

M-H, Huang C-C, Hung P-L, et al. Paraneoplastic neurological disorders in children with benign ovarian tumors. **Brain Dev** 2014 Mar;36(3):248-53).

COMMENTARY. Paraneoplastic disorders (PND) present with multiple manifestations resembling subacute encephalitis, peripheral neuropathy, cerebellar ataxia, opsoclonus myoclonus with neuroblastoma, and other symptoms. Patients with anti-NMDAR encephalitis may present with psychosis, memory deficits, seizures, speech problems, involuntary movements, and breathing disorders. Approximately 50% cases have ovarian tumors, mostly teratoma. In the present study, 5 of 6 (83%) patients with PND and ovarian teratoma had complete remission of symptoms after tumor removal. The authors recommend immunotherapy in all patients following tumor removal, despite apparent recovery after surgery [1].

References.

1. Hsu M-H, et al. *Brain Dev.* 2014 Mar;36(3):248-53.

DEMYELINATING DISORDERS

POTASSIUM CHANNEL KIR4.1-SPECIFIC ANTIBODIES AND ACQUIRED DEMYELINATING DISEASE

Researchers at Technische Universitat, Munich, and other centers in Germany, studied the prevalence of KIR4.1-IgG by ELISA in 47 children with acquired demyelinating disease (ADD), in 22 with other neurologic diseases, 22 with autoimmune disease, and in 18 healthy controls. Serum antibodies to KIR4.1 were identified in 57% of children with ADD but none with other neurologic disease, autoimmune disease or healthy controls. KIR4.1-IgG titers were predominantly found in children with MS or clinically isolated syndrome and in 1 in 3 with demyelinating encephalitis, similar to the prevalence in adults with ADD. KIR4.1-IgG titers were significantly higher in children with ADD compared with control groups ($p < 0.0001$); they were not age-dependent and did not correlate with myelin oligodendrocyte glycoprotein (MOG) antibody responses. MOG-IgG occurs before age 10 y in ADD whereas KIR4.1 antibodies are found in older children and adult patients with MS. (Kraus V, Srivastava R, Kelluri SR, et al. Potassium channel KIR4.1-specific antibodies in children with acquired demyelinating CNS disease. **Neurology** 2014 Feb 11;82(6):470-3).

COMMENTARY. KIR4.1, a potassium channel expressed on oligodendrocytes and astrocytes, contributes to the maintenance of the electrochemical gradient by removing potassium from the extracellular space. Mutations of the KIR4.1 gene cause EAST syndrome characterized by epilepsy, ataxia, sensorineural deafness, and tubulopathy [1]. The prevalence of KIR4.1-IgG in children with MS is similar to adults.

References.

1. Bockenbauer D, et al. *N Engl J Med.* 2009 May 7;360(19):1960-70.