**Diagnosis of IBrainD or Primary CNS vasculitis**

**Childhood CNS Vasculitis or Inflammatory Brain Disease (Suspected)**

**Clinical Presentation:**
Child presents with newly acquired neurological deficits (focal and/or diffuse) and/or psychiatric symptoms

**Evaluation — First Step:**
**Clinical Assessment:**
General assessment for underlying disease/illness; Detailed neurological tests; Psychiatric/neurocognitive assessment (when applicable)

**Blood Tests:**
Blood inflammatory markers (e.g. ESR, CRP, CBC); Auto-antibody testing (e.g. antinuclear antibody, antineutrophil cytoplasmic antibody, IgG)

**Diagnostic Imaging:**
Cerebral MRI (with gadolinium enhancement, T2, and FLAIR); MRA with post-gadolinium vessel-wall imaging

**Examples of IBrainD diagnosed at this step:**
- ADEM, MS, Optic Neuritis
- Angiography-positive (large vessel) childhood CNS vasculitis
- Examples of IBrainD diagnosed at this step:
  - Anti-NMDA Receptor Encephalitis, Limbic Encephalitis, Hashimoto Encephalitis, NMO

**Evaluation — Second Step:**
**CSF analysis:**
Opening pressure; CSF cell count; Infectious work-up (bacterial, fungal, and viral cultures and serology); Oligoclonal banding; Neuronal antibodies (e.g., anti-NMDAR, anti-AMPA, anti-AQP4, anti-LGI1)

**Blood Tests:**
Anti-NMDAR, anti-AMPA, anti-AQP4, anti-LGI1

**Diagnostic Imaging:**
Conventional angiography

**Examples of IBrainD diagnosed at this step:**
- Anti-NMDA Receptor Encephalitis, Limbic Encephalitis, Hashimoto Encephalitis, NMO

**Evaluation — Third Step:**
**Brain biopsy**
Lesional or nonlesional; nondominant frontoparietal lobe

**Examples of IBrainD diagnosed at this step:**
- Rasmussen’s Encephalitis, Neurosarcoïdosis

**Other Diagnosis**

- Noninflammatory vasculopathies
- Mimic of IBrainD
- Secondary IBrainD (vasculitic and nonvasculitic)

- Angiography-negative (small vessel) childhood CNS vasculitis

- Angiography-negative (small vessel) childhood CNS vasculitis

- Examples of IBrainD diagnosed at this step:
  - Rasmussen’s Encephalitis, Neurosarcoïdosis