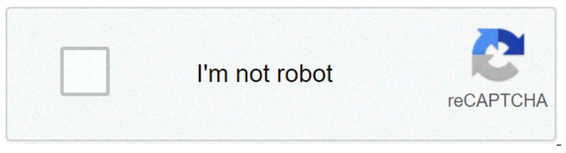

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Sickle cell blood cells

Sickle cell trait means having one gene for a condition called sickle cell disease (SCD). This in itself does not normally cause problems and sickle cell trait is not considered as a disease. It is extremely rare for it to cause problems or complications, which mainly occur under conditions of severe physical stress (explained below).Sickle cell trait is important because your children can inherit the sickle cell gene. If both parents have sickle cell trait or sickle cell disease, their children could get a double dose of the sickle cell gene, which would give them the serious condition called SCD. This is relevant if you are pregnant or wanting to start a family.Anyone can have sickle cell trait but it is most common in people whose family origin is Black African, Black Caribbean or Black British. It also occurs in people who originate from the Middle East, India and Eastern Mediterranean areas. In other populations, sickle cell trait is unusual but can occur.In England, about 240,000 people carry a sickle cell gene.You inherit haemoglobin genes from both parents. One sickle cell gene gives you sickle cell trait; two sickle cell genes give you SCD.This means that: if both parents have sickle cell trait, their children have: a 1 in 2 chance of having sickle cell trait, a 1 in 4 chance of having SCD, and a 1 in 4 chance of having no sickle genes (as shown in the diagram above).If one parent has SCD and the other has sickle cell trait, their children have: a 1 in 2 chance of having sickle cell trait, and a 1 in 2 chance of having SCD.If one parent has sickle cell trait and the other has no sickle genes, their children have: a 1 in 2 chance of having sickle cell trait, and a 1 in 2 chance of having normal haemoglobin genes.Actually, the inheritance of SCD is slightly more complicated than that. This is because there are other haemoglobin genes which can interact with sickle cell trait. If you have one of these genes plus a sickle gene, you also get SCD (the combination behaves like two sickle cell genes). Examples of these interacting genes are HbC, beta thalassaemia, Hb Lepore and HbO Arab.The result of these genes is that an important body chemical called haemoglobin is altered. This is what affects the red blood cells and makes them change into a sickle shape. With sickle cell trait, you have only small amounts of altered haemoglobin, so the blood cells are not much affected (unless conditions are very extreme). With sickle cell disease, there is more altered haemoglobin and the red blood cells easily change into sickle shape, which causes various problems. These are described in more detail in our separate leaflet called Sickle Cell Disease.Sickle cell trait is diagnosed in the same way as SCD. It involves a blood test. The test may be offered to pregnant women and if necessary their partners. Babies may also need the test.To read about sickle cell trait diagnosis, see the separate leaflet called Sickle Cell Disease (Sickle Cell Anaemia).It is extremely uncommon for people with sickle cell trait to have any symptoms. Complications of sickle cell trait are also uncommon. Most of the complications which do occur are due to extreme physical conditions, and so are usually preventable. The possible complications are:These can occur in conditions where oxygen is extremely low - such as flight in an unpressurised aircraft, very high altitude or problems during a general anaesthetic. Also, if you become severely lacking in fluid in the body (dehydrated).In these situations, sickle cells can form and can block small blood vessels. This may cause episodes of pain in bones, muscles or the spleen. If in the spleen, it is called splenic infarction and may cause pain in the tummy (abdomen) or chest. These problems may be severe enough to make you very unwell and to need treatment in hospital. They will usually clear up with treatment and when oxygen and hydration levels are back to normal.To avoid possible complications, you shouldAvoid flying in unpressurised aircraft.Avoid visiting very high altitudes.Keep your fluid intake up, especially if you have a fever or are exercising.Sometimes people can become seriously ill as a result of extreme physical training. This can particularly occur if it is a forced pace, under very hot conditions, if they are unused to the training, or if they do not drink enough fluid. Anyone can become ill under these conditions, but people with sickle cell trait are probably at higher risk.In these conditions, people with sickle cell trait may get pain episodes or pain in the spleen (as explained above).How can complications of severe exercise be prevented?The following are a few recommendations which may be useful to prevent complications occurring:Tell your trainer, sports association, doctor and nurse that you have sickle cell trait, and make sure they are aware of the guidelines.Avoid extremes such as heat, very high altitude, exertion to the point of collapse, or sudden increase in exercise without proper training. You can still take part in sports if you take precautions. There are professional athletes who have sickle cell trait.When exercising, drink enough fluid. Have breaks for drinks, rest and cooling.Sports where you can pace yourself are safer than a forced pace.If you have symptoms such as pain or difficulty breathing, stop, report your symptoms, rest and rehydrate. Get medical advice if symptoms don't improve quickly.Build up your training gradually.If you also have asthma, make sure it is treated and well controlled.Extreme activities may not be suitable - you will need medical advice.As with any condition affecting the blood, it is healthier not to smoke.There are some other conditions which seem slightly more common in people with sickle cell trait. However, there is still uncertainty as to how far these conditions are linked to sickle cell trait. Possibly, you may be more prone (compared with people without the trait) to:Blood in the urine (haematuria) - this may be noticeable, or in a tiny quantity found only on urine tests. It may affect about 1 in 50 people with sickle cell trait. If you have blood in the urine, it should be investigated (tests done) to see if there is any particular cause. Treatment is needed in some cases.A form of kidney cancer. If you develop weight loss, loin pain, fever, and/or tummy pain, along with blood in the urine, always see a doctor straightaway. You should be referred urgently for investigation.Bladder or kidney infections (urinary tract infections).A blood clot in the leg or lung (deep vein thrombosis (DVT) or pulmonary embolus).To help prevent these problems, advice is to avoid dehydration by drinking enough fluid. To prevent a DVT or a pulmonary embolus, keep mobile and move your legs when on a long journey or flight. See separate leaflets called Deep Vein Thrombosis, Preventing DVT When You Travel and Pulmonary Embolism for more details.If you have sickle cell trait, you are more at risk of severe complications if you catch malaria. If you visit any area where there is a risk of malaria, it is absolutely essential that you take antimalarial tablets. You must also take precautions to avoid being bitten by mosquitoes.Symptoms of sickle cell trait in childrenYounger children are extremely unlikely to have any symptoms. They will usually be no more prone to becoming ill than children who do not have sickle cell trait. Older children and young people who do heavy physical exercise may have the same complications as adults (see above). Back to Previous Page [PDF-1.43 MB] Download and print this page pdf icon[PDF – 296 KB] People with sickle cell disease (SCD) are at greater risk than the general population for forming blood clots. A blood clot in one of the large veins, usually in a person’s leg or arm, is called a deep vein thrombosis (DVT). If a DVT is not treated, it can get bigger or break off and travel to the lungs. A blood clot in the lung is called pulmonary embolism (PE) and can cause death. Call your provider as soon as possible if you experience any of the DVT symptoms listed below. Swelling of your leg or arm Pain or tenderness in your leg or arm not caused by an injury Skin on your leg or arm that is warm to the touch, with swelling or pain Redness of the skin on your leg or arm, with swelling or pain Get medical attention immediately if you experience any of the PE symptoms listed below. Sudden, severe, and unusual shortness of breath Sudden, severe, and unusual chest pain Faster-than-normal or irregular heartbeat Coughing up blood Talk to your provider about any factors that might increase your risk for blood clots. Some of these factors are: Other risk factors include: Birth control that contains estrogen Being overweight Hormone replacement therapy that contains estrogen Family history of blood clots Trauma, particularly when the vein is injured Smoking Immobility or sitting for long periods Learn more about blood clots » Talk to your provider to get the facts about anticoagulants (commonly known as “blood thinners”). The lifesaving benefits of these medicines often outweigh the potential risks. Still, it’s important to learn about both before you start taking them. Anticoagulants are medicines that lower your risk for developing future blood clots. Here is what you need to know: If you have a blood clot for the first time without any factors that might increase your risk, your provider will prescribe you anticoagulants for life. If you have a blood clot for the first time caused by a temporary event such as a hospitalization, surgery, or being unable to move because of a fracture, your provider will prescribe you anticoagulants for 3–6 months. If you have frequent blood clots as a result of factors that might increase your risk, your provider will prescribe you anticoagulants for life. Although effective in lowering your chance of developing future blood clots, anticoagulants come with risks. For example, they can cause you to bleed more than usual when you have a cut. Learn more about anticoagulants »pdf iconexternal icon Learn more about the safe use of medicines » This information is based on ASH SCD Guidelines: Cardiopulmonary and Kidney Diseaseexternal icon. Endari was granted FDA approval to treat sickle cell, a condition that affects about 100,000 Americans.Share on PinterestAbout 100,000 people in the United States are affected by sickle cell disease.But there haven’t been any new treatments for the inherited blood disorder in decades — until now.The U.S. Food and Drug Administration (FDA) recently approved Endari (L-glutamine oral powder), which was created for people with sickle cell disease, ages 5 years and older.Researchers hope the drug can reduce severe complications associated with the ailment. Sickle cell disease affects mostly minority groups.One in every 13 African-American babies is born with sickle cell trait.That’s the gene associated with the disease, the U.S. Centers for Disease Control and Prevention (CDC) reported.Those with the condition have abnormal hemoglobin, or sickle-shaped cells.Those cells can cause blockages of blood to organs and tissues, debilitating pain, and life-threatening complications.People with sickle cell disease have an average life expectancy of 40 to 60 years. Until the approval of Endari, there was only one drug approved for people with the disease, said Dr. Richard Pazdur, acting director of the Office of Hematology and Oncology Products in the FDA’s Center for Drug Evaluation and Research, in a statement. Tiffany Rattler, a board member of the nonprofit SOS Parent and Guardian Support Group based in Texas, was pleased to hear that the disease finally has a new therapeutic option.“This is an extremely significant development, as individuals living with this particular chronic condition have had only one approved medication option before now,” Rattler told Healthline.Like other chronic conditions, a one-size treatment does not fit all with sickle cell.“The more disease-modifying treatment options available, the more likely we are to see improvements in health outcomes and quality of life for individuals living with sickle cell,” Rattler said.Whether or not the drug is an ideal therapy choice depends on the patient, but the drug at least gives people another option, Rattler added. “If the medical community continues investing resources in evidence-based research regarding these therapies, and continues to support education and awareness by sharing findings with the public, we can continue to improve the lives of individuals with sickle cell disease,” she said.Dr. James G. Taylor VI, director of the Center for Sickle Cell Disease at the Howard University College of Medicine is optimistic about the drug but does have reservations.“My biggest disappointment with the studies that led to this FDA approval is that they are not yet published in the medical literature. This information is critical for physicians like myself who are likely to prescribe this to patients,” he told Healthline.The drug seems to have a rational basis for preventing complications, but there are limited results from the phase III study and they are not published. That said, the drug could offer new hope for patients with the disease who face a shortage of qualified doctors and staggering treatment costs, Taylor said.Endari was studied in a randomized trial of people with sickle cell disease, ranging in age from 5 to 58 years.All of the study participants had two or more painful crises within a 12-month span prior to enrolling in the trial. People were assigned to take either Endari or a placebo. Scientists monitored them over the course of 48 weeks.The researchers found that the people who were given Endari had fewer hospital visits for pain, fewer hospitalizations for disease-related pain, and shorter hospital stays.They also noted that 8 percent of the participants taking Endari experienced acute chest syndrome — a serious complication of the disease — compared with 23 percent in the placebo group. As part of its approval, Endari received classification as an orphan drug, a designation that means it’s purpose is to treat rare diseases, which qualifies it for some development incentives.Emmaus Medical, Inc. produces the medication.The FDA noted that constipation, cough, nausea, headache, abdominal pain, pain in the extremities, back pain, and chest pain are common side effects of the drug. Facebook Twitter Linkedin Pinterest Sickle cell disease is an inherited blood disorder. It is marked by flawed hemoglobin. That’s the protein in red blood cells that carries oxygen to the tissues of the body. So, sickle cell disease interferes with the delivery of oxygen to the tissues.Red blood cells with normal hemoglobin are smooth, disk-shaped, and flexible, like doughnuts without holes. They can move through the blood vessels easily. Cells with sickle cell hemoglobin are stiff and sticky. When they lose their oxygen, they form into the shape of a sickle or crescent, like the letter C. These cells stick together and can’t easily move through the blood vessels. This can block small blood vessels and the movement of healthy, normal oxygen-carrying blood. The blockage can cause pain.Normal red blood cells can live up to 120 days. But, sickle cells only live for about 10 to 20 days. Also, sickle cells may be destroyed by the spleen because of their shape and stiffness. The spleen helps filter the blood of infections. Sickled cells get stuck in this filter and die. With less healthy red blood cells circulating in the body, you can become chronically anemic. The sickled cells also damage the spleen. This puts you are at greater at risk for infections.What causes sickle cell disease? Sickle cell is an inherited disease caused by a defect in a gene.A person will be born with sickle cell disease only if two genes are inherited—one from the mother and one from the father.A person who inherits just one gene is healthy and said to be a "carrier" of the disease. A carrier has an increased chance of having a child with sickle cell disease if he or she has a child with another carrier. For parents who are each carriers of a sickle cell gene, there is a 1 in 4, or a 25 % chance of having a child with sickle cell disease.What are the risk factors for sickle cell disease? Having a family history of sickle cell disease increases your risk for the disease. In the United States, it mainly affects African Americans.What are the symptoms of sickle cell disease? The following is a list of symptoms and complications associated with sickle cell disease. However, each person may experience symptoms differently. Symptoms and complications may include:Anemia. Because sickled cells are short-lived or destroyed, there are less red blood cells available in the body. This results in anemia. Severe anemia can make you feel dizzy, short of breath, and tired.Pain crisis, or sickle crisis. This occurs when the flow of blood is blocked to an area because the sickled cells have become stuck in the blood vessel. The pain can occur anywhere, but most often occurs in the chest, arms, and legs. Infants and young children may have painful swelling of the fingers and toes. Interruption in blood flow may also cause tissue death.Acute chest syndrome. This occurs when sickling occurs in the chest. This can be life-threatening. It often occurs suddenly, when the body is under stress from infection, fever, or dehydration. The sickled cells stick together and block the flow of oxygen in the tiny vessels in the lungs. It resembles pneumonia and can include fever, pain, and a violent cough.Splenic sequestration (pooling). Crises are a result of sickle cells pooling in the spleen. This can cause a sudden drop in hemoglobin and can be life-threatening if not treated promptly. The spleen can also become enlarged and painful from the increase in blood volume. After repeated episodes, the spleen becomes scarred, and permanently damaged. Most children, by age 8, do not have a working spleen either from surgical removal, or from repeated episodes of splenic sequestration. The risk of infection is a major concern of children without a working spleen. Infection is the major cause of death in children younger than age 5 in this population.Stroke. This is another sudden and severe complication of people with sickle cell disease. The misshapen cells can block the major blood vessels that supply the brain with oxygen. Any interruption in the flow of blood and oxygen to the brain can result in severe brain damage. If you have one stroke from sickle cell anemia, you are more likely to have a second and third stroke.Jaundice, or yellowing of the skin, eyes, and mouth. Jaundice is a common sign and symptom of sickle disease. Sickle cells do not live as long as normal red blood cells and, therefore, they are dying faster than the liver can filter them out. Bilirubin (which causes the yellow color) from these broken down cells builds up in the system causing jaundice.Priapism. This is a painful obstruction of the blood vessels in the penis by sickle cells. If not promptly treated, it can result in impotence.The symptoms of sickle cell disease may look like other blood disorders or medical problems. Always consult your health care provider for a diagnosis.How is sickle cell disease diagnosed? Along with a complete medical history and physical exam, you may have blood and other tests.Many states routinely screen newborns for sickle cell so that treatment can begin as soon as possible. Early diagnosis and treatment can reduce the risk of complications.Hemoglobin electrophoresis is a blood test that can determine if a person is a carrier of sickle cell, or has any of the diseases associated with the sickle cell gene.How is sickle cell disease treated? Your doctor will consider your age, overall health and other factors when determining the best treatment for you.Early diagnosis and prevention of complications is critical in sickle cell disease treatment. Treatment aims to prevent organ damage including strokes, prevent infection, and treat symptoms. Treatment may include:Pain medications. This is for sickle cell crises.Drinking plenty of water daily (8 to 10 glasses). This is to prevent and treat pain crises. In some situations, intravenous fluids may be required. Blood transfusions. These may help treat anemia and prevent stroke. They are also used to dilute the sickled hemoglobin with normal hemoglobin to treat chronic pain, acute chest syndrome, splenic sequestration, and other emergencies.Vaccinations and antibiotics. These are used to prevent infections.Folic acid. Folic acid will help prevent severe anemia.Hydroxyurea. This medication helps reduce the frequency of pain crises and acute chest syndrome. It may also help decrease the need for blood transfusions. The long-term effects of the medication are unknown.Regular eye exams. These are done to screen for retinopathy. Bone marrow transplant. Bone marrow transplants can cure some people with sickle cell disease. The decision to have this procedure is based on the severity of the disease and ability to find a suitable bone marrow donor. These decisions need to be discussed with your doctor and are only done at specialized medical centers.What are the complications of sickle cell disease? Any and all major organs are affected by sickle cell disease. The liver, heart, kidneys, gallbladder, eyes, bones, and joints can suffer damage from the abnormal function of the sickle cells and their inability to flow through the small blood vessels correctly. Problems may include the following:Increased infectionsLeg ulcersBone damageEarly gallstonesKidney damage and loss of body water in the urineEye damageMultiple organ failureLiving with sickle cell diseaseSickle cell disease is a life-long condition. Although the complications of sickle cell disease may not be able to be prevented entirely, living a healthy life-style can reduce some of the complications.It is important to eat a healthy diet with lots of fruits, vegetables, whole grains, and protein, and drink lots of fluids.Do not take decongestants because they cause constriction of blood vessels and could trigger a crisis.Other factors that may trigger a crisis include high altitudes, cold weather, swimming in cold water, and heavy physical labor.Avoid infections by getting an annual flu shot, washing your hands frequently, avoiding those who are sick, and getting regular dental exams.Key pointsSickle cell disease is an inherited blood disorder marked by defective hemoglobin.It inhibits the ability of hemoglobin in red blood cells to carry oxygen.Sickle cells tend to stick together, blocking small blood vessels causing painful and damaging complications.Sickle cell disease is treated with pain medications as needed, drinking 8 to 10 glasses of water each day, blood transfusions, and medications.Next stepsTips to help you get the most from a visit to your health care provider:Before your visit, write down questions you want answered.Bring someone with you to help you ask questions and remember what your provider tells you.At the visit, write down the names of new medicines, treatments, or tests, and any new instructions your provider gives you.If you have a follow-up appointment, write down the date, time, and purpose for that visit.Know how you can contact your provider if you have questions.