Self-Care Toolkit

16 of 90 pages Parents' Handbook for Sickle Cell Disease

5 of 120 pages A parent's guide to manage sickle cell disease

4 of 20 pages Your Child and Sickle Cell Disease

2 of 12 pages Sickle Cell and Thalassemia: School Health and Safety

1 of 29 pages Sickle Cell Disease: Information for School Personnel

Accessibility: Content hard to find during a crisis

Tools that separate related content can create challenges for individuals seeking information in a time of crisis. The tool below, *A parent's guide to managing sickle cell disease*, contains 5 pages of content on acute pain distributed across the 120 page document.

is when your child's spleen can suddenly get very big and lead to worsening of their anaemia. You may be shown how to feel your child's spleen when he is well, so that you can tell if it is getting bigger and could need medical attention.

Painful episode

Pain is a very common symptom. The classic sickle cell episode or 'painful crisis' occurs when the very small blood vessels become blocked by 'sickled' red blood cells. This episode usually lasts several days and your child will need to be given regular painkillers and plenty of fluids and occasionally may need admission to hospital. Dactylitis may be the first painful episode that you see and usually occurs between the ages of 6 and 18 months when a finger or other parts of the hand, or a toe or other parts of the feet become swollen and painful. After this age, pain may occur in the arms, legs or back. Children may get less severe episodes of pain lasting only an hour or two. For further information about pain and how to manage sickle cell pain at home see page 42. It is worthwhile remembering that not all pain is due to sickle cell disease.

Bedwetting (nocturnal enuresis)

Bedwetting is normal in all children up until the age of about 7 years. It may take longer for a child with sickle cell disease to become dry at night. Because of tiredness from the anaemia the child may sleep very deeply at night and not wake up in time to go to the toilet. In addition, in sickle cell disease the kidneys are not able to produce concentrated urine, the bladder

p 19 What is a painful episode?

Managing sickle cell pain at home

Most episodes of sickle cell pain can be managed at home. In young babies and toddlers it may be difficult to know whether they are in pain or not. It is likely that you will notice that your child is not behaving as normal. She may be fretful and miserable, persistently crying, or only crying when moved. As your child gets older, you will find that she gets better at being able to tell you where the pain is. Sometimes, as in dactylitis (hand foot syndrome), you may see swelling of the part which hurts or it may feel warm to your touch.

Giving painkillers (analgesics)

It is a good idea to keep a supply of paracetamol (Calpol, Disprol) medication at home. If your child is in pain, give the paracetamol regularly every 4–6 hours as recommend on the bottle or by your doctor or pharmacist, but do not exceed the amount recommended.

Your doctor may also prescribe ibuprofen (Junifen) which helps relieve inflammation. This can be given with the paracetamol. The doctor may prescribe other painkillers for your child to use at home and as your child gets older, she will know which one works best for her.

Aspirin should not be given to a child under the age of 16 years.

If your child is getting no relief from the painkillers, you should call your GP or take her to the Accident & Emergency department. As well as giving regular pain relief, it is a good idea to also try some of the other treatments described below.

Extra fluids

Children with sickle cell disease should always be encouraged to drink plenty of fluids even when they are well. Dehydration (not enough water in the body) is known to be one of the causes of sickle cell pain.

Warm baths

Let your child soak in a warm bath for a while. Check that it is not too hot and do not let it get cold because this can trigger another pain episode. When a person is in pain sometimes gentle exercise in the warm water feels good and relieves anxiety.

Using warm moist towels or heat pads

Moist towels – Soak a face towel in warm water and wring it out then use it to gently massage the painful area. This can be very soothing and will often relieve pain. Do not let the towel get cold as this will make the pain worse. Heat pads (bought from the chemist) can be put on the painful area. They are either electric or non-electric.

Electric pads – Electric pads will have a temperature dial which needs to be set at the required heat temperature, but always check the manufacturer's instructions.

Non-electric pads – These may need to be heated in a bowl of warm water. Again, check the manufacturer's instructions.

p 42 Managing SC pain

In these situations you will need to get your child to the hospital straight away. Some hospitals have an arrangement so that you can take your child straight to the children's ward. If your hospital does not have this arrangement, take her to the Accident and Emergency department.

If you are unsure whether your child needs to go to hospital, always seek advice from your GP, specialist nurse or hospital doctor.

Some medical problems

Please remember that sickle cell disease is very variable and your child may never get any of the following problems or she may have some at different times in her life. Sometimes it is possible to manage a medical problem at home either on your own or with the help of your specialist nurse or GP. Sometimes it will be necessary for your child to go to hospital because she needs medical attention. If you are unsure whether your child needs to go to hospital, always seek advice from your GP, specialist nurse or hospital doctor.

Painful episodes

This is the most commonly seen medical problem in children with sickle cell and is caused by red blood cells becoming sickle-shaped and then blocking small blood vessels. One of the first signs of sickle cell disease may be painful swelling of the fingers or other parts of the hand, or the toes or other parts of the foot. This is known as dactylitis (hand foot syndrome) and may occur from about 6 months of age. If your child has been crawling or walking and then suddenly seems reluctant to do this, it may be because of dactylitis. She will need to be given regular painkillers and plenty of fluids. However if the pain remains moderate or severe and it cannot be managed at home effectively or if she is not able to drink or is vomiting she will need admission to hospital (see page 42 for how to manage sickle cell pain at home). The swelling usually goes down after a few days. It is unusual for children to have dactylitis after about 18 months of age.

Blockage of blood vessels can occur in any part of the body – muscles and bones, stomach or chest – and will cause pain in that part.

Pain may be mild but sometimes it can be very severe and will be very frightening for your child. Pain can sometimes be triggered by your child getting cold or hurting herself or by a viral infection, for example, flu, but many times there is no obvious cause. If the pain is severe and the painkiller you are using at home has not worked, it may be best for your child to go to hospital where she can be given stronger pain medicine.

Of course, like everyone else, she may have pain that is not due to sickle cell.

This can be confusing but as she grows up you/she will learn how to tell the difference.

p 44 Painful episodes

Medical Emergencies

Situations when your child needs to be seen by a doctor straight away

Fever Oral temperature of 38.5ºC or above

Very severe headache, dizziness or stiff neck

Breathing difficulty Pain or trouble breathing

Pain, if severe and not responding to the pain medicine you have at home

Colour Very pale palms, or lips

Spleen Sudden enlargement

Penis Painful erection lasting for more than 2 hours

Change in behaviour Appearing confused or drowsy or unable to speak

Fits, convulsions Body spasms and loss of consciousness

Weakness, particularly if not associated with pain and affecting one side of the body.

p 48 Pain as a medical emergency

Common reasons for hospital admission

Your child may be admitted to hospital for:

- Pain relief
- Other medications
- Intravenous fluids
- Medical investigations
- Blood transfusion
- An operation

Pain relief

You will probably be able to manage painful episodes at home using paracetamol and ibuprofen or any other painkiller that your doctor has prescribed. If these medicines do not control the pain, your child will need a stronger painkiller and it is likely that she will need to be admitted to hospital. A stronger painkiller, such as morphine, can be given by mouth but if your child is unwell and not drinking, it can be given in a drip into a vein (intravenous) or under the skin (subcutaneous). You should let the doctors and nurses know which painkillers she has already taken and at what time. Your child may know the best painkillers for her and which ones helped in previous painful episodes.

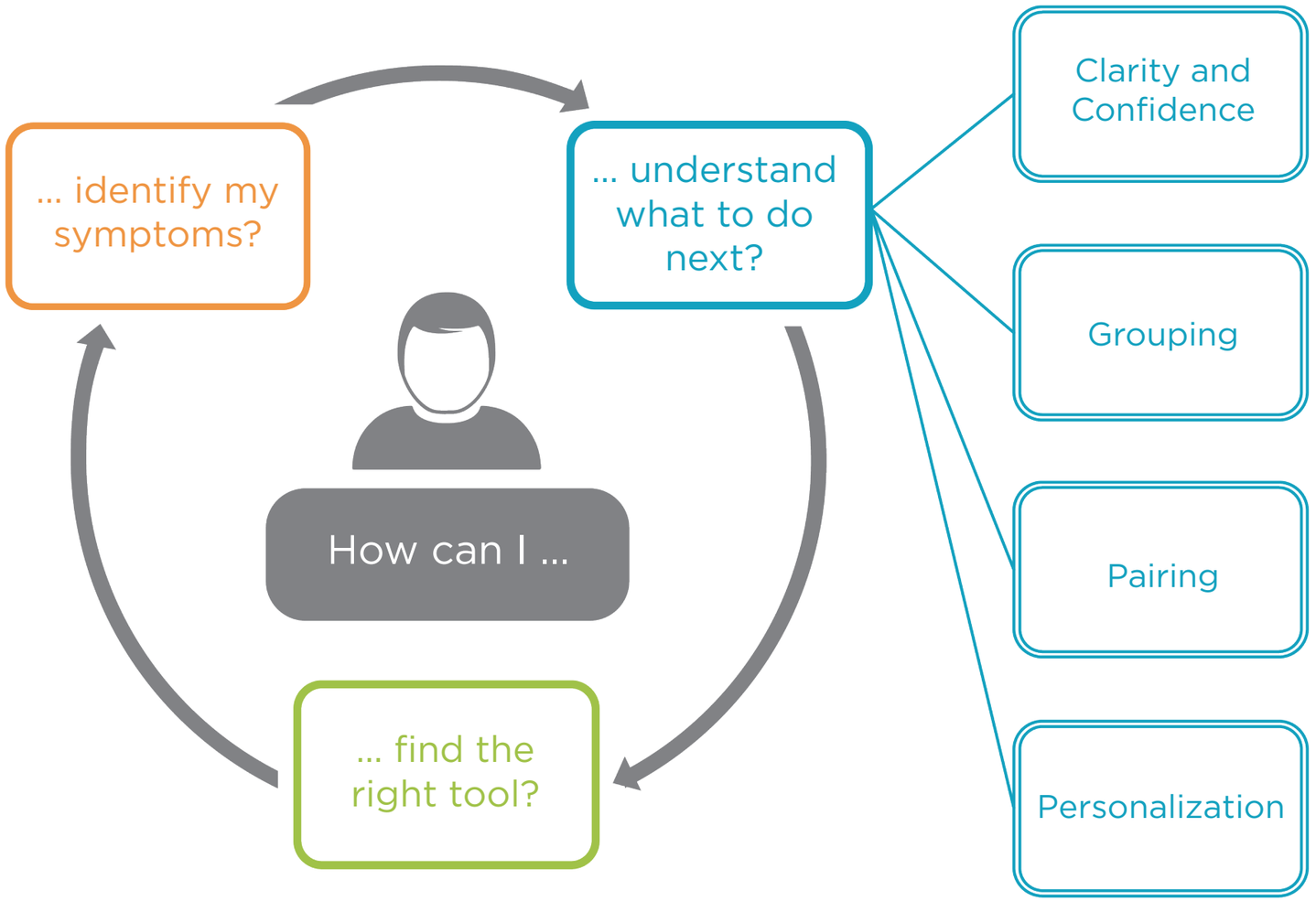
Some parents are concerned about using very strong painkillers, such as morphine, because they can be associated with drug dependency (when the body becomes too used to the medication and cannot do without it). Pain experts suggest that dependency is extremely unlikely to occur if the painkiller is used properly in the early stages of the painful episode and the effect is closely monitored. What is important is that enough pain medication should be given in the early stages to help your child cope better with the pain.

Many hospitals use a series of drawings of faces or bodies to help children indicate where the pain is and whether it is mild or severe. When children are very young, they may not be able to say how bad their pain is and their parents will have to help the doctors and nurses by telling them how severe they think their child's pain is. Older children may prefer to use numerical scales to score the severity of their pain from 1 to 5 or 1 to 10, with 1 indicating 'the least pain' and 5 or 10 indicating 'the worst pain ever', depending on the type of scale used.

When a child is old enough, she can control (within safe limits) the amount of painkiller she gets through a special pump. This method is known as patient controlled analgesia (PCA); the drug is given through a drip in the vein. Generally from about 6–8 years old, most children can start taking part in managing their own pain relief. Children will often turn the pump into a game, using the button on the PCA machine to 'zap' the pain away. This will help your child to feel that she has some control over the pain and will make her feel less frightened and powerless.

Many hospitals have a pain control team to help people with all types of pain. The team is usually made up of experts, such as anaesthetists, psychologists and specialist nurses, who will advise on

p 50 Pain relief


... identify my symptoms?
... understand what to do next?
How can I ...
... find the right tool?
Clarity and Confidence
Grouping
Pairing
Personalization

Actionability: Patients are most often told to seek medical help

Is this a desired outcome? Out of 396 action items identified across 26 patient and caregiver facing acute pain tools, more than half (52%) of recommended actions are to seek urgent medical help.

Communication
54 actions

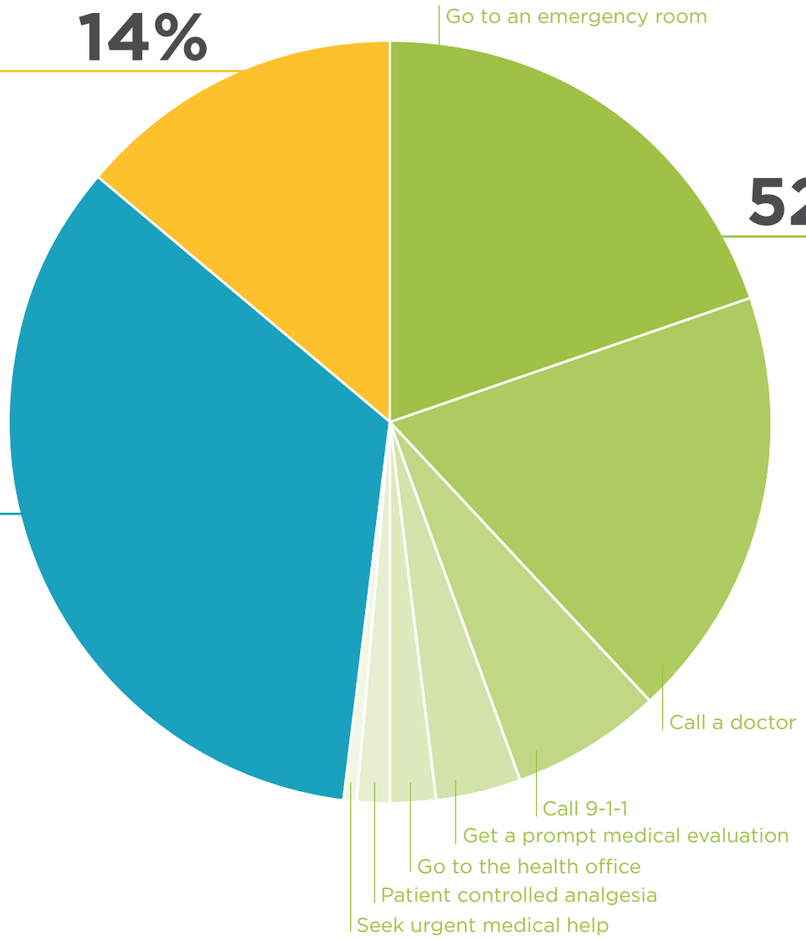
14%

Communicate with doctors
Notify parents
Plan with the school

Condition management
136 actions

34%

Avoid triggers
Make a plan for home
Manage medication
Identify symptoms
Prevent future crises



Seek medical help
206 actions

52%

Go to an emergency room
Call a doctor
Call 9-1-1
Get a prompt medical evaluation
Go to the health office
Patient controlled analgesia
Seek urgent medical help

Actionability: Clarity and Confidence

Poor example from *What You Should Know About Sickle Cell Disease*

General advice does not provide enough detail to be actionable

Detailed steps help people take action and follow through

Exercises for Younger Children

Exercise #1: The Rag Doll

"Pretend that you are a robot (or wooden doll), all stiff and straight. Your arms and legs don't bend at all. They just stay straight."

"Now you are a rag doll, all floppy, with no bones." (Lift your child's arm up, shake it a little to make sure it's really loose.) "All loose and floppy."

"When you need to relax, pretend to be the wooden doll, then the rag doll."

Exercise #2: Spaghetti

"Pretend that you are spaghetti in a package that has not yet been opened. You are all stiff and straight." (Have your child hold this for a few moments.)

"Now you're cooked spaghetti, all over the plate. Are you covered with sauce or meatballs?"

"When you need to relax, pretend to be spaghetti in the package, then the cooked spaghetti."

Good example from *A Parents' Handbook for Sickle Cell Disease*

Actionability: Grouping

Knowledge

Action

Knowledge

6. **Watch for signs of stroke.** Some children living with SCD may have learning difficulties due to health problems associated with stroke (blockage of blood vessels in the brain that then causes brain damage). Strokes may be difficult to detect when they affect a small portion of the brain, but they are extremely important to watch for because they are relatively common in the early school years among children with sickle cell disease. Teachers should be aware that declines in academic achievement, inability to maintain attention, difficulties with organization, and mild delays in vocabulary development may be due to small brain injuries caused by strokes. Moreover, teachers are in a unique position to notice changes in school performance that might indicate a stroke and should not simply assume that poor attention in the classroom is due to a lack of the child's motivation or desire to do well in school. Teachers should contact parents when changes in learning or a child's attentiveness are detected so that the child's doctor can be notified. Formal neurocognitive and educational testing may be necessary to determine any learning difficulties caused by stroke. The testing may help school personnel in developing the best teaching strategies for the student. Many students with SCD may qualify for a 504 plan or individualized education plan (see section 3, #2 for more about 504 or IEP). For more information, see the pull out box on stroke.

Poor example from *Tips for Supporting Students with Sickle Cell Disease*

Background knowledge and action steps are separated for quick reference

Action step is embedded in knowledge content

- Do not eat raw or undercooked eggs. Raw eggs might be "hiding" in homemade hollandaise sauce, Caesar and other homemade salad dressings, tiramisu, homemade ice cream, homemade mayonnaise, cookie dough, and frostings.
- Do not eat raw or unpasteurized milk or other dairy products (cheeses). Make sure these foods have a label that says they are "pasteurized".

5. ***Avoid Reptiles***—Salmonella (mentioned previously) is present in some reptiles and can be especially harmful to people with SCD. Make sure children and adults stay away from turtles, snakes, and lizards.

Emergency Guide: When To See the Doctor

It is very important that every person with SCD have a plan for how to get help immediately—at any hour—if there is a problem. Be sure to find a medical facility that will have access to your medical records or keep a copy that you can bring.

Go to an emergency room or urgent care facility *right away* for:

- Fever above 101°F.
- Difficulty breathing.
- Chest pain.
- Abdominal (belly) swelling.
- Severe headache.
- Sudden weakness or loss of feeling and movement.
- Seizure.
- Painful erection of the penis that lasts more than 4 hours.

Call a doctor *right away* for:

- Pain anywhere in the body that will not go away with treatment at home.
- Any sudden problem with vision.

Knowledge

Action

Good example from *Living well with Sickle Cell Disease, Self-Care Toolkit*

Actionability: Pairing

Symptoms paired with recommended actions help people make decisions and take appropriate next steps

If (student name) develops symptoms at school, follow the steps below:

TRUST the student's complaints. ALWAYS respond.

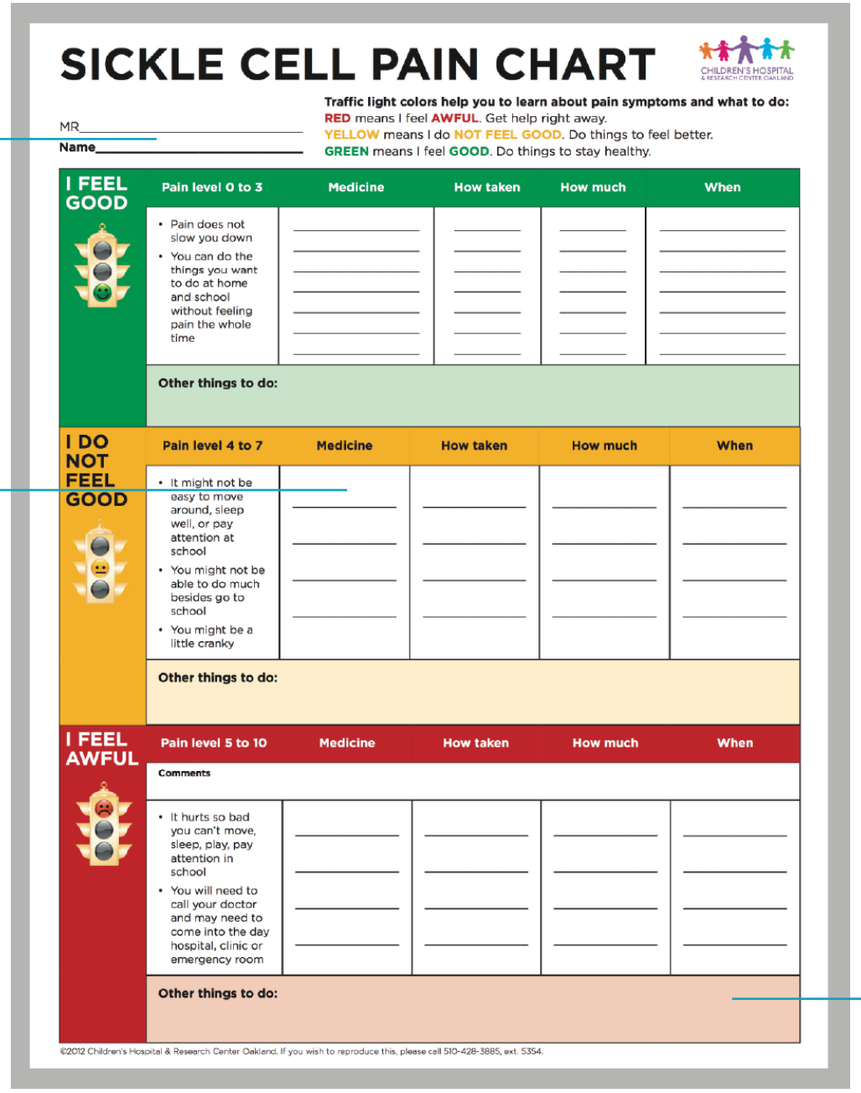
IF YOU SEE THIS	*DO THIS*
NONURGENT: Mild Symptoms • Minor localized pain in extremity • Minor injury • Feeling of something wrong • Tiredness, mild fatigue	SEND TO HEALTH OFFICE: Do NOT doubt student's complaint ➢ Provide oral hydration (at least 4-8oz/hour) ➢ Allow to rest ➢ Contact parent/guardian ➢ Administer pain medication as ordered ➢ Do NOT put ice on minor injuries ➢ Observe and reassess frequently ➢ Return to class if feeling better ➢ Send home if pain persists/prohibits active learning
URGENT: Moderate symptoms • Moderate fever ≥ 100°F (37.8°C) • Swelling/tenderness in extremity • Mild to moderate pain • Increased fatigue	SEND TO HEALTH OFFICE with escort: Do NOT doubt student ➢ Provide oral hydration (at least 4-8oz/hour) ➢ Allow to rest ➢ Contact parent/guardian to transport student for medical care or home ➢ Administer pain medication as ordered ➢ Elevate affected extremity ➢ Do not put ice on painful area ➢ Observe student closely. Do NOT leave unattended.

Good example from *Sample Emergency Care Plan - Student with Sickle Cell Disease*

Actionability: Personalization

Patient can “own“ the tool by writing in name

Patients can create their own plan for personal use and sharing with others



SICKLE CELL PAIN CHART

CHILDREN'S HOSPITAL & RESEARCH CENTER OAKLAND

MR_____

Name_____

Traffic light colors help you to learn about pain symptoms and what to do:
RED means I feel **AWFUL**. Get help right away.
YELLOW means I do **NOT FEEL GOOD**. Do things to feel better.
GREEN means I feel **GOOD**. Do things to stay healthy.

I FEEL GOOD	Pain level 0 to 3	Medicine	How taken	How much	When
	• Pain does not slow you down • You can do the things you want to do at home and school without feeling pain the whole time				
	Other things to do:				

I DO NOT FEEL GOOD	Pain level 4 to 7	Medicine	How taken	How much	When
	• It might not be easy to move around, sleep well, or pay attention at school • You might not be able to do much besides go to school • You might be a little cranky				
	Other things to do:				

I FEEL AWFUL	Pain level 5 to 10	Medicine	How taken	How much	When
	Comments				
	• It hurts so bad you can't move, sleep, play, pay attention in school • You will need to call your doctor and may need to come into the day hospital, clinic or emergency room				
	Other things to do:				

Good example from *My Pain Plan*

Space for “other things to do“ gives flexibility and recognizes people have many ways to cope in crisis

Usability:

Current state

- Most tools intended for patients and non-medical audiences require them to decipher content because of they do not follow known information design principles.

Implication

- Individuals miss important messages while scanning or searching the content.

Opportunity

- There is an opportunity to deliberately apply communication design principles to bring clarity and prioritization to the content of a tool. This will enable individuals to easily navigate material to locate information based off of their needs.

Accessibility

Current state

- Most tools (18 out of 26) intended for patient and non-medical audiences are not written at the target reading grade level (grade 5-6).

- When tools provide definitions of medical terms, patient comprehension is supported and the tool becomes more self evident. When illustrations are included they provide additional information and visual communication when presenting new materials.

Implication

- Tools do not meet their intended audience's comprehension needs. Important information may be missed or misunderstood due to the complexity of the content. In the context of an acute pain crisis, this can be annoying at best, dangerous at worst.

Opportunity

- There is an opportunity to design a tool that supports comprehension for the majority of patient and non-medical audiences by creating content at the appropriate target reading grade level (grades 5-6). This tool should also self evident, providing users with the definitions of medical terms.

Actionability

Current state

- Patients & non-medical audience are hearing a lot from their tools about going to the emergency room, and less about how to prevent themselves from needing to go to the emergency room.

- Information about self management is related to information about acute pain and emergency room visits, but this connection is not always clear within tools.

- Most tools intended for patients and non-medical audiences (31 of 42 tools) do not facilitate interactivity; instead, they serve as a reference.

Implication

- Most tools are not useful on a regular basis. They support passive use — e.g. occasionally needing to look something up — rather than acting as a utility or communication device for patients in acute pain crisis.

Opportunity

- There is an opportunity to design a tool that can serve as a companion for patients. This tool might be used for tracking, reflection, information storage, care plan communication, and potentially provide analytics that help anticipate a future pain crisis. The tool would serve patients holistically and support preventative care in addition to during times of acute pain.

What's next:

- developing tool prototype (pain passport)
- gather feedback on prototypes through field work
- continue evaluation of tools in other content areas (transition, self-management)

Tool data sources

U.S. Department of Health and Human Services
National Institutes of Health
National Heart, Lung, and Blood Institute

UNIVERSITY OF ILLINOIS
Hospital & Health Sciences System
Changing medicine. For good.

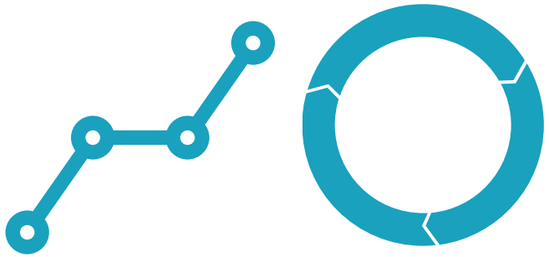
TEXAS
Health and Human
Services

Texas Department of State
Health Services

Process:

1. Tool collection + review (filtering for acute pain content)

2. Creation of cataloging instrument + entry of tools



3. Data analysis, synthesis of findings + model creation