



Article:

Joesph Wiencek and Garrett Booth
Sickle Cell Trait in the Blood Supply.
J Appl Lab Med 2017;1:773-775.
<http://jalm.aaccjnls.org/content/1/6/773>

Guest: Dr. Joesph Wiencek is a second year clinical chemistry fellow in the Department of Pathology at Vanderbilt University Medical Center in Nashville, Tennessee.

Randye Kaye:

Hello, and welcome to this edition of "JALM Talk" from *The Journal of Applied Laboratory Medicine*, a publication of the American Association for Clinical Chemistry. I'm your host, Randye Kaye.

It's estimated that approximately 1% of the 14 million blood donations performed each year in the USA are from sickle cell trait donors. These carriers are not aware of their sickle cell trait status and present unknowingly. The current guidelines from the American Board of Blood Banking and the World Health Organization that include a questionnaire regarding medical history, a limited physical examination, and measurement of blood hemoglobin concentration before blood donation, are not sufficient to determine presence of sickle cell trait.

Further, American Board of Blood Banking accredited laboratories are not required to perform testing for silent hemoglobin variants such as sickle cell trait on donated blood. Due to this, sickle cell trait blood donations may be given to sickle cell disease patients or other vulnerable patient populations.

In the May 2017 issue of JALM, a Laboratory Reflections: Technical Tips article entitled "Sickle Cell Trait in the Blood Supply," discusses the identification of a leuko-reduced red blood cell unit containing sickle cell trait blood before it was released to a sickle patient in crisis.

The practice in this particular laboratory is to test for sickle cell trait before releasing blood, using Sickledex. This test utilizes the relative insolubility of sickle cell hemoglobin protein to distinguish it from normal hemoglobin. And the positive result, in this case, prevented the blood being released to a sickle cell patient which would have exacerbated their crisis.

The presence of hemoglobin S was subsequently confirmed by cation-exchange high-performance liquid chromatography. The first author of this article is Dr. Joesph Wiencek, a second year clinical chemistry fellow in

the Department of Pathology at Vanderbilt University Medical Center in Nashville, Tennessee and he is our guest for today's podcast. Welcome, Dr. Wiencek.

Dr. Joesph Wiencek: Thank you, Randye and also to JALM for the invitation to discussing this exciting topic.

Randye Kaye: This case certainly highlights the need to address the question "should we require a universal adoption of screening blood donors for hemoglobin variants?" Could you expand on the some of the limiting factors that might prevent or hinder the universal adoption?

Dr. Joesph Wiencek: As many of us know, blood transfusions are one of the most common medical procedures performed in the United States. And with over several million blood donations performed each year, there certainly continues to be a lot of debate in adding universal hemoglobinopathy screening.

Getting back your question, when discussing this topic, I commonly come across several arguments, such as the extra added cost to screen for hemoglobin variants, the issue of screening every donor, and the potential of lack of evidence that these products are inferior and, yes, all of these are excellent questions. But we also have to consider that there are, one, inexpensive diagnostic tests available to consider such as the Sickledex, that could be performed at the site of collection. And two, blood collection centers within the United States can and do test for hemoglobin variants, but not for every donor and not at each donation.

I believe future guidelines might be helpful to add clarity to this discussion. Lastly, to date, there are pathologic cases reported from recipients of sickle cell trait blood. And for this reason, I would have to argue, do we really need to set up a large study, and how would we recruit volunteers to be willingly ready to sign up for a study like that? I believe it would be difficult to convince someone to take part in a study like this.

So yes, there's limited information available, but documented cases with severe outcomes need to be highlighted in future practice guidelines.

Randye Kaye: Now, there's some ongoing debate that sickle cell trait is a benign condition. Do you have any insights into this current discussion?

Dr. Joesph Wiencek: Now, this is a fascinating question. For those of our listeners that do not know, sickle cell trait carriers may present to donor collection services with an unremarkable past medical history, a normal hemoglobin concentration, and if they were born before the implementation of new

born screening for hemoglobinopathy, may not even have an awareness of their carrier status.

Now with that being said, there have also been several cases reported describing sudden deaths in previously healthy individuals with sickle cell trait. These deaths were said to be the result of the sudden cardiac arrhythmia, heatstroke, or rhabdomyolysis, and because of this information, the U.S. Air Force pilots, and NCAA athletes, who undergoes significant physical stresses, are required to undergo hemoglobinopathy screening.

Interestingly, at Vanderbilt, we identified several student athletes, per year, with sickle trait and other hemoglobinopathy.

Randy Kaye: Wow. So, do you think there should be a requirement to A), add Sickledex at the point of care, B), or to ask additional questions about the known family history of sickle cell trait or hemoglobinopathy question to the donor questionnaire?

Dr. Joesph Wiencek: The requirement of a Sickledex at the point of care would be no doubt an extra cost; however, the donor and collection center could obtain valuable information in the process. However, with that being said, the Sickledex has its caveat. Number one, it will not be able to pick up every hemoglobinopathy variant that could cause transfusion-related harm and two, like any test, there's always a risk for false negatives and false positives.

On the other hand, centers could advocate for the use of high-performance liquid chromatography, but the cost and times run specimens will not be feasible for a large scale process such as a blood donation. I think a simpler and maybe a more cost effective approach would be to add a question or two the universal donor health questionnaire, and it would be nice if future guidelines for donor acceptance criteria could address these concerns as well.

Randy Kaye: Okay, that makes sense to me. In your opinion though, if patients are identified as sickle cell trait carriers, should they be allowed to donate red blood cells, platelets, which can have up to two milliliters of RBCs, or plasma?

Dr. Joesph Wiencek: This is such an important question as blood products are already in a limited supply. When you donate whole blood, you have to wait up to 56 days or approximately eight weeks until the next donation. When you donate plasma or platelets, you can donate within approximately a week. By identifying sickle cell trait carriers, I think blood collection centers could advocate for these people to donate more platelets and plasma.

The idea would not be to deter them from donating, rather recruiting them to donate more of a certain type of blood product.

Randye Kaye: I see. Thank you. If the donated blood product is still acceptable for use, what patient or patients should it be given to?

Dr. Joesph Wiencek: I really think this is a difficult question because you have to consider the circumstances of when you might need a transfusion. Most likely, the patient is not well, under stress, or in critical condition. So, I don't really think there is an ideal patient for one of these units should be given to.

Randye Kaye: So, if a blood collection center identifies a donor with a hemoglobin variant, what do you think the next step should be?

Dr. Joesph Wiencek: Personally, I think it would be beneficial to let the donor know that they are a carrier or not. For many people born before the implementation of newborn screening, this could be the only point in time that they would obtain this type of medical information.

Randye Kaye: That was Dr. Joesph Wiencek from Vanderbilt University Medical Center talking about the JALM Laboratory Reflections: Technical Tips article "Sickle Cell Trait in the Blood Supply" for this podcast. Thanks for tuning in for "JALM Talk." See you next time and don't forget to submit something for us to talk about.