Physicians find success treating a child's case of rare encephalitis

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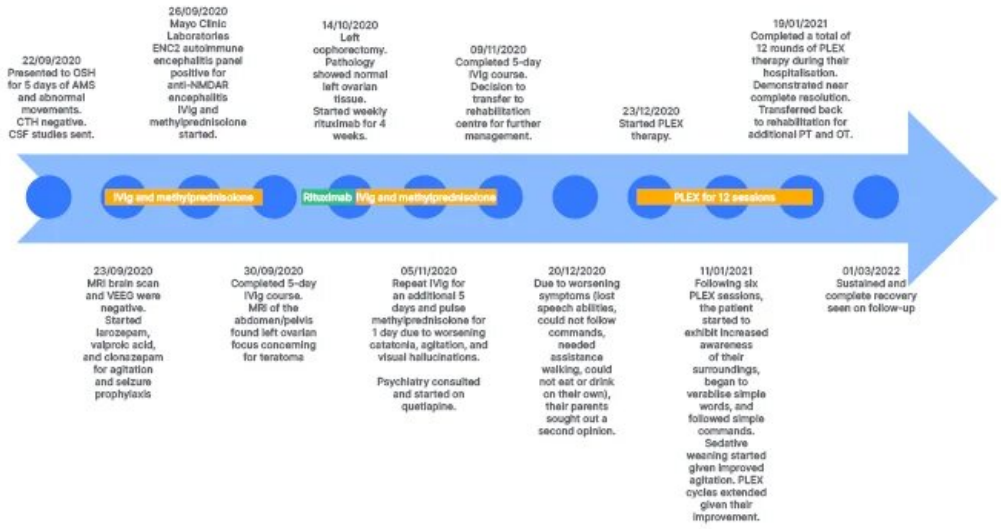

Patient treatment timeline. AMS: altered mental state; CSF: cerebrospinal fluid; CTH: computed tomography of the head; ENC2: encephalopathy, autoimmune/paraneoplastic evaluation of spinal fluid; IVIg: intravenous immunoglobulin; LP: lumbar puncture; NMDAR: N-methyl-D-aspartate receptor; OSH: outside hospital; OT: occupational therapy; PLEX: plasma exchange; PT: physical therapy; VEEG: video electroencephalography. Credit: *EMJ Neurology* (2022). DOI: 10.33590/emjneurol/22-00096

The prognosis for the sick child, who hadn't responded to standard treatment, was dire. But a team of Rutgers physicians believed there might be hope despite the conventional wisdom against pursuing any further treatment.

What happened over the course of the next few weeks in that autumn of 2020—described as a case study in the *EMJ Neurology*—was notable and representative of a newer approach in successfully treating a mysterious disease, the physicians said.

The study highlights the medical case of a 5-year-old girl suffering from anti-NMDAR (N-methyl-D-aspartate receptor) encephalitis, a rare, difficult-to-diagnose malfunction of the brain. Unresponsive to treatments, the child had been transferred to a rehabilitation center and been in a catatonic state for three months when a team of Rutgers physicians were called in to help.

The autoimmune disease believed to be triggered by both environmental and genetic factors has also been the topic of a bestselling book by New York Post writer Susannah Cahalan. She described her medical ordeal suffering from anti-NMDAR encephalitis and recovery in her 2012 memoir, "Brain on Fire." The title of the book, and the 2016 Netflix movie that followed, comes from a phrase used by Cahalan's treating physician to describe the catastrophic brain inflammation that ultimately rendered the reporter trance-like until she was cured.

"With autoimmune diseases, the body attacks a specific system it mistakenly identifies as foreign," said Vikram Bhise, an author of the case study and an associate professor of pediatrics and neurology and director of the Division of Child Neurology and Neurodevelopmental Disabilities at Rutgers Robert Wood Johnson Medical School and The Bristol-Myers Squibb Children's Hospital at Robert Wood Johnson University Hospital. "In the case of anti-NMDAR encephalitis, the body

attacks the NMDA receptors in the brain. This causes a massive malfunction exhibited by a combination of psychiatric, cognitive and motor problems." (NMDA receptors are brain structures that play an important role in learning and memory.)

Bhise and two other Rutgers physicians entered the case after the child's mother requested a second opinion and the family's attending physician reached out to Bhise. The mother told the Rutgers team that after a rapid phase of degeneration of mental state and body function, the child had remained immobile and unresponsive.

Generally, time is of the essence in treating autoimmune diseases and the standard of care dictates that no treatment is useful if too much time has passed, Bhise said. Most of the time, any damage caused by the disease can't be undone.

Bhise arranged for the child to be admitted to the pediatric intensive care unit at The Bristol-Myers Squibb Children's Hospital at Robert Wood Johnson University Hospital, and decided to try one more treatment.

"I said, 'You know, a lot of time has gone by. But I think you still have to try these things,'" Bhise recalled.

The child had been given a course of steroids, pooled antibodies and a long-term immunosuppressant. Bhise and his team decided to administer a series of blood plasma exchanges designed to reset the immune system by cleaning out all the inflammation in the bloodstream.

They saw progress almost instantly.

"Within one or two exchanges, the mom said, 'Hey, I think something's a little different,'" Bhise said. "I mean, no one knew this child better than her mom."

As they continued with the treatment, ultimately with nearly a dozen more plasma exchanges, the child improved steadily until she had made a full recovery.

"I think the lesson that we've learned here is that you can still treat this disease after time has passed," Bhise said. "You shouldn't stop trying. This is important to know so that other folks in the field do not prematurely give up when they see children—and probably adults as well—with difficult-to-treat anti-NMDAR encephalitis."

Other Rutgers physicians who were members of the medical team and authors of the case study included Yisha Cheng, a resident physician in pediatric medicine and a 2020 graduate of Rutgers Robert Wood Johnson Medical School; and Dalya Chefitz, a physician in the department of pediatrics and director of the division of pediatric hospital medicine at The Bristol-Myers Squibb Children's Hospital at Robert Wood Johnson University Hospital.

More information: Yisha Cheng et al, Never Too Late to Treat NMDAR Encephalitis: A Paediatric Case Report and Review of Literature, *EMJ Neurology* (2022). DOI: 10.33590/emjneurol/22-00096

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