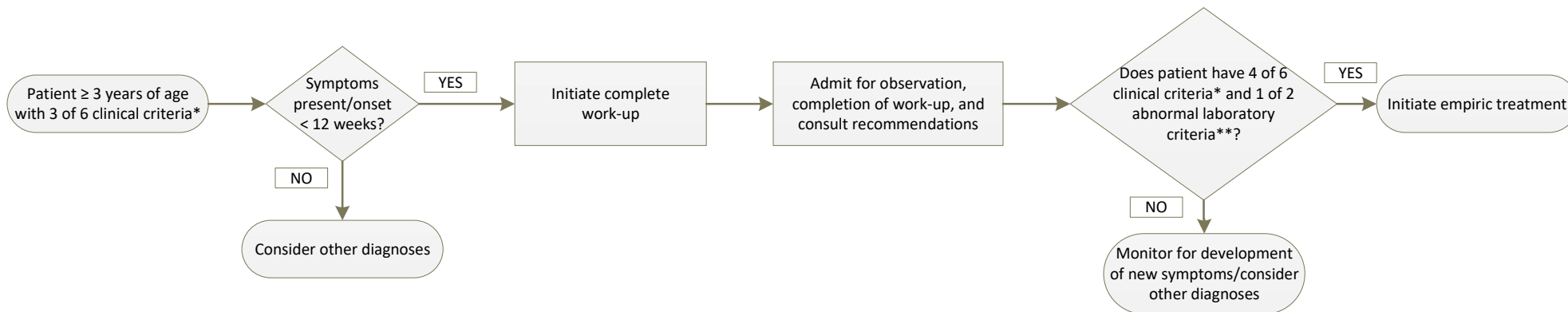


Autoimmune Encephalitis

Clinical Practice Guideline



*Clinical Criteria

- 1) Abnormal psychiatric behavior or cognitive dysfunction
- 2) Speech dysfunction (pressured speech, verbal reduction, mutism)
- 3) Seizures
- 4) Movement disorder, dyskinesia
- 5) Decreased level of consciousness
- 6) Autonomic instability (more than one set of vitals, more than one vital sign involved)

-requires trends of vital signs over time, and most typically develops later in the course. This will be most used in determining need for treatment and appropriate placement (ICU vs. acute care)

**Laboratory Criteria

- 1) Abnormal EEG (focal or diffuse slowing, epileptic activity, or extreme delta brush)
- 2) CSF with pleocytosis or oligoclonal bands

Exclusion Criteria

- 1) Age < 3 years
- 2) History of behavioral/psychiatric symptoms > 12 weeks
- 3) Less than 3 clinical criteria
- 3) Alternate diagnosis more likely

Work-Up

Consults

- Neurology
- Psychiatry
- Rheumatology
- *ID if fever and CSF pleocytosis and /or MRI concerning for infection*

Labs

- ANA with Rfx ENA/DNA
- Anti-thyroid Perox antibodies
- Anti-Thyroglobulin antibodies
- CBC with differential
- CMP
- CRP
- ESR
- MOG Ab
- Urinalysis
- Urine drug screen
- CSF- opening pressure, cell counts, protein, glucose, gram stain/culture, oligoclonal bands/IgG index, NMDA Ab (ARUP- *see lab ordering guide*), freeze tube

Imaging/Procedures

- EEG (length to be determined by neurology)
- MRI brain with and without contrast
- Pelvic ultrasound (females), testicular ultrasound (males)

Other

- Busch-Francis catatonia Rating Scale (BFCRS)- *psychiatry to perform*
- Vitals signs every 4 hours

Lab Ordering Guide

CSF NMDA only (no serum)

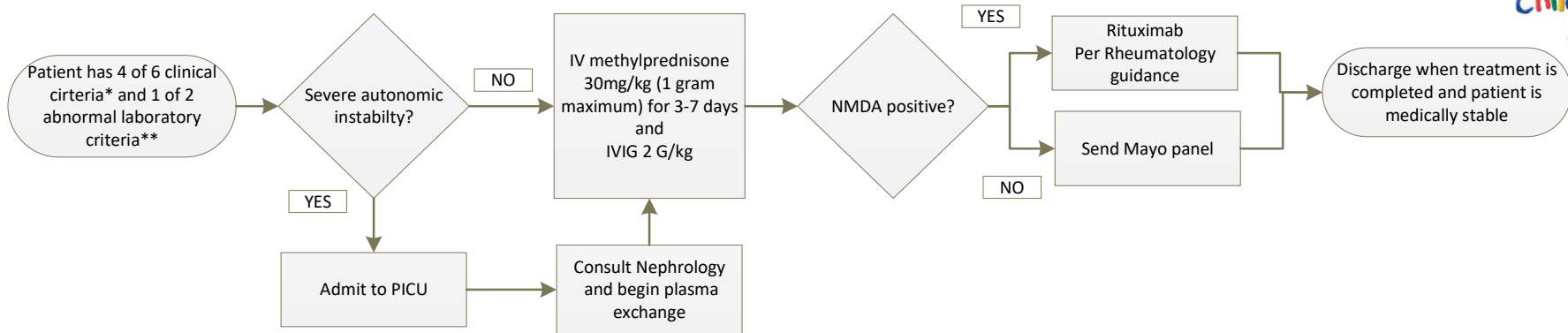
- Lab miscellaneous: ARUP test # 2005165 NMDA receptor Ab IgG CSF with reflex titer

If NMDA is negative and suspicion is high:

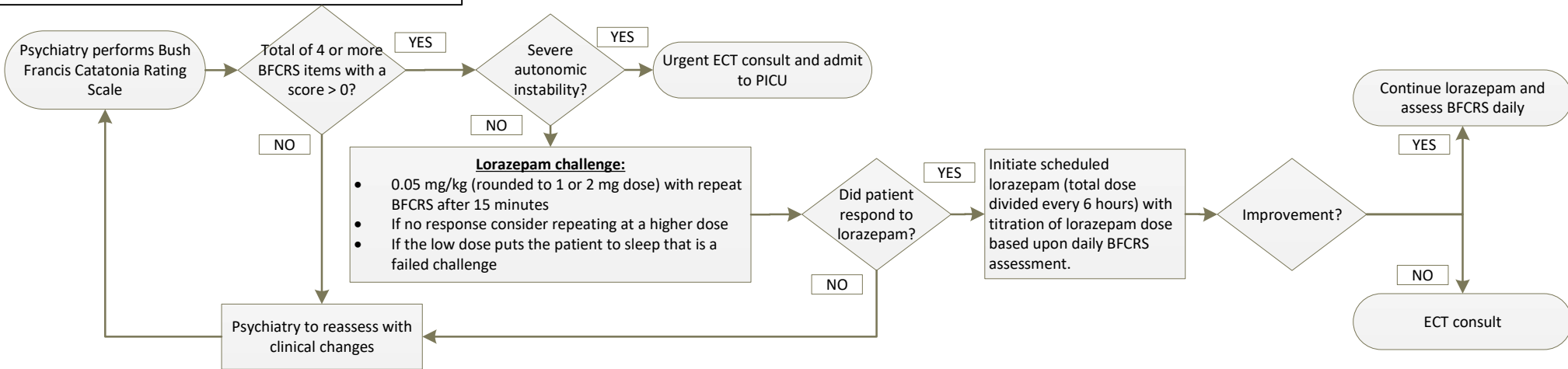
Encephalitis panel- Mayo

- Lab miscellaneous: Mayo # ENCI Encephalopathy AI evaluation, spinal fluid

Empiric Treatment



Catatonia Management Bush Francis Catatonia Rating Scale (BFCRS)



References:

Albert DV, Pluto CP, Weber A, et al. Utility of Neurodiagnostic Studies in the Diagnosis of Autoimmune Encephalitis in Children. *Pediatr Neurol.* 2016;55:37-45.

Dalmau J, Lancaster E, Martinez-herandez E, Rosenfeld MR, Balice-gordon R. Clinical experience and laboratory investigations in patients with anti-NMDAR encephalitis. *Lancet Neurol.* 2011;10(1):63-74.

De bruijn MAAM, Aarsen FK, Van oosterhout MP, et al. Long-term neuropsychological outcome following pediatric anti-NMDAR encephalitis. *Neurology.* 2018;90(22):e1997-e2005.

Graus F, Titulaer MJ, Balu R, et al. A clinical approach to diagnosis of autoimmune encephalitis. *Lancet Neurol.* 2016;15(4):391-404.

Gresa-arribas N, Titulaer MJ, Torrents A, et al. Antibody titres at diagnosis and during follow-up of anti-NMDA receptor encephalitis: a retrospective study. *Lancet Neurol.* 2014;13(2):167-77.

Titulaer MJ, Mccracken L, Gabilondo I, et al. Treatment and prognostic factors for long-term outcome in patients with anti-NMDA receptor encephalitis: an observational cohort study. *Lancet Neurol.* 2013;12(2):157-65.

Ho ACC, Mohammad SS, Pillai SC, et al. High sensitivity and specificity in proposed clinical diagnostic criteria for anti-N-methyl-D-aspartate receptor encephalitis. *Dev Med Child Neurol.* 2017;59(12):1256-1260.

Meirenske Wang R, Guan HZ, Ren HT, Wang W, Hong Z, Zhou D. CSF findings in patients with anti-N-methyl-D-aspartate receptor-encephalitis. *Seizure.* 2015;29:137-42.

Wilson JE, Carlson R, Duggan MC, et al. Delirium and Catatonia in Critically Ill Patients: The Delirium and Catatonia Prospective Cohort Investigation. *Crit Care Med.* 2017;45(11):1837-1844.