Distinguishing between two very similar pediatric brain conditions

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Slight differences in clinical features can help physicians distinguish between two rare but similar forms of autoimmune brain inflammation in children, a new study by UT Southwestern scientists suggests. The findings, published online in *Pediatric Neurology*, could provide patients and their families with a better prognosis and the potential to target treatments specific to each condition in the future.

About half of all cases of encephalitis—a rare type of brain inflammation that affects about 1 of every 200,000 people in the U.S. each year—can be traced to an infection. For a portion of other cases in which the cause isn't initially clear, researchers have discovered a link with the patients' own immune systems inappropriately targeting and damaging the brain.

The most common forms of immune-related pediatric encephalitis are acute disseminated encephalomyelitis (ADEM) and autoimmune encephalitis (AE). Although these are two distinct disorders, explains UTSW pediatric critical care fellow Molly E. McGetrick, M.D., their presentation—including disorientation and other signs of altered mental status, seizures, or motor and sensory abnormalities—is largely the same in children, hindering an accurate diagnosis. In addition, the rarity of AE and ADEM makes amassing data to help distinguish these conditions more difficult.

"Despite their similarities in presentation, patients with AE tend to have a more prolonged and protracted condition that requires more therapies than those with ADEM," McGetrick explains. "Being able to definitively distinguish between these conditions could help doctors guide patients and their families on what to expect."

To reveal the unique features of each condition, McGetrick and her colleagues searched medical records spanning a decade ending in December 2019 for pediatric patients diagnosed at UT Southwestern with encephalitis or encephalomyelitis. They identified 75 patients diagnosed with immune-related encephalitis: 23 with ADEM and 52 with AE.

When the researchers compared patient histories, lab and imaging results, and outcomes, they found slight differences between the two conditions. For example, patients with ADEM had a shorter time from symptom onset to diagnosis compared with those with AE, and those with ADEM universally had abnormal magnetic resonance imaging findings compared with just 61 percent of those with AE. AE patients were more likely than those with ADEM to have markers of elevated inflammation present in their blood and cerebrospinal fluid results. AE patients also tended to have longer hospital stays (21 days versus 13 days for ADEM patients) and were more likely to leave the hospital with a neurological disability that required significant physical and occupational therapy.

McGetrick notes that currently ADEM and AE patients are given similar therapies, including corticosteroids to reduce the body's inflammatory

reaction to autoantibodies, intravenous immunoglobulins to bind and neutralize pathologic autoantibodies, or plasmapheresis to remove autoantibodies from the body over a series of sessions. Many times, symptoms for both conditions will resolve with these treatments, but they can take longer for AE and recur in some individuals. The more researchers can learn about the distinguishing characteristics of these conditions, she says, the more they may be able to target specific treatments for each condition, improving the outlook for these patients.

"One of the biggest take-home messages from this study is that we still have a lot to learn about these conditions," McGetrick says. "The more we know, the brighter the future will ultimately be for these patients."

More information: Molly E. McGetrick et al. Clinical Features, Treatment Strategies, and Outcomes in Hospitalized Children With Immune-Mediated Encephalopathies, *Pediatric Neurology* (2020). DOI: 10.1016/j.pediatrneurol.2020.11.014

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