

NIH News in Health

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Battling Bent Blood Cells Progress in Sickle Cell Disease

With every beat of your heart, blood carries oxygen from your lungs throughout your body. This life-sustaining process happens automatically, whether you're awake or asleep.

But for people with sickle cell disease, it often goes awry. People with this disease have an abnormal type of hemoglobin, the oxygen-carrying molecule in red blood cells.

Normally, red blood cells are flexible and shaped like a disc. But the hemoglobin in people with sickle cell disease causes abnormally shaped red blood cells. Most commonly, they're a crescent (or sickle) shape.

These inflexible, bent cells can stick to the blood vessel walls. The resulting clumps slow or stop the flow of blood. This may lead to pain and organ damage.

"Sickle cell disease can potentially block blood supply to any organ in the body," explains Dr. Swee Lay Thein, a blood disorder expert at NIH.

Most people with sickle cell disease used to die before reaching adulthood. But with modern



treatments, people in the U.S. now live into their 40s, 50s, or even 60s. And researchers are working on cutting-edge therapies, such as fixing the broken **gene** that causes the disease.

"I think treatment is going to look very different in the next 20 years," says Dr. Allison King, an expert on sickle cell disease in children at Washington University in St. Louis.

One Gene, Many Symptoms • Sickle cell disease is caused by changes in a single gene. But everyone has two copies of the gene. You inherit one copy from each parent.

More than two million people in the U.S. carry one abnormal copy, called sickle cell trait. They don't usually have any symptoms. But if

you inherit two copies, the result is sickle cell disease.

About 100,000 people in the U.S. live with sickle cell disease. Most are African American. But every baby born in the U.S. is tested for sickle cell disease at birth. This helps doctors start treatment as early as possible.

Children don't usually show symptoms until they're between six and 12 months old, Thein explains. Babies' red blood cells have a different type of hemoglobin, called fetal hemoglobin. As they grow, the body switches to producing adult hemoglobin. Then, the sickling cycle begins.

A normal red blood cell lives for around three to four months. But in people with sickle cell disease, the cells usually live for just two to three weeks. This leads to anemia, a condition in which your blood has low amounts of red blood cells or hemoglobin. Anemia lowers oxygen in the body, which can cause fatigue, dizziness, and headaches.

Pain is another common symptom.

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Definitions

Gene

A stretch of DNA you inherit from your parents that defines features, like your risk for certain diseases.

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It can be so severe that people end up in the hospital. These pain episodes are called a “sickle cell crisis.”

“People’s lives are disrupted by these episodes,” says Thein. “For children, that means missing school. Adults might miss a lot of work.”

Blocked blood flow to the brain can cause strokes, even in children. Strokes and clogs in the blood vessels in the lungs are some of the most dangerous complications of the disease, Thein says.

New Drug Options • The most common treatment for sickle cell disease is a drug called hydroxyurea. It coaxes the adult body to make fetal hemoglobin. This increases the number of functional red blood cells. Hydroxyurea doesn’t work for everyone. But three new treatments have been approved in the last few years. Some of the newer drugs prevent

Definitions

Stem Cells

Immature cells that have the potential to develop into many different cell types in the body.

sickled cells from sticking to the blood vessels.

Thein’s team is testing a drug to stop blood cells from bending in the first place. “That would be the most effective thing—to stop the sickling process,” she explains.

Even though drugs help many people, taking medication daily can be hard, King explains. Low levels of oxygen in the brain in people with sickle cell disease can affect memory. King and her colleagues are testing ways to use technology, such as smartphone apps, to help people take their drugs as prescribed.

Some people with sickle cell disease may need to have regular or emergency blood transfusions, in which they receive donated blood.

Fixing the Blood Cells • Currently, the only cure for sickle cell disease is a bone marrow transplant. Bone marrow is the spongy tissue containing the **stem cells** that give rise to blood cells.

In a bone marrow transplant, the stem cells in the patient’s bone marrow that produce blood cells are first destroyed. Then, stem cells from a donor without sickle cell disease are transferred into the patient. These new stem cells will produce blood cells that don’t sickle.

The procedure is risky. It’s considered too dangerous for adults. But the main problem, explains Dr. Matthew Hsieh, a transplant researcher at NIH, is that most children don’t have a matched donor. If certain proteins on the donor’s cells are different than the child’s, the transplant can fail.

Hsieh and others have developed new methods to transplant bone marrow from people who aren’t a perfect match. They’re also developing ways to prepare the bone marrow for a transplant that could make the procedure safer for adults.

Researchers are also testing an



Wise Choices

Tips for Living With Sickle Cell Disease

- **Check in with your doctor regularly.** Most people with sickle cell disease should see their doctor every three to 12 months.
- **Get recommended vaccinations.** People with sickle cell disease have a higher risk of infection. Learn more at www.cdc.gov/vaccines/schedules/easy-to-read/index.html.
- **Adopt a healthy lifestyle.** Getting enough sleep, eating healthy, and avoiding alcohol and tobacco can make you feel better and reduce pain.
- **Manage pain.** Work with a pain specialist to come up with an individual treatment plan.
- **Know and avoid your triggers.** Many things can set off pain. Common ones include exhaustion and dehydration.

approach called gene therapy. In gene therapy, a patient’s own stem cells are collected. Then, they’re altered in the lab to fix a gene. Finally, they’re given back to the patient.

A recent gene therapy study at NIH successfully added a working copy of the hemoglobin gene into stem cells. Another study will see if adult cells can be altered to produce fetal hemoglobin.

If you’re living with sickle cell disease, talk with your health care provider to develop a care plan that’s right for you. Maintaining a healthy lifestyle can help you prevent or control some of its complications (see the Wise Choices box for tips).

If you’re interested in joining a clinical trial, call 1-800-411-1222 or visit www.clinicaltrials.gov. ■

NIH News in Health

ISSN 2375-6993 (Print) ISSN 1556-3898 (Online)

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Web Links

For more about sickle cell disease and an **extended Q&A**, see “Links” in the online article: newsinhealth.nih.gov/2020/09/battling-bent-blood-cells

A Biting Problem

Taming TMJ Troubles

Your jaw works hard every day so you can laugh, talk, smile, and eat. When it's working properly, you may not give it much thought. But if your jaw starts to hurt, it can take the joy out of simple, everyday things.

The jaw joint is one of the most complex joints in the human body. For most people, it moves effortlessly up and down, side to side, and in and out, transitioning from one movement to the next seamlessly. But, more than 10 million people in the U.S. live with jaw pain and dysfunction.

Doctors call these conditions temporomandibular disorders. They're more commonly called temporomandibular joint (TMJ) disorders.

"Temporomandibular disorders—and how people respond to them—vary widely," explains Dr. Dena Fischer, a dental health expert at NIH. "For example, some experience discomfort, others tension, and still others severe pain."

Some people get symptoms in the muscles that move the jaw. For others, it's in a disc within the jaw joint that's damaged. You can also develop arthritis, or joint **inflammation**. You can even have more than one kind of disorder at the same time.



Wise Choices Tackling Jaw Pain

Simple, self-care practices to try:

- Avoid hard foods.
- Use ice packs on the joint.
- Try over-the-counter pain medications, like ibuprofen, for a short amount of time.
- Avoid extreme movements, like wide yawning and chewing gum.
- Learn relaxation and stress reduction techniques.

TMJ disorders sometimes start after an injury. But for most people, there's no obvious cause. In addition to pain, other symptoms can include stiffness, limited jaw movement, painful clicking or popping in the joint, or changes in the way the teeth fit together.

If you have any of these symptoms, talk with your health care provider. To diagnose a TMJ disorder, they'll ask you questions about your symptoms and examine your head, neck, face, and jaw. They'll also check your dental and medical history. They may use imaging tests, like X-rays, too.

Experts recommend starting with simple, self-care practices for jaw pain (see the Wise Choices box for tips). "For a lot of people, the pain will resolve over time," Fischer explains. "Your doctor may also recommend trying a bite guard. These are plastic splints that fit over the teeth."

Sometimes, TMJ disorders can become chronic—causing pain or discomfort that lasts more than three months. Aggressive treatments include surgery, splints that change the bite, and even adjusting or removing teeth. But whether these treatments help hasn't been scientifically studied, explains Fischer.

For some people, they may make things worse. "And once you have surgery, you can't put things back the way they were before," she says.

If you have symptoms that last more than three months, your dentist or health care provider may refer you to a specialist. Doctors who specialize in muscles and bones, arthritis, pain, and the nervous system may be able to help.



But better treatments are needed. NIH-funded researchers have been studying the role that genes play in who develops a TMJ disorder and how long it lasts. In a large study, researchers identified several genes that are more common in people who have severe jaw pain. They're now testing whether early treatment can help people with certain genes lower their risk of developing a chronic disorder.

"We hope that having a better understanding of why temporomandibular disorders develop will ultimately help us prevent them and find new treatments," Fischer says. ■



Definitions

Inflammation

Heat, swelling, and redness caused by the body's protective response to injury or infection.



Web Links

For more about TMJ disorders, see "Links" in the online article: newsinhealth.nih.gov/2020/09/biting-problem



Health Capsules

For links to more information, please visit our website and see these stories online.

Protein May Reverse Age-Related Memory Loss

Exercise is important for your body and your brain at any age. A new study reveals how physical activity may slow the decline in learning and memory as you get older.

Researchers looked at the brains of young and old mice. Some had access to a running wheel in their cage, while others did not. The team found changes in the part of the brain called the hippocampus. This area is important for learning and memory.

Older mice that were physically active had more brain cells called neurons in the hippocampus and

made fewer errors on learning and memory tasks.

The team then injected older, less active mice with blood plasma from other mice. Older mice given plasma from active mice showed increases in neurons, learning, and memory performance similar to active mice.

Researchers linked these brain benefits to a protein called GPLD1. When injected with the gene for GPLD1, older mice performed similarly to the active mice on memory tasks and had more neuron growth.

The team looked at the protein in

people, too. Older adults who were more active had higher levels of GPLD1 in their blood than inactive adults. Because GPLD1 is produced in the liver, more research is needed to determine how it works on the brain. This research could lead to ways to treat or protect against age-related decline in the brain.

“Through this protein, the liver is responding to physical activity and telling the old brain to get young,” says Dr. Saul Villeda, of the University of California, San Francisco, who led the study. ■

Communicating Clearly While Wearing a Face Covering

Wearing a face covering is an important part of keeping you and others healthy right now. But they can also make talking to those around you more difficult. Face coverings can muffle sound. They can also hide important clues about the speaker’s message and emotions. This can make it hard to understand speech, especially for those with hearing loss.

Millions of people in the U.S have hearing loss, including half of those older than 75. Now, more than ever, it’s important to make an extra effort to communicate. Speak more

clearly and louder than you normally would, without shouting. Reduce background noise when possible. Be aware that physical distance can also make hearing more difficult.

Make sure the person you’re speaking with understands you. Ask and adapt if needed. You can also offer to use another method—a smartphone, paper and pen, or whiteboard—to get your message across. When it’s essential that you understand spoken details, like at a doctor’s appointment, consider bringing a friend or family member to help.

Face coverings can make communication challenging for everyone. But people with hearing problems often rely on lip-reading to understand what’s being said. Consider using a clear face covering to make your mouth visible, instead of a cloth covering. With a little extra effort and problem solving, we can all communicate clearly while staying safe.

For more tips, go to: www.nidcd.nih.gov/about/nidcd-director-message/cloth-face-coverings-and-distancing-pose-communication-challenges-many. ■



Featured Website

Know the Science

www.nccih.nih.gov/health/know-science/make-sense-health-research

Learn how to better understand health research in the news. Find tips on reading a scientific paper and interpreting the findings.

The website walks you through important questions to ask about the goals of the research, possible bias, and study design.

Know the Science: 9 Questions To Help You Make Sense of Health Research

Introduction

Almost every day, new findings on medical research are published, some of which may include complementary health approaches.

Research studies about medical treatments and practices published in scientific journals are often the sources of news stories and can be important tools in helping you manage your health.

But finding scientific journal articles, understanding the studies they describe, and interpreting the results can be challenging.

One way to make it easier to understand information you find in a scientific journal is to share the information with your health care providers and get their opinions. Once you understand the basics and terminology of scientific research, you have one more tool to help you make better, informed decisions about your health.

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