Routine sickle cell disease screening among migrants may help save lives

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Hematologists are zeroing in on the escalating humanitarian crisis in southern Europe by assessing the burden of blood disorders among refugees and identifying strategies to facilitate more timely identification and treatment of refugees with sickle cell disease (SCD). A study from an Italian research team published today in *Blood* suggests SCD is common among refugees and screening efforts should be increased.

SCD is an inherited disorder that affects the production of hemoglobin, a protein in red blood cells that delivers oxygen to cells throughout the body. This abnormal hemoglobin can cause red blood cells to become rigid and sickle shaped. The cells stick together and can block the flow of blood and oxygen throughout the body, leading to intense pain and other serious issues such as stroke, infection, pulmonary complications, and even death. Experts stress that, for migrants with known or undiagnosed SCD, the extreme conditions during their travels—dehydration, psychological stress, and exposure to very high or low temperatures—can trigger severe pain crises. Delays in care can lead to severe organ complications and death.

Researchers led by Lucia De Franceschi, MD, of the University of Verona in Italy, found that the vast majority of migrants arriving to Italy are from areas where SCD is highly prevalent, notably sub-Saharan countries, which consistently report the highest burden of SCD globally. In fact, an estimated 80 percent of all newborns with SCD are born in this region, according to the Centers for Disease Control and Prevention.

Even though SCD is endemic to these areas and has been identified as a global health priority by the African Union and the World Health Organization, there is currently no screening program.

Based on their analysis of 2014-2017 data from 13 Italian reference centers, SCD was relatively common in refugees, and it was primarily identified when individuals went to emergency departments for acute sickle cell-related events. De Franceschi reports the rates of anemia were higher among refugees than in the native Italian SCD population, which could be due to malnutrition.

A total of 70 patients with hemoglobin disorders were identified, the majority of whom were male. Half were adults with the median age of 21 years and the other half were children. Most came from West African countries Senegal and Nigeria, as well as Morocco, Egypt, and Tunisia in North Africa, and Syria in the Middle East. During this same four-year period, researchers report a total of 624,688 refugees arrived on the Italian coast.

"Our data underscore the need to more systematically screen refugees coming from areas where SCD is endemic and suggest a strong national network might help to spur action," said De Franceschi, whose work is supported by University of Verona (FUR) and the scientific Italian society for studying hemoglobinopathies (SITE). "Early identification of cases can help prevent severe SCD-related events and life-threatening complications and redirect patients to comprehensive SCD centers for specialty clinical management, follow-up, and timely initiation of treatment."

The group pilot tested a program that screened all refugees at a single refugee center using one of the new rapid point-of-care screening devices (SickleSCAN BioMedomics, Inc.), the results of which were then validated using the gold standard laboratory test. Front-line health

providers in refugee centers and emergency department personnel were trained to recognize the signs and symptoms of SCD and intervene to provide necessary care.

De Franceschi said this approach has allowed them to offer treatment with hydroxyurea and maintain routine clinical follow-up of newly identified SCD patients. De Franceschi said that, based on their findings, efforts should focus on:

- Routine screening for SCD in refugees from countries endemic for SCD within 10-14 days from their arrival to identify potentially vulnerable patients
- A structured, collaborative national network
- Educating ED physicians to identify and treat acute SCD-related events

"We hope our data might open international political and social discussions about the accessibility of health care for refugees to treat both acute and chronic complications related to SCD," De Franceschi said.

In a commentary of the study published today in Blood, authors Russell E. Ware, MD, PhD, and Courtney D. Thornburg, MD, MS, write that based on this study incoming migrants should be routinely tested for SCD and so should their offspring.

More information: Lucia De Franceschi et al. Access to emergency department for acute events and identification of sickle cell disease in refugees, *Blood* (2019). DOI: 10.1182/blood-2018-09-876508

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