Rho
Giving flight to research

Sickle Cell Anemia and Neuropsychological Function

A recent clinical trial sheds new light on this under-studied disease

Executive Summary

This article describes a recent clinical trial, conducted by Rho, that compared neuropsychological function in adults with sickle cell anemia with healthy controls. The study is noteworthy because it:

- was the first large, well-controlled study assessing cognitive function in neurologically asymptomatic adults with sickle cell anemia (SCA)
- showed specific areas of lower neurocognitive performance among adults with SCA when compared with controls
- helped describe and quantify an underlying problem in an under-studied population
- helped establish which neuropsychological measures should be used in future neurocognitive research in adults with SCA

Research Details

For many years, sickle cell anemia (SCA) was thought of as a childhood disease due to the high mortality rate among children with the blood disorder. With improved therapies and a better understanding of the disease, life expectancy estimates for people with SCA now extend beyond 50 years, and SCA is more commonly viewed as a chronic disease. Unfortunately, the long-held perception of SCA as a childhood condition has resulted in fairly limited research among adults with the disease.

In recent years, researchers with the National Heart Lung and Blood Institute (NHLBI) have been trying to change that trend by expanding research to better understand SCA in both children and adults. Scientists and researchers with Rho have been privileged to partner with the NHLBI in these efforts through the Basic and Translational Research Program (BTRP) for sickle cell disease.

Research among adult SCA sufferers is often focused on the cumulative toll the disease takes on the body over time. SCA is associated with a number of debilitating symptoms and complications, including acute pain episodes, chronic pain, organ failure, stroke, and difficulty fighting off infections. Another condition of growing interest to SCA researchers is neurocognitive impairment.

In a recent study conducted by the NHLBI's BTRP for sickle cell disease, adults with SCA were compared with controls without SCA to determine whether the two groups showed differences in neuropsychological function. The multi-center trial was conducted at 12 medical centers in the US with an enrollment of 212 participants (160 adult SCA patients and 52 controls). To assess neuropsychological condition and function, study participants were administered a wide-ranging battery of neurocognitive measures as well as an MRI evaluation.

One key element of the study was that the researchers wanted to study only adults with SCA who had not shown any previous history of cognitive impairment. Patients with a medical history of brain injury, neurocognitive impairment, depression, stroke, other chronic health disorders, or those using medications that might affect neurocognitive function were excluded from the study. In effect, the SCA participants in the study were asymptomatic when it came to any existing neuropsychological problems.

The researchers found that adults with SCA showed poorer performance on neurocognitive tests when compared with controls. They also found that SCA was associated with a potential age-related decline in cognitive performance and that MRI results do not adequately explain the differences in performance.

History of SCA Research

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These findings are important because they may help physicians and fellow researchers better understand SCA and how it impacts the brain over time. Moreover, the research helped quantify a previously unstudied health problem of SCA. The study also helped researchers identify the key tools for measuring neuropsychological function in future studies. The researchers hope that this research will open a door to future neuropsychological research among SCA sufferers and, ultimately, ways to treat this aspect of the disease.

The complete results of the study are available in the May 12, 2010 issue of the Journal of the American Medical Association (JAMA, Vol. 303, No. 18, pp 1823-1831).

Study Results

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