**Table 5. Comparison of adult autoimmune encephalitis (AE) criteria with proposed classification criteria for pediatric AE**

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| Features of AE | Adult AE criteria29 | Proposed pediatric AE criteria |
| **1. POSSIBLE AE** |
| Clinical and Paraclinical evidence of neuroinflammation | Subacute onset (rapid progression of < 3 months) of working memory deficits (short-term memory loss), altered mental status, or psychiatric symptoms≥ 1 of the following clinical or paraclinical features:* New focal CNS findings
* Seizures not explained by a previously known seizure disorder or other condition
* Presence of inflammatory changes in CSF (CSF leukocytosis > 5 cells/mm3)
* MRI features suggestive of encephalitis (brain MRI hyperintesnse signal on T2/FLAIR sequences highly restricted to one or both medial temporal lobes (limbic encephalitis) or in multifocal areas involving grey matter, white matter or both compatible with demyelination or inflammation)
 | Acute or subacute onset of neurologic and/or psychiatric symptoms over ≤ 3 months in a previously healthy child≥ 2 of the following clinical features:* Altered mental status/level of consciousness, or EEG with slowing or epileptiform activity (may be focal or generalized)
* Focal neurologic deficits
* Cognitive difficulties\*
* Acute developmental regression
* Movement disorder (excluding tics)
* Psychiatric symptoms
* Seizures not explained by a previously known seizure disorder or other condition

Paraclinical testing may not be available |
| AE serology | Not available | Not available |
| Exclusion of other etiologies | Reasonable exclusion of alternative causes, including infectious | Reasonable exclusion of alternative causes, including infectious |
| **2. PROBABLE ANTIBODY-NEGATIVE AE** |
| Clinical and Paraclinical evidence of neuroinflammation | Subacute onset (rapid progression of < 3 months) of working memory deficits (short-term memory loss), altered mental status, or psychiatric symptoms.Exclusion of well-defined syndromes of autoimmune encephalitis (e.g. limbic encephalitis, Bickerstaff’sbrainstem encephalitis, acute disseminated encephalomyelitis).≥ 2 of the following:* MRI abnormalities suggestive of autoimmune encephalitis
* CSF pleocytosis (white blood cell count of more than 5 cells per mm3), CSF-specific oligoclonal bands or elevated CSF IgG index, or both
* Brain biopsy showing inflammatory infiltrates and excluding other disorders
 | Acute or subacute onset of neurologic and/or psychiatric symptoms over ≤ 3 months in a previously healthy child.≥ 2 of the following:* Altered mental status/level of consciousness, or EEG with slowing or epileptiform activity (may be focal or generalized)
* Focal neurologic deficits
* Cognitive difficulties\*
* Acute developmental regression
* Movement disorder (excluding tics)
* Psychiatric symptoms
* Seizures not explained by a previously known seizure disorder or other condition

≥ 1 of the following:* Presence of inflammatory changes in CSF (CSF leukocytosis > 5 cells/mm3, and/or CSF oligoclonal banding)
* MRI features suggestive of encephalitis
* Brain biopsy showing inflammatory infiltrates and excluding other disorders
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| AE serology | Absence of autoantibodies associated with autoimmune encephalitis identified in serum and/or CSF | Absence of autoantibodies associated with autoimmune encephalitis identified in serum and/or CSF |
| Exclusion of other etiologies | Reasonable exclusion of alternative causes, including infectious | Reasonable exclusion of alternative causes, including infectious |

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| **3. DEFINITE AE** |
| Clinical and Paraclinical evidence of neuroinflammation | Subacute onset (rapid progression of < 3 months) of working memory deficits (short-term memory loss), altered mental status, or psychiatric symptoms | Acute or subacute onset of neurologic and/or psychiatric symptoms over ≤ 3 months in a previously healthy child≥ 2 of the following:* Altered mental status/level of consciousness, or EEG with slowing or epileptiform activity (may be focal or generalized)
* Focal neurologic deficits
* Cognitive difficulties\*
* Acute developmental regression
* Movement disorder (excluding tics)
* Psychiatric symptoms
* Seizures not explained by a previously known seizure disorder or other condition

≥ 1\*\* of the following:* Presence of inflammatory changes in CSF (CSF leukocytosis > 5 cells/mm3, and/or CSF oligoclonal banding)
* MRI features suggestive of encephalitis
* Brain biopsy showing inflammatory infiltrates and excluding other disorders
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| AE serology | Presence in the serum and/or CSF of well-characterized autoantibodies associated with autoimmune encephalitis | Presence in the serum and/or CSF of well-characterized autoantibodies associated with autoimmune encephalitis |
| Exclusion of other etiologies | Reasonable exclusion of alternative causes, including infectious | Reasonable exclusion of alternative causes, including infectious |
| \* Severe cognitive dysfunction that is not attributable to a traditional psychiatric syndrome as documented by a qualified clinician (e.g. neurologist, psychiatrist, neuropsychologist), or a significant drop in IQ (>20 points)\*\* When CSF antibodies against N-methyl-D-aspartate receptor (NMDAR), gamma-aminobutyric acid A (GABA-A), glutamic acid decarboxylase 65 (GAD65) are present, further paraclinical markers of neuroinflammation are not required to diagnose definite AE. When only serum antibodies are present, one or more paraclinical marker of neuroinflammation is required.  |