



## SICKLE CELL ANEMIA HYDROXUREA ORDERS

Patient: \_\_\_\_\_

Age \_\_\_\_ Wt: \_\_\_\_\_ kg Date of Wt (dd/mm/yyyy) \_\_\_\_\_

Allergies: \_\_\_\_\_

The following orders will be carried out by a licensed healthcare professional **ONLY ON THE AUTHORITY OF AN APPROVED PRESCRIBER. Where choice occurs, check as appropriate.**

Refer to APPHON Guidelines for Management of Sickle Cell Disease

### INDICATIONS:

Frequent painful crises (3 or more episodes/year), acute chest syndrome (2 or more episodes/year), chronic hypoxemia (pulse oximetry less than 94% on 2 consecutive visits), severe anemia (Hgb less than 70 g/L or exceptionally low fetal Hgb), elevated WBC (greater than  $20 \times 10^9/L$ ), elevated LDH (greater than 2 x upper limit of normal), elevated Transcranial Doppler (TCD) velocities, inability to transfuse for stroke prevention.

### EXCLUSION CRITERIA:

Pregnancy or inability to use reliable contraception if sexually active (male and female), severe renal impairment, active liver disease or infants younger than 9 months.

### MONITORING:

- History and physical examination: at baseline, every 4 weeks while escalating then every 4 to 8 weeks
- CBC, differential, platelets, reticulocyte: at baseline, every 4 weeks while escalating then every 4 to 8 weeks
- Ferritin, Iron, TIBC, Folate and B<sub>12</sub>: baseline then every 6 months
- Bilirubin (total), ALT and creatinine: baseline then every 3 to 6 months
- HbF: baseline then every 3 to 6 months
- Pregnancy test (if menstruating): baseline then if menses are delayed 2 weeks. Consider hormonal contraception.

Other: \_\_\_\_\_

### TREATMENT:

**Hydroxyurea** \_\_\_\_\_ mg (20 mg/kg/dose) PO once daily with water on an empty stomach.

Reduce initial dose for pre-existing renal dysfunction or myelosuppression. Available as 500 mg capsules. Various recipes for a compounded suspension are available depending on willingness of pharmacy to prepare using cytotoxic precautions. If unable to swallow a full capsule, empty contents of capsule in a water, stir well & administer immediately.

The dose may be increased by 5 mg/kg/dose every 8 weeks to a maximum tolerated dose up to 35 mg/kg/dose. Check compliance if patient requires high doses. Maintain ANC above  $2 \times 10^9/L$ .

If patient experiences toxicity, hold hydroxyurea until lab values normalize. Check weekly and restart at previous dose or decrease dose by 2.5 mg/kg/dose if prolonged recovery more than 2 weeks or if toxicity recurs.

### HEMATOLOGIC TOXICITY:

ANC less than  $1 \times 10^9/L$ , Hgb decrease from baseline/previous level more than 20% or Hgb less than 70 g/L with associated low reticulocyte count, platelets less than  $80 \times 10^9/L$ , absolute reticulocyte count less than  $0.08 \times 10^{12}/L$  (unless Hgb 80 g/L or more).

### RENAL AND HEPATIC TOXICITY:

Serum creatinine increase 50% above baseline, ALT greater than 2 x upper limit of normal.

DATE (dd/mm/yyyy)	Time (24hr/hh:mm)	Prescriber Signature	Printed Surname/Registration#
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DATE (dd/mm/yyyy)	Time (24hr/hh:mm)	Verified By (Signature)	Printed Surname
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