# **Encephalitis**

**Encephalitis** is <u>inflammation</u> of the <u>brain</u>.<sup>[5]</sup> Severity is variable.<sup>[1]</sup> Symptoms may include <u>headache</u>, <u>fever</u>, <u>confusion</u>, a <u>stiff neck</u>, and <u>vomiting</u>.<sup>[1]</sup> Complications may include <u>seizures</u>, <u>hallucinations</u>, trouble speaking, <u>memory problems</u>, and problems with hearing.<sup>[1]</sup>

Causes of encephalitis include <u>viruses</u> such as <u>herpes</u> <u>simplex virus</u> and <u>rabies</u> as well as <u>bacteria</u>, <u>fungi</u>, or <u>parasites</u>. Other causes include <u>autoimmune</u> <u>diseases</u> and certain medications. [2] In many cases the cause remains unknown. Risk factors include a weak immune system. [2] Diagnosis is typically based on symptoms and supported by <u>blood</u> tests, <u>medical</u> imaging, and analysis of cerebrospinal fluid. [2]

Certain types are preventable with <u>vaccines</u>. [5] Treatment may include <u>antiviral medications</u> (such as <u>acyclovir</u>), <u>anticonvulsants</u>, and <u>corticosteroids</u>. [1] Treatment generally takes place in hospital. [1] Some people require <u>artificial respiration</u>. [1] Once the immediate problem is under control, <u>rehabilitation</u> may be required. [2] In 2015, encephalitis was estimated to have affected 4.3 million people and resulted in 150,000 deaths worldwide. [3][4]

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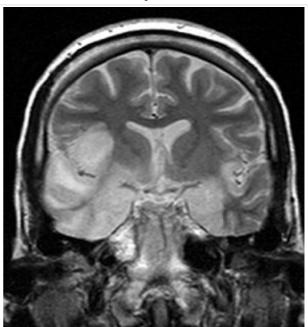
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#### **Encephalitis**



MRI scan image shows high signal in the temporal lobes and right inferior frontal gyrus in someone with HSV encephalitis.

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Specialty	Neurology, Infectious disease
Symptoms	Headache, fever, confusion, stiff neck, vomiting <sup>[1]</sup>
Complications	Seizures, trouble speaking, memory problems, problems hearing <sup>[1]</sup>
Duration	Weeks to months for recovery <sup>[1]</sup>
Types	Herpes simplex, West Nile, rabies, Eastern equine encephalitis, others <sup>[2]</sup>
Causes	Infection, autoimmune, certain medication, unknown <sup>[2]</sup>
Diagnostic method	Based on symptoms, supported by blood tests, medical imaging, analysis of cerebrospinal fluid <sup>[2]</sup>
Treatment	Antiviral medication, anticonvulsants,

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	corticosteroids, artificial respiration <sup>[1]</sup>
Prognosis	Variable <sup>[1]</sup>
Frequency	4.3 million (2015) <sup>[3]</sup>
Deaths	150,000 (2015) <sup>[4]</sup>

# Signs and symptoms

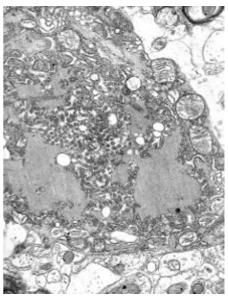
Adults with encephalitis present with acute onset of <u>fever</u>, <u>headache</u>, <u>confusion</u>, and sometimes seizures. Younger children or <u>infants</u> may present with irritability, <u>poor appetite</u> and <u>fever</u>. <u>[6] Neurological</u> examinations usually reveal a <u>drowsy</u> or confused person. <u>Stiff neck</u>, due to the irritation of the <u>meninges</u> covering the brain, indicates that the patient has either meningitis or meningoencephalitis. [7]

#### Cause

#### Viral

Viral encephalitis can occur either as a direct effect of an acute <u>infection</u>, or as one of the <u>sequelae</u> of a <u>latent infection</u>. The majority of viral cases of encephalitis have an unknown cause, however the most common identifiable cause of viral encephalitis is from <u>herpes simplex</u> infection.<sup>[8]</sup> Other causes of acute viral encephalitis are rabies virus, poliovirus, and measles virus.<sup>[9]</sup>

Additional possible viral causes are <u>arboviral flavivirus</u> (St. Louis <u>encephalitis</u>, <u>West Nile virus</u>), <u>bunyavirus</u> (La Crosse strain), <u>arenavirus</u> (lymphocytic choriomeningitis virus), <u>reovirus</u> (Colorado tick virus), and <u>henipavirus</u> infections. [10][11] The <u>Powassan virus</u> is a rare cause of encephalitis.



Rabies virus

#### **Bacterial** and other

It can be caused by a <u>bacterial</u> infection, such as bacterial <u>meningitis</u>, [13] or may be a complication of a current infectious disease syphilis (secondary encephalitis). [14]

Certain parasitic or protozoal infestations, such as <u>toxoplasmosis</u>, <u>malaria</u>, or <u>primary amoebic</u> <u>meningoencephalitis</u>, can also cause encephalitis in people with <u>compromised</u> <u>immune systems</u>. <u>Lyme</u> <u>disease</u> or <u>Bartonella henselae</u> may also cause encephalitis.

Other bacterial pathogens, like <u>Mycoplasma</u> and those causing <u>rickettsial disease</u>, cause inflammation of the meninges and consequently encephalitis. A non-infectious cause includes acute disseminated encephalitis which is demyelinated.<sup>[15]</sup>

# Limbic encephalitis

Limbic encephalitis refers to inflammatory disease confined to the <u>limbic system</u> of the brain. The clinical presentation often includes <u>disorientation</u>, <u>disinhibition</u>, <u>memory loss</u>, <u>seizures</u>, and behavioral anomalies. <u>MRI</u> imaging reveals T2 <u>hyperintensity</u> in the structures of the medial temporal lobes, and in some cases, other limbic structures. Some cases of limbic encephalitis are of autoimmune origin. [16]

#### **Autoimmune encephalitis**

Autoimmune encephalitis signs can include <u>catatonia</u>, <u>psychosis</u>, <u>abnormal</u> movements, and autonomic <u>dysregulation</u>. Antibody-mediated anti-N-methyl-D-aspartate-receptor encephalitis and <u>Rasmussen</u> <u>encephalitis</u> are examples of autoimmune encephalitis. [17] <u>Anti-NMDA receptor encephalitis</u> is the most common autoimmune form, and is accompanied by <u>ovarian teratoma</u> in 58 percent of affected women 18–45 years of age. [18]

#### **Encephalitis lethargica**

<u>Encephalitis lethargica</u> is identified by high fever, <u>headache</u>, delayed physical response, and <u>lethargy</u>. Individuals can exhibit upper body <u>weakness</u>, <u>muscular pains</u>, and <u>tremors</u>, though the cause of encephalitis lethargica is not currently known. From 1917 to 1928, an epidemic of encephalitis <u>lethargica</u> occurred worldwide.<sup>[19]</sup>

# **Diagnosis**

People should only be diagnosed with encephalitis if they have a decreased or altered level of consciousness, lethargy, or personality change for at least twenty-four hours without any other explainable cause. <sup>[20]</sup> Diagnosing encephalitis is done via a variety of tests: <sup>[21]</sup>

- Brain scan, done by MRI, can determine inflammation and differentiate from other possible causes.
- <u>EEG</u>, in monitoring brain activity, encephalitis will produce abnormal signal.
- <u>Lumbar puncture</u> (spinal tap), this helps determine via a test using the cerebral-spinal fluid, obtained from the lumbar region.
- Blood test
- Urine analysis
- Polymerase chain reaction (PCR) testing of the cerebrospinal fluid, to detect the presence of viral DNA which is a sign of viral encephalitis.



Spinal tap on a newborn

### Prevention

Vaccination is available against tick-borne<sup>[23]</sup> and <u>Japanese encephalitis</u><sup>[24]</sup> and should be considered for at-risk individuals. Post-infectious <u>encephalomyelitis</u> complicating <u>smallpox vaccination</u> is avoidable, for all intents and purposes, as smallpox is nearly eradicated.<sup>[25]</sup> Contraindication to <u>Pertussis</u> immunization should be observed in patients with encephalitis.<sup>[26]</sup>

### **Treatment**

Treatment (which is based on supportive care) is as follows:<sup>[27]</sup>

- Antiviral medications (if virus is cause)
- Antibiotics, (if bacteria is cause)
- Steroids are used to reduce brain swelling
- Sedatives for restlessness
- Acetaminophen for fever
- Occupational and physical therapy (if brain is affected post-infection)

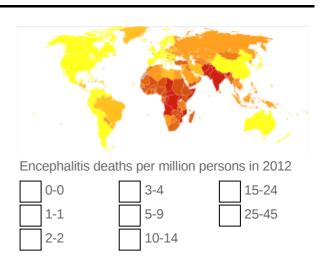
<u>Pyrimethamine</u>-based maintenance therapy is often used to treat Toxoplasmic Encephalitis (TE), which is caused by <u>Toxoplasma gondii</u> and can be life-threatening for people with weak immune systems.<sup>[28]</sup> The use of <u>highly active antiretroviral therapy</u> (HAART), in conjunction with the established pyrimethamine-based maintenance therapy, decreases the chance of relapse in patients with HIV and TE from approximately 18% to 11%.<sup>[28]</sup> This is a significant difference as relapse may impact the severity and prognosis of disease and result in an increase in healthcare expenditure.<sup>[28]</sup>

# **Prognosis**

Identification of poor prognostic factors include <u>cerebral edema</u>, <u>status epilepticus</u>, and <u>thrombocytopenia</u>. [29] In contrast, a normal <u>encephalogram</u> at the early stages of diagnosis is associated with high rates of survival. [29]

# **Epidemiology**

The <u>number of new cases a year</u> of acute encephalitis in Western countries is 7.4 cases per 100,000 people per year. In tropical countries, the incidence is 6.34 per 100,000 people per year. The number of cases of encephalitis has not changed much over time, with about 250,000 cases a year from 2005 to 2015 in the US. Approximately seven per 100,000 people were hospitalized for encephalitis in the US during this time. In 2015, encephalitis was estimated to have affected 4.3 million people and resulted in 150,000 deaths worldwide. Herpes simplex encephalitis has an incidence of 2–4 per million of the population per year.



# **Terminology**

Encephalitis with <u>meningitis</u> is known as <u>meningoencephalitis</u>, while encephalitis with involvement of the spinal cord is known as encephalomyelitis.<sup>[2]</sup>

The word is from <u>Ancient Greek</u> ἐγκέφαλος, *enképhalos* "brain",<sup>[32]</sup> composed of ἐν, *en*, "in" and κεφαλή, *kephalé*, "head", and the medical suffix *-itis* "inflammation".<sup>[33]</sup>

### See also

Rasmussen's encephalitis

- Bickerstaff's encephalitis
- La Crosse encephalitis
- Wernicke's encephalopathy
- Meningitis
- Cerebritis
- Encephalomyelitis
- Zika Virus
- Naegleriasis (primary amoebic meningoencephalitis/PAM)
- World Encephalitis Day

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# **Further reading**

- Steiner, I.; Budka, H.; Chaudhuri, A.; Koskiniemi, M.; Sainio, K.; Salonen, O.; Kennedy, P. G. E. (1 May 2005). "Viral encephalitis: a review of diagnostic methods and guidelines for management". *European Journal of Neurology*. 12 (5): 331–343. doi:10.1111/j.1468-1331.2005.01126.x (https://doi.org/10.1111%2Fj.1468-1331.2005.01126.x). PMID 15804262 (https://pubmed.ncbi.nlm.nih.gov/15804262).
- Basavaraju, Sridhar V.; Kuehnert, Matthew J.; Zaki, Sherif R.; Sejvar, James J. (September 2014). "Encephalitis Caused by Pathogens Transmitted through Organ Transplants, United States, 2002–2013" (https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4178385). Emerging Infectious Diseases. 20 (9): 1443–51. doi:10.3201/eid2009.131332 (https://doi.org/10.3201%2Feid2009.131332). PMC 4178385 (https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4178385). PMID 25148201 (https://pubmed.ncbi.nlm.nih.gov/25148201).

National Center for Biotechnology Information. "Encephalitis" (https://www.ncbi.nlm.nih.gov/pubmedhealth/PMHT0024785/). PubMed Health. National Library of Medicine. Retrieved 2015-08-05.

## **External links**

WHO: Viral Encephalitis (http://www.who.int/topics/encephalitis\_viral/en/)

### Classification ICD-10: A83 (htt D p://apps.who.int/cla ssifications/icd10/br owse/2016/en#/A8 3)-A86 (http://apps. who.int/classificatio ns/icd10/browse/20 16/en#/A86), B94.1 (http://apps.who.int/ classifications/icd1 0/browse/2016/en#/ B94.1), G05 (http:// apps.who.int/classifi cations/icd10/brows e/2016/en#/G05) · ICD-9-CM: 323 (htt p://www.icd9data.co m/getICD9Code.as hx?icd9=323) • MeSH: D004660 (ht tps://www.nlm.nih.g ov/cgi/mesh/2015/M B\_cgi?field=uid&ter m=D004660) · DiseasesDB: 22543 (http://www.d iseasesdatabase.co m/ddb22543.htm) MedlinePlus: **External** resources 001415 (https://ww w.nlm.nih.gov/medli neplus/ency/article/ 001415.htm) • eMedicine: emerg/163 (https://e medicine.medscap

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verview)

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