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

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**3.0 Guideline**

**Indications for Hydroxyurea**
- In infants 4 months of age, children, and adolescents with SCA, offer treatment with hydroxyurea regardless of clinical severity to reduce SCD-related complications. Refer to NH guideline.
- 2 or more hospitalizations for Vaso-Occlusive Crises (VOC) episodes in a 12-month calendar period
- 5 or more days in the hospital requiring transfusions
- Regardless of VOC episodes, hydroxyurea is offered due to VOC pain managed at home
- Abnormal Transcranial Doppler (TCD) in patient refusing transfusion therapy
- Chronic hypoxemia
- Low hemoglobin <7.5 g/dL
- High conditional TCD velocities
- High cardiac index on screening
- Blood vessels occlusive
- Poor growth and development

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

**Prior to starting hydroxyurea ensure that the following is reviewed:**
- Details of VOC episodes – number and severity
- Evidence of organ damage – TCD velocities, prolactin, hypothyroidism, academic performance
- Does the patient have sleep apnea?
- Psychosocial issues which might impact compliance with treatment regimen such as transportation, and finances for drug and range
- Document growth and development
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- Document discussion, history and physical including height and weight, oxygen saturations, and laboratory reports

**Ongoing monitoring of bloodwork**
- Monitor CBC, CRP, and ferritin weekly
- CBC, CRP, and ferritin, every 6 months
- Hb analysis
- ALT, AST, and LDH
- Hemoglobin >7 g/dL

**If Hematologic Toxicity Occurs**
- Discontinue hydroxyurea until counts recover (usually 5-7 days)
- Restart at same dose or hold dose and that is maximum therapeutic dose

**Patient and Family Involvement**
- Clinical effect
- Review of VOC episodes with family
- Side effects
- Hematologic toxicity and beneficial effects
- Keep patient fully engaged in management. This improves compliance and instills confidence in taking the drug
- Celebrate beneficial effects, and be open when toxicities occur. Show graphs of response e.g. MCV >107%
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