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Preventing and Treating Complications of Sickle Cell Disease in Pediatric Patients

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by

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Preventing and Treating Complications of Sickle Cell Disease in Pediatric Patients

Sickle cell anemia, also known as sickle cell disease, affects over 90,000 people in the United States alone with those of African descent being at greater risk (Dobson & Byrne, 2014). Worldwide, it affects over 30 million people (Khoury, Musallam, Mroueh, & Abboud, 2011). It is usually detected during first year of life. After that, it is a lifelong disease (Hollins et al., 2012). Patients with sickle cell disease in the United States are expected to live until they are approximately 48 years old (BMJ Group, 2011). It is classified as an autosomal recessive disorder of the blood. A child inherits abnormal hemoglobin from both their mother and father which causes their red blood cells to become distorted or sickled (Shiel, 2014). The hemoglobin that is abnormal is known as hemoglobin S or Hb S.

Although the terms sickle cell anemia and sickle cell disease are often used interchangeably, sickle cell anemia is actually the most common type of sickle cell disease. Sickle cell anemia is the most common type of sickle disease and is caused by the homozygous state or Hb SS (Musumadi, Westerdale, & Appleby, 2012). Hb SS makes the red blood cell more "susceptible to be modulated by stress, hypoxia, and by the inflammatory response" (Odièvre, Verger, Silva-Pinfo, & Elion, 2011, p. 533).

Sickle cell disease can cause many problems in individuals. Some of these problems include anemia, pain, stroke, and severe infections (Thompson & Steinberg, 2012). These individuals are frequent patients in the hospital. They often seek treatment after experiencing a sickle cell crisis. One of the main focuses for treating sickle cell disease is hydration. This also helps with the prevention of sickle cell crises. However, there are many more options for treating sickle cell disease which depends on what the patient's presenting symptoms are.

In order to effectively treat and manage sickle cell disease and prevent sickle cell crisis, it is imperative that a nurse understands the possible complications associated with the disease. After analyzing which complications the patient is at risk for or are already having, the nurse can then properly intervene. Proper nursing interventions should be based on evidence based practice. It is especially important to be able to think critically when caring for pediatric clients as they may not be able to convey their symptoms as most adults would. Pediatric patients may also be more susceptible for more complications or more severe complications because their bodies are not fully developed. In order to prevent further complications in pediatric patients and lower the mortality rate, nurses must use the most effective interventions and must also intervene quickly.

Pain

Pain is one of the most common symptoms experienced by patients with sickle cell disease. It is the main reason sickle cell patients come to the hospital for treatment. When deoxygenated, the abnormal hemoglobin causes the red blood cell to become sickled or crescent shaped. This is due to crystallization and polymers being formed (Musumadi et al., 2012). When the cell becomes sickled, it leads to the occlusion of small blood vessels because the red blood cells are not able to easily pass through the blood vessels. The limitation of blood flow can lead to ischemia, or insufficient blood supply to the organs. A painful vaso-occlusive crisis occurs when the sickle cells become attached to endothelium and causes inflammation and tissue damage. Pain experienced with sickle cell disease can be either acute or chronic or a mixture of both. Pain may also be classified as neuropathic.

Acute Pain

The type of pain associated with the majority of hospitalizations is acute. This may also be referred to as an acute crisis. It often occurs after increased emotional stress, cold weather, asthma, extreme temperature changes, dehydration, and/or infection (Myers & Eckes, 2012). It may also be brought on by alcohol intoxication, pregnancy, hypoxia, acidosis, and hypovolemia (Maakaron & Taher, 2013). A crisis warrants for prompt medical management (Myers & Eckes, 2012). Acute pain experienced with sickle cell disease can be compared with pain associated with cancer. Although there are some precipitating factors, pain is often unpredictable (Dobson & Byrne, 2014). The most common first sign of acute crisis presents as dactylitis, or pain and inflammation in the fingers and toes. Pain may be present in any part of the body especially the joints, abdomen, bones and soft tissue. It can last for several hours (Maakaron & Taher, 2013). In order to manage acute pain, nurses should encourage their patients to routinely follow up with their healthcare provider and, if possible, the same healthcare provider should be seen each time (Ballas, 2011).

Because sickle cell disease affects the blood flow to various organs, the patient should also be seen by a multidisciplinary team. This is a way to make sure the patient receives proper assessments and is treated in the most effective way. Specialist may collaborate in order to come up with the best plan of care for the patient. While in the hospital, the patient may need to receive medication to control their pain. Intravenous medications can be challenging to give to sickle cell disease patients because of dehydration, decreased blood flood, multiple accesses and veins that are difficult to cannulate. It has been proposed to administer patient controlled analgesics, or PCA, subcutaneously rather than through an intravenous line (Myers & Ekes, 2012). Although there is not much recent research, some hospitals have been noted to use diamorphine in subcutaneous infusions in order to manage pain in sickle cell disease. It can be

used as an alternate to intermittent injections as both have the same effectiveness. Subcutaneous infusions have been found to be better than using non-steroidal anti-inflammatory drugs, (NSAIDs). NSAIDs should not be used for long periods as they worsen complications of sickle cell disease (Okpala & Tawil, 2002). Although PCA may be better than NSAIDs, it still has many side effects and complications including respiratory distress and acute chest syndrome. Patients' respiratory statuses should be assessed frequently when being administered any type of opioids.

There are pain managements that do not have any side effects reported. Guided imagery is being tested as a way to manage acute pain episodes in sickle cell disease patients (Dobson & Byrne, 2014). Guided imagery can be defined as "any of various techniques…used to guide another person or oneself in imagining sensations—especially in visualizing an image in the mind—to bring about a desired physical response" (National Center for Complementary and Alternative Medicine, 2013). Although it is typically used to reduce stress and anxiety, it can also be used to lower blood pressure and manage pain (American Cancer Society, 2008). This type of management can be very beneficial because there are no possible side effects, allergic reactions, or interactions with medications or foods. One downfall of guided imagery use with children is that the child must be able to learn how to use guided imagery. In a study performed on children aged six to eleven, it was found that guided imagery decreased the number of pain episodes while also decreasing the pain intensity. The children did not use as many pain medications. Many of the children did not have to use pain medications at all. If a child did use pain medication, they only used ibuprofen versus all of the opioids that they had prescribed to them. Due to the decrease in pain, the children were able to attend school more (Dobson & Byrne, 2014). Guided imagery should be used as the main prevention and treatment of pain

experienced in children with sickle cell disease. It has been proven effective in decreasing pain. If a child can use guided imagery, it can help to prevent further complications that are induced or worsened by pain medications. The child should have access to pain medication on an as needed basis for pain that becomes too severe or is unrelieved by guided imagery. Guided imagery is also cost effective because pain medication does not have to be ordered as frequently and complications do not have to be treated. However, patients should still routinely follow up with their health care providers for evaluation. If their pain becomes too severe, other pain management options should be explored for that patient. Bed rest is also important during a crisis whether or not the patient is using guide imagery and/or other pain management options (Thompson & Steinberg, 2012).

Chronic Pain

Chronic pain is characterized as three or more months of continuous pain (Ballas, 2011). It is often felt in the bones and joints (Maakaron & Taher, 2013). There are two main causes of chronic pain. One cause is from factors that can be seen such as necrosis or leg ulcers. The second cause is frequent, severe acute crises especially when left untreated which leads to blood vessel damage. Both of these result in a nagging, achy, and/or deep pain that does not go away (Ballas, 2011).

In order to prevent a patient from having chronic pain or to at least prolong chronic pain from happening, patient should be seen and treated in a hospital for each acute crisis that they incur. The patient should also try to prevent acute crises from happening by staying hydrated and practicing stress relief exercises. Some studies have also shown that there is a correlation between vitamin D deficiency and chronic pain (Osunkwo et al., 2012). Vitamin D is a fat soluble vitamin that can be acquired from some foods and ultraviolet rays. A serum 25-

hydroxyvitamin D level of less than twelve nanograms per milliliter is associated with vitamin D deficiency. Children are recommended to have an intake of 400 to 600 IU or 10 to 15 micrograms of vitamin D daily (Office of Dietary Supplements, 2011). People with sickle cell disease are more at risk to develop vitamin D deficiency (Rovner et al., 2008). Patients whose vitamin D levels are low and are not receiving vitamin D supplementation tend to have more pain days than those that receive high doses of vitamin D (Osunkwo et al., 2012). More recent studies have not found any correlations between vitamin D deficiency and chronic pain in sickle cell disease patients. However, all the patients in this particular study did not receive a high dose of vitamin D; they all just experienced some form of vitamin D deficiency (Jackson, Krauss, DeBaun, Strunk, & Arbeláez, 2012). With this being said, there is a possibility that the patients' pain days may have decreased if they had received a vitamin D supplementation during this study. Even so, high doses of vitamin D as a preventative and/or treatment for chronic pain needs to be further studied in order to get a conclusive result before implementation. Although supplementing vitamin D may not treat chronic pain experienced in those with sickle cell disease, nurses should still encourage their patients to have an adequate intake of vitamin D or get supplementation in order to avoid complications of vitamin D deficiency such as bone fragility, rickets, hypertension and type II diabetes (Kovacs & Stöppler, 2013). Chronic pain prevention and treatment also includes the ones associated with acute pain such as pain medications and guided imagery. Patients should follow up with their healthcare providers to find a regimen that works best for them. During hospital stays, patients should be monitored carefully especially when using opioids. Nurses need to report any signs and symptoms of adverse effects so that changes may be made if necessary.

Neuropathic Pain

Patients may also experience neuropathic pain. Neuropathic pain can be "defined as pain primarily initiated by dysfunction of the peripheral or central nervous system" (Brandow, Farley, & Panepinto, 2014, p. 512). Neuropathic pain is "persistent pain resulting from damage to the peripheral or central nervous system (CNS) or abnormal communication within the nervous system" (Wang, Wilkie, & Molokie, 2010, p. 403). Symptoms associated with neuropathic pain include tingling, numbness, and burning and shooting pain (Cassoobhoy, 2013). It may be caused by tissue damage as result of vaso-occlusion. It can also be due to the treatment of chronic pain from the activation of neuroglia (Ballas, 2011). Neuroglia are "[n]onneuronal cellular elements of the central and peripheral nervous system; thought to have important metabolic functions" (Farlex, 2012).

In order to prevent patients from experiencing neuropathic pain, nurses should be cautious when administering medications for pain. Nurses should also make sure their patients stay hydrated as a way to prevent tissue damage which can lead to neuropathic pain. Whenever a patient experiences an acute crisis, they should be treated quickly in order to prevent irreversible tissue damage (Ballas, Gupta, & Adams-Graves, 2012). Treating neuropathic pain with pain medications should be avoided as they can worsen it. Possible treatment options for neuropathic pain include electrical stimulation, physical therapy, acupuncture, and relaxation therapy (Cassoobhoy, 2013). Nurses should communicate with both the patient and the healthcare provider in order to determine the best treatment for the patient. If the patient has to take pain medication for acute or chronic pain, the nurse should educate the patient on signs and symptoms of worsening neuropathic pain. The patient should be instructed to notify their healthcare provider if they start experiencing worsening neuropathic in order to avoid serious disabilities (Cassoobhoy, 2013).

Infection

Another possible complication associated with sickle cell disease is infection. Patients with sickle cell disease are at greater risk for acquiring infections due to spleen damage. Pneumonia is one of the main causes of death among infants and young children with sickle cell disease (Centers for Disease Control and Prevention, 2014). In order to prevent infection, a child will begin taking penicillin twice daily before they are two months old. This prophylactic dosing will continue until the child is at least five years old (Adis Medical Writers, 2013). Vaccines should also be given and kept up to date. This especially includes those for influenza, meningococcal disease, pneumonia, and any other that are recommended by the health care provider. Proper hand washing done by the patient, family, and nurse can also prevent the patient from getting an infection. The nurse should also provide education on food safety to avoid salmonella. This includes washing fruits and vegetables, thoroughly cooking meat, only eating pasteurized dairy products, and not eating raw eggs or products with raw eggs. Reptiles may also have salmonella so patients should be advised to stay away from turtles, snakes, and lizards. Patients and parents should be encouraged to contact or visit their health care provider if they have any signs of infection (Centers for Disease Control and Prevention, 2014). Children presenting with a fever should be seen by their health care provider as they may have an infection or are at a greater risk for acquiring an infection (Bansil, Kim, Tieu, & Barcega, 2013). Infections need to be treated as early as possible with antibiotics in order to prevent further complications (Centers for Disease and Control and Prevention, 2014).

Acute Chest Syndrome

Another complication of sickle cell disease is acute chest syndrome. Acute chest syndrome is a group of symptoms associated with sickle cell disease. It is characterized by

severe pain, pulmonary infiltrates, chest pain, leukocytosis, tachycardia, and tachypnea (Myers & Eckes, 2012). Pediatric patients with acute chest syndrome may also experience fever, cough, and wheezing. It is another reason why people with sickle cell disease are hospitalized and also the leading cause of death among patients with sickle cell disease (Khoury et al., 2011). Pediatric patients are more likely to get acute chest syndrome especially between the ages of two and four. Patients are also at risk three days following surgery (Nasser & Hassan, 2011). Acute chest syndrome happens during acute crisis when the bone marrow is involved which causes fat embolisms inside of the lungs (Myers & Eckes, 2012). Fat embolisms are the number one cause of acute chest syndrome in adults (Nasser & Hassan, 2011). After a bone marrow infarction, bone marrow contents can create pulmonary emboli (Khoury et al., 2011). If a sickle cell patient is experiencing bone pain, they likely have a bone marrow infarction and should be monitored for acute chest syndrome (Maakaron & Taher, 2013). Acute chest syndrome may also be a result of infections such as those caused by Mycoplasma pneumoniae and Streptococcus pneumoniae. Infection is the number one cause of acute chest syndrome in children (Nasser & Hassan, 2011). Children should be monitored for signs and symptoms of infections. Nurses should take precautions in order to decrease the amount of germs spread to prevent the child from acquiring an infection. If the patient is taking hydroxyurea, they should have their white blood cell count monitored as it may lowered which puts them at risk for infection (Centers for Disease Control and Prevention, 2014). Antibiotics should be used to treat the infection (Thompson & Steinberg, 2012).

Morphine use in the treatment for acute crisis has been identified as a risk for developing acute chest syndrome (Birken et al., 2013). Having too much of an opioid analgesic can cause acute chest syndrome due to it depressing the respiratory system (Ballas, Kesen, et al., 2012).

Taking this into account, it is imperative that nurses monitor their patients who are receiving morphine or any opioid analgesic. Routine respiratory assessments should be done in order to prevent acute chest syndrome. If possible, opioid analgesics should only be used when needed. They should not be used in mass amounts or as a preventative pain management as they can precipitate acute chest syndrome. Other treatments, such as guided imagery, should be explored for pain as to avoid further respiratory complications. If it is absolutely necessary that a patient receives an opioid analgesic such as morphine, it should be used carefully. If the patient's respiratory rate falls below the normal range, the opioid analgesic should be discontinued. Pediatric patients should have respirations between twelve and sixty depending on their age (New York State Health Department, n.d.). Opioid antagonist such as Narcan should be kept on hand in order to reverse any possible adverse effects. Patients receiving opioids or patients that recently had opioids prior to acquiring acute chest syndrome should be encouraged to do deep breathing (Thompson & Steinberg, 2012).

For preventive measures, studies have proven that "specified patient monitoring with criteria for the initiation of supplemental oxygen; scheduled incentive spirometry; patient ambulation after the first 24 hours; opioid type and dosing schedule, maintenance and exacerbation asthma orders, and opioid titration and weaning guidelines" will decrease the rate of patients with acute chest syndrome when being managed for pain (Reagan, DeBaun, & Frei-Jones, 2011, p. 263). One of the main focuses for treating acute chest syndrome is maintaining ventilation by incentive spirometer for mild cases or mechanical ventilation for severe cases (Ballas, Kesen, et al., 2012). The use of the incentive spirometer is especially important for patients who are on bed rest or who had a recent surgery (Centers for Disease Control and Prevention, 2014). Pain management, oxygen therapy, and blood transfusions may also be

indicated for treatment (Thompson & Steinberg, 2012). Corticosteroids, however, should be avoided as they may prolong hospital stay and increase the risk for readmission. Bronchodilators may be used for patients that are wheezing or also have asthma (Khoury et al., 2011). The nurse needs to keep track of the patient's intake and output due to the possibility of fluids building up in the lungs (Thompson & Steinberg, 2012).

Renal Complications

Children with sickle cell disease may also suffer from renal complications due to sickling. One common renal complication is hyposthenuria (Musumadi et al., 2012). Hyposthenuria is the "[e]xcretion of urine of low specific gravity due to an inability of the tubules of the kidneys to produce concentrated urine" (Dictionary.com, 2002). This can cause the patient to excrete large amounts of urine which leads to dehydration. Dehydration may cause the patient to go into a vaso-occlusive crisis (Musumadi et al., 2012). It is important for the nurse to assess their patient of signs and symptoms of dehydration. The nurse should also educate their patient on proper hydration. Patients may also experience glomerular hyperfiltration during their adolescent years (Adis Medical Writers, 2013). As a nurse, one needs to monitor their patient's kidney function. Glomerular hyperfiltration may result in renal failure later in life (Paula, Nascimento, Sousa, Bastos, & Barbosa, 2013). Non-steroidal anti-inflammatory may prevent glomerular hyperfiltration; however, it should not be used because it can cause adverse effects in relation to the kidneys. Patients with sickle cell disease may also have hematuria and proteinuria, which has the possibility of leading to renal failure. The patient should have urinalyses done to determine if there is any blood or protein in the urine. If a patient presents with hematuria, the nurse has to make sure the patient stays on bed rest and maintains adequate hydration. In order to decrease proteinuria, the patient will be prescribed angiotensin-converting

enzyme inhibitors or angiotensin receptor blockers. Both have been proven to reduce proteinuria and should be used even if the patient does not have hypertension (Lerma & Batuman, 2012). However, if the patient is receiving angiotensin-converting enzyme inhibitors, serum potassium level should be monitored because they are at risk for developing hyperkalemia (Becker, 2011). The administration of supplemental vitamin E may also be used due to its antioxidant effects. If the patient does progress to end stage renal failure, a kidney transplant may be considered. Transplant recipients should have their fluid intake and output strictly monitored as they are more at risk to have acute sickle cell crises (Lerma & Batuman, 2012). Nurses need to make sure their patients receiving transplants maintain adequate hydration in order to avoid these crises especially after episodes of vomiting and/or diarrhea.

Complications of the Eyes

Sickle cell patients also have the possibility of acquiring vision problems. Cells that are sickled can become lodged in the blood vessels of the retina (Thompson & Steinberg, 2012). This can lead to partial or complete blindness. In severe cases, retinal hemorrhaging can occur and the retina can become detached which typically leads to permanent blindness (Shiel, 2014). Retinal detachment can happen unexpectedly. Early detection is key. Sickle cell patients should have routine visits with an ophthalmologist (Thompson & Steinberg, 2012). Laser treatments are an option if it is detected early (Shiel, 2014).

Cerebrovascular System

Cells that are sickled may obstruct the flow of blood to the brain. This can potentially lead to the patient experiencing a stroke (Centers of Disease Control and Prevention, 2014). Because strokes can be fatal, it is important that nurses know the signs and symptoms associated with stroke and also teach parents. Signs and symptoms include weakness or numbness in the

extremities, sudden changes in speech, seizures, and loss of consciousness. If a child exhibits any of these symptoms, interventions must be done immediately in order to prevent death (Mayo Clinic, 2011). Even if death is avoidable, the child can still have long term effects such as learning problems, disability, paralysis, and brain damage (National Heart, Lung, and Blood Institute, 2012). Children with sickle cell disease should be examined in order to be determined if they are at risk for stroke. This can be done using a transcranial Doppler ultrasound (Centers for Disease Control and Prevention, 2014). Abnormal mean velocity flow may be present in those at risk for stroke (Lakhkar, Lakhkar, & Vaswani 2012).

One possible way to prevent stroke is through blood transfusions. Blood transfusions should be done on a monthly basis for patients who are at risk for or who have already had a stroke (Adis Medical Writers, 2013). They have been proven to prevent recurrent strokes in patients that have already had a stroke. However, the patient would need blood transfusions for the rest of their life due to the high probability of having a stroke after cessation (Verduzco & Nathan, 2009). Continuing blood transfusions puts the patient at risk for iron overload which can be life threatening (Centers for Disease Control and Prevention, 2014). It can cause liver damage, heart damage, lung damage, and damage to the endocrine organs. This may lead to cirrhosis, heart failure, pulmonary hypertension, diabetes, and delayed puberty (Mir & Logue, 2014). Even so, the benefits outweigh the risks until better options are made available. If a patient is receiving blood transfusions, iron levels should be monitored. The patient should also receive iron chelation therapy in order to prevent complications of iron overload (Centers for Disease Control and Prevention, 2014).

Erythrocytapheresis has been viewed as one alternative option to blood transfusions and it appears to have fewer risks associated with it. Erythrocytapheresis is "the withdrawal of blood,

separation and retention of red blood cells, and retransfusion of the remainder into the donor" (Saunders, 2007). It is safer than blood transfusions and it can also be used to prevent and treat other complications caused by sickle cell disease (Ullrich et al., 2008). Erythrocytapheresis has been noted to be effective for the treatment of acute chest syndrome as well as maintaining respiratory status (Velasquez, Mariscalco, Goldstein, & Airewele, 2009).

Curing Sickle Cell

Although there are many different options for the prevention and treatment of the complications associated with sickle cell disease, there are no methods to prevent acquiring sickle cell disease. There are a few curative procedures. One of the main methods to cure sickle cell disease is stem cell transplant (Thompson & Steinberg, 2012). Stem cells are extracted from peripheral blood, bone marrow or cord blood. Complications of stem cell transplantation include anemia, seizures, electrolyte imbalances, infertility, and infection. Due to the various life threatening complications, stem cell transplantation is only used for certain cases. In order to prevent these complications, the patient should be advised to have a sibling or parent to be their donor if possible. The patient should also undergo a myeloablative or nonmyeloablative preparative regimen before having the stem cell transplant (Thompson, Ceja, & Yang, 2012). Patients should be monitored for signs and symptoms of infection. Precautions should be taken in order to prevent infection including good hand washing, providing patient with a private room, and wearing appropriate personal protective equipment. Labs should also be drawn in order to monitor electrolytes.

Conclusion

Sickle cell disease is a disorder that affects millions worldwide. Children are often admitted to the hospital repeatedly due to complications. Even though sickle cell disease cannot

be prevented, there are ways to prevent and treat the complications associated with it. Pain can be severe and affect a person's daily life. Patients taking morphine or other pain medications have been known to stay at the same pain level and have recurrent pain episodes. Guided imagery should be used as the first line for prevention and treatment of pain episodes. Acute chest syndrome can be caused by infection and some of the treatments for pain. These patients should be encouraged to use incentive spirometers and take antibiotics. All sickle cell patients should take precautions to avoid acquiring infection in order to prevent vaso-occlusive crises and acute chest syndrome. Patients should also have their renal functioning assessed frequently. They should also follow up with an ophthalmologist regularly. Transcranial Doppler ultrasounds should also be performed in order to determine patients at risk for stroke. Patients having severe complications of sickle cell disease may be considered for stem cell transplantation. It is imperative to prevent all these complications in order to lengthen the life expectancy for patients and also improve their quality of life.

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