

Clinical diagnosis of autoimmune encephalitis: facts and misfacts

Professor Russell Dale
University of Sydney, Australia



disclosures

- Receive hourly pay for IDMC role for Roche: fingolimod v ocrelizumab RCT
- No COI
- Nil else to disclose

Learning objective

Be aware of the clinical diagnostic criteria of autoimmune encephalitis in adults, and children.

Autoantibody biomarkers have revolutionised autoimmune neurology, and defined treatable neurological conditions.

Identify the pitfalls and vulnerabilities of making a diagnosis of autoimmune encephalitis.

Key messages

There is a clinical diagnostic criteria for autoimmune encephalitis in adults (Graus et al, Lancet Neurology 16)

There is a clinical diagnostic criteria for autoimmune encephalitis, specific to the paediatric patient (Celluci et al, N2 20)

Despite the criteria, there are pitfalls and potential for error, which include:

- Consider whether the clinical phenotype aligns with the suspected diagnosis
- Consider whether the radiological phenotype aligns with the suspected diagnosis
- Provide evidence of CNS inflammation (MRI or CSF)- be suspicious of the AE diagnosis in the absence of CNS inflammation evidence.
- Understand the limitations of autoantibody biomarker testing, including specificity, and tissue to be tested
- Appreciate the presence of seronegative autoimmune encephalitis, but the pitfalls of this diagnosis
- Be aware of the differential diagnosis, and exclude important alternative causes, when possible

References

Graus F, Titulaer MJ, Balu R, Benseler S, Bien CG, Cellucci T, Cortese I, Dale RC, Gelfand JM, Geschwind M, Glaser CA, Honnorat J, Höftberger R, Iizuka T, Irani SR, Lancaster E, Leypoldt F, Prüss H, Rae-Grant A, Reindl M, Rosenfeld MR, Rostásy K, Saiz A, Venkatesan A, Vincent A, Wandinger KP, Waters P, Dalmau J. A clinical approach to diagnosis of autoimmune encephalitis. *Lancet Neurol.* 2016 Apr;15(4):391-404.

Tania Cellucci, Heather Van Mater, Francesc Graus, Eyal Muscal, William Gallentine, Marisa S. Klein-Gitelman, Susanne M. Benseler, Jennifer Frankovich, Mark P. Gorman, Keith Van Haren, Josep Dalmau, Russell C. Dale. Clinical approach to the diagnosis of autoimmune encephalitis in the pediatric patient. *Neurol Neuroimmunol Neuroinflamm* Mar 2020, 7 (2) e663; DOI:10.1212/NXI.0000000000000663

Dalmau J, Graus F. Diagnostic criteria for autoimmune encephalitis: utility and pitfalls for antibody-negative disease. *Lancet Neurol.* 2023 Jun;22(6):529-540.