Atlantic Provinces Pediatric Hematology Oncology Network

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Quick Reference Guideline for Sickle Cell Disease in Children and Adolescents: Diagnosis, Guidelines for Care, and Protocols for Management of Acute and Chronic Complications

APPHON/ROHPPA supportive care guidelines are developed by Atlantic Provinces health professional specialists using evidence-based or best practice references. Format and content of the guidelines will change as they are reviewed and revised on a periodic basis. Care has been taken to ensure accuracy of the information. However, any physician or health professional using these guidelines will be responsible for verifying doses and administering medications and care according to their own institutional formularies and policies and acceptable standards of care.

Appropriate sickle cell disease care includes early screening, extensive health maintenance with appropriate prophylactic measures, parental education, psychosocial support, and medical assessment with monitoring for the development of chronic organ dysfunction. In addition, management of life threatening acute illness should occur in a setting where knowledge and resources about sickle cell disease is available and where physicians have ready access to baseline information about the patient.

The following guideline summary is the recommendations from the full guideline: **Sickle Cell Disease in Children and Adolescents: Diagnosis, Guidelines for Care, and Protocols for Management of Acute and Chronic Complications.**

The **purpose** of these recommendations is to provide clinical institutions and other organizations with a framework on which to build their own institutional protocols and to encourage standardization of protocols across regions to enhance consistency of care for patients and families with Sickle Cell anemia. They are intended to serve as general guidelines, recognizing that deviations from them will be appropriate in individual cases. This guideline addresses only the more common pediatric complications and should not be used as a substitute for hands-on-care by providers with experience in the management of sickle cell disease.

Target users of these guidelines are all health professionals within the Atlantic Provinces caring for sickle cell children and youth.

Summary of Guideline Recommendations:

Recommendation	
I. Screening and Diagnosis	
	Screen at risk groups and/or new born screening ERs to keep a list of Sickle Cell patients
	New born screening-peripheral smear
	Hemoglobin electrophoresis for at risk patients and for positive or unreliable screening
II. Initial Evaluation	
	All children positive for Sickle cell disease should see Pediatric hematologist ASAP; exception sickle cell trait.
	Utilize : : **Baseline Studies at Diagnosis and Follow-up schedule (Appendix 1)** **: Sickle Cell Anemia Monitoring Checklist (Appendix 2)**
	Components of evaluation: : History and Physical : Baseline lab tests and other investigations

Recommendation	
	: Immunization history : Preventive measures : Patient/Parent education (See Appendix 3 for information sheet) : Family support
III. Follow-up	
	Utilize : : **Baseline Studies at Diagnosis and Follow-up schedule (Appendix 1)** **: Sickle Cell Anemia Monitoring Checklist (Appendix 2)**
IV. Preventive Measures	
	• **3 months and older with asplenia or hyposplenia** -antibiotic prophylaxis (*penicillin VK*): • **3 months of age and younger with asplenia or hyposplenia** antibiotic prophylaxis including coverage against E.coli and Klebsiella sp. Recommend cefixime 8 mg/kg/day once or twice daily. • **If allergic to penicillin** refer to allergist • Children **with asplenia or hyposplenia** who are **not high risk for overwhelming post-splenectomy infection and who have received their pneumococcal vaccination:** **a)** antibiotic prophylaxis for at least 2 years post-splenectomy **AND** b) Can stop antibiotic prophylaxis at age 5 years in consultation with a hematologist. • Children at **high risk for pneumococcal infection** should receive life-long antibiotic prophylaxis. • **Families non-compliant with antibiotic prophylaxis** – have a stand-by supply of prophylactic antibiotics to give child if febrile or suspected of a fever and seek medical attention immediately. • Give **Vaccines** as per recommendations : Pneumococcal (Table 1, pg 6) : Meningococcal (Table 2, pg 7) : Haemophilus Influenza type B (Hib), Table 3, pg 8) : Influenza (Table 4, pg 8)
V. Management of Fever	
	Definition of a fever • 37.5° C axillary • 38.3° C oral • 38.5° C rectal **Fever is an urgent priority**. Every “at risk” patient should be treated as affected by Sickle Cell Disease until proven otherwise. : **Initiate Sickle Cell Fever Orders (Appendix 5)** **: Contact Pediatric Hematologist** on call ***See guideline page 9-14 for more information on evaluation, treatment, management, and discharge criteria.***

Recommendation	
VI. Pain	
	Pain management at home : increase fluids, quiet activity, enhance body warmth (baths, heating pads), massage : medications: acetaminophen ; add morphine if required
	Immediately go to the Emergency Department if your child has: • Chest pain or abnormal breathing • Abdominal pain • Pain with fever or redness or swelling • Significant headache • Sudden enlargement of the spleen • If pain is not controlled by the measures named above **Admit if:** • Requires 2 or more IV opioid boluses in 1 hour • Febrile • Dehydrated • Unwell ***See full guideline page 14-16 for more information on evaluation, treatment, management, and discharge criteria.***
VII. Acute Chest Syndrome	
	2nd Most Common Complication; Most Common Cause of Death **Identify: Symptoms of Presentation** • Fever* • Cough* • Chest Pain • Shortness of Breath** • Pain in arms and legs** • Chills • Wheezing • Hemoptysis • Productive Cough *** most common presentation of young children** **** most common presentation of adults** **Physical Signs** • Hypoxemia (frequent) • Pulmonary rales • Dullness to percussion **or** • Normal chest exam
	Precipitating Causes • Infections: Pneumococcus, *Mycoplasma pneumoniae;* Legionnaire's disease; Chlamydia; Viruses – parvovirus, RSV; Influenza, para-influenza • Hypoventilation/atelectasis – pain, opioid use • Pulmonary embolism • Fat embolism (due to bone ischemia) causing pulmonary vascular occlusion • Pulmonary edema – intravenous fluids • Bronchospasm (asthma) • Smoking

Recommendation	
	• High altitude • Flight in unpressured airplane • Vasoconstrictive drugs
	Begin Acute Chest Syndrome protocol only after completion of "Sickle Cell Anemia & Fever, Initial Evaluation and Treatment Protocol" and "Sickle Cell Anemia Inpatient Treatment Protocol". • Admit to pediatric or hematology ward • Call Pediatrician or Hematologist as soon as the patient arrives on the floor ***See full guideline page 17-18 for more information on evaluation, treatment, management, and discharge criteria.***
VIII. Surgery	
	• **Consult Pediatrician or Hematologist in advance** and on admission. • Children with sickle cell anemia are at risk for post-operative complications, particularly acute chest syndrome. • The need for pre-op transfusion is determined by the Hematologist/Pediatrician in advance and on an individual basis. Simple or partial exchange transfusion should be strongly considered for all children with Hgb SS or Sβ°thal prior to any procedure requiring general anesthesia. Aim for hemoglobin of 100 g/L and Hgb S less than 30%. Except in cases of emergencies, pre-op transfusions are scheduled 2 to 7 days prior to surgery. • Consult respiratory, physiotherapy, anesthesiology, and surgery.
	See full guideline page 18-21 for information on how to proceed with: **:** Preoperative preparation : Anesthetic management : ENT/Dental cases or Hernia repair : Adenoidectomy : Tonsillectomy and adenectomy : Abdominal surgery (Cholecystectomy or Splenectomy) or other major surgery – Laparoscopic or Open : Peri-operative nursing care : Discharge criteria and Follow-up
IX. Acute Splenic Sequestration	
5.1	**Identify:** : An acute illness associated with Hgb 20 g/L or more below patient's baseline value with acutely enlarged spleen. : Mild to moderate thrombocytopenia is often present. : Reticulocytosis equal to or greater than baseline is usually present. If reticulocyte count is decreased, consider coexistent aplastic crisis (see guideline page 23). **Consult Pediatric Hematology**

Recommendation	
	See full guideline page 21-22 for more information evaluation, treatment, management, and discharge criteria.
X. Aplastic Crisis	
	Identify: : An acute illness associated with Hgb below patient's baseline value with a substantially decreased reticulocyte count (often less than 1%). : Most cases are caused by the acute infection with human parvovirus. : If acute enlargement of spleen is present, consider coexistent splenic sequestration. : Aplasia is usually limited to 7-10 days, but because this may exceed the patient's mean erythrocytic survival time, profound anemia may ensue. **Consult Pediatric Hematology** ***See full guideline page 23/24 for more information on evaluation, treatment, management, and discharge criteria.***
XI. Acute Stroke or Neurologic Event	
	Identify: acute neurological changes present **Consult Hematology and Neurology** ASAP. ***See full guideline page 24-25 for more information on evaluation, treatment, management, and discharge criteria.***
XII. Priapism	
5.1	**XII. PRIAPISM** **Identify:** prolonged (greater than 30 minutes) painful erection of the penis : common in children/adolescents with Sickle cell disease : often starting during the early morning hours. **Two forms:** a) stuttering episodes which last less than 2-4 hours but are often recurrent and may precede a severe episode **b)** severe episodes that last more than 2-4 hours and may eventually result in impotence
	Contact physician (hematologist and/or pediatrician) if: :greater than or equal to 2 episodes within one month :greater than or equal to 4 episodes within one year :episode lasting longer than 2-4 hours seek emergent medical attention. ***See full guideline page 25-28 for more information on evaluation, treatment, management of mild and severe cases, and discharge criteria.***
XIII. Cholelithiasis	
	Identify: • Hemolysis causing increased bilirubin, resulting in gallstone formation - Most evident in children with Hgb SS, Hgb SC and Hgb SB° thalassemia.

Recommendation		
	• Abdominal pain (children with sickle cell disease may have many causes): ▪ Gallstones ▪ Vaso-occlusive crisis ▪ Acute hepatic crisis ▪ Liver sequestration ▪ Splenic sequestration ▪ Acute chest syndrome ▪ Cholecystitis ▪ Pancreatitis ▪ Appendicitis ▪ Urinary tract infection ▪ Pelvic inflammatory disease ***See full guideline page 28-29 for more information on evaluation, treatment, management, and discharge criteria.***	
XIV. Chronic Transfusion		
	Goal: to suppress erythropoiesis to maintain the percentage of the patient's cells (i.e. hemoglobin S) at less than 30%.	
	Aim: Maintain Hgb S less than 30% and a pre-transfusion Hgb of 80-100 g/L; achieved by providing simple transfusions every 3-4 weeks.	
	Indications for chronic transfusions 1. Stroke 2. Transient ischemic attacks 3. Abnormal Transcranial Doppler (greater than 200 cm/sec x 2) 4. Severe recurrent pain episodes 5. Severe, recurrent acute chest syndrome 6. Recurrent splenic sequestration in young children 7. Severe chronic anemia with cardiac failure 8. Pulmonary hypertension **Consult Pediatric Hematologist** ***See full guideline page 29-32 for more information regarding chronic transfusion therapy.***	
XV. Hydroxyurea therapy		
	Hydroxyurea should be initiated and monitored only by Hematologists with expertise in chemotherapy and sickle cell disease and after written documentation of patient education and consent.	
	AIM- increase Hgb F and decreased WBC - Effects expected after taken daily for 3-6 months.	
	Inclusion Criteria: 1. Diagnosis: Hgb SS or S β-thalassemia 2. greater than or equal to 5 years of age	

Recommendation		
	3. greater than or equal to 3 severe vaso-occlusive pain events per year, or 4. greater than or equal to 2 episodes of acute chest syndrome, or 5. Any combination of greater than or equal to 3 episodes of acute chest syndrome and severe pain per year **Exclusion Criteria** 1. Pregnancy 2. Inability to use reliable contraception if sexually active (men ***and*** women) 3. Inability to comply with daily dosing and laboratory monitoring 4. Serum creatinine greater than 76 µmol/L (1 mg/dL) **Criteria to start treatment** Hgb decrease from previous level of no more than 20%, ANC greater than 2.5 x 109/L, platelets greater than 150 x 109/L, Reticulocyte count greater than 100 x 10^9/L. ***See full guideline page 32-34 for more information regarding use of hydroxyurea in Sickle Cell Disease, including monitoring for toxicity.*** ***Refer to Appendix 4 for pre-printed orders for hydroxyurea treatment.***	

For more information see full APPHON/ROHPPA guideline: **Guideline for Sickle Cell Disease in Children and Adolescents: Diagnosis, Guidelines for care and Protocols for Management of Acute and Chronic Complications**

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