

Transcript Details

This is a transcript of an educational program. Details about the program and additional media formats for the program are accessible by visiting: <https://reachmd.com/programs/project-oncology/sickle-cell-disease-care-examining-hydroxyurea-and-chronic-transfusions/36449/>

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Sickle Cell Disease Care: Examining Hydroxyurea and Chronic Transfusions

Announcer:

You're listening to *Project Oncology* on ReachMD. On this episode, we'll hear from Dr. Parul Rai, who's a physician in the Department of Hematology at St. Jude's Hospital in Memphis, Tennessee. She'll be discussing disease-modifying therapies for patients with sickle cell disease.

Here's Dr. Rai now.

Dr. Rai:

So hydroxyurea and chronic transfusions are one of the oldest and most established disease-modifying therapies that are available for patients with sickle cell disease. While some studies have shown that maybe because hydroxyurea improves your hemoglobin—so decreases your anemia—maybe it improves the heart size, so the heart doesn't get that dilated. The heart muscle thickness also gets a little better if you're using hydroxyurea. But there have been studies that have shown that patients who are on hydroxyurea who started on hydroxyurea at a younger age still have that fibrosis that I was talking about in the heart muscle. So it's not really helping with that. So there's some changes in the heart that happen over time. They do kind of slow down, and they're not as bad in patients on hydroxyurea, but other changes like fibrosis, we've not really seen any improvement with hydroxyurea.

Now, transfusions, on the other hand, have also been studied for a long time, and mainly transfusions are used for patients to prevent stroke, so who are at high risk of having a stroke and have abnormal transcranial Doppler velocities. To prevent a stroke from happening, we put those patients on transfusions, and that has also been established in the phase 3 STOP study to show that those patients who have abnormal transcranial Doppler velocities and are placed on transfusions don't have strokes as compared to patients in whom you don't put them on transfusions. Transfusions are also used for secondary stroke prevention, meaning if you've had a stroke, the chance of having another stroke are high, so for those patients, if you've had a stroke, they're put on lifetime transfusions.

However, neither of these therapies until now have shown to improve the cardiac injury, so we don't know if patients are on hydroxyurea over time, does their heart get better? We don't know that yet.

So hydroxyurea is something that we offer to every patient who—around nine months of age, we start them on hydroxyurea, if they have SS and S/beta-thalassemia genotypes, which are the more severe types of sickle cell disease, which we call sickle cell anemia. So all patients who are nine months of age, we start hydroxyurea. We strongly recommend starting hydroxyurea in those patients. And we start at a low dose, around 20 mg/kg, and slowly increase the dose to the maximum-tolerated dose, and we keep them on that. And we regularly follow them, initially monthly and then every three months, when the dose is stable to make sure that there are no complications from that and no issues with that.

Transfusions, are usually given if they need it for any acute complications, so if they come into the hospital and they have some acute chest pain because of which they're needing oxygen and they're not stable from a respiratory point, then we give blood to these patients, like acute simple transfusions. Or if the hemoglobin suddenly drops because of some infection and they need blood, they're not hemodynamically stable, we give transfusions at that time. Chronic transfusion, meaning monthly transfusion, is mostly in pediatrics, given to patients to prevent stroke, so if they have high risk for stroke given the abnormal blood velocity, transcranial Doppler velocity. So to prevent a stroke from happening, those patients are put on transfusions, or if they've had a stroke, then we put them on transfusions.

Announcer:

That was Dr. Parul Rai talking about managing sickle cell disease with hydroxyurea and chronic transfusion. To access this and other episodes in our series, visit *Project Oncology* on ReachMD.com, where you can Be Part of the Knowledge. Thanks for listening!