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Prevention and management of acute complications in children with sickle cell disease – a CPS Pedscases Podcast

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Introduction

Hi everyone, my name is Audrey Slater. I am a fourth-year medical student at the Université de Montréal, Quebec. This podcast will review the Canadian Paediatric Society Position Statement from November 2022 “Acute complications in children with sickle cell disease: Prevention and management” written by Dr. Carolyn E. Beck, a hospitalist pediatrician at SickKids hospital, Dr. Evelyne D. Trottier, a pediatric emergency physician at CHUSJ, Dr. Melanie Kirby-Allen, a haematologist at SickKids Hospital and Dr Yves Pastore, a haematologist at CHUSJ. This podcast has been reviewed by the authors.

Sickle cell disease (SCD) is a chronic, multisystem blood disease. Complications arise when the sickling of red blood cells causes hemolysis and vascular occlusion, leading to hemolytic anemia, pain syndromes, and organ damage. During this podcast, we will discuss how to approach the prevention and management of acute complications of sickle cell disease in children.

After this podcast, the listeners should be able to:

1. Understand how to prevent mortality and morbidity associated with SCD
2. Recognize and manage the common complications of SCD
3. Know the basic principles of transfusion for patients with SCD

Background and prevention

SCD is the most common human genetic disease, affecting at least 6000 individuals in Canada. It is caused by genetic mutations in the beta-globin gene required to create red blood cells. Affected individuals can be homozygous for the mutated beta globin gene (HbSS), or have heterozygosity for hemoglobin S (HbAS) and another beta-globin gene mutation such as beta-thalassemia ($HbSB^0$ thalassemia). In Canada, it is the disorder most frequently diagnosed among all other diseases screened for in newborn

screening. As a result of immigration, SCD is also becoming increasingly prevalent in many areas of Canada; therefore, it becomes even more important for a range of providers to understand the disease and its main complications. With prevention, treatment strategies, and comprehensive care, a significant decrease in mortality in children has been observed, but unfortunately morbidity persists due to the common complications of sickle cell disease.

The main clinical manifestations of SCD include chronic hemolytic anemia, pain syndromes (also known as vaso-occlusive episodes) and major organ complications. Individuals with SCD are also at greater risk of fulminant bacterial infections, due to decreased splenic function.

Newborn screening is an essential first step. It reduces SCD-related infant mortality rates, and allows earlier referral, parental education, preventive strategies, and genetic counseling. Furthermore, enhanced vaccination against encapsulated bacteria is imperative for children with SCD. The following vaccines should be administered:

- 20-valent pneumococcal conjugate and polysaccharide vaccines against *Streptococcus pneumoniae*
- Conjugated quadrivalent meningococcal and serogroup B vaccines against *Neisseria meningitidis*
- Extra booster dose against *Haemophilus influenzae* type B
- Immunization against hepatitis A and B
- Annual influenza vaccines
- If the child will be travelling, vaccines against *Salmonella typhi* and malaria prophylaxis are recommended.

Children with SCD should also receive antibiotic prophylaxis, consisting of daily prophylactic penicillin VK or amoxicillin from 2 months to 5 years old. That period should be extended in cases of surgical splenectomy, history of invasive bacterial infections, or in those whose immunizations are not up to date.

Additional treatment includes hydroxurea, proven to significantly reduce acute chest syndrome (ACS), vaso occlusive episodes (VOE), transfusions, hospitalization, and mortality. It should be offered to all children 9 months of age or older, after discussing the risks and benefits with the parent. While it is the standard of care for children with SCD types HbSS and HbSβ^0 thalassemia, it can also be considered for symptomatic HbSC. It is important to remember hydroxurea should be held in cases of cytopenia, and hematology consulted.

Now that we have a basic understanding of Sickle Cell Disease treatments and preventive measures, lets go through some clinical cases to discuss acute complications and their management.

Vaso-occlusive episode (VOE)

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Charles, a 8-year-old boy with SCD, presents to the emergency department (ED) with a 1-day history of leg and arm pain. He was given acetaminophen and ibuprofen at home but continues to rate his pain 9/10. On exam, he has diffused leg tenderness without erythema, edema, or fever, and his vital signs are normal.

Take a moment to think about what your working diagnosis is for Charles' case. While you may have many causes of leg pain in a child in mind for a differential diagnosis, in a child with sickle cell disease, we always need to consider this could be a vaso-occlusive episode.

Management of VOE includes prompt pain management. Intranasal fentanyl followed by a dose of oral morphine should be administered promptly, within 30 to 60 minutes of arrival. Intranasal fentanyl, quickly administered with a rapid onset of action, significantly reduces the time needed to provide the first opioid dose and the number of intravenous line insertions. This should be combined with ibuprofen and acetaminophen if not given recently at home by the family. If pain control is achieved with oral opioid, children may be managed as outpatients after observation for a minimum of 2 to 3 hours.

Charles' pain remains elevated despite oral opiates and is now in both legs. What are the next steps?

Our differential diagnosis should be explored. For example, osteomyelitis and avascular necrosis or thrombosis should be considered and investigated depending on the presentation, as children with SCD are at greater risk for such complications. However, as there is no edema or erythema and as the pain is in multiple locations, deep vein thrombosis and infection are thought to be less likely.

For his pain, we should start by administering a morphine IV bolus, followed by morphine IV infusion with intermittent bolus doses or patient-controlled analgesia. Charles will be admitted to the hospital as his pain is difficult to control. He should receive round the clock acetaminophen and NSAIDs in addition to opioids. In addition to pharmacological pain management, there are other important principles to consider.

Psychological support and physical comfort measures, such as heat packs and distraction, should also be provided. Constipation related to opioids should be prevented with regular dosing of PEG 3350. To maintain total fluid intake, oral hydration is encouraged, and IV fluids are administered, though overhydration beyond maintenance volumes should be avoided due to the increased risk of acute chest syndrome. Incentive spirometry, a seated position, and walking as soon as possible should also be promoted to limit the risk of pulmonary complications. Supplemental oxygen should be initiated if required to maintain a target oxygen saturation of 95%; however, avoiding oxygen if not indicated is important due to the theoretical risk of hypoventilation leading to ACS.

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When Charles' pain is under control, the next step would be to wean off his opioid infusion and transition to regular oral dosing. Pain medication education should be offered to his parents for his return home. When weaning pain control at home, parents are advised to first wean the opioid, then NSAID, and finally acetaminophen, with appropriate doses to manage his VOE in the following days after discharge.

For further resources on how to treat pain for children with SCD, also visit the site of Solution for Kids In Pain, https://kidsinpain.ca/youth-in-pain/. This site also includes a summary tool for healthcare providers done in collaboration with the CPS.

Acute chest syndrome (ACS)

Two days into Charles' admission for VOE, he develops respiratory distress, fever, and chest pain. A chest x-ray (CXR) reveals a left lower lobe infiltrate.

Knowing that he has sickle cell disease, what do you think is his leading diagnosis?

This scenario is describing acute chest syndrome (ACS), defined by a new pulmonary infiltrate in the presence of fever and respiratory signs or symptoms. Causes of ACS include infection, pulmonary infarction, or fat embolism, and because they are difficult to differentiate, all have the same management principles. As up to half of the children with ACS present during hospitalization, most commonly for VOE, patients admitted for pain should be monitored for signs of ACS.

Let's review the investigation and management principles that should be applied for Charles.

First, he should receive oxygen therapy to maintain an oxygen saturation of at least 95%. Investigations should include a chest x-ray, blood culture, complete blood count, reticulocyte count, and crossmatch. A nasopharyngeal viral swab and *Mycoplasma pneumoniae* PCR should be considered for infectious etiology. Empiric therapy, a broad-spectrum third-generation cephalosporin for typical bacteria and a macrolide for *Mycoplasma pneumoniae,* should be started immediately, before receiving tests results. The macrolide can be discontinued if the *Mycoplasma* PCR is negative. A Hematology consultant should be involved in Charles' care. If he clinically deteriorates, antibiotic coverage should be broadened and, based on clinical severity, a simple or exchange transfusion could be considered. It is particularly important to remember that patients with sickle cell disease are at high risk of developing alloimmunisation; therefore, blood transfusion should always be discussed with the hematology team and blood bank specialists. As patients with ACS may rapidly deteriorate, always anticipate the possible need for transfer to an intensive care environment.

Fever

Malik, a 4-year-old boy with SCD, presents to the ED with rhinorrhea and a temperature of 38.7°C. He is alert and non-toxic, is a little bit congested but no other apparent focus of infection. His mother assures you that she administers his prophylactic antibiotics daily, and that his immunizations are up to date.

Let's discuss the initial investigations for Malik.

As individuals with SCD rapidly develop functional asplenia, they are particularly at risk for invasive bacterial infections, such as sepsis, meningitis, or osteomyelitis. Therefore, febrile episodes, defined as oral temperature of 38.0°C or rectal temperature of 38.5°C, should warrant medical evaluation, investigation and treatment. Within 30-60 minutes of arrival, a complete blood count, reticulocyte count, bilirubin, and blood culture are drawn, and a third-generation cephalosporin (either IV or IM) is administered. The investigation of the source of the fever should include a chest x-ray, and further infectious work-up as clinically indicated, such as an influenza swab.

When considering febrile patients with sickle cell disease, there are several criteria that determine if they are a "low risk" patient, which can help guide if hospital admission is required.

Low-risk criteria for SCD and fever are the following:

- Well-appearing and hemodynamically stable
- Fever less than 40°C
- Older than 6 months
- WBC between 5 and 30 $x10^9$/L, platelet count more than 100 $x10^9$/L and not significantly lower than baseline, Hb more than 60 g/L and not > 20 g/L lower than baseline. If the baseline hemoglobin is unknown, the patient should be admitted.
- No respiratory distress or CXR abnormality
- No clinical findings suggestive of meningitis, osteomyelitis, septic arthritis, ACS, or splenic sequestration
- No history of pneumonoccal sepsis or meningitis
- No significant pain or dehydration
- Initial visit for the episode
- Safe for discharge and ability to follow-up

Patients who do not meet low risk criteria should be admitted and continued on intravenous antibiotics.

Given that Malik meets criteria for low risk patient, he may be discharged home, after receiving ceftriaxone and arranging subsequent daily follow-up to ensure clinical improvement and negative blood cultures.

<u>Splenic sequestration</u>

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Six months later, Malik returns to the ED with sudden weakness and abdominal fullness. He is tachycardic, tachypneic, with normal blood pressure, and has a palpable spleen at 5 cm below the costal margin (normal for him is 1 cm). Malik's mother, who was taught to palpate the spleen in clinic, fortunately noticed his enlarged spleen at home. Labs reveal a Hb 65 g/L (baseline 92 g/L), reticulocytosis, but no thrombocytopenia.

This clinical presentation is concerning for splenic sequestration, which is a complication that most commonly occurs before age 5, but can happen at any age. Splenic sequestration is a potentially fatal complication of SCD. Patient's should receive 5 mL/kg to a maximum of 10 mL/kg transfusion of packed RBCs. Caution should be applied with blood transfusion volume in splenic sequestration, as sequestered blood can recirculate rapidly following transfusion, increasing risk for hyperviscosity. Close monitoring of hemoglobin is required, as it should not rise above 100 to 110 g/L. If sequestration episodes reoccur, secondary prevention, such as elective splenectomy or chronic transfusion therapy are discussed with hematology.

Aplastic episode

Olivia, a 18-month-old girl with SCD, presents with mild fever, pallor and fatigue. Her Hb is 43 g/L, with reticulocyte count markedly reduced.

This case highlights that when a child with SCD presents with an acute complication, it is important to always observe whether the reticulocyte count is elevated. In the case of low reticulocyte count, aplastic episode secondary to parvovirus B19 infection ("fifth disease"), or other viral suppression should be suspected. Close follow up is indicated even if the anemia is not yet severe as it can continue to worsen. Aplastic episodes are more common in children with SCD, as with other inherited hemolytic anemias, following parvovirus B19 infection. Signs and symptoms of aplastic episodes are secondary to severe anemia, typically associated with an inappropriately low reticulocyte count, mild thrombocytopenia and leukopenia. Management for Olivia should include blood culture and serology for parvovirus B19, ceftriaxone, and transfusion with RBC 10 mL/kg. An important point to always remember is to discuss with hematology and blood bank specialists when transfusing a patient with SCD.

Patients with aplastic crises should be monitored closely until their reticulocyte count begins to rise. More than one transfusion may be required while awaiting bone marrow recovery. Recurrences rarely occur due to development of immunity against parvovirus B19.

Stroke

At age 7, Olivia re-presents in your ED with right-sided weakness, speech disturbance, and decreased level of consciousness. The most recent of her annual transcranial Doppler (TCD) assessments was abnormal (cerebral blood flow velocities >200

cm/second), and she was due to begin a prophylactic blood transfusion program as primary stroke prevention intervention.

You recognize that at the top of your differential diagnosis should be stroke and complete a stat CT scan. The patient's brain imaging shows an acute ischemic stroke (AIS). While there are a variety of possible causes of pediatric stroke, SCD is the most common, primarily affecting children between 2 and 9 years old. Ischemic strokes are the typical presentation, caused by vasculopathy. Annual screening should be done with transcranial doppler in search of vasculopathy in children between 2 and 16 years old with SCD.

Let's review the initial management of stroke in a patient with sickle cell disease.Oxygen should be started immediately to maintain saturation of at least 95%. IV hydration should be started at a maintenance rate. Head imaging should be ordered stat, typically a CT scan due to accessibility of resources; however, if an MRI is available urgently, it would be the preferred imaging method. If CT is negative but the clinical suspicion for stroke remains, an MRI with MR angiogram should be done. Patients are often admitted to the ICU for an exchange transfusion, with the aim of decreasing her HbS level to less than 30%, in close collaboration and consultation with the hematology team. A neurology consult should also be made.

For patients with remaining deficits after acute stroke treatment, ongoing management should include comprehensive rehabilitation services, a chronic transfusion program to prevent stroke recurrence, and collaborative management with a specialized centre, meaning the patient should be discussed with and/or seen by a tertiary care center.

Principles of transfusion

Throughout the podcast we have discussed the indications for blood transfusions. Now, let's discuss and review the principles of transfusion for a person with SCD.

Transfusion in patients with sickle cell disease involves more risks than the general population. First, due to various factors such as red blood cell deformity, SCD patients' blood has a higher viscosity. As a principle, high blood viscosity is associated with vascular complications; therefore, particular attention should be taken not to increase the post-transfusion hemoglobin above 100g/L. Second, patients with SCD are at greater risk for alloimmunization. For this reason, liasing with the blood bank to optimize phenotypic matching and reduce risk for alloantibody development and other transfusion reactions is absolutely crucial. Whenever possible, it is also important to discuss the indication for blood transfusion with the patient's hematologist, to ensure that the benefits of transfusing outweigh the potential risks.

As for the indications of transfusion, it should be considered when patients present with acute anemia more severe than their baseline (as they have chronic anemia) and complications of SCD. Decisions should be based on baseline hemoglobin, current CBC

and reticulocyte count, and clinical status and context. Acute sequestration with severe anemia, and aplastic episodes are the clinical scenarios with the highest strength of recommendation for simple transfusion. It should also be considered in cases of severe anemia with hemoglobin lower than 60 g/L or a decrease of more than 20 g/L from baseline, as well as severely symptomatic ACS. **Importantly, uncomplicated painful episodes do not require transfusion.**

Exchange transfusion, during which the recipient's blood is removed during the donor infusion, can also be considered. It is the preferred option in SCD patients with stroke. In ACS, its advantage over simple transfusion is debated. A pediatric hematologist should always be consulted. When requesting blood, inform blood bank that this patient has SCD and request extended crossmatch to avoid alloimmunization.

Conclusion

With the increased prevalence of sickle cell disease due to immigration and early diagnosis through newborn screening, it is increasingly important that healthcare practitioners be familiar with the disease and its main complications. Listeners should now be comfortable with the presentation and management of vaso-occlusive episodes, acute chest syndrome, fever, splenic sequestration, aplastic episodes, and stroke. We have also discussed the basic risks and indications of transfusion in children with SCD. For most cases, an expert team should be consulted to optimize care and follow-up of such patients.

Thanks for listening!

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