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
- YES

Discuss and decide on alternative therapy with child and caregivers

Hematology team to obtain agreement from child and caregivers

Does child or/and caregiver agree to initiate hydroxyurea?

- NO
- YES

Prior to initiating hydroxyurea please ensure the following is completed:

- Bloodwork
- Patient education and ensure there is an ongoing discussion
- Develop a treatment plan
- Write prescription
- Initial dose at home

Discusses alternate therapies if any are available

HMTD or clinical target achieved?

- NO
- YES

Reduce and/or hold dose

- NO
- YES

Consider dose escalation. Refer to tip sheet

Continue with same dose

Indications for hydroxyurea:

- Prior to 1 year of age, children, and adolescents with SCD, 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
- ≥3 acute chest crises requiring transfusion
- 5 or 6 months of life, severe anemia on clinical grounds due to VOC pain managed at home
- Abnormal Thyroid Ultrasonography (TUS) in patient refractory to treatment therapy
- Chronic kidney disease
- Low hemoglobin ≤70 g/L
- High conditional TCD velocities
- Poor growth and development

Prior to starting hydroxyurea ensure that the following is reviewed:

- Details of VOC episodes – number and severity
- Evidence of organ damage – TCD velocities, proteinuria, hematocrit, academic performance
- Does the patient have sleep apnea?
- Psychosocial issues which might impact compliance with treatment regimen such as transportation, and finances for drug or travel
- Document growth and development
- Through clinical examination
- Document discussion, history and physical including height and weight, oxygen saturations, and laboratory reports

Document discussion

Thorough physical examination

Details of VOC episodes

Poor growth and development

High conditional TCD velocities

Abnormal Trans Cranial Doppler (TCD) in patient refractory to treatment therapy

Chronic kidney disease

Low hemoglobin ≤70 g/L

If Hematologic Toxicity Occurs

- Discontinue hydroxyurea until counts recover (usually 5-7 days)
- Restart at same dose
- Reduce to previous dose and that is maximum therapeutic dose

Threshold for Dose Reductions

- Neutrophil ANC ≥ 1.0 x 10^9/L
- Ratio count ≤ 1.5 x 10^12/L
- Platelets ≥ 120 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
- Reduce to previous 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 potentially aggressive management. This improves compliance and instills confidence in taking the drug
- Collect and benefit from, and be open when toxicities occur. Show graphs of response e.g. MCV ≥ 10%
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