Appendix A: Standardized Phrases Used for Electronic Medical Record Documentation (For Patients on Hydroxyurea)

Topic	Text
Discussion of the risks and benefits of continued therapy (female patients)	*** was advised to avoid pregnancy when on HU as the effects on a fetus are unclear, although there have been a number of babies conceived when women were on HU without apparent harm. The medication would be stopped as soon as a pregnancy is recognized.
Discussion of the risks and benefits of continued therapy (male patients)	*** was advised to avoid conceiving a pregnancy while on hydroxyurea since the effects on a fetus are unclear, and that there are some reports about reduced sperm production in men on hydroxyurea.
Review of labs and discussing results with the patient, a parent or guardian, or any other appropriate family member and patient adherence	Today we reviewed hydroxyurea therapy, including CBC which demonstrates adequate neutrophil and platelet count. More recent HbF% ***; repeat in *** months. Adherence to taking the medication on a daily basis and to follow-up lab visits was emphasized to optimize benefits and avoid bone marrow toxicity.

*** indicates name of patient; CBC, complete blood count; HU, hydoxyurea

Appendix B: Standardized Phrases Used for Electronic Medical Record Documentation (For Patients NOT on Hydroxyurea)

Topic	Text
Discussion of the importance of hydroxyurea with the patient, a parent or guardian, or any family member	Today we reviewed hydroxyurea therapy, which has been studied and approved for use in sickle cell anemia (HbSS/Sβ^0thal). We discussed the primary benefits of reduction of acute pain episodes and acute chest syndrome and in some cases improvement in anemia, with no clear evidence yet that there is significant impact on the development or progression of chronic complications like lung or kidney damage. We reviewed a primary mechanism of action to increase HbF, which may take 2-3 months or more to achieve an effect or clinical improvement. We discussed starting with 500 mg/day, and checking the CBC in 1-2 weeks. Depending on the neutrophil and platelet count, we would consider increasing the dose to 1000 mg/day, and check a CBC on the new dose in 2 weeks. The dose would be increased as tolerated until there is clinical improvement and/or there is evidence of bone marrow toxicity. Once we reach a stable dose, we would check a CBC every 2-3 months to monitor for hematological toxicity. Adherence to taking the medication on a daily basis and to follow-up lab visits was emphasized to optimize benefits and avoid bone marrow toxicity.
Review of the reasons for the patient's declining hydroxyurea--includes a discussion of the risks and benefits of treatment versus non treatment	We reviewed potential side effects. I noted that it would be unexpected to cause significant nausea or GI upset, is not expected to cause hair loss or other severe side effects like harsh chemotherapy treatments even though it is considered a form of oral chemotherapy that is used for certain bone marrow

	conditions. It is not associated with an increased risk of leukemia in patients with sickle cell disease.
Review of the patient's medical reasons for ineligibility (female patients)	*** was advised to avoid pregnancy when on HU as the effects on a fetus are unclear, although there have been a number of babies conceived when women were on HU without apparent harm. The medication would be stopped as soon as a pregnancy is recognized.
Review of the patient's medical reasons for ineligibility (male patients)	*** was advised to avoid conceiving a pregnancy while on hydroxyurea since the effects on a fetus are unclear, and that there are some reports about reduced sperm production in men on hydroxyurea.”

*** indicates name of patient; CBC, complete blood count; HU, hydoxyurea