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
4. Alternate diagnosis more likely

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**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
- 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

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Does patient have 4 of 6 clinical criteria* and 1 of 2 abnormal laboratory criteria**?

YES

Initiate empiric treatment

NO

Monitor for development of new symptoms/consider other diagnoses

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Symptoms present/onset < 12 weeks?

YES

Initiate complete work-up

NO

Consider other diagnoses

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Patient ≥ 3 years of age with 3 of 6 clinical criteria*

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**Work-Up**

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

Labs
- ANA with Rfx ENA/DNA
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**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

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This guideline does not take into account individual patient situations, and does not substitute for clinical judgment

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Empiric Treatment

- Patient has 4 of 6 clinical criteria* and 1 of 2 abnormal laboratory criteria**
  - Severe autonomic instability?
    - YES: IV methylprednisolone 30mg/kg (1 gram maximum) for 3-7 days and IVIG 2 G/kg
    - NO: Admit to PICU
  - NMDA positive?
    - YES: Rituximab Per Rheumatology guidance
    - NO: Send Mayo panel
  - Discharge when treatment is completed and patient is medically stable

Catatonia Management
Bush Francis Catatonia Rating Scale (BFCRS)

- Psychiatry performs Bush Francis Catatonia Rating Scale
  - Total of 4 or more BFCRS items with a score > 0?
    - YES: Urgent ECT consult and admit to PICU
    - NO: Severe autonomic instability?
      - YES: Lorazepam challenge:
        - 0.05 mg/kg (rounded to 1 or 2 mg dose) with repeat BFCRS after 15 minutes
        - If no response consider repeating at a higher dose
        - If the low dose puts the patient to sleep that is a failed challenge
      - NO: Psychiatry to reassess with clinical changes
  - Did patient respond to lorazepam?
    - YES: Initiate scheduled lorazepam (total dose divided every 6 hours) and assess BFCRS daily
    - NO: Continue lorazepam and assess BFCRS daily
  - Improvement?
    - YES: ECT consult
    - NO: Psychiatry to reassess with clinical changes

References:


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