

SUPPLEMENT 2

RELEVANT EXTRACTS FROM DSM-5

The following summarizes the neurocognitive disorders in DSM-5. For the complete DSM-5 see Diagnostic and Statistical Manual of Mental Disorders, 5th edn. 2013, American Psychiatric Association. Washington, DC.

DSM-5 NEUROCOGNITIVE DISORDERS (PAGE 591)

The neurocognitive disorders (NCDs) begin with delirium, followed by the syndromes of major NCD, mild NCD and their etiological subtypes. These subtypes are listed as

- Alzheimer's disease
- Vascular NCD
- NCD with Lewy bodies,
- NCD due to Parkinson's disease
- Frontotemporal NCD
- NCD due to traumatic brain injury
- NCD due to HIV infection
- Substance/medication induced NCD
- NCD due to Huntington's disease
- NCD due to prion disease
- NCD due to other medical conditions
- NCD due to multiple etiologies
- Unspecified NCD

The NCD category encompasses the group of disorders in which the primary clinical deficit is in cognitive function, and that are acquired rather than developmental.

The NCDs are unique among DSM-5 categories that these are syndromes for which the underlying pathology, and frequently the etiology as well, can be potentially determined. The various underlying disease entities have all been the subject of extensive research, clinical experience, and expert consensus on diagnostic criteria. The DSM-5 criteria for these disorders have been developed in close consultation with the expert groups for each of these disease entities and align as closely as possible with the current consensus criteria for each of them. The potential ability of biomarkers is also discussed in relation to diagnosis. Dementia is subsumed under the newly named entity *major neurocognitive disorder*, although the term *dementia* is not precluded from the etiological subtypes in which that term is standard. Furthermore, DSM-5 recognizes a less severe level of cognitive impairment, *mild neurocognitive disorder*, which can also be a focus of care, and which in DSM-4 was subsumed under the title "Cognitive Disorder Not Otherwise Specified". Diagnostic criteria are provided for both these syndromic entities, followed by diagnostic criteria for the different etiological subtypes. Several of the NCDs frequently coexist with one another, and their relationships may be multiply characterized under different chapter subheadings, including "Differential Diagnosis" (e.g. NCD due to Alzheimer's disease vs. vascular NCD), "Risk and Prognostic Factors" (e.g. vascular pathology

increasing the clinical expression of Alzheimer's disease), and /or "Comorbidity" (e.g. mixed Alzheimer's disease-vascular pathology).

The term *dementia* is retained in the DSM-5 for continuity and maybe used in settings where physicians and patients are accustomed to this term. Although dementia is the customary term for disorders like the degenerative dementias that usually affect older adults, the term neurocognitive disorder is widely used and often preferred for conditions affecting younger individuals, such as impairment secondary to traumatic brain injury or HIV infection. Furthermore, the major NCD definition is somewhat broader than the term dementia, in that individuals with substantial decline in a single domain can receive this diagnosis, most notably the DSM-4 category of "Amnesic Disorder", which would now be diagnosed as major NCD due to another medical condition and for which the term *dementia* would not be used.

The criteria for the various NCDs are all based on defined cognitive domains. Table 1 provides for each of the key domains a working definition, examples of symptoms or observations regarding impairments in everyday activities and examples of assessments. The domains thus defined, along with guidelines for the clinical thresholds, form the basis on which NCDs, their levels, and their subtypes may be diagnosed.

DSM-5 DELIRIUM (PAGE 596)

The introductory paragraphs are as follows:

THE DIAGNOSTIC CRITERIA

- A. A disturbance in attention (i.e. reduced ability to direct, focus, sustain, and shift attention) and awareness (reduced orientation to the environment).
- B. The disturbance develops over a short period of time (usually hours to a few days), represents a change from baseline attention and awareness, and tends to fluctuate in severity during the course of the day.
- C. An additional disturbance in cognition (e.g. memory deficit, disorientation, language, visuospatial ability, or perception).
- D. The disturbances in Criteria A and C are not better explained by another pre-existing established or evolving neurocognitive disorder and do not occur in the context of a severely reduced level of arousal, such as coma.
- E. There is evidence from the history, physical examination, or laboratory findings that the disturbance is a direct physiological consequence of another medical condition, substance intoxication or withdrawal (i.e. due to a drug of abuse or to a medication), or exposure to a toxin, or is due to multiple etiologies.

Specific categories are listed for

Substance intoxication delirium
Substance withdrawal delirium
Medication-induced delirium
Delirium due to another medical condition
Delirium due to multiple etiologies

Specify if:

Acute (Lasting a few hours or days)
Persistent: Lasting weeks or months

Specify if:

Hyperactive: the individual has a hyperactive level of psychomotor activity that may be accompanied by mood lability, agitation, and /or refusal to cooperate with medical care.

Hypoactive: The individual has a hypoactive level of psychomotor activity that may be accompanied by sluggishness and lethargy that approaches stupor.

Mixed level of activity: The individual has a normal level of psychomotor activity even though attention and awareness are disturbed. Also includes individuals whose activity level rapidly fluctuates.

PREVALENCE (DSM-5, PAGE 600)

The prevalence of delirium is 10-30% in older individuals presenting to emergency departments, where the delirium often indicates a medical illness.

The prevalence of delirium when individuals are admitted to hospital ranges from 14-24% and estimates of the incidence of delirium during hospitalization range from 6 to 56% in general hospital populations. Delirium occurs in 15-53% of older individuals postoperatively and in 70-87% of those in intensive care. Delirium occurs in up to 60% of individuals in nursing homes or post-acute care settings and in up to 83% of all individuals at the end of life.

DSM-5 MAJOR AND MILD NEUROCOGNITIVE DISORDERS (PAGE 602)

MAJOR NEUROCOGNITIVE DISORDER

DIAGNOSTIC CRITERIA

- A. Evidence of significant cognitive decline from a previous level of performance in one or more cognitive domains (complex attention, executive function, learning and memory, language, perceptual motor, or special cognition) based on:
 1. Concern of the individual, a knowledgeable informant, or the clinician that there has been a significant decline in cognitive function; and
 2. A substantial impairment in cognitive performance, preferably documented by standardized neuropsychological testing or, in its absence, another quantified clinical assessment.
- B. The cognitive deficits interfere with independence in everyday activities (i.e. at a minimum, requiring assistance with complex instrumental activities of daily living such as paying bills or managing medications).
- C. The cognitive deficits do not occur exclusively in the context of delirium
- D. The cognitive deficits are not better explained by another mental disorder (e.g. major depressive disorder, schizophrenia).

Specify whether due to:

Alzheimer's disease

Frontotemporal degeneration

Lewy body disease
Vascular disease
Traumatic brain injury
Substance/medication use
HIV infection
Prion disease
Parkinson's disease
Huntington's disease
Another medical condition
Multiple etiologies
Unspecified

Specify:

Without behavioral disturbance: if the cognitive disturbance is not accompanied by any clinically significant behavioral disturbance

With behavioral disturbance (specify disturbance): if the cognitive disturbance is accompanied by a clinically significant behavioral disturbance (e.g. psychotic symptoms, mood disturbance, agitation, apathy, or other behavioral symptoms).

Specify current severity:

Mild: Difficulties with instrumental activities of daily living (e.g. housework, managing money).

Moderate: Difficulties with basic activities of daily living (e.g. feeding, dressing).

Severe: Fully dependent

MILD NEUROCOGNITIVE DISORDER

DIAGNOSTIC CRITERIA

- A. Evidence of modest cognitive decline from a previous level of performance in one or more cognitive domains (complex attention, executive function, learning and memory, language, perceptual motor, or special cognition) based on:
1. Concern of the individual, a knowledgeable informant, or the clinician that there has been a mild decline in cognitive function; and
 2. A modest impairment in cognitive performance, preferably documented by standardized neuropsychological testing or, in its absence, another quantified clinical assessment.
- B. The cognitive deficits do not interfere with capacity for independence in everyday activities (i.e. complex instrumental activities of daily living such as paying bills or managing medications are preserved, but greater effort, compensatory strategies, or accommodation may be required).
- C. The cognitive deficits do not occur exclusively in the context of delirium
- D. The cognitive deficits are not better explained by another mental disorder (e.g. major depressive disorder, schizophrenia).

Specify whether due to:

Alzheimer's disease
Frontotemporal degeneration
Lewy body disease
Vascular disease

Traumatic brain injury
Substance/medication use
HIV infection
Prion disease
Parkinson's disease
Huntington's disease
Another medical condition
Multiple etiologies
Unspecified

Specify:

Without behavioral disturbance: if the cognitive disturbance is not accompanied by any clinically significant behavioral disturbance

With behavioral disturbance (specify disturbance): if the cognitive disturbance is accompanied by a clinically significant behavioral disturbance (e.g. psychotic symptoms, mood disturbance, agitation, apathy, or other behavioral symptoms).

DIAGNOSTIC FEATURES

Major and mild NCDs exist on a spectrum of cognitive and functional impairment. Major NCD corresponds to the condition referred to in DM-4 as dementia, retained as an alternative in this volume. The core feature of NCDs is acquired cognitive decline in one or more cognitive domains (Criterion A) based on both 1) a concern about cognition, on the part of an individual, a knowledgeable informant, or the clinician, and 2) performance on an objective assessment that falls below the expected level or that has been observed to decline over time. Both a concern and objective evidence are required because they are complimentary. Where there is an exclusive focus on objective testing, a disorder may go unrecognized in high functioning individuals whose currently "normal" performance actually represents a substantial decline in abilities, or an illness may be incorrectly diagnosed in individuals whose currently "low" performance does not represent a change from their own baseline or is a result of extraneous factors like test conditions or a passing illness. Alternatively excessive focus on subjective symptoms may fail to diagnose illness in individuals with poor insight, or whose informants deny or fail to notice their symptoms, or it may be overly sensitive in the so called worried well.

A cognitive concern differs from a complaint in that it may or may not be voiced spontaneously. Rather, it may need to be elicited by careful questioning about specific symptoms that commonly occur in individuals with cognitive deficits. For example, memory concerns include difficulty in remembering a short grocery list or keeping track of the plot of a television program; executive concerns include difficulty resuming a task when interrupted, organizing tax records, or planning a holiday meal. At the mild NCD level, the individual is likely to describe these tasks as being more difficult or as requiring extra time or effort or compensatory strategies. At the major NCD level, such tasks may only be completed with assistance or may be abandoned altogether. At the mild NCD level, individuals and their families may not notice such symptoms or may view them as normal, particularly in the elderly; thus careful history taking is of paramount importance. The difficulties must represent changes rather than lifelong patterns: the individual or informant may clarify this issue, or the clinician can infer change from prior experience with the patient or from occupational

or other clues. It is also critical to determine that the difficulties are related to cognitive loss rather than to motor or sensory limitations.

Neuropsychological testing, with performance compared with norms appropriate to the patient's age, educational attainment, and cultural background, is part of the standard evaluation of NCDs and is particularly critical in the evaluation of mild NCD. For major NCD, performance is typically 2 or more standard deviations below appropriate norms (3rd percentile or below). For mild NCD, performance typically lies in the 1-2 standard deviations range (between 3rd and 16th percentiles). However, neuropsychological testing is not available in all settings, and neuropsychological thresholds are sensitive to the specific test(s) and norms employed, as well as to test conditions, sensory limitations, and intercurrent illness. A variety of brief office-based or "bedside" assessments, as described in table 1, can also supply objective data in settings where such testing is unavailable or infeasible. In any case, as with cognitive concerns, objective performance must be interpreted in light of the individual's prior performance. Optimally, this information would be available from a prior administration of the same test, but often it must be inferred based on appropriate norms, along with the individual's educational history, occupation, and other factors. Norms are more challenging to interpret in individuals with very high or very low levels of education and in individuals being tested outside their own language or cultural background.

Criterion B relates to the individual's level of independence in everyday functioning. Individuals with major NCD will have impairment of sufficient severity so as to interfere with independence, such that others will have to take over tasks that the individuals were previously able to complete on their own. Individuals with mild NCD will have preserved independence, although there may be subtle interference with function or a report that tasks require more effort or take more time than previously.

The distinction between major and mild NCD is inherently arbitrary, and the disorders exist along a continuum. Precise thresholds are therefore difficult to determine. Careful history taking, observation, and integrating with other findings are required, and the implications of diagnosis should be considered when an individual's clinical manifestations lie at a boundary.