Clinical Validation Guidelines

ENCEPHALOPATHY

Definition: a diffuse alteration in brain function or structure caused by disease, injury, drugs or chemicals. The alteration can be acute (mental abilities or cognition) or chronic (brain structure).

Common neurological symptoms of encephalopathy can include loss of memory and cognitive ability, changes in personality, inability to concentrate, lethargy and progressive loss of consciousness.

Diagnostic Criteria [1,2]: To clinically validate the following diagnoses the listed criteria must be met AND the diagnosis must be documented by a physician in the medical records.

(G93.40) Chronic (structural) encephalopathy (ALL of the following Criteria #1 and Criteria #2 is needed to make the diagnosis):

1) Irreversible alteration in brain function due to permanent brain damage, **WITH**
2) Diffuse (generalized) or focal changes in brain function/mental status that are slowly progressive.

Causes include trauma, HIV-related, toxic heavy metals (lead, arsenic, mercury), Korsakoff (alcohol), anoxia and hereditary enzyme deficiencies.

(G93.40) Acute (functional) encephalopathy (ALL of the following Criteria #1- #6 are needed to make the diagnosis):

1) Documented change in baseline GCS score (GCS <15) or baseline level of orientation (AAO < 3-4).
2) Absence of a structural abnormality (no identifiable mass/neoplasm, stroke, seizure-activity or post-ictal states)
3) Caused by an identifiable underlying condition, **AND**
4) Symptoms are persistent (no "waxing and waning" of the altered mental function), **BUT**
5) Reversible and resolves when the underlying cause is corrected
6) Does not meet criteria for chronic encephalopathy (see above).

(G93.41) Metabolic Encephalopathy (ALL of the above Criteria #1- #6 are needed to make the diagnosis), **AND:**

- Alteration is due to a metabolic disturbance (infections, fevers, dehydration, electrolyte imbalance or hypoxemia).

(G92.9) Toxic Encephalopathy (ALL of the above Criteria #1- #6 are needed to make the diagnosis), **AND:**

- Alteration is due to medications, illicit drugs or chemicals.

(G92.8) Toxic –Metabolic encephalopathy (ALL of the above Criteria #1- #6 are needed to make the diagnosis), **AND:**

- Alteration is due to a failure in metabolism or excretion which results in an accumulation of toxic levels of naturally occurring metabolites and waste products (e.g. acute organ failure).

Common Causes of Encephalopathy:

- Sepsis, UTI or other infection
- Overdose or incorrect usage of medication or illicit drug
- Dehydration
- Electrolyte imbalance (hyponatremia)
- Organ failure

Clinical Indicators of Encephalopathy:

- Comparisons of present mental function to baseline mental function (e.g. nursing notes)
- If no toxic or metabolic factors are evident or are not the likely cause of the patient's mental status change, encephalopathy is unlikely the diagnosis.
- CT/MRI and other imaging expected to be unremarkable since the brain abnormality is functional, not structural.
- Unlikely to be encephalopathy if mental status does not improve during hospitalization.

Differential Diagnoses for Encephalopathy:

- **(F10.221)** Intoxication delirium is likely to be consistent with a toxic encephalopathy due to alcohol but requires provider specification of the toxic encephalopathy. Documentation should state "intoxication delirium caused by toxic encephalopathy due to alcohol."
- **(F10.231)** Alcohol dependence with withdrawal delirium (delirium tremens) is not a toxic encephalopathy since the toxic substance has actually been withdrawn.
- **(K76.82)** Hepatic encephalopathy is a chronic condition. An elevated ammonia level in the presence of neurological impairment (e.g. altered mental status, combativeness, somnolence) and the presence of underlying liver disease, such as liver cirrhosis, would indicate this diagnosis.
- **(G40.419):** Post-ictal State: when encephalopathy is due to a postictal state it is not coded separately since it is considered to be integral to the seizure. Therefore this diagnosis will

fall under Generalized epilepsy and epileptic syndromes, intractable, without status epilepticus.

- **(F05.9)** Delirium (ALL of the above Criteria #1-#4 are needed to make the diagnosis)**:**
  1) Presence of a disturbance in attention (i.e. reduced ability to direct, focus, sustain and shift attention) and awareness (reduced orientation to the environment) that is:
     a. Acute (develops over hours or days) and demonstrates a change from baseline, AND
     b. Fluctuates over the course of the day
  2) There is an additional disturbance in cognition (related to memory, disorientation, language, and perception).
  3) The disturbance cannot be better explained by another pre-existing, evolving or already determined neurocognitive disorder and does not occur in the context of severely reduced level of arousal (e.g. coma).
  4) The history, physical exam or laboratory findings provide evidence that the disturbance is a direct physiological consequence of another medical condition (substance intoxication or withdrawal, exposure to a toxin, or due to multiple etiologies).

Common Causes of Delirium:

- Pain, infection, nutrition, constipation, hydration, medication and environment

Clinical Indicators of Delirium:

- The Confusion Assessment Method (CAM) and the Confusion Assessment Method for the Intensive Care Unit (CAM-ICU) are tools used to identify delirium in patients.
- Delirium describes the same set of clinical circumstances as encephalopathy however delirium is a symptom whereas encephalopathy is a medical condition. If delirium is documented in the presence of diagnostic criteria for acute encephalopathy, the later cannot be assumed to be present without provider documentation of the encephalopathy (e.g. “delirium due to metabolic encephalopathy.”)

(F01-F03) Dementia (ALL of the above Criteria #1- #4 are needed to make the diagnosis)**:**

1) There is a decline from baseline cognitive function that involves ONE or more of the following cognitive domains:
   a. Learning/memory
   b. Language
   c. Executive function
   d. Complex attention
   e. Perceptual-motor skills

    f. Social cognition
2) The decline is progressive (no acute changes in mental function)
3) The decline affects daily functioning and independence
4) Sun-downing (presence of waxing and waning symptoms of altered mental status) occurs. This can manifest as agitation, anxiety or confusion during the late afternoon or evening.

Common Causes of Dementia:

- The most common form of dementia in older adults is Alzheimer's Disease which is most often diagnosed after the age of 65 years.
- Other forms of dementia include: vascular dementia, dementia with Lewy bodies, Pick's disease and Parkinson's.

Clinical Indicators of Dementia:

1) Diagnosis is based the patient's medical history, physical exam and mental status exams.
2) Labs and imaging tests are performed to rule out other conditions.
3) Common mental status exams used to diagnose dementia include the Mini-Mental State Exam (MMSE) and the Mini-Cog Test.
4) Patients with dementia who present with acute cognitive dysfunction such as encephalopathy or delirium should demonstrate a clear difference between baseline mental function and acute presentation. When the acute issue is resolved, the documentation should demonstrate a return to baseline function.
5) Nursing and case management notes can be reviewed to clarify baseline function.

References:

1) Tang, C., Pinson, R. (2023). Encephalopathy. CDI Pocket Guide (16th Edition), Pages 136-141.
2) Prescott, L., James, M. (2023). Encephalopathy. ACDIS Pocket Guide: The Essential CDI Resource. Pages 137-145.
    a. Dementia, pages 130-136.
    b. Delirium, pages 465-468.