Your Questions Answered

MULTIPLE SCLEROSIS THERAPIES

Q A lesion was found at the base of my skull four years ago, and my last MRI revealed a blockage consistent with Arnold Chiari Malformation Type 1. Can you tell me something about this disease?

DR. JASON ROSENBERG RESPONDS:

A Arnold Chiari Malformation Type 1 (CIM) is a developmental malformation of the area toward the back of the skull. When this part of the skull is undersized, the cerebellum can be pushed downward, out of the skull and into the upper neck. This may block the normal flow of cerebrospinal fluid between the neck and skull and cause such common CIM symptoms as headaches, dizziness, and visual problems. Less common symptoms include fainting spells, clumsiness, and difficulty swallowing. Typically, CIM symptoms worsen gradually over time, but they sometimes occur suddenly.

A fluid-filled cavity called a syrinx may also form in the spinal cord itself, resulting in scoliosis, progressive numbness of the arms, or spasticity (stiffnes) and weakness in the legs.

Mild CIM typically has no symptoms and is often discovered during a brain scan being done for another reason. MRI scanning can detect CIM, associated syrinx, and any resulting blockage of cerebrospinal fluid.

A common treatment for CIM is surgery, which involves widening the opening at the base of the skull, expanding the covering membrane, and opening the bony ring of the upper spinal vertebra. Without clear symptoms, there is no real reason for concern or follow-up scans, provided that there is no associated syrinx. In your case it would be important to see a neurologist to determine whether the CIM was present on prior scans, and whether it could have been caused by downward pressure on the cerebellum from your lesion.

Jason Rosenberg, M.D., is assistant professor of neurology and director of the Johns Hopkins Headache Center at Bayview in Baltimore, MD.

DO YOU HAVE A QUESTION TO ASK THE EXPERTS?
Send it to neurologynow@lwwny.com.

MENIERE'S DISEASE

Q My niece was just diagnosed with Rett syndrome. How does this differ from autism?

DR. JEFFREY L. NEUL RESPONDS:

A Rett syndrome is a childhood neurodevelopmental disease that mostly affects girls and is characterized by loss of spoken language and hand use. After normal development, there is gradual regression of mental and physical abilities, usually between six and 18 months of age. Many children become socially withdrawn, avoid eye contact, and dislike being held. After the regression period, these children often regain interest in social interaction. Eye contact returns as a form of communication. But children with Rett syndrome may also develop repetitive hand movements, such as hand wringing or clapping, that remain after the regression period. They may have difficulty walking, abnormal breathing patterns, and an increased risk of developing seizures. A mutation in a specific gene, MECP2, is found in the majority of Rett syndrome cases.

Joanne Picard, in walker, is the daughter of Simone Chalifoux, President of RSSA (Rett Syndrome Society of Alberta).

Autism is more common in boys. It is also characterized by impaired social interaction and communication, but autistic children often do not regain interest in interaction. A small number of people with autism also have a mutation in MECP2.

Neither disorder has a cure. Treatments for both are focused on symptoms, such as improving social interaction in autism and treating seizures in Rett syndrome.

Jeffrey L. Neul, M.D., Ph.D., is assistant professor of pediatrics in neurology at Baylor College of Medicine in Houston, TX, assistant medical director of the Blue Bird Circle Rett Clinic, and principal investigator of the current study "Analysis of the Dopamine System in Rett syndrome," supported by the NIH/NINDS.

THE RETT SYNDROME SOCIETY OF ALBERTA

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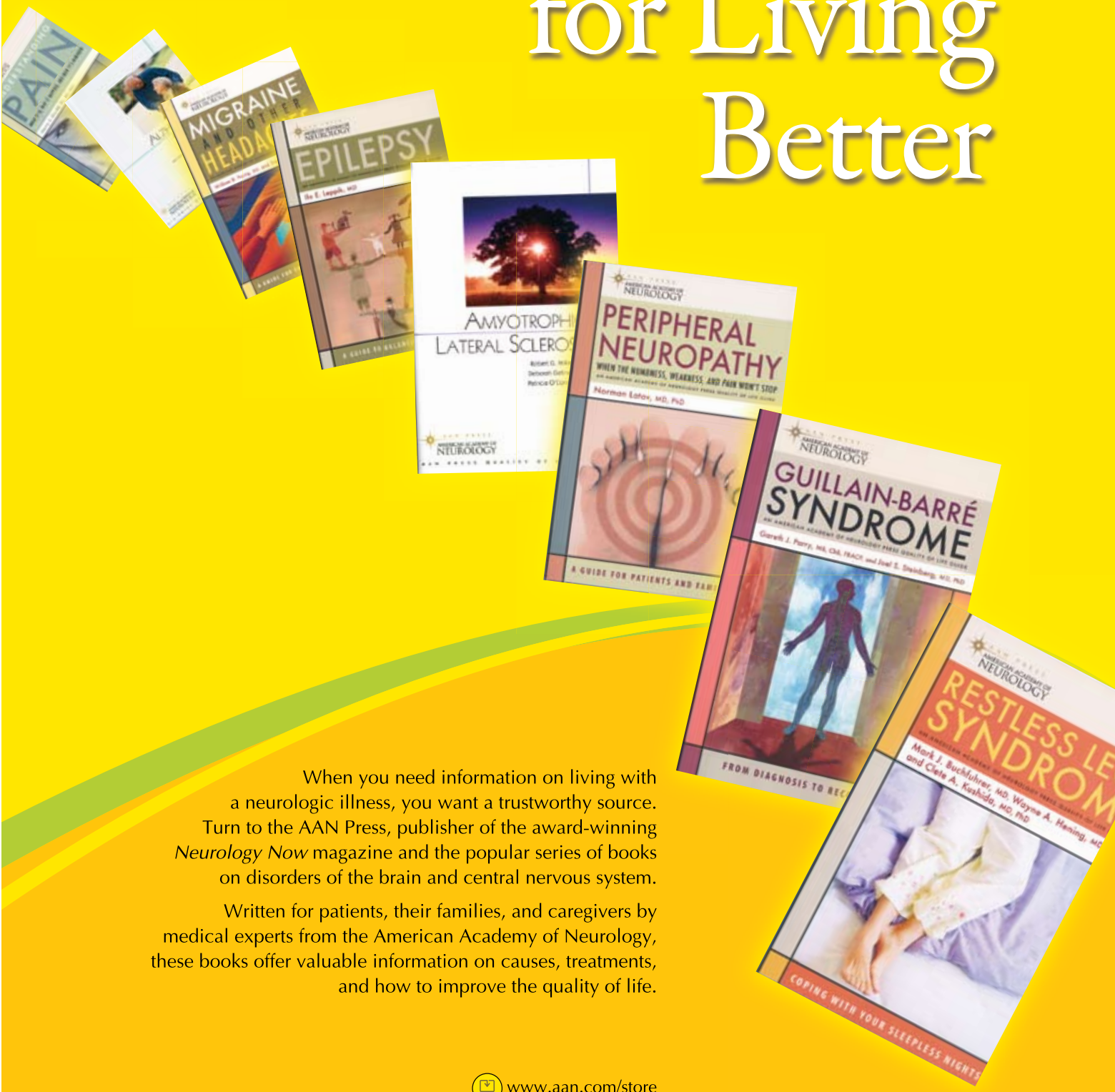

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