

## Tabel over anatomisk klassifikation, klinisk præsentation og differentialdiagnoser ved Autoimmun Encefalitis.

Modificeret fra Abboud H, et al. J Neurol Neurosurg Psychiatry 2021;0:1–12. doi:10.1136/jnnp-2020-325300

| Anatomisk klassifikation                        | Klinisk syndrom  | Associerede antistoffer   | Differential diagnoser  | Yderligere undersøgelser   |
|---|--|---|---|--|
| <b>Limbisk encefalitis</b>                      | -Kognitiv<br>-Psikiatrisk<br>-Epileptisk                                       | Hu, CRMP5/CV2, Ma2, NMDAR, AMPAR, LGI1, CASPR2, GAD65, GABABR, DPPX, mGluR5, AK5, Neurexin-3α | HSV, VZV, HHV6  | CSF virus PCR, CSF VZV/HSV IgG/IgM   |
| <b>Kortikal/subkortikal encefalitis</b>         | -Kognitiv<br>-Epileptisk   | PCA-2 (MAP1b), NMDAR, GABA A/B R, DPPX, MOG   | ADEM, AHL, tumefaktiv MS, PML, CJD, lupus cerebritis, Behcet, neurosarcoidosis, neurosyfilis, lymfom, anoxisk skade, Krampe-relaterede forandringer | MOG-IgG, CSF JCV PCR, CSF RTQuIC, ANA/ ENA, HLA-B51, ACE/IL2R, CT thorax/FDG PET (sarkoidose), Wasserman/RPR, CSF cytologi og flow cytometri |
| <b>Striatl encefalitis</b>                      | -Bevægeforstyrrelse  | CRMP5/CV2, DR2, NMDAR, LGI1, PD10A  | CJD, toksisk encefalopati, anoxisk skade, hyperglykæmisk skade, uræmi   | RTQuIC, CSF virus PCR, Toksikologisk og metabolisk screening   |
| <b>Diencefal encefalitis</b>                    | -Autonom<br>-Søvnforstyrrelser   | Ma 1–2, IgLON5, DPPX, AQP4  | Neurosarcoidosis, Behcet, Wernicke, Mb. Whipple   | ACE/IL2R, CT thorax/FDG PET (sarkoidose), HLA-B51, thiamin   |
| <b>Hjernestamme encefalitis</b>                 | -Kognitiv<br>-Bevægeforstyrrelse<br>-Kranio-bulbær                             | Ri, Ma 1–2, KLHL11, IgLON5, DPPX, AQP4, MOG, GQ1b   | Rhombencephalitis (listeria), viral infektion, CLIPPERS, neurosarcoidose, Behcet, lymfoma, PML, pontin myelinolyse, Erdheim-Chester, Mb. Whipple    | CSF dyrkning, CSF virus PCR, HLA-B51, CSF cytologi og flow cytometri, CSF JCV PCR, knogleskanning  |
| <b>Cerebellitis eller cerebral degeneration</b> | -Ataktisk  | Hu, Ri, Yo, Tr, CASPR2, KLHL11, NIF, mGluR1, GAD65, VGCC                                      | Viral or post-viral cerebellitis, coeliaki, Miller Fisher syndrome, vitamin E mangel, MSA-C, SCA  | Viral PCR, coeliaki screening, anti-GQ1b, vitamin E, DaT/PE2I skanning   |
| <b>Meningoencefalitis</b>                       | -Kognitiv<br>-Epileptisk<br>-Meningeal   | GFAP  | Tuberkulose, neurosarcoidose, Behcet, bacterial or viral infection, leptomeningeal carcinomatose, GPA, IgG4 - related disease                       | Mikrobiom 16S/18S, ACE/IL2R, CT thorax/FDG PET (sarkoidose), HLA-B51, CSF dyrkning, CSF virus PCR, CSF cytologi og flow cytometri            |
| <b>Encefalomyelitis</b>                         | -Bevægeforstyrrelse inkl PERM og SPSP<br>-Spinal presentation<br>-Optikospinal | GAD65, amfifysin, glycine receptor, PCA-2 (MAP1B), GABA A/B R, DPPX, CRMP5/CV2, AQP4, MOG     | ADEM  | MOG-IgG, CSF virus PCR   |

Forkortelser: ADEM, akut dissemineret encefalomyelitis; AHL, akut hæmorrhagisk leukoencefalitis; ANA, antinuclear antibody; AQP4, aquaporin-4; CJD, Creutzfeldt-Jacob disease; CLIPPERS, chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids; DaT scan, dopamine transporter scan; ENA, extractable nuclear antigens; GFAP, glial fibrillar acidic protein; GPA, gliomatose med polyangitis; GQ1b, anti-ganglioside Q1B; HHV6, human herpes virus-6; HSV, herpes simplex virus; JCV, John Cunningham virus; MOG, myelin oligodendrocyte glycoprotein; MS, multipel sklerose; MSA-C, cerebellar multi system atrofi; NMOSD, neuromyelitis optica spectrum disorder; PERM, progresive encephalomyelitis with rigidity and myoclonus; PML, progressiv multifocal leukoencefalopati; SCA, spinocerebellar ataxia; SPSP, Stiff-Person Syndrome Disorder; VZV, varicella zoster virus.