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Sickle Cell Disease (SCD)

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Section 1: Introduction

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More than 20 million people worldwide suffer from sickle cell disease. Sickle cell disease is an inherited red blood cell disorder caused by a gene mutation and is the most common genetic disorder in the United States. Sickle cell disease generally affects people of African, Mediterranean, Middle Eastern, and Hispanic descent. Sickle cell disease can cause many health complications including severe pain, stroke, blood clots, and increased risk of infections. Even though sickle cell disease is the most common genetic disorder in the United States, many healthcare providers do not have adequate knowledge in treating this disease. It is important for nurses to be aware of the serious complications and best treatment options for patients with sickle cell disease to best care for this specific patient population. In this course, participants will learn the common types of sickle cell disease and the sickle cell trait, how sickle cell disease is diagnosed and treated, and the symptoms and complications of sickle cell disease.

Section 2: What is Sickle Cell Disease?

References: 1, 2, 4, 12, 13, 14

Sickle cell disease is a genetic disorder that involves the red blood cells throughout the body. Red blood cells contain hemoglobin which deliver oxygen from the lungs throughout the body to produce energy. The energy creates waste, called carbon dioxide, which the red blood cells deliver back to the lungs to excrete. Healthy red blood cells are round shaped, but sickle red blood cells are shaped like the letter “C”. Sickle cells get their name because their shape looks like a farm tool called a “sickle”. Sickle cells often get stuck in small blood vessels because of their abnormal shape which can cause a blockage of blood flow leading to many complications. Sickle cells also have a shorter lifespan than healthy red blood cells. Healthy red blood cells have a lifespan of about 120 days,

but sickle cells only have a lifespan of 10 to 20 days. This shortened lifespan can cause a reduced number of red blood cells and decreased oxygenation throughout the body.

The Sickle Cell Trait

Sickle cell disease is a genetic disorder where children inherit two sickle cell traits from each of their parents. The sickle cell trait is known as hemoglobin S or “Hb S”. A child who inherits the sickle cell trait from one parent and a normal hemoglobin gene from the other parent will have the sickle cell trait. A child who inherits a sickle cell trait from each parent can have sickle cell disease. About 300 million people worldwide have the sickle cell trait, and it is more common in African and Middle Eastern descent.

People with the sickle cell trait may have some sickle cells, but typically do not have any symptoms or complications like those with sickle cell disease. Rarely, the sickle cell trait can cause complications such as chronic kidney disease, organ damage, and increased risk of infections. Those that have the sickle cell trait can pass it to their children and can pass along sickle cell disease to their children if both parents have the sickle cell trait. When a couple is considering having children, it is important to know if both parents have the sickle cell trait. Nurses should educate patients with the sickle cell trait about genetic counseling when considering having children to help patients understand the risks and available options.

Early diagnosis of sickle cell disease is important to prevent life threatening complications such as infection, blood clots, stroke, pain, and organ damage. The sickle cell trait can be detected on a simple blood test and is part of routine newborn screenings in the United States. Testing can also be done during pregnancy through sampling either the amniotic fluid or the placenta to determine different genetic abnormalities. These tests can be done as early as 8 to 10 weeks into the pregnancy.

Different Types of Sickle Cell Disease

There are several different types of sickle cell disease. The type of sickle cell disease depends on which of the different abnormal hemoglobin genes are inherited from the parents. The most common types of sickle cell disease include HbSS, HbSC, and HbS beta thalassemia. HbSS is the most common and the most severe type of sickle cell disease. About 65% of people with sickle cell disease have HbSS. HbSS requires both parents passing on the abnormal hemoglobin “S” gene to their child. HbSS is commonly referred to as “sickle cell anemia” because it causes chronic anemia due to the high number of sickle cells present in the body. HbSC is the type of sickle cell disease where the child inherits the abnormal hemoglobin “S” gene and the abnormal hemoglobin “C” gene. About 25% of people with sickle cell disease have HbSC and usually present with less severe symptoms and complications. HbS beta thalassemia affects about 10% of people with sickle cell disease and is when the child inherits the abnormal hemoglobin “S” gene and the abnormal beta thalassemia gene. There are two different types of beta thalassemia genes: “zero” and “plus”. People with HbS beta zero usually present with more severe symptoms and complications than people with HbS beta plus.

There are several types of rare sickle cell disease including HbSD, HbSO, and HbSE. All types include the abnormal hemoglobin “S” gene and either abnormal hemoglobin “D”, “O”, or “E” gene. These rare types all vary in severity of symptoms and complications.

Section 2 Personal Reflection

As a nurse, how could you support patients who discover their child has a severe form of sickle cell disease?

Section 2 Key Words

Sickle cell disease - An inherited genetic disorder involving the red blood cells that can cause blockages in small blood vessels.

Red blood cells - Cells that contain hemoglobin to help carry oxygen throughout the body.

Hemoglobin - A protein in red blood cells that helps carry oxygen throughout the body.

Sickle cells - Red blood cells that are "C" shaped due to a genetic mutation.

Sickle cell trait - One inherited mutated hemoglobin gene causing some sickle cells to be present in the body, but minimal symptoms and complications when compared to sickle cell disease.

Sickle cell anemia - The most common and most severe form of sickle cell disease where the child inherits an abnormal hemoglobin "S" gene from each parent, also known as "HbSS".

HbSC - A milder type of sickle cell disease where the child inherits the abnormal hemoglobin "S" gene and the abnormal hemoglobin "C" gene and affects about 25% of people with sickle cell disease.

HbS beta thalassemia - A type of sickle cell disease where the child inherits the abnormal hemoglobin "S" gene and the abnormal beta thalassemia gene and affects about 10% of people with sickle cell disease.

Section 3: Complications of Sickle Cell Disease

References: 1, 2, 3, 4, 5, 14, 15, 16

Patients with sickle cell disease deal with many chronic complications. Some symptoms can be mild, but many can be life threatening. It is important that

patients are aware of different factors that can trigger serious complications and recognize initial signs and symptoms for early treatment. Complications of sickle cell disease include:

- Infection
- Anemia
- Kidney and liver problems
- Acute chest syndrome
- Blood clots
- Stroke
- Avascular necrosis
- Dactylitis
- Priapism
- Leg ulcers
- Vision loss
- Organ damage
- Chronic pain
- Vitamin deficiency
- Delayed growth and puberty
- Cardiac problems
- Problems during pregnancy

Infection

Patients with sickle cell disease are at an increased risk for infection that can lead to serious complications. Sickle cells can cause dysfunction in the spleen, which plays an important role in fighting off infections in the body. If the spleen is not

able to fight off bacteria and viruses, the body may be more susceptible to infections. Patients with sickle cell disease are at higher risk for infections such as influenza, meningitis, pneumonia, salmonella, and staphylococcus. Infection is a leading cause of death in patients with sickle cell disease in lower income countries due to the decreased accessibility of preventative measures such as good sanitation and preventative medications. Fever is often the first sign of an infection and is especially important to watch for in children with sickle cell disease. According to the Centers for Disease Control and Prevention (CDC), infection is the leading cause of death in young children with sickle cell disease.

Anemia

Patients can suffer from chronic anemia due to the shorter lifespan of sickle cells. Anemia is an inadequate amount of red blood cells causing decreased oxygenation throughout the body. Patients with anemia may exhibit symptoms such as fatigue, dizziness, dyspnea, tachycardia, and delayed puberty. Children with sickle cell related anemia usually do not exhibit these symptoms as they are able to compensate for the lack of oxygenated blood with an increased heart rate. Adults cannot compensate as well and may need frequent blood transfusions and close monitoring to treat chronic anemia caused by sickle cell disease.

Kidney and Liver Problems

The restriction of blood flow caused by sickle cells can impact all organs in the body. The kidneys are often impacted by sickle cell disease. Restricted blood flow to the kidneys can cause organ damage, chronic kidney disease, and cancer. Symptoms of kidney damage include polyuria, bedwetting, increased blood pressure, fatigue, dyspnea, and nausea. Patients can have acute kidney damage or chronic kidney issues throughout their lives. Over time, patients with sickle cell disease are at higher risk of kidney failure and cancer. If patients have chronic kidney damage, they may need to be treated with dialysis and frequent

monitoring. It is important that patients with sickle cell disease are educated to closely monitor their urine output and blood pressure and initiate early treatment if exhibiting symptoms of kidney damage.

Sickle cells can also restrict blood flow to the liver which can cause liver damage. Lack of oxygen to the liver can happen suddenly and repeatedly. Children often have an easier time recovering from liver damage, but adults may acquire liver failure from repeated liver damage. Patients with sickle cell disease can suffer from acute sickle cell hepatic crisis when restricted blood flow from sickle cells causes acute pain in the liver.

Cholelithiasis is also a common complication of sickle cell disease. Cholelithiasis is the formation of gallstones caused by a buildup of bilirubin which is a waste that is released when red blood cells break down and is excreted through the liver. An excess amount of bilirubin can build up in the body because sickle cells break down faster than healthy red blood cells. The combination of an excess amount of bilirubin and liver damage can cause cholelithiasis.

Acute Chest Syndrome

Acute chest syndrome is a life-threatening complication caused when sickle cells restrict oxygenated blood from the lungs. Acute chest syndrome can cause dyspnea, chest pain, cough, and lead to acute respiratory distress and injury to the lungs. Acute chest syndrome is a medical emergency and accounts for 25% of deaths in adults with sickle cell disease. Long term complications of acute chest syndrome can include chronic lung disease and pulmonary hypertension.

Blood Clots and Stroke

Because sickle cells block blood flow, patients with sickle cell disease are at increased risk of blood clots and stroke. Deep vein thrombosis (DVT) is a blood clot commonly found in the legs, pelvis, and arms. DVTs can form when sickle cells

get trapped in small blood vessels. Symptoms of a DVT include swelling, pain, tenderness, and redness. DVTs travel throughout the body and cause further complications. If a DVT travels to the lungs, it can cause a pulmonary embolism which can be life threatening. Symptoms of a pulmonary embolism include tachycardia, dyspnea, chest pain, dizziness, arrhythmias, and cough.

Patients with sickle cell disease are also at an increased risk of stroke. A stroke can occur when blood flow to the brain is restricted. Symptoms of a stroke include face dropping, weakness, numbness, confusion, headaches, and difficulty with speech and vision. Children with sickle cell disease are at a much higher risk of stroke than adults. Children most commonly experience “silent strokes”. Silent strokes do not show any physical symptoms and can increase the risk for future strokes. A transcranial doppler ultrasound (TCD) can be used to detect if children are at a higher risk of having a stroke. This test measures the speed of blood flow in the brain and is recommended for children ages 2 to 16 years old with sickle cell disease. A stroke is a medical emergency and requires prompt treatment. It is important that patients with sickle cell disease understand the increased risk of stroke and take appropriate preventative measures.

Avascular Necrosis

Avascular necrosis is the death of bone tissue caused by sickle cells blocking blood flow to the bones. When bones do not get enough oxygen due to sickle cell disease, the tissue begins to die and can cause severe pain. Avascular necrosis can affect multiple joints at once and most commonly causes pain in the hip joint. Symptoms of avascular necrosis include pain and limited mobility. Patients with avascular necrosis may need chronic pain medication, surgery, or joint replacement.

Other Complications

Various types of pain can be caused by sickle cell disease. One type of sickle cell pain is dactylitis. Dactylitis is painful swelling in the hands and feet that is caused by sickle cells blocking blood flow and is common in infants and young children. Dactylitis often develops suddenly and can last up to a few weeks at a time. Priapism is a prolonged painful erection and can be caused by sickle cells restricting blood flow. Priapism can be a medical emergency if lasting more than four hours. Painful leg ulcers can also be a complication of sickle cell disease. Sickle cells restricting blood flow in the legs can cause painful open sores. This can also lead to an increased risk of infection.

Vision loss, delayed growth and puberty, vitamin deficiency, cardiac issues, and problems during pregnancy can also be complications of sickle cell disease. Sickle cells can restrict blood flow to vessels in the eyes causing vision loss. Children with sickle cell anemia can have vitamin deficiencies and delayed growth and puberty due to the decreased amount of red blood cells. Cardiac problems such as pulmonary hypertension and coronary artery disease can be caused by restricted blood flow. High blood pressure, blood clots, miscarriage, and premature births can all be complications of pregnancy in patients with sickle cell disease.

Section 3 Personal Reflection

For patients with sickle cell disease, how might their quality of life and daily activities be affected by complications of sickle cell disease?

Section 3 Key Words

Anemia - An inadequate amount of red blood cells causing chronic decreased oxygenation throughout the body.

Acute sickle hepatic crisis - A complication of sickle cell disease where restricted blood flow from sickle cells causes acute pain in the liver.

Cholelithiasis - The formation of gallstones in the gallbladder caused by a buildup of bilirubin.

Bilirubin - A waste that is released when red blood cells break down and is excreted through the liver.

Acute chest syndrome - A life-threatening complication of sickle cell disease caused when sickle cells restrict oxygenated blood from the lungs.

Deep vein thrombosis - A blood clot commonly found in the legs, pelvis, and arms that can form when sickle cells get trapped in small blood vessels.

Pulmonary embolism - A life threatening blood clot found in the lungs that can be caused by sickle cell disease.

Transcranial doppler ultrasound (TCD) - A test that measures the speed of blood flow in the brain to detect if children with sickle cell disease are at risk of having a stroke.

Avascular necrosis - A complication of sickle cell disease causing the death of bone tissue due to sickle cells blocking blood flow to the bones.

Dactylitis - A complication of sickle cell disease that causes painful swelling in the hands and feet due to sickle cells blocking blood flow.

Priapism - A complication of sickle cell disease that causes a prolonged painful erection due to sickle cells restricting blood flow.

Section 4: Sickle Cell Crisis

References: 1, 2, 3, 4, 5, 8, 17, 18

A serious complication of sickle cell disease is a sickle cell crisis. The term “sickle cell crisis” is used to describe several different conditions including:

- Vaso-occlusive crisis
- Aplastic crisis
- Hemolytic crisis
- Splenic sequestration crisis

Vaso-occlusive Crisis

A vaso-occlusive crisis is one of the most common complications of sickle cell disease. Also known as an “acute pain crisis”, a vaso-occlusive crisis is sudden severe pain caused by sickle cells restricting blood flow in different parts of the body. Patients experiencing a vaso-occlusive crisis often describe pain as sharp, stabbing, and throbbing. Patients often experience pain without warning in multiple locations at once, most commonly in the extremities, back, and chest. Patients can experience a vaso-occlusive crisis repeatedly, and often suffer from chronic pain related to sickle cell disease.

Patients must be aware of potential triggers to avoid vaso-occlusive crises. A vaso-occlusive crisis can be brought on by an unknown cause, but triggers can include:

- High altitude
- Dehydration
- Infection
- Increased stress
- Temperature changes
- Smoking
- High intensity exercise

It is important for patients to be aware of preventative measures to avoid vaso-occlusive crises. Nurses should encourage patients to drink plenty of fluids especially when experiencing warm weather or during exercise. Dehydration can cause oxygen to shift out of the bloodstream which can trigger a pain crisis. Patients should avoid high altitudes, extreme heat or cold and always dress appropriately for the weather. Extreme weather and high altitudes can cause a decrease in oxygen which can trigger a pain crisis. Patients should also be educated on the importance of avoiding tobacco use, prevention measures for infection, and reducing stress to help prevent a pain crisis.

A vaso-occlusive crisis can last several hours to several days. The pain is often so severe that patients will need to be hospitalized for monitoring and treatment. However, management of a vaso-occlusive crisis is often inadequate due to the lack of knowledge surrounding sickle cell disease and pain crises. According to Medical News Today, 20% of physicians in 2015 reported feeling uncomfortable treating patients with sickle cell disease. It is important that healthcare providers are educated on the complications of sickle cell disease and the current effective treatment strategies for this patient population.

Aplastic Crisis

An aplastic crisis occurs when the production of red blood cells suddenly drops leading to life-threatening anemia. An aplastic crisis is a common complication of sickle cell disease and can be life-threatening. The cause of aplastic crises is often attributed to an infection of parvovirus B19 or can be from an unknown cause. Symptoms of an aplastic crisis are similar to anemia including fatigue, dizziness, dyspnea, and tachycardia. Aplastic crises can cause sudden severe pain and splenic sequestration. Close monitoring and blood transfusions are needed to treat an aplastic crisis. With the appropriate treatment, patients can recover within 10 to 14 days after beginning treatment.

Hemolytic Crisis

A hemolytic crisis is similar to aplastic crisis, but instead of a rapid decrease in production of red blood cells, a large number of red blood cells are rapidly destroyed and the body cannot compensate. Hemolysis is the destruction of red blood cells in the body and can cause life-threatening anemia. Symptoms of a hemolytic crisis are similar to anemia including fatigue, dizziness, dyspnea, tachycardia, and blood in urine. Close monitoring, blood transfusions, and supplemental oxygen may be needed to treat hemolytic crises.

Splenic Sequestration Crisis

A splenic sequestration crisis is a life-threatening complication of sickle cell disease. The spleen is an organ that helps filter out waste from the blood and fight infections. Splenic sequestration can occur at any age but is most common in children under 5 years old. Sickle cells can get stuck in the spleen causing enlargement and a decreased amount of oxygenated blood flow throughout the body. This can lead to hypovolemic shock and death. Patients must be aware that a splenic sequestration crisis is a medical emergency that may require hospitalization for monitoring and treatment.

Section 4 Personal Reflection

As a nurse, what are some ways you can support your patients with sickle cell disease when dealing with a vaso-occlusive crisis?

Section 4 Key Words

Vaso-occlusive crisis - A common complication of sickle cell disease where patients experience sudden severe pain caused by sickle cells restricting blood flow throughout the body.

Aplastic crisis - The sudden drop of hemoglobin levels causing life-threatening anemia.

Hemolytic crisis - The sudden destruction of a large number of red blood cells causing life-threatening anemia.

Splenic sequestration crisis - The sudden enlargement of the spleen causing a sudden drop in hemoglobin levels.

Section 5: Treatment for Sickle Cell Disease

References: 1, 3, 4, 5, 6, 8, 11, 19

Treatment for sickle cell disease focuses on managing pain and preventing serious complications. There is currently no cure for sickle cell disease, but research is ongoing to continue to develop stem cell and gene therapy in the hopes to find a definitive cure. The goals of sickle cell disease treatment include:

- Acute and chronic pain management
- Management of sickle cell anemia
- Prevention of severe complications such as sickle cell crises, stroke, and organ damage
- Infection prevention

Blood transfusions, intravenous hydration, pain medications, infection prevention, and preventative screenings are all part of routine management of sickle cell disease. A blood transfusion is the process of intravenously transferring donated healthy red blood cells to the patient to treat many different blood disorders including sickle cell disease. Patients with sickle cell disease should be regularly monitored by healthcare providers and ensure they are adhering to appropriate preventative measures to limit the complications of sickle cell disease. According to current research, treatment recommendations for sickle cell disease include:

- Regular appointments with a primary healthcare provider
- Transcranial doppler testing in patients with sickle cell anemia ages 2 to 16 years to assess for risk of stroke
- Long-term blood transfusions for patients with increased risk of stroke
- Administer blood transfusions preoperatively in patients with sickle cell anemia
- Early initiation of opioid medications for severe pain in vaso-occlusive crises
- A combination of pain medications and physical therapy to treat avascular necrosis
- Annual vision testing to assess for vision loss

Pain Management

Pain management is a crucial part of sickle cell disease treatment. Adequate management of acute and chronic pain can allow patients with sickle cell disease to improve their quality of life. Many healthcare providers do not have adequate knowledge to properly treat a vaso-occlusive crisis and patients with sickle cell disease often suffer from inadequate pain management.

For mild pain during a vaso-occlusive crisis, patients can focus on hydration and the use of non-pharmacological therapies and non-prescription pain medications such as acetaminophen and ibuprofen. Non-pharmacological pain management can include massage, rest, and distraction. Research shows that prompt initiation of pain medications for a vaso-occlusive crisis can provide the best outcomes for patients with sickle cell disease. Patients experiencing severe pain during a vaso-occlusive crisis may need to be hospitalized for opioid medications, intravenous fluids, and blood transfusions. Opioid medications including morphine and oxycodone should be used as first-line therapy for severe pain. The National Heart, Lung, and Blood Institute recommends that pain medications should be started within 30 to 60 minutes of triage for patients who are hospitalized with a

vaso-occlusive crisis. Many hospitals have a sickle cell disease protocol for addressing vaso-occlusive crises and closely monitor patients during hospitalization. Patients should be frequently reassessed during a vaso-occlusive crisis to ensure their pain is being adequately treated.

There are several new medications approved to treat a vaso-occlusive crisis. Hydroxyurea (Droxia, Hydrea, Siklos) is now regularly used to reduce the frequency of vaso-occlusive crises. Hydroxyurea can also reduce the need for frequent blood transfusions to treat sickle cell anemia, lower the risk of acute chest syndrome, and lower the risk of dactylitis in children. Hydroxyurea is recommended for patients with sickle cell disease who meet criteria including:

- Experiencing at least 6 vaso-occlusive crises per year
- History of acute chest syndrome
- History of stroke or a high risk of stroke
- Severe sickle cell anemia
- Severe unrelieved chronic pain

A potential side effect of hydroxyurea is that it can increase the risk of infection. Patients taking hydroxyurea must be closely monitored by a healthcare provider to ensure adequate pain management and for signs of infection. Crizanlizumab-tmca (Adakveo) is another medication approved by the Food and Drug Administration (FDA) in 2019 for sickle cell disease in patients older than 16 years of age. Crizanlizumab-tmca helps to reduce pain during a vaso-occlusive crisis by preventing sickle cells from blocking blood flow and improving oxygenation. Crizanlizumab-tmca does have some side effects including nausea, joint pain, back pain, and fever. L-glutamine (Endari) is another medication approved by the FDA in 2017 and helps reduce the frequency of vaso-occlusive crises.

Chronic pain is another complication of sickle cell disease. Patients suffering from chronic pain relating to sickle cell disease can use a combination of opioid medications, non-prescription medications, and non-pharmacological pain

interventions to treat chronic pain. Non-pharmacological interventions such as physical therapy, acupuncture, massage, and support groups can be beneficial in managing chronic pain. Non-prescription pain medications such as ibuprofen and acetaminophen can help treat chronic pain, but often these medications do not match the severity of the pain. Opioid medications can be effective in treating severe chronic pain, but patients must be aware of serious side effects. The chronic use of opioid medications puts patients at a high risk of developing dependence and tolerance to these medications, as well as opioid misuse. Opioids are highly addictive medications and can have serious complications if not monitored closely.

It is important for healthcare providers to educate patients on their options for treating pain related to sickle cell disease. Patients should be aware of the benefits and risks of different pain medications and the importance of seeking medical treatment early when experiencing a vaso-occlusive crisis.

Blood Transfusion Treatment

Blood transfusions can be beneficial in treating various complications of sickle cell disease. Blood transfusions are first-line therapy for sickle cell anemia and can be effective treatment for:

- Risk of stroke
- Vaso-occlusive crisis
- Aplastic crisis
- Hemolytic crisis
- Splenic sequestration crisis
- Acute chest syndrome
- Preoperative prophylaxis
- Complications in pregnancy

- Organ failure
- Unresolved priapism

Patients must be hospitalized to receive blood transfusion treatments and often need frequent blood transfusions to help manage symptoms. Patients receiving frequent blood transfusions must be monitored closely to watch for side effects. Adverse effects of blood transfusions can include infection, excess iron buildup which can cause damage to organs, and an immune response to the donor blood.

Infection Prevention

Infection prevention is an important part of managing sickle cell disease especially in children. In addition to watching for early signs of infection, healthcare providers can educate patients on other preventative measures such as handwashing and prophylactic medications. A daily dose of penicillin in children with sickle cell disease up to 5 years of age and in adults with sickle cell disease who have a history of pneumonia or a splenectomy. It is important that patients are educated on the importance of vaccination compliance. Children should receive all recommended vaccinations according to the American Academy of Pediatrics and vaccinations against pneumonia, meningitis, hepatitis B, and influenza. Adults should adhere to vaccination recommendations including annual influenza and pneumonia vaccinations.

New Treatment Options

Current research is creating many new opportunities for sickle cell disease treatment. Voxelotor (Oxbryta) is a medication approved by the FDA in 2019 for patients older than 12 years of age. This medication helps lower the risk of anemia and improve blood flow throughout the body by preventing red blood cells from forming the sickle shape. Voxelotor and other medications for sickle cell disease

have contributed to improving the quality of life for patients who suffer from this lifelong disease.

The newest development in sickle cell disease treatment is the approval of gene therapy for patients older than 12 years of age. Exagamglogene autotemcel (Casgevy) and lovotibeglogene autotemcel (Lyfgenia) are two medications that have the potential to greatly reduce the occurrence of a vaso-occlusive crisis and other complications of sickle cell disease. Gene therapy involves restoring an abnormal or missing gene to improve how the cells function. These new genes are developed in a laboratory setting and are a new form of sickle cell disease treatment. The hope is that continued research and future gene therapies may one day be able to provide a cure for sickle cell disease.

Stem cell transplant is another new treatment for sickle cell disease. Stem cell transplant, or bone marrow transplant, involves replacing the bone marrow affected by sickle cell disease with healthy bone marrow from a donor. Bone marrow is the soft tissue in bones where red blood cells are created. Replacing sickle cell bone marrow with healthy donor cells can ensure that the body does not continue to produce sickle cells. This can greatly decrease complications of sickle cell disease. There are strict criteria for a stem cell transplant due to the potential serious adverse effects. Results of stem cell transplants have been shown to be better in pediatric patients with sickle cell disease. Indications for a stem cell transplant include:

- Severe sickle cell anemia
- Stroke
- Recurrent acute chest syndrome
- Recurrent vaso-occlusive crisis
- Recurrent priapism
- Organ failure

It is important that patients are educated on the serious risks of stem cell transplant. Adverse events can include severe infection, rejection of donor cells, organ damage, infertility, and effects on existing conditions such as cardiac, lung, and kidney disease. Stem cell transplants can also be very expensive and are not guaranteed to work. Patients must undergo extensive testing to find a match for the transplant, and must undergo prophylactic treatment to prepare the body for the donor cells. The benefits and risks should be carefully considered when determining if stem cell transplant is a viable treatment option for sickle cell disease.

Section 5 Personal Reflection

What benefits versus risks should be considered when determining appropriate pain management for a patient with sickle cell disease?

Section 5 Key Words

Blood transfusion - The process of intravenously transferring donated healthy red blood cells to the patient to treat many different blood disorders including sickle cell disease.

Opioid medications - Prescription medications used to treat severe pain and include morphine, hydrocodone, and fentanyl.

Hydroxyurea (Droxia, Hydrea, Siklos) - A medication used to reduce the frequency of vaso-occlusive crises, the need for frequent blood transfusions to treat sickle cell anemia, lower the risk of acute chest syndrome, and lower the risk of dactylitis in children.

Crizanlizumab-tmca (Adakveo) - A medication approved in 2019 to reduce pain during a vaso-occlusive crisis by preventing sickle cells from blocking blood flow and improving oxygenation.

L-glutamine (Endari) - A medication approved in 2017 to help reduce the frequency of vaso-occlusive crises.

Voxelotor (Oxbryta) - A medication approved in 2019 to help lower the risk of anemia and improve blood flow throughout the body by preventing red blood cells from forming the sickle shape.

Gene therapy - The process of restoring an abnormal or missing gene to improve how the cells function.

Stem cell transplant - The process of replacing the bone marrow affected by sickle cell disease with healthy bone marrow from a donor to ensure that the body does not continue to produce sickle cells; also known as bone marrow transplant.

Bone marrow - The soft tissue in bones where red blood cells are created.

Section 6: Case Study #1

A nurse is caring for a 30-year-old patient who is pregnant with their first child. The patient is concerned that the child will have sickle cell disease because the patient carries the sickle cell trait. The patient reports no symptoms of sickle cell disease throughout their life and states they were told the sickle cell trait showed up on a routine blood test when they were a child. The patient states “I do not want to give my baby a lifelong disease. I have seen other people suffer from sickle cell disease and I do not want my baby to suffer.”

1. What education can the nurse provide to the patient regarding how sickle cell disease is inherited?
2. What is important for the patient to understand regarding the sickle cell trait versus sickle cell disease?
3. What options can the nurse provide to the patient to test the baby for sickle cell disease?

Section 7: Case Study #1 Review

This section will review the case studies that were previously presented in each section. Responses will guide the clinician through a discussion of potential answers as well as encourage reflection.

1. What education can the nurse provide to the patient regarding how sickle cell disease is inherited?

   Children must inherit two sickle cell traits from their parents. If the child inherits the sickle cell gene from one parent and a normal hemoglobin gene from the other parent, they will have the sickle cell trait. If the child inherits a sickle cell gene from each parent, they can have sickle cell disease. It is important to know if both parents carry the sickle cell trait. If the patient is the only parent who carries the sickle cell trait, their child will not inherit sickle cell disease.

2. What is important for the patient to understand regarding the sickle cell trait versus sickle cell disease?

   Having the sickle cell trait does not mean you have sickle cell disease. People with the sickle cell trait may have some sickle cells, but typically do not have any symptoms or complications like those with sickle cell disease. Those that have the sickle cell trait can pass it to their children. There are different types of sickle cell disease depending on the type of sickle cell gene inherited from each parent. For example, if both parents pass on hemoglobin “S” gene, the child will have HbSS which is the most severe form of sickle cell disease. Other types of sickle cell disease may not be as severe.

3. What options can the nurse provide to the patient to test the baby for sickle cell disease?

Early diagnosis of sickle cell disease is important to prevent life threatening complications such as infections, stroke, and organ damage. The sickle cell trait can be detected on a simple blood test and is part of routine newborn screenings in the United States. Testing can also be done during pregnancy through sampling either the amniotic fluid or the placenta to determine different genetic abnormalities. These tests can be done as early as 8 to 10 weeks into the pregnancy.

Section 8: Case Study #2

A patient arrives in the emergency department complaining of dizziness, shortness of breath, and weakness. Upon further examination, the nurse discovers that the patient is tachycardic and hypotensive. The patient states they have a history of sickle cell disease and a deep vein thrombosis (DVT) several years ago.

1. What complication of sickle cell disease is most likely for this patient?
2. What education should the nurse provide to the patient about potential complications of sickle cell disease?

Section 9: Case Study #2 Review

This section will review the case studies that were previously presented in each section. Responses will guide the clinician through a discussion of potential answers as well as encourage reflection.

1. What complication of sickle cell disease is most likely for this patient?

   The patient is most likely experiencing anemia due to the shorter lifespan of sickle cells creating an inadequate amount of red blood cells in the body. Symptoms of anemia include fatigue, dizziness, dyspnea, tachycardia, and delayed puberty. The patient should also be assessed for a DVT since they

have a previous history and patients with sickle cell disease are at an increased risk for DVTs. Deep vein thrombosis (DVT) is a blood clot commonly found in the legs, pelvis, and arms. DVTs can form when sickle cells get trapped in small blood vessels and can travel throughout the body and cause further complications. If a DVT travels to the lungs, it can cause a pulmonary embolism which can be life threatening.

2. What education should the nurse provide to the patient about potential complications of sickle cell disease?

   The nurse should educate the patient about signs of anemia including fatigue, dizziness, dyspnea, tachycardia, and delayed puberty. The patient should know that anemia can turn into a sickle cell crisis and become life threatening. The patient should seek medical treatment immediately and may require frequent blood transfusions and close monitoring to treat chronic anemia. The patient should also be aware of the risk for another DVT. The patient should know the signs of a DVT including swelling, pain, tenderness, and redness.

Section 10: Case Study #3

A patient with sickle cell disease arrives in the emergency room complaining of severe pain in their back and chest. The patient states they took two doses of ibuprofen at home without any pain relief. The nurse notes that this is the patient's fourth time in 3 months to the emergency room for severe pain.

1. What type of sickle cell crisis is the patient experiencing?
2. What education can the nurse provide to the patient regarding preventative measures for a sickle cell crisis?

Section 11: Case Study #3 Review

This section will review the case studies that were previously presented in each section. Responses will guide the clinician through a discussion of potential answers as well as encourage reflection.

1. What type of sickle cell crisis is the patient experiencing?

   The patient is experiencing a vaso-occlusive crisis or an “acute pain crisis”. A vaso-occlusive crisis is sudden severe pain caused by sickle cells restricting blood flow in different parts of the body. Patients experiencing a vaso-occlusive crisis often describe pain as sharp, stabbing, and throbbing. Patients often experience pain without warning in multiple locations at once, most commonly in the extremities, back, and chest. Patients can experience a vaso-occlusive pain crisis repeatedly, and often suffer from chronic pain related to sickle cell disease.

2. What education can the nurse provide to the patient regarding preventative measures for a sickle cell crisis?

   Patients must be aware of potential triggers to avoid vaso-occlusive crises. A vaso-occlusive crisis can be brought on by an unknown cause, but many times triggers include high altitude, dehydration, infection, increased stress, temperature changes, smoking, and high intensity exercise. It is important for patients to be aware of preventative measures to avoid vaso-occlusive crises. The nurse can encourage the patient to drink plenty of fluids especially when experiencing warm weather or during exercise. Dehydration can cause oxygen to shift out of the bloodstream which can trigger a pain crisis. Patients should avoid high altitudes, extreme heat or cold and always dress appropriately for the weather. Extreme weather and high altitudes can cause a decrease in oxygen which can trigger a pain crisis. Patients should also be educated on the importance of avoiding tobacco

use, prevention measures for infection, and reducing stress to help prevent a pain crisis.

Section 12: Case Study #4

A nurse is caring for a patient with sickle cell disease. The patient is diagnosed with “HbSS” sickle cell disease and has had many complications over the past few years. The patient has just learned that they are at an increased risk of stroke and will need to come back every month for a blood transfusion. The patient states “I am so tired of being in the hospital for pain management and blood transfusions. I wish there was a magic pill that I could take that would cure me of my sickle cell disease.”

1. What medications for sickle cell disease may reduce the patient’s need for hospitalizations and blood transfusions?
2. What education should the nurse provide to the patient about new treatment options for sickle cell disease?

Section 13: Case Study #4 Review

This section will review the case studies that were previously presented in each section. Responses will guide the clinician through a discussion of potential answers as well as encourage reflection.

1. What medications for sickle cell disease may reduce the patient’s need for hospitalizations and blood transfusions?

   Blood transfusions, intravenous hydration, pain medications, infection prevention, and preventative screenings are all part of routine management of sickle cell disease. Hydroxyurea (Droxia, Hydrea, Siklos) is now regularly used to reduce the frequency of vaso-occlusive crises. Hydroxyurea can also

reduce the need for frequent blood transfusions to treat sickle cell anemia, lower the risk of acute chest syndrome, and lower the risk of dactylitis in children. Crizanlizumab-tmca (Adakveo) helps to reduce pain during a vaso-occlusive crisis by preventing sickle cells from blocking blood flow and improving oxygenation. Crizanlizumab-tmca does have some side effects including nausea, joint pain, back pain, and fever. L-glutamine (Endari) was approved by the FDA in 2017 and helps reduce the frequency of vaso-occlusive crises. Voxelotor (Oxbryta) helps lower the risk of anemia and improve blood flow throughout the body by preventing red blood cells from forming the sickle shape.

2. What education should the nurse provide to the patient about new treatment options for sickle cell disease?

   The newest development in sickle cell disease treatment is the approval of gene therapy for patients older than 12 years of age. Exagamglogene autotemcel (Casgevy) and lovotibeglogene autotemcel (Lyfgenia) are two medications that have the potential to greatly reduce the occurrence of a vaso-occlusive crisis and other complications of sickle cell disease. Stem cell transplant is another new treatment for sickle cell disease. Stem cell transplant, or bone marrow transplant, involves replacing the bone marrow affected by sickle cell disease with healthy bone marrow from a donor. There are strict criteria for a stem cell transplant due to the potential serious adverse effects and it is important that patients are educated on the serious risks. Adverse events can include severe infection, rejection of donor cells, organ damage, infertility, and effects on existing conditions such as cardiac, lung, and kidney disease. Stem cell transplants can also be very expensive and are not guaranteed to work. Patients must undergo extensive testing to find a match for the transplant, and must undergo prophylactic treatment to prepare the body for the donor cells. The benefits and risks should be carefully considered when determining if stem cell transplant is a viable treatment option for sickle cell disease.

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