

Chronic Lymphocytic Inflammation with Pontine Perivascular Enhancement Responsive to Steroids (CLIPPERS)

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Background

Chronic Lymphocytic Inflammation with Pontine Perivascular Enhancement Responsive to Steroids (CLIPPERS) is an extremely rare, treatable condition with only around 140 cases reported since it was first characterised in 2010. CLIPPERS is a type of encephalomyelitis, which is a general term describing inflammation of the brain and spinal cord. CLIPPERS has been reported in mostly middle-aged and elderly patients and occasionally in children with slightly more males than females being affected.

What are the symptoms?

The symptoms of CLIPPERS usually develop over weeks or months, with very few patients reporting that symptoms occur over a matter of days. Whilst the symptoms of CLIPPERS are varied, the most common symptoms include difficulty with speech (dysarthria), limb weakness, abnormal or uncoordinated movements (ataxia), double vision (diplopia), cognitive impairments, seizures, abnormal facial sensations such as numbness and tingling, and headaches.

Other symptoms can include bouts of sudden uncontrollable and inappropriate laughing or crying (pseudobulbar affect), ringing in the ears (tinnitus), a shaking or tremor in the muscles, uncontrolled eye movements (nystagmus), distortion of taste or smell, and nausea. It can sometimes affect other organs such as the lungs, skin (bumps) or the covering of the eyes (conjunctivitis).

What is the cause?

CLIPPERS is likely an inflammatory condition, although the underlying cause is unknown. However, in some patients, it is preceded by viral infections such as COVID-19, Varicella Zoster virus (VZV) and Hepatitis B virus.

How is it diagnosed?

There is currently no unified diagnostic standard for diagnosing CLIPPERS and no single laboratory or imaging test that can confirm a CLIPPERS diagnosis. Instead, diagnosis is based on the symptoms and various laboratory and imaging tests which are needed to support the diagnosis and to rule out many other conditions with overlapping symptoms. These can include:

- Blood tests

- Tests of the fluids around the brain and spine (via a lumbar puncture)
- Magnetic resonance imaging (MRI) of the brain
- Taking a small sample of the brain (biopsy)

How is it treated?

Most patients with CLIPPERS recover significantly within weeks of starting steroids, an anti-inflammatory medicine. These can be taken either intravenously (through the vein) or orally. However, depending on the severity of the symptoms and the length of these symptoms before treatment, some people might not completely recover. Prolonged treatment with steroids (for at least one year) is usually necessary to prevent symptoms from recurring or worsening. Occasionally, steroid alternatives (like azathioprine) are used when such prolonged treatments are needed. Regular follow-up monitoring with MRI is conducted to assess for recurrence and check for progression to lymphoma (a form of blood cancer).

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