Guy's and St Thomas' NHS
NHS Foundation Trust

Evelina Children's Hospital
St Thomas' Hospital
Lambeth Palace Road
London SE1 7EH
Tel: 020 7188 7188

GSTFT Clinical Practice Guideline

The Use of Hydroxycarbamide in Sickle Cell Disease

Date:	**1 May 2010**
Review date:	**1 May 2012**
Authors:	**Dr Baba Inusa**
Speciality:	**General Paediatrics & Haemoglobinopathies**
Directorate:	**Children's and Genetics**
Pages:	**3**

Hydroxycarbamide in Sickle Cell Disease

Indications:

Adults and children with Sickle Cell Disease who have:

i) >3 admissions with painful crises in the previous 12 months, or

ii) >1 admission with painful crisis in the previous 12 months, and are symptomatic in the community, or

iii) >2 life threatening complications of the disease, such as acute chest syndrome, or

iv) other indications (such as secondary stroke prevention, pulmonary hypertension) must be discussed with Dr Jo Howard or Dr Baba Inusa

Exclusions:

i) Pregnancy or not practicing active contraception
ii) Active hepatitis

Regimen details:

Commence at 15mg/kg to nearest 500mg (250mg in children)

Assess clinical response and if sub-optimal, increase by 2.5mg/kg every 4 weeks until:
Neutrophils < 1.5 x 10^9/l or
Platelets < 80 x 10^9/l or
Retics <1% or
Hb >3g/dl from baseline

Then stop hydroxycarbamide until full blood count has recovered.
Restart at 2.5mg/kg (or 1 capsule – 500mg) lower. This is the maximum tolerated dose (MTD)

Administration:

Orally
Available as 500mg strength tablets-other streghths 250mg being explored by pharmacy

Frequency: Continuous

Extravasation: N/A

Anti- emetics: Nausea and vomiting are rare side effects

Regular investigations:

Day 1

Fbc
Haemoglobin electrophoresis and HbF%
U+Es
LFTs
Reticulocytes
Urate
Alpha genotype if not known, LDH

Day 14 (and every 14th day until dose stable)

Fbc
Hb F%
U+Es
LFTs
Reticulocytes

Once stable every 8 weeks

Fbc
Hb F%
U+Es
LFTs
Reticulocytes, LDH

Toxicities: Bone marrow suppression and cytopenias.
Hyperpigmentation of nails and skin
Nausea and vomiting
Skin rash
Alopecia
Diarrhoea
Teratogenicity

Dose Modifications

Haematological Toxicity

Neutrophils (x 10^9/L)		**Platelets** (x 10^9/L)	**Dosage**
≥ 1.5 x 10^9/L	&	≥ 80 x 10^9/L	100% dose
< 1.5 x 10^9/L	or	< 80x 10^9/L	Stop treatment and recheck FBC twice weekly until N>1.5 and Plt >80. Restart treatment at 2.5mg/kg or 500mg daily lower.

If cytopenias do not resolve within one week, or if patients has neutropenic sepsis, consider use of G-CSF

Renal Impairment No dose reduction necessary

Hepatic Impairment Use with caution