
	Document Scope: Departmental	
	Document Type: Clinical Practice Guideline Approved on 2021-07-20 Next Review Date: 2024-07-19	
	<b>Sickle Cell Disease: Guidelines to Hydroxyurea</b>	Version: 3

## 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

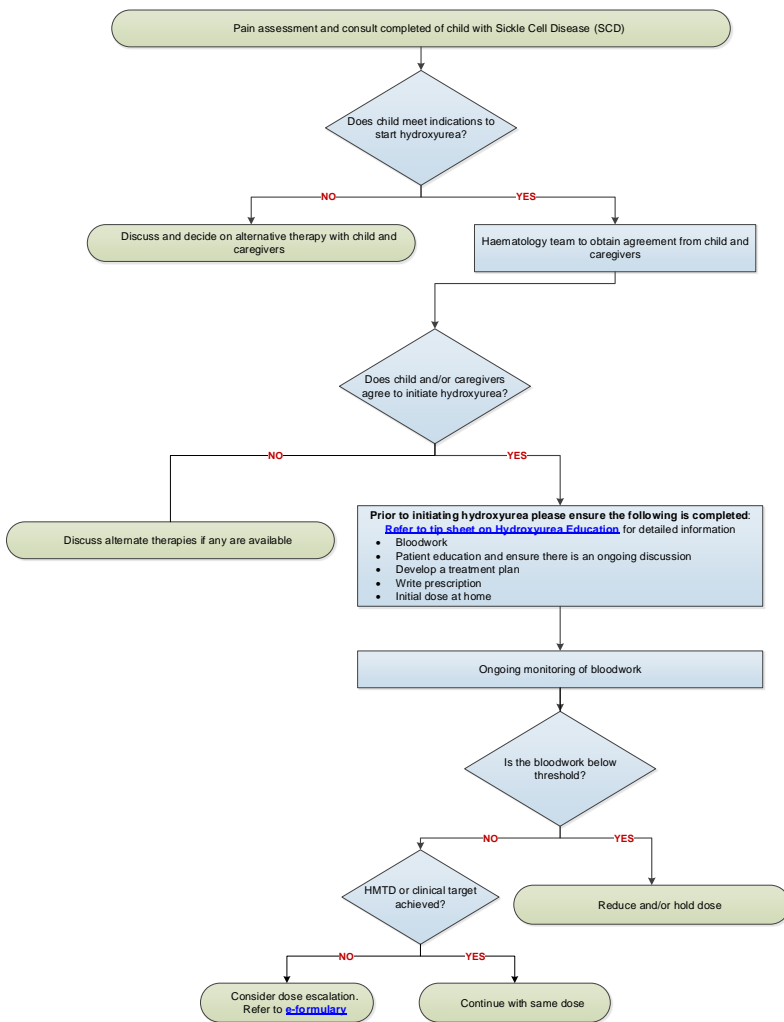
- **Hydroxyurea (HU)** is a chemotherapy medicine that has been used to treat many disorders, including sickle cell disease (SCD). Research has shown that patients with sickle cell disease who take hydroxyurea are admitted to hospital because of painful events only half as often as patients who do not 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.

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## Sickle Cell Disease: Guidelines to Hydroxyurea

### 3.0 Guideline

Sickle Cell Disease: Hydroxyurea Therapy



**Indications for hydroxyurea**

- In infant  $\geq 9$  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](#)
- $\geq 2$  hospitalizations for Vaso-Occlusive Crises (VOC) episodes in a 12 month calendar period
- $\geq 1$  acute chest crises requiring transfusion
- Significant # of days missed from school/work due to VOC pain managed at home regardless of # of hospital admissions
- Abnormal Trans Cranial Doppler (TCD) in patient refusing transfusion therapy
- Chronic hypoxemia
- Low hemoglobin  $< 70\text{g/L}$
- High conditional TCD velocities
- Presence of silent infarcts on screening
- Neurocognitive decline
- 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, hypoxemia, academic performance
- Does the patient have sleep apnea?
- Psychosocial 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 reports

**Blood work Monitoring**

- Bi-monthly CBC, diff, retics
- Q6 months CBC, diff, retics, Hb analysis (HB F%), Bili, LDH, creatinine, ALT, AST, BUN

**Threshold for Dose Reductions**

- Neutrophil ANC  $< 2.0 \times 10^3/\text{L}$
- Retic count  $< 80 \times 10^3/\text{L}$
- Platelets  $< 80 \times 10^3/\text{L}$
- Hemoglobin  $< 70 \text{g/L}$


**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 patient/family 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, HB F%.

## PRINTABLE VERSION

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## 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

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### Attachments:

[Hydroxyurea Education and Discussions Tip Sheet.pdf](#)

[Revision History.docx](#)

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