New study suggests sickle cell disease may affect brain function in adults

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Sickle cell disease may affect brain function in adults who have few or mild complications of the inherited blood disease, according to results of the first study to examine cognitive functioning in adults with sickle cell disease. The multicenter study, funded by the National Heart, Lung, and Blood Institute (NHLBI), part of the National Institutes of Health, compared brain function scores and imaging tests in adult patients with few sickle cell complications with results in similar adults who did not have the blood disease.

Researchers report that the brain function scores in sickle cell patients were, on average, in the normal range. However, twice as many patients as healthy adults (33 percent versus 15 percent) scored below normal levels. Those who were more likely to score lower were older and had the lowest levels of hemoglobin, the protein in red blood cells that carries oxygen in the blood, compared to sickle cell participants who scored higher. Findings from brain magnetic resonance imaging scans did not explain differences in scores.

Researchers at 12 sites within the NHLBI-supported Comprehensive Sickle Cell Centers conducted the study. Their results are published in the May 12 issue of the Journal of the American Medical Association. An editorial accompanies the article.

"This study suggests that some adult patients who have sickle cell disease may develop cognitive problems, such as having difficulty organizing their thoughts, making decisions, or learning, even if they do not have

severe complications such as stroke related to sickle cell disease," said NHLBI Acting Director Susan B. Shurin, M.D. "Such challenges can tremendously affect a patient's quality of life, and we need to address these concerns as part of an overall approach to effectively managing sickle cell disease."

Researchers tested cognitive functioning of 149 adult sickle cell disease patients (between the ages of 19 and 55) and compared them to 47 healthy study participants of similar age and education levels from the same communities. All of the participants were African-American.

More sickle cell disease patients scored lower on measures such as intellectual ability, short-term memory, processing speed, and attention, than participants in the healthy group. The sickle cell disease participants did not have a history of end-organ failure, stroke, high blood pressure, or other conditions that might otherwise affect brain function.

"We need to study whether existing therapies, such as blood transfusions, can help maintain brain function, or perhaps even reverse any loss of function," noted Elliott P. Vichinsky, M.D., of the Children's Hospital & Research Center Oakland, principal investigator of the study and the lead author of the paper. "These effects were found in patients who have clinically mild sickle cell disease, which raises the question of whether therapies should be given to all patients to help prevent these problems from developing."

Researchers involved in this study are recruiting patients with sickle cell disease into a clinical trial to determine whether blood transfusions may help preserve cognitive function. Participants will receive transfusions every three or four weeks for six months as part of the clinical study. Information about this study can be found at www.clinicaltrials.gov, search for NCT00850018.

Sickle cell disease affects about 70,000 Americans. At one time, many children died from the disease, but new therapies have enabled sickle cell disease patients to live well into middle age or beyond. As more people with sickle cell disease are living into adulthood, health care providers are uncovering previously unrecognized complications.

Studies of brain function in children who have sickle cell disease have suggested that some children with the disease, even if they have not suffered a stroke, have experienced silent brain injury. Others without obvious changes on brain scans may have some level of cognitive dysfunction that seems to worsen with age. Stroke is a common complication of sickle cell disease, and can lead to learning disabilities, lasting brain damage, long-term disability, paralysis, or death.

Sickle cell disease involves an altered gene that produces abnormal hemoglobin. Red blood cells with sickle hemoglobin that have too little oxygen become C-shaped in addition to becoming stiff and sticky. These crescent-shaped cells can clump to block blood flow, causing severe pain and potential organ damage. In the United States, the disease mainly affects those of African descent, but it is also found in other ethnic groups, including those of Hispanic and Middle Eastern descent.

More information: Paper: JAMA. 2010;303[18]:1823-1831.
Sickle Cell Disease: www.nhlbi.nih.gov/health/dci/D … /Sca/SCA_WhatIs.html

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