

What else should I know about hydroxyurea?

If you are a man or woman considering having children you should not take hydroxyurea. Please talk to your doctor.

If you are of childbearing age and are taking hydroxyurea, you must use an acceptable form of contraception.

- Hydroxyurea can be taken safely.
- Close monitoring of your blood count is mandatory.
- It is not a treatment for acute pain.
- It must be taken every day to be effective.
- If you experience side effects from the hydroxyurea let your medical provider know so that changes can be made instead of stopping the medication entirely.



References

1. Steinberg MH, McCarthy WF, et al. Am J Hematol 2010; 85:403-8.
2. Voskaridou E, Christoulas D, et al. Blood 2010; 115: 2354-63.
3. Lopes de Castro Lobo C, Pinto JF, et al. Br J Haematol 2013.

Where can I find more information?

Contact Cayenne Wellness Center and Children's Foundation at 818-940-0079 or our website at: www.cayennewellness.org.

You may also refer to the websites below:

Printed Material:

by NIH: National Heart, Lung, and Blood Institute <http://www.nhlbi.nih.gov/health/health-topics/topics/sca/treatment>

by NCBI. Hydroxyurea for the Treatment of Sickle Cell Disease. <https://www.ncbi.nlm.nih.gov/books/NBK38503/>

by NICHQ: Hydroxyurea Resources. <http://sicklecell.nichq.org/resources/scd-hydroxyurea-resources>

Videos:

Sickle Cell Hydroxyurea Video "Ask Me Why?" <https://www.youtube.com/watch?v=mgp5PI-DsDI>

Hydroxyurea: The Best Hope for Sickle Cell Anemia Patients. <https://www.youtube.com/watch?v=IS165Ys5Dps>



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Rev 10/16

HYDROXYUREA:

MANAGEMENT OF SICKLE CELL DISEASE



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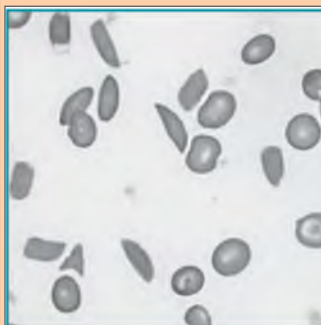
What is hydroxyurea?

Hydroxyurea is a medication that has been shown to reduce painful crisis, reduce hospitalizations, and may improve both the length and quality of life in patients with sickle cell disease. Hydroxyurea, also known by the brand names Hydrea or Droxia, was originally developed as chemotherapy, but later was shown to be effective at increasing the production of hemoglobin F (fetal) in red blood cells. Hemoglobin F interferes with the formation of stiff sickled cells, keeping red blood cells soft and flexible.

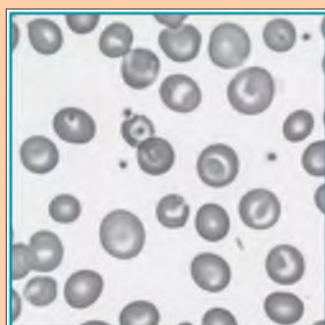
What are the benefits of treatment?

Hydroxyurea is not a cure for sickle cell disease, however research has shown that individuals who continue taking and responding to hydroxyurea have these benefits:

- Fewer pain crisis or painful episodes
- Preventing acute chest syndromes
- Reduced need for blood transfusions
- Fewer hospitalizations
- Less damage to vital organs over time
- Association with increased lifespan¹⁻³



Blood cells **before** taking hydroxyurea



Blood cells **after** taking hydroxyurea

Hydroxyurea treatment for sickle cell disease. St. Jude Children's Research Hospital, 2008. (Accessed October 30, 2014, at <http://www.stjude.org/SJFile/i7600.pdf>.)

Is hydroxyurea safe?

Hydroxyurea is a chemotherapy drug which was used for many years to treat people with certain types of cancer. Hydroxyurea has been used safely in patients with sickle cell disease for over 20 years and has had FDA approval for use in sickle cell disease since 1998.

Due to the impressive benefits from hydroxyurea seen in multiple clinical trials, it has been studied in infants as young as 9 months old. It is now recommended to start at age 9 months in babies with sickle cell disease even before their first pain crisis.

Hydroxyurea should only be prescribed and monitored by an experienced medical provider.

What are the side effects?

- Reduced blood counts which may increase the risk for serious infections and bleeding. Your medical provider will monitor for these side effects at least monthly and may need to adjust your dose.
- Occasional cosmetic side effects include: nail darkening, skin darkening, and thinning of hair. Bald spots are not typically seen.
- Common mild side effects include diarrhea, nausea or vomiting. These tend to improve with time. Nausea is avoided by taking the dose in the evening rather than morning.
- May cause risk of birth defects in a fetus conceived by a mother or father who is taking hydroxyurea.

How Hydroxyurea Works

Hemoglobin F (or fetal hemoglobin) protects against sickle cell symptoms. It prevents hemoglobin S from sticking together and decreases the sickling of red blood cells. Patients with high hemoglobin F levels have fewer symptoms and do better than those patients with low hemoglobin F levels. Hydroxyurea increases hemoglobin F and keeps the red blood cells round.

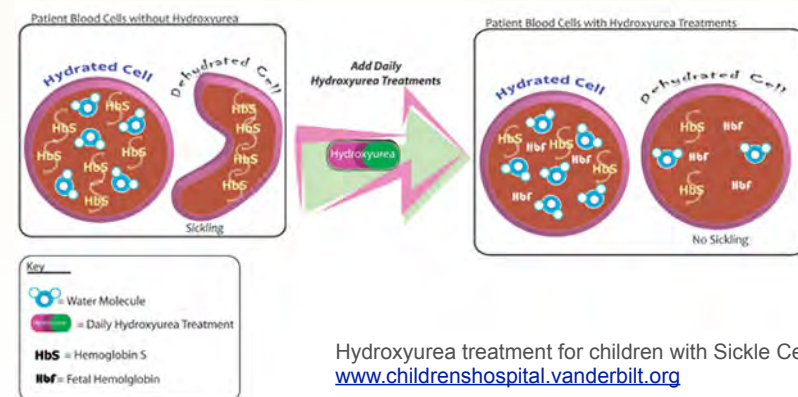
What should I do if I want to begin taking hydroxyurea?

Before starting hydroxyurea the following tasks need to be done:

- Discuss risks and benefits of hydroxyurea with your hematologist.
- Complete the necessary tests before treatment begins (blood counts, liver and kidney function tests, fetal hemoglobin level, and HIV testing).
- Discuss contraception options.
- Carefully consider your commitment to this medication. It must be taken every day to be effective. Hydroxyurea does not work immediately, and cannot be taken on an "as needed" basis. It may take 6 to 12 months before you notice any benefit from the medication, and even then it will be in the form of fewer crises per year rather than preventing all crises. You should be able to commit to taking this medication, the required laboratory monitoring, and dose adjustments for one year when considering therapy.
- It is not safe to stop and restart hydroxyurea without the supervision of your medical provider. You cannot "take a drug holiday" and restart hydroxyurea at the previously tolerated dose.

CONCERNS ABOUT CHEMOTHERAPY:

Hydroxyurea is not known to cause cancer in patients with sickle cell disease, but concerns have been raised because it is chemotherapy. In two studies of more than 550 patients followed for nearly 20 years there has been no evidence of an increased risk of cancer.^{1,2}



Hydroxyurea treatment for children with Sickle Cell. www.childrenshospital.vanderbilt.edu