

## **Bickerstaff brainstem encephalitis (BBE)**

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### **Background**

Bickerstaff brainstem encephalitis (BBE) is a rare autoimmune inflammatory disorder of the brainstem and nervous system.

It is named after Edwin Bickerstaff, who described the condition in 1951, and is part of the spectrum of anti-GQ1b antibody-associated diseases, along with Miller Fisher syndrome and Guillain-Barré syndrome.

### **How is it caused?**

Although the exact pathological mechanism is not fully understood, BBE typically occurs after an infectious trigger: usually upper respiratory or gastrointestinal tract infections.

### **What are the symptoms?**

Symptoms generally appear acutely over days to weeks and include:

- Ophthalmoplegia (eye movement paralysis)
- Ataxia (loss of coordination)
- Altered sensorium or level of consciousness (ranging from confusion to coma)

Additional symptoms include:

- Diplopia (double vision)
- Limb weakness
- Facial weakness
- Dysphagia (difficulty swallowing)

### **How is it diagnosed?**

Diagnosis of BBE is based upon the clinical presentation and exclusion of alternate causes of brainstem dysfunction. When present, anti-GQ1b antibodies in the serum provide confirmation, but their absence does not rule out the diagnosis.

Cerebrospinal fluid (CSF) findings are heterogeneous and may demonstrate evidence of inflammation (i.e. elevated protein and/or white blood cells) and in some cases the MRI (magnetic resonance imaging) brain scan may show lesions in the brainstem.

### **How is it treated?**

The illness can be treated by administering immunotherapies, beginning with intravenous immunoglobulin (IVIG) and, in severe cases, plasma exchange.

Corticosteroids such as methylprednisolone may also be used.

### **What are the outcomes?**

With supportive care, most patients recover completely within months. Recovery in childhood is faster than in adulthood. With appropriate treatment, mortality is low. However, some patients may have persistent neurological sequelae and may require physical therapy and neurorehabilitation.

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