Sickle Cell Disease: Guidelines to Hydroxyurea

1.0 Policy statement

Patients admitted on hydroxyurea shall not be given hydroxyurea prescription on discharge. **Haematology is to prescribe in order to monitor blood work.**

2.0 Definitions

- **Hydroxyurea (HU)** is a chemotherapy medicine that has been used to treat many disorders, including sickle cell disease (SCD). Research has shown that patients with sickle cell disease who take hydroxyurea are admitted to hospital because of painful events only half as often as patients who do not take hydroxyurea, have fewer acute chest crises and have less need for blood transfusions if they are admitted to hospital. Please see [www.aboutkidshealth.ca](http://www.aboutkidshealth.ca).

- **Maximum Therapeutic Dose (MTD)** is maximum dose or clinical efficacy achieved.

- **Transcranial Doppler (TCD)** is a non-invasive ultrasound used to screen for strokes measuring the rate of blood flow through the large vessels on both sides of the brain.

- **Vaso-Occlusive Crises (VOC)** are blockages of the blood vessels anywhere in the body by deformed red blood cells. This causes a lack of oxygen in the affected area of the body. Symptoms depend on where the blood vessels are blocked.
3.0 Guideline

**Pain assessment and consult completed of child with Sickle Cell Disease (SCD)**

**Does child meet indications to start hydroxyurea?**

**NO**
- Discuss and decide on alternative therapy with child and caregivers
  - Hematology team to obtain agreement from child and caregivers

**YES**
- Prior to initiating hydroxyurea please ensure the following is completed:
  - Refer to tip sheet on Hydroxyurea Education
  - Patient education and ensure there is an ongoing discussion
  - Bloodwork

**Does child and/or caregivers agree to initiate hydroxyurea?**

**NO**
- Discuss alternate therapies if any are available

**YES**
- Prior to initiating hydroxyurea please ensure the following is completed:
  - Refer to tip sheet on Hydroxyurea Education
  - Patient education and ensure there is an ongoing discussion
  - Bloodwork

**Ongoing monitoring of bloodwork**

**In the bloodwork below threshold?**

**NO**
- RMTD or clinical target achieved?
  - Reduce and/or hold dose

**YES**
- Consider dose escalation. Refer to tip sheet
- Continue with same dose

**Reduction for Dose Reductions**
- Neutrophil ANC < 1.8 x 10^9/L
- Ratio count: < 0.9 x 10^9/L
- Platelets < 60 x 10^9/L
- Hemoglobin < 7.0 g/L

**If Hematologic Toxicity Occurs**
- Discontinue hydroxyurea until counts recover (usually 5-7 days)
- Restart at same dose. If threshold is again reached, reduce to previous dose and that is maximum therapeutic dose

**Patient and Family Involvement**
- Clinical effect
- Review VOC episodes with family
- Side effects
- Hematologic toxicity and beneficial effects
- Keep potentially negative management feedback.
- Improve compliance and instills confidence in taking the drug
- Celebrate beneficial effects, and be open when toxicities occur. Show graphs of response e.g. MCV, Hb F%.

**Indications for hydroxyurea**
- In initial 4 months of age, children, and adolescents with SCA, offer treatment with hydroxyurea regardless of clinical severity to reduce SCD-related complications. Refer to NIH guidelines:
  - < 2 hospitalizations for Vaso-Occlusive Crises (VOC) episodes in a 12 month calendar period
  - < 5 acute chest crises requiring transfusion
  - Absence of clinical target (threshold due to VOC pain managed at home)
  - Abnormal Trans-Cranial Doppler (TCD) in patient failing translation therapy
  - Chronic hypoxemia
  - Low hemoglobin < 7.0 g/L
  - High conditional TCD velocities
  - Previously uncontrolled attacks on screening
  - Poor growth and development

**Prior to starting hydroxyurea ensure that the following is reviewed:**
- Evidence of organ damage – TCD velocities, proteinuria, hypoxemia, academic performance
- Does the patient have sleep apnea?
- Psychosocial issues which might impact compliance with treatment regimens such as transportation, and finances for drug coverage
- Document growth and development
- Through physical examination
- Document discussion, history and physical including height and weight, oxygen saturations, and laboratory results

**4.0 Related Documents**

- **Dissolve and Dose Drug Administration ==>**
- **Chemotherapy At Home: Safely Handling and Giving Medicines**
- **Chemotherapy At Home: Safely Giving Your Child Capsules**
- **Hydroxyurea Education and Discussions Tip Sheet**
5.0 References


7. The effect of prolonged administration of Hydroxyurea on morbidity and mortality in adult patients with sickle cell syndromes; results of a 17-year, single center trial (LaSHS). Blood 2010;115(12):2354-2363


9. Ware, RE. How I use hydroxyurea to treat sickle cell disease. Blood 1 July 2010.Vol 115, Number 26

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Attachments:

- Hydroxyurea Education and Discussions Tip Sheet.pdf
- Revision History.docx
- scd_hydroxyurea_v4_2021.pdf