

# Rhombencephalitis

Rhombencephalitis (RE) is a syndrome of multiple causes and multiple outcomes. Most authors now use the terms “rhombencephalitis” and “[brainstem encephalitis](#)” interchangeably even though anatomically they are slightly different.

## Etiology

The etiologic categories of RE include infections, autoimmune diseases, and paraneoplastic syndromes (PNS). [Listeria monocytogenes](#) is the most common cause of infectious RE. Listeria RE primarily occurs in healthy young adults. It usually occurs as a biphasic time course with a flu-like syndrome followed by brainstem dysfunction; 75% of patients have a cerebrospinal fluid (CSF) pleocytosis, and almost 100% have an abnormal brain MRI scan. Positive CSF and blood cultures are the most specific for diagnosis.

Enterovirus is probably the second most common infectious cause of RE; however, 95% of cases have occurred in the Asian-Pacific region and there is no specific treatment. Herpes simplex virus (HSV) is the third most common infectious cause of RE, and about 80% of cases are caused by HSV1 and 20% by HSV2. About 50% only had involvement of the brainstem whereas the other 50% also had supratentorial involvement of the temporal and frontal lobes. Mortality with acyclovir treatment was 22% versus those not on acyclovir 75%. Epstein-Barr virus (EBV) and human herpesvirus 6 (HHV6) have caused a few cases. The most common autoimmune etiology is Behçet disease. Over 90% of those with Behçet RE had abnormal MRI scans and 94% had a CSF pleocytosis. Treatment is with corticosteroids and immunosuppressive agents, but only 25% have complete recovery. Paraneoplastic causes are the third category of RE. Brain MRIs are usually normal; there is usually a CSF pleocytosis but the protein is usually normal. Often anti-neuronal antibodies can be found.

## Treatment

Treatment primarily is with ampicillin.

Because Listeria and HSV are the most common treatable acute causes of RE, we recommend empiric therapy with ampicillin and acyclovir for all cases after samples have been obtained from CSF and blood for cultures and the polymerase chain reaction (PCR). Antibiotics can be changed based upon MRI, culture results, PCR results, and antibody studies <sup>1)</sup>.

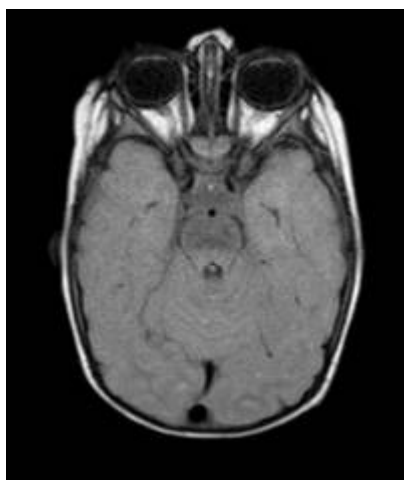
## Outcome

Prognosis is poor and treatment is only partially beneficial.

## Case report

Medina-Flores,R.; Germanwala,A.; Molina,J. T.; Meltzer,C. C.; Wiley,C. A.October 2003: a 59-year-old woman with sudden onset of diplopia. Listerial [rhombencephalitis](#) Brain pathology (Zurich, Switzerland) 2004 ;14(2):225-226

1 year old child [diarrhoea](#) and vomiting later persistent convulsive seizure with cervical hyperextension and hyperextension of the left arm, First treated with [diazepam](#) and [midazolam](#) with subsequent resolution of the crisis. Later empirical treatment with [levetiracetam](#) + [cefotaxime](#), but he continued to have seizures



Increased [signal intensity](#) in [T2](#) and [FLAIR sequences](#) associated with [restricted diffusion](#) foci in the [white matter](#) of the [cerebellar hemispheres](#), posterior region of the [pons](#) with extension to the [dentate nucleus](#), bilateral [thalamus](#), bilateral [periventricular](#) white matter and both [centrum semiovale](#).

No foci of pathological contrast uptake were identified during intravenous contrast administration.

The findings may correspond to signs of cerebellitis, encephalitis and rhomboencephalitis, probably related to an infectious cause, with changes in signal intensity also associated with the seizure episode. An ischemic cause should be considered as a differential finding as a cause of bilateral involvement.

1)  
Jubelt B, Mihai C, Li TM, Veerapaneni P. Rhombencephalitis / brainstem encephalitis. Curr Neurol Neurosci Rep. 2011 Dec;11(6):543-52. doi: 10.1007/s11910-011-0228-5. Review. PubMed PMID: 21956758.

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