

## **POTASSIUM CHANNEL ANTIBODY ENCEPHALOPATHY**

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Voltage-gated potassium channel (VGKC) antibodies were first described in acquired neuromyotonia which is a syndrome of spontaneous and continuous muscle fiber contraction resulting from hyperexcitability of motor nerves, and may be part of a spectrum of diseases that includes cramp fasciculation syndrome. The patients complain of muscle stiffness, cramps, myokymia (visible undulation of the muscle), pseudomyotonia and weakness. Increased sweating is common and may be a significant problem. Myokymia characteristically continues during sleep and general anaesthesia (unlike muscle stiffness in stiff person syndrome). Sensory symptoms are not uncommon. Antibodies to VGKCs are present in approximately 40% of patients, and are more commonly present in the 20% of patients with an associated thymoma.

In a few patients, there are CNS symptoms such as insomnia, amnesia, hallucinations, delusions, and personality change (usually called Morvan's Syndrome). This is a rare condition which can resolve spontaneously or following treatment. About 40% have a thymoma or other tumour; and only about 60% of all patients have VGKC antibodies.

An increasing number of patients are being identified with a VGKC-antibody-associated encephalopathy, usually described as "limbic encephalitis". These patients do not usually have a tumour although the exact frequency is not yet determined. They usually present with memory loss and seizures but the full clinical spectrum is not yet clear. Some of them appear to have a monophasic illness that may be associated with a preceding infection (compare Guillain Barre syndrome). In the UK there are around 40 new patients identified each year. Some at least of the patients respond well to immunosuppressive treatments, the antibodies fall often to normal values, and the condition does not relapse on cessation of treatment.

Questions and controversies: What is the full clinical spectrum of diseases associated with VGKC antibodies, and are they pathogenic or merely markers for a CNS disease process. How often are VGKC-antibody associated disorders paraneoplastic? What are the antibody characteristics that determine which patients get CNS symptoms, or are there other factors that might distinguish patients with neuromyotonia from those with CNS symptoms. What is the best way to treat these patients, and do they always improve clinically?