

Diagnosis of IBrainD or Primary CNS vasculitis

Childhood CNS Vasculitis or Inflammatory Brain Disease (Suspected)

Other Diagnosis

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

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

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

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

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

Evaluation—Third Step:

Brain biopsy

Lesional or nonlesional; nondominant frontoparietal lobe

• **Angiography-negative (small vessel) childhood CNS vasculitis**
• **Examples of IBrainD diagnosed at this step:**
Rasmussen's Encephalitis, Neurosarcoidosis

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