SCU
SICKLE CELL UNIT

A Patient's Guide to Hydroxyurea

HOW DOES HYDROXYUREA WORK?

Haemoglobin (Hb) is the oxygen carrying protein in red blood cells. Most persons have Hb A in their red blood cells but persons with sickle cell disease (SCD) have inherited a gene from their parents that causes their body to make a different type of haemoglobin called Hb S. As blood travels through the body, haemoglobin in red blood cells picks up oxygen from the lungs and carries it to all other parts of the body. In SCD blood vessels may become blocked depriving different parts of the body of oxygen. This causes pain and over time can also damage organs and affect their proper functioning.

Babies born with SCD do not show symptoms of the disease right away. This is because when babies are born, they have a special type of haemoglobin called Hb F. This type of haemoglobin protects the baby from the complications of SCD. Over the first 6 months of life, as Hb F levels decrease and Hb S levels increase, the infant eventually starts having symptoms related to SCD.

Hydroxyurea (HU) is a drug known to increase the amount of Hb F in the red blood cells. Persons with SCD who take HU show an increase in Hb F levels in their red blood cells over time and this may protect them from some of the complications of the disease. There are likely to be other ways in which HU helps prevent sickle cell problems, but these are not clearly understood.

WHAT ARE THE POSSIBLE BENEFITS OF HU THERAPY?

Hydroxyurea is the only drug approved for the treatment of sickle cell disease. **HU has been shown to reduce the frequency of many of the complications such as painful crises, acute chest syndrome, the need for transfusions and hospitalization by at least half with minimal toxicity.**

WHAT ARE THE RISKS?

There are certain risks and discomforts that may accompany HU therapy. HU can reduce the white cell count and decrease the body's ability to fight infections. It can occasionally cause upset stomach, vomiting, skin rashes and hair loss. Experience tells us though that these bad effects are **uncommon** and happen in **less than 1 %** of all patients taking the drug. **They also usually go away quickly when the drug is stopped.**

WHAT ARE THE POSSIBLE SIDE EFFECTS?

Frequency	Side effect
Common (> 20%of persons)	**Early**: Decreased white blood cell count – increased risk of infections Decreased platelets – increased risk of bleeding
Occasional (5–20% of persons)	**Immediate**: stomach upset **Early**: mouth ulcers, anaemia, hair loss
Rare (<5% persons)	**Early**: skin rashes, dizziness, headaches, confusion, hallucinations, seizures **Delayed**: skin colour and nail changes, bleeding, fever & infections **Late**: possible cancer risk, possible birth defects

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IS HYDROXYUREA RIGHT FOR YOU OR YOUR CHILD?

YES if you have Hb SS or HbSβ^0 thalassemia **and one or more of the following:**

- Recurrent painful crises
- Recurrent acute chest syndrome
- Severe symptomatic anaemia
- In children following a stroke if transfusion programs are not offered

NO if you are pregnant, wish to have children in the near future or have active liver disease.

BEFORE HYDROXYUREA IS STARTED…

There are a number of things that have to be done before hydroxyurea is started:

- Baseline blood and urine tests will be done.
- All post pubertal females will be expected to do a pregnancy test and if not already on contraception, one will be recommended for them. Contraception will also be discussed with male patients and their partners.
- Your immunization status will be reviewed and updated (this helps to reduce the risk of infections).
- Once hydroxyurea is started you will also be expected to take folic acid daily.

HU is usually started at a low dose and slowly increased over time if the patient remains well. Additionally, simple blood tests can sometimes inform the doctor if a potential side effect is developing before the patient has any symptoms at all. It is for these reasons that people who take HU need to be seen regularly for a check up and for blood tests.

AFTER HYDROXYUREA IS STARTED…

- Initially, you will be seen every 4 weeks and blood tests will be done at those visits.
- Keep your appointments – though problems might prevent you from attending clinic from time to time, persons who regularly miss appointments may not be able to continue taking hydroxyurea.
- Report possible problems.
- Tell your doctor immediately if you think you may be pregnant.

Live a healthy life style: eat well, keep well hydrated, exercise, and do not drink alcohol or smoke