Encephalopathy

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The diagnosis and documentation of encephalopathy can be confusing and controversial. Clinicians struggle with, for example, the distinction between delirium and encephalopathy, and with recognizing encephalopathy when a patient with dementia seems to have an acute alteration in baseline mental status.

A specific, authoritative definition of encephalopathy is hard to come by. Non-specific descriptions abound online: any brain disease, disorder or disease of the brain, dysfunction of the brain, degeneration of brain function, and brain damage or malfunction. The National Institute of Neurological Disorders and Stroke (NINDS) has described encephalopathy as a term for “any diffuse disease of the brain that alters brain function or structure” and says the “hallmark of encephalopathy is an altered mental status.” It then proceeds to list a hodge-podge of acute and chronic causes.

Categories of encephalopathy

There are 2 distinct categories of encephalopathy: acute and chronic. Many sources confuse and confound these categories, lumping them together as one. The chronic encephalopathies are characterized by chronic mental status alteration that, in most cases, is slowly progressive (anoxic encephalopathy being an exception). They result from permanent, usually irreversible, structural changes within the brain itself. Some may be halted or reversed by early detection and treatment. Examples of the chronic encephalopathies include anoxic brain injury; chronic traumatic encephalopathy; heavy metals (lead, arsenic, mercury, etc); HIV-related; hereditary enzyme deficiencies; Korsakoff; and spongiform.

Acute encephalopathy is characterized by an acute or subacute global, functional alteration of mental status due to systemic factors. It is reversible when these abnormalities are corrected, with a return to baseline mental status. Acute encephalopathy may be further identified as toxic, metabolic, or toxic-metabolic. Toxic encephalopathy describes acute mental status alteration due to medications, illicit drugs, or toxic chemicals. Metabolic encephalopathy is caused by any of a large number of metabolic disturbances. Toxic-metabolic encephalopathy describes a combination of toxic and metabolic factors. Causes of acute toxic and metabolic encephalopathy include acute organ failure such as hepatic and renal; alcohol; dehydration; electrolyte
imbalance; fever; hypertension; hypoxemia; illicit drugs; infections including sepsis; medications; toxic chemicals; and Wernicke (thiamine deficiency).

Acute intra-cranial processes (such as stroke or traumatic lesions) alone should not be classified as acute encephalopathy but are more correctly considered an alteration of consciousness (stupor or coma) or concussion.

In contrast to the generic term “encephalopathy,” the acute toxic and metabolic encephalopathies as a group are well defined and well described. The 2013 Neurocritical Care Society Practice Update states that “acute encephalopathy is synonymous with acute confusional state, acute organic brain syndrome or delirium...[it] describes the clinical presentation of a global cerebral dysfunction induced by systemic factors.”

**Delirium vs. acute encephalopathy**

Delirium and acute encephalopathy are essentially 2 different terms describing the same condition. Delirium represents the mental manifestation while encephalopathy identifies the underlying pathophysiologic process. This is why the American Psychiatric Association's Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5), classifies acute toxic and metabolic encephalopathic states as delirium and does not use encephalopathy in its definitions.

The coding classifications (ICD-9 and ICD-10) use “encephalopathy” to classify what DSM-5 calls delirium. ICD relegates delirium to a symptom of lesser importance. To permit correct coding for these cases, the term encephalopathy is needed to capture a true picture of the patient's condition. Clinicians may continue to follow DSM definitions using delirium but should also incorporate the necessary ICD terminology to prevent understating the severity of illness of patients. Examples include:

- Toxic encephalopathy due to phenytoin, causing delirium
- Delirium due to metabolic encephalopathy

In fact, DSM-5 acknowledges this imperative in a “coding note” for delirium: “Include the name of the other [underlying] medical condition in the name of the delirium (e.g., 293.0 [F05] delirium due to hepatic encephalopathy). The other medical condition should also be coded and listed separately immediately before the delirium [hepatic encephalopathy first, delirium second].”

**Acute encephalopathy complicating dementia**

A common clinical conundrum is presented by the patient with preexisting dementia who is admitted with an altered mental status. Patients with dementia often experience transient, episodic mental status fluctuation, and it may be difficult to determine if any change in baseline has actually occurred. On the other hand, patients with dementia are particularly vulnerable to acute encephalopathic changes with any metabolic or toxic stress.
When a significant alteration of mental status has occurred and systemic toxic or metabolic factors are present, it is a rather simple matter to diagnose toxic or metabolic encephalopathy when mental status returns to baseline as these abnormalities are corrected. If delirium due to these underlying medical conditions is diagnosed, toxic or metabolic encephalopathy should also be identified as its cause (see DSM-5 coding note above).

The chronic encephalopathies (such as Korsakoff, anoxic, or traumatic) are distinctly different from acute toxic or metabolic encephalopathy. They result from permanent, usually irreversible, structural changes within the brain itself.

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