Deciding on the HU dose

For the first several months, the doctor will give your child a small amount of HU and check their blood levels monthly:

- If their levels of white blood cells and platelets stay normal, the doctor will slowly give your child more HU over many months
- If their levels of white blood cells or platelets fall too low, the doctor will give your child less HU

Your child will have checkups

After several months without problems, your child will only need to see their doctor for checkups and blood work every two to three months. They will see the doctor more often if they have a problem.

How can I learn more?

Call your child's sickle cell doctor or nurse. They can answer any of your questions about HU.

Doctor's/Nurse's Name: _____

Phone Numbers: _____

_____

Patient's Portal Link: _____

Visit these websites:

To watch a video about a family living with sickle cell disease:
http://www.youtube.com/watch?v=iKQmQHh4E2w

To get a quick overview about HU:
https://www.stjude.org/content/dam/en_US/shared/www/patient-support/hematology-literature/hydroxyurea-treatment-for-sickle-cell-disease.pdf

To get more in-depth information about HU:
https://www.nhlbi.nih.gov/sites/default/files/media/docs/sickle-cell-disease-report%20020816_0.pdf

To learn about HU research results:
https://consensus.nih.gov/2008/sicklecellstatement.htm

To find out how to prevent sickle cell disease symptoms:
https://www.vanderbilthealth.com/sicklecell/43988

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TREATING SICKLE CELL DISEASE:

Is hydroxyurea right for your child?

PSCRC

The Pacific Sickle Cell Regional Collaborative works to ensure that people with sickle cell disease receive quality care, no matter where they live or seek care.
PacificSCD.org

What is sickle cell disease?

Sickle cell disease changes the shape and texture of hemoglobin, which is a protein in red blood cells.

People with sickle cell disease have hemoglobin that makes red blood cells a different shape:

Normal hemoglobin A makes red blood cells round and soft

Sickle cell disease hemoglobin S makes red blood cells hard, sticky, and banana or sickle-shaped

The banana-shape of sickle cells block blood flow, which can cause:

- Pain
- Damage to organs, muscles, and tissues
- Damage to blood vessels due to:
  - Too many platelets (cells that help us stop bleeding when we get hurt)
  - Too many white blood cells (cells that protect us against infections)

What is hydroxyurea?

Hydroxyurea (HU) is a medicine that treats sickle cell disease. It can't cure your child of sickle cell disease, but it can help your child feel better if they take it every day. It can help them stay healthy as they get older.

How does it work?

- HU helps red blood cells travel more easily through the body.
- Helps red blood cells stay round and soft
- Lowers platelets and white blood cells to normal levels

What does it do?

HU works in two ways:

1. HU helps red blood cells make more hemoglobin F. This type of hemoglobin lowers the chances that red blood cells will change into the banana or sickle shape.
2. HU helps decrease the number of sticky white blood cells.

Research shows people who take Hydroxyurea:

- Live longer
- Have less pain
- Need fewer blood transfusions and hospital stays
- Have fewer cases of acute chest syndrome
- Have less damage to their brain, lungs, kidneys, and spleen

Who is it for?

It's for people 9 months and older who have sickle cell disease:

- It helps babies and young children experience fewer health problems due to sickle cell disease.
- It helps children and adults feel better, especially if they've had:
  - Severe pain
  - Severe anemia (a low number of red blood cells)
  - Acute chest syndrome
  - Problems with their internal organs

Don't take HU during pregnancy or if you are planning to get pregnant. Doctors don't know how HU might affect an unborn baby.

Is hydroxyurea safe?

Yes. HU is safe when a healthcare provider who is a specialist in sickle cell disease gives it to you and checks your blood levels often OR if the healthcare provider works closely with a sickle cell specialist.

What are the possible side effects?

There have not been serious side effects in people with sickle cell disease who have taken HU.*

Short-term side effects:

Your child may get a stomach ache. To prevent this, have them take HU during a meal.

Boys may make less sperm and their sperm may move slowly. This usually returns to normal when they stop taking HU.

Long-term side effects:

Doctors aren't yet sure what the long-term side effects are. In the past, doctors thought that people who take HU might be more likely to get cancer, but they haven't found this to be true. There does not appear to be an increased risk of cancer, even in people taking HU for more than 20 years.

How does treatment work?

Your child will take HU by mouth once a day. They can drink it as a liquid or swallow it as a capsule.

Your child needs to take HU every single day. This is because their body makes new red blood cells every day, so their daily amount of HU helps their body prevent sickle cells.

It may take 4-6 months before they feel better on HU.

*Nevitt SJ, Jones AP, Howard J. Hydroxyurea (hydroxycarbamide) for sickle cell disease. Cochrane Database Syst Rev. 2017;4(4):CD002202. Published 2017 Apr 20. doi:10.1002/14651858.CD002202.pub2