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 alternate therapies if any are available

YES

Hematology team to obtain agreement from child and caregivers

Does child and/or caregivers agree to initiate hydroxyurea?

NO

Prior to initiating hydroxyurea please ensure the following is completed:

Discuss alternate therapies if any are available

YES

Ongoing monitoring of bloodwork

In the bloodwork below threshold?

HMTD or clinical target achieved?

NO

NO

Reduce and/or hold dose

NO

YES

Consider dose escalation. Refer to a hematologist

YES

Continue with same dose

Indications for hydroxyurea:

- In infants 4-6 months of age, children, and adolescents with SCD, offer treatment with hydroxyurea regardless of clinical severity to reduce SCD-related complications. Refer to NIH guidelines.
- A 2 hospitalizations for Vaso-Occlusive Crises (VOC) episodes in a 12-month calendar period
- 5 or more short episodes requiring transfusion
- Significant risk of splenic sequestration
- Admission for or mild to moderate level of Splenic Infarction
- Abnormal Transcranial Doppler (TCD) in patient refusing transfusion therapy
- Chronic hypoxemia
- Low hematocrit < 35%
- High conditional TCD velocities
- Fibrinogen levels elevated on screening
- Bone pain on screening
- Poor growth and development

To be discussed with child and caregivers

Prior to starting hydroxyurea ensure that the following is reviewed:

- Details of VOC episodes – number and severity
- Evidence of organ damage – TCD velocities, proteinuria, hypoxemia, academic performance
- Does the patient have sleep apnea?
- Psychological issues which might impact compliance with treatment regimen such as transportation, and finances for drug coverage
- Document growth and development
- Thorough physical examination
- Document discussion, history and physical including height and weight, oxygen saturation, and laboratory results

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

Prior to initiating hydroxyurea please ensure the following is completed:

Refer to e-formulary

Dose child meet indications to start hydroxyurea?

Yes

Discuss alternate therapies if any are available

NO

NO YES

Does child and/or caregivers agree to initiate hydroxyurea?

NO

Prior to initiating hydroxyurea please ensure the following is completed:

Refer to tip sheet on Hydroxyurea Education

YES

Initial dose at home

Write prescription

Develop a treatment plan

Patient education and ensure there is an ongoing discussion

YES

Consider dose escalation. Refer to hematologist

Ongoing monitoring of blood work

In the bloodwork below threshold?

HMTD or clinical target achieved?

NO

NO

Reduce and/or hold dose

NO

YES

Consider dose escalation. Refer to hematologist

YES

Continue with same dose

References

- Monitoring
  - CBC, U & E, urine
  - 6 months CBC, diff, retic, LDH analysis
  - Bili, JDH, creatinine, ALT, AST, BUN

- Threshold for Dose Reductions
  - Neutrophil ANC 2.0 x 10⁹/L
  - Retic count < 1.0 x 10⁹/L
  - Platelets < 150 x 10⁹/L
  - Hemoglobin > 70 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 of VOC episodes with family
  - Side effects
  - Hematologic toxicity and beneficial effects
  - Keep potentially regenerative management. This improves compliance and instills confidence in taking the drug
  - Celebrate beneficial effects, and be open when side effects occur. Show graphs of response e.g. MCV, MCH%
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

Revised by:

- Melina Cheong, RN, Nurse Practitioner, Division of Haematology/Oncology
- Melanie Kirby, MD, Staff Physician, Division of Haematology/Oncology
- Marina Strzelecki, Clinical Pharmacist, Pharmacy

Attachments:

Hydroxyurea Education and Discussions Tip Sheet.pdf

Revision History.docx

scd_hydroxyurea_v4_2021.pdf