From DSM to DM-ID

Robert J. Fletcher, Jarrett Barnhill, Jane McCarthy & André Strydom

From *DSM* to *DM-ID*

Robert J. Fletcher [a], Jarrett Barnhill [b], Jane McCarthy [c], and André Strydom [d]

aNational Association for the Dually Diagnosed, Kingston, New York, USA; bDepartment of Psychiatry, University of North Carolina School of Medicine, USA; cDepartment of Forensic and Neurodevelopmental Sciences, King's College London, UK; dUniversity College London Division of Psychiatry, UK

ABSTRACT

Recognizing the diagnostic challenges that clinicians face when attempting to arrive at an accurate psychiatric diagnosis for individuals with intellectual and developmental disabilities (IDD) co-occurring with mental illness (MI), in 2007 the National Association for the Dually Diagnosed (NADD), in association with the American Psychiatric Association (APA), published Diagnostic Manual—Intellectual Disability (*DM-ID*): A Textbook of Diagnosis of Mental Disorders in Persons with Intellectual Disability (Fletcher, Loschen, Stavrakaki, & First, 2007). The *DM-ID* was designed as a companion to the *DSM-IV-TR* and aimed to assist clinicians to arrive at a more accurate *DSM-IV-TR* diagnosis for individuals with IDD. In 2013, the American Psychiatric Association published the *DSM-5*, thus necessitating revision of the *DM-ID* to incorporate the changes from the *DSM-IV-TR* to the *DSM-5*. The authors discuss the need for and development of the original *DM-ID* and changes in the DSM-5. The authors then offer insight into several chapters in the DM-ID-2 across the lifespan of individuals with IDD, looking at the changes in the *DSM-5* and how these impact the ascertainment of mental disorders in individuals with IDD.

Individuals with intellectual and developmental disabilities (IDD) can experience the same psychiatric disorders as people in the general population. While estimates of the prevalence of mental disorders among people with IDD varies, research indicates that the prevalence is higher with people who have IDD than in the general population. Prevalence estimates range from 30% to 70% of individuals with IDD having mental illness or behavioral problems (Szymanski & King, 1999). The range of findings can be attributed to a variety of factors including differences in population sampling and methodologies used in identifying psychiatric disorders in persons with IDD. Two of the larger studies are: Cooper, Smiley, Morrison, Williamson, and Allan (2007), who revealed a rate of 40.9% with a population-based study (*N* = 1023) employing a comprehensive individualized clinical assessment; and National Core Indicators (NCI), which has identified a rate of 55%

(N = 13,466) (National Core Indicates, 2016) based on patient charts from thirty states in the U.S.

Recognizing the diagnostic challenges that clinicians are faced with when attempting to arrive at an accurate diagnosis for individuals with IDD coexisting with mental illness (MI), the National Association for the Dually Diagnosed (NADD), in association with the American Psychiatric Association (APA), published *Diagnostic Manual—Intellectual Disability (DM-ID): A Textbook of Diagnosis of Mental Disorders in Persons with Intellectual Disability* in 2007 (Fletcher, Loschen, Stavrakaki, & First, 2007). The challenges stem, to a great extent, from the difficulty or inability of individuals with IDD to describe their own symptoms. Diagnosis for an individual within the population without IDD generally relies upon the person's description of his or her experiences and feelings. Individuals with IDD have limited receptive and expressive language, thus limiting their ability to describe their symptoms. They may also lack the self-reflection to describe internal states. Furthermore, individuals with IDD who are experiencing mental illness may present in very different ways than their peers without IDD. Accurate diagnosis can be further stymied by diagnostic overshadowing, in which the diagnosis of IDD can overshadow coexisting mental disorders and predispose practitioners to overlook the presence of psychopathology or attribute the symptoms of psychopathology to the IDD (Reiss, Levitan, & Szysko, 1982). Also, some people with IDD tend to try to hide their disability under a "cloak of competence," while others may try to please the evaluator by providing the answer the individual thinks the evaluator wants ("acquiescence bias"). The *DM-ID* provides guidance for assessing and diagnosing specific disorders in individuals with IDD and provides information on recognizing challenging behaviors of individuals with IDD and how to differentiate between behavioral problems and psychiatric disorders. The *DM-ID* was designed as a companion to the *DSM-IV-TR* and aimed to assist clinicians to arrive at a more accurate *DSM-IV-TR* diagnosis for individuals with IDD.

Work on the *DM-ID* began almost 10 years before its publication, when Dr. Robert Fletcher, Founder and CEO of NADD, submitted a proposal to the NADD Board of Directors to develop a companion to the *DSM-IV* to facilitate a more accurate *DSM-IV* diagnosis for people with IDD. Experts were recruited for work groups for each diagnostic category. Approximately 60 experts participated in this project. The editors for the *DM-ID* were Robert Fletcher, Earl Loschen, Chrissoula Stavrakaki, and Michael First. The *DM-ID* covers all major diagnostic categories of mental disorders as defined in the *DSM-IV-TR*. Each work group reviewed the existing research concerning the disorder(s) on which they were working, with emphasis on how the disorder manifests in individuals who have IDD. The Cochrane system was used to evaluate the research reviewed (Cochrane Library, 2001).

Based upon the research and the work group's expert consensus, modifications and adaptations of the *DSM-IV-TR* diagnostic criteria were proposed which included addition of symptom equivalents, omission of symptoms, changes in symptom count, modification of symptom duration, modification of age requirements, addition of explanatory notes, and criteria sets that do not apply. In addition, advice about working with the individual and with respondents in order to achieve an accurate diagnosis was provided.

During the summer of 2006, prior to publication, field trials were held to assess the clinical usefulness of the *DM-ID*. The results were reported briefly in the introduction to the *DM-ID* and more thoroughly in an article published in the *Journal of Clinical Psychiatry* in 2009 (Fletcher et al., 2009). Sixty three clinicians, from eleven different countries, were recruited to participate in the research. These clinicians were asked to use the *DM-ID* with a minimum of 20 clients and to provide feedback about the clinical usefulness of the *DM-ID*. A clinical survey was developed. Part I, completed once by each clinician, provided information about the training and experience of each clinician who participated in the field trials and sought the clinician's assessment of the usefulness of the *DSM-IV-TR* when used with individuals who have IDD, as well as the clinician's reasons for this assessment. Part II was completed for each patient after the clinician had used the *DM-ID* to arrive at a diagnosis. Demographic information about the client was collected, followed by information about the *DSM-IV-TR* diagnosis and the *DM-ID* diagnosis arrived at. Finally, three yes/no questions about use of the *DM-ID* were answered: (1) "Did the *DM-ID* allow you to come up with a more specific diagnosis than you would have with *DSM-IV-TR*?" (2) "Did the *DM-ID* allow you to arrive at a psychiatric diagnosis that you think is appropriate for this patient?" and (3) "Did you find the *DM-ID* allowed you to avoid using the NOS (not otherwise specified) category." Three questions on a five-point scale were also asked: (1) "Was the *DM-ID* easy to use (user-friendly) to arrive at a psychiatric diagnosis for this patient?" (2) "Did you find the *DM-ID* clinically useful in the diagnosis of this patient?" and (3) "For the diagnosis used for this patient, do you feel that the number of adapted criteria were too few/excessive?"

Eight hundred and forty-five surveys on use of the *DM-ID* with specific patients were completed. Overall, response to the use of the *DM-ID* was positive, with 67.9% of respondents rating the *DM-ID* as "easy" or "very easy" to use and 83.1% of respondents indicating that the *DM-ID* allowed them to arrive at an appropriate psychiatric diagnosis for the patient. Over one-third of clinicians (36.5%) indicated that the *DM-ID* allowed them to arrive at a more specific diagnosis than the *DSM-IV-TR*.

The publication of the *DSM-5* (American Psychiatric Association, 2013) necessitated that the *DM-ID* be updated. NADD began putting together work groups to revise the *DM-ID* during the summer of 2012. The editors of the

revised *DM-ID* are Robert Fletcher, Jarrett Barnhill, and Sally Ann Cooper. One hundred and four experts were recruited to work in 26 work groups. A chairperson was identified for each work group. Work has been proceeding on the various chapters, and publication is anticipated for the summer of 2016.

Changes from *DSM-IV* to *DSM-5* reflect developments in genetic research and neuroimaging as well as efforts to promote ease of use. The disorders included in *DSM-5* have been reordered into a revised organizational structure, reflecting the fact that mental disorders do not always fit completely within the boundaries of a single disorder and that some symptom domains involve multiple diagnostic categories. *DSM-5* recognizes developmental issues utilizing a lifespan approach and including descriptions of how the disorder presentation changes across the lifespan. The multi-axial approach has been dropped. A number of disorders that had been distinct in *DSM-IV*, such as autistic disorder, Asperger's disorder, and pervasive developmental disorder, have been consolidated in *DSM-5* and the *DM-ID-2* into autism spectrum disorder (ASD). Trauma- and stressor-related disorders in the *DSM-5* and *DM-ID-2* is an umbrella diagnostic area that now includes reactive attachment disorder, disinhibited social engagement disorder, post-traumatic stress disorder (PTSD), acute stress disorder, and adjustment disorder. Disorders previously referred to as "dementias" are now designated as major or mild neurocognitive disorders.

It would be impossible, in the space of this article, to review all the challenges faced in developing the *DM-ID-2*. In the next sections, we look at a sample of disorders discussed in the *DSM-5* and the *DM-ID-2*, beginning with those often seen early in life (designated as neurodevelopmental disorders in the *DSM-5*) and ending with challenges encountered late in life (neurocognitive disorders). In between these early-lifetime and late-lifetime challenges, we consider a group of disorders that have a serious impact on the lives of individuals with IDD: trauma- and stressor-related disorders. The specific disorders found in this article are intended to illustrate issues of diagnostic limitations, lack of research in the population with IDD, and important changes in the conceptualization of these disorders.

Neurodevelopmental Disorders

The *DSM-5* reconfigures "Disorders with Onset during Childhood and Adolescence" (found in *DSM-IV-TR* and *DM-ID*) and stereotypic movement disorders and tic disorders to create neurodevelopmental disorders. Neurodevelopmental disorders share three basic features: an age of onset during the developmental period, diverse etiologies, and a large number of overlapping symptoms that co-occur in what appear to be discrete syndromes. But intellectual disability (intellectual developmental disorder) or IDD is included as a discrete "syndrome" within neurodevelopmental disorders.

Herein lies a problem: IDD is a subset of neurodevelopmental disorders, yet it is frequently listed among the exclusion criteria in the *DSM-5* (American Psychiatric Association, 2013). The central focus of the *DM-ID-2* is on co-occurring IDD and mental disorders. The presence of IDD shapes the presentation and course of many neurodevelopmental disorders and by doing so creates several cognitive dissonances. In many circumstances, the clinicians will have to judge how to modify inclusion, specifiers, and exclusion criteria to match up with heterogeneous populations of individuals with IDD. Resolving these dissonances is one of the major challenges for the authors of this section.

There are several additional changes in diagnostic criteria contained in both the *DSM-5* and the upcoming *DM-ID-2* beyond those mentioned earlier, such as the removal of the exclusion of ADHD in the context of ASD; the realigning of impulse control and disruptive behavior disorders, the creation of disruptive mood dysregulation disorder (DMD) for individuals with affect dysregulation and ADHD (previously diagnosed as bipolar disorder), and the creation of trauma- and stress-related disorders (American Psychiatric Association, 2013).

The *DSM-5* modifies the diagnostic criteria for IDD in a manner that shifts the emphasis from IQ scores to social-support needs. Severity of ID now depends upon the level of social-supports needs as measured by functional domain criteria (conceptual, social, and practical domains). Yet even this shift toward an emphasis on adaptive criteria does not resolve the problems we face in capturing the multidirectional relationships between ID, other neurodevelopmental disorders, and late-onset psychiatric disorders. The presence of ASD further complicates this process.

Remaining Foundational Issues for People with IDD

(1) Age of Onset—This is a more complex issue than it first seems. For many people with IDD, the presence of specific neurodevelopmental disorders can be "overshadowed" by baseline global cognitive and adaptive deficits. In addition, the majority of referrals for individuals with IDD involve behavioral/psychiatric complaints rather than neurodevelopmental disorders. As a consequence, the various combinations of IDD, challenging behavior, and late-onset psychiatric disorders can overshadow the "age" of onset for many neurodevelopmental disorders. It may be more useful to describe the age of recognition along with patterns of comorbidity in order to avoid the ambiguity associated with the determination of an age of onset (Barnhill, 2014).

(2) Parameters of the Developmental Period—Most mental health consultations and diagnoses are "point in time" events. As a result, the clinician has little opportunity to follow the ongoing development,

especially the interactions between the person's special vulnerabilities (including neurodevelopmental disorders), levels of resilience, and experiences across the life cycle. For example, the expression of many neurodevelopmental disorders changes in response to many interrelationships with IDD and ASD. These intertwining developmental trajectories influence and are influenced by the social ecology, changing academic or occupational demands, availability of learning experiences, impact of accessibility/utilization of services, and the vicissitudes of interventions for co-occurring mental disorders (Gardner, Griffiths, & Hamlin, 2012). Each of these impact brain neuroplasticity secondary to new learning that blurs the endpoint of the neurodevelopmental period. From this perspective, most diagnoses are hypotheses that may change over time (Barnhill, 2011; Piek, Dawson, Smith, Gasson, 2008).

(3) Diagnosis and Discrepancy Criteria—The concept of diagnosis relies upon measuring that the gap between expected and actual performance is larger than that "normally associated with ID." In many neurodevelopmental disorders, the diagnosis is either based on standardized assessment scores or a judgement call by the clinician. Clinical judgement can be both a blessing and a curse. For example, the presence of IDD changes its developmental trajectory as well as the risk for co-occurring mental disorders. As a result, many standardized measures are not sufficiently normed. Test scores and clinical judgements can be undermined by the severity of ID; comorbidity with ASD or genetic/metabolic disorders; and heterogeneity found in many cognitive, social communication, attentional, executive functional, and motor skills (Barnhill, 2003; Fletcher, Loeschen, Stavrakaki & First, 2007).

(4) Behavioral Phenotypes and Neurodegenerative Disorders—Diagnosis is only as good as the quality of observation data and current scientific evidence. This evidence changes in response to new technologies, genetic discoveries (behavioral phenotypes or metabolic disorder), and the development of new treatments for evolving brain disorders. Recent evidence suggests that many late-onset psychiatric disorders are preceded by unrecognized neurodevelopmental disorders (Barnhill, 2012). Genome-wide array studies (GWAS) suggest that ID, ASD, specific learning and attention deficit–hyperactivity disorders, and some forms of epilepsy share genetic profiles (Guilmarte et al., 2009). Early recognition and diagnosis of at-risk infants and children permit early intervention of many neurodevelopmental and neurodegenerative disorders (Gresham & Vellutino, 2010). Longitudinal assessment is useful in tracking the changing developmental trajectory as well as monitoring the efficacy of treatment intervention.

Diagnostic Issues Needing More Study—The DM-ID-2 and Beyond

Neurodevelopmental disorders appear as distinct syndromes that are largely based on phenomenology and not neurobiological criteria. Each syndrome represents a variety of complex signs and symptoms that frequently co-occur with other neurodevelopmental, behavioral, and primary psychiatric disorders. For example, motor disorders are divided into three heterogeneous disorders: developmental co-ordination, stereotypic movement (with and without self-injury), and tic disorders. Each can be comorbid with attention deficit hyperactivity, specific learning, communication, or autism spectrum, intellectual disability, and obsessive-compulsive and related disorders (Barnhill, 2011). When severe profound ID (SPID) or ASD/SPID are present, these boundaries are likely more diffuse. We are left to choose between making inappropriate diagnoses, excluding this population (nihilistic), or providing our best clinical judgement. Barnhill (2003, 2011) proposed an alternative, a pattern of *DSM-IV-TR* diagnosis that focused on defining neurobiological endo-phenotypes based on observable clusters of behaviors, patterns of comorbidity, trauma history (Aupperle, Melrose, Stein, & Paulus, 2012), issues related to attachment temperament, and ethological features. In these articles, the author algorithm resembles the Research Domain Criteria now considered to be an alternative to diagnostic classification systems for researchers (Adam, 2013).

Unfortunately, many of our best practices and evidence-based medicine are based on lumping (large studies, statistical analyses, and meta-analyses) at the expense of defining specific endo-phenotypes or using data from single-case designed studies. Bridging the gaps between these conceptual models may provide useful insights that allow for more individualized treatment planning.

Implications and Speculations

Since the *DM-ID-2* is designed for people with IDD, several new applications warrant consideration.

The use of the functional domains to assign a level of severity for IDD makes it reasonable device for classifying neurodevelopmental disorders in a similar manner. Currently, discrepancy criteria include a domain for clinical judgement as well as the gap between actual and expected performance based on standardized instruments. Many of these instruments use age-based normative data that may have lessening degrees of validity and reliability for people with severe-profound IDD. Perhaps the best examples are communication and specific learning disabilities (SLD). For nonverbal individuals with profound IDD, the domains of comprehension, expression, and pragmatics need to be expanded. The global deficits associated with (SPID)

limit the individual's verbal and conceptual skills to the extent a more basic level of analysis that assesses functional neurobiological substrates such as the ability to use basic shape recognition, cued responses for previously learned skills or capacity to respond to picture communication systems. Even individuals with mild-moderate IDD may require modifications. Under these conditions, it may be more useful to augment test score with the pattern of scatter on the functional domains criteria (conceptual, social and practical) rather than discrepancy/performance criteria based on chronological or developmental age. This night add some structure to clinical judgment.

The structure of the *DSM-5* (symptoms, specifiers, exclusion criteria) can provide a methodology to differentiate primary and secondary neurodevelopmental disorders. Perhaps more useful is their application for creating diagnostic algorithms that can reduce the heterogeneity of neurodevelopmental disorders. For an example, the diagnosis for a person with chronic schizophrenia might read: chronic schizophrenia in the context of mild ID, behavioral phenotype associated with Velocardiofacial syndrome, history of childhood developmental co-ordination and communication disorder (social pragmatics) and physical abuse during early childhood. Such a diagnostic scheme can provide more useful information for monitoring symptoms change and implementing changes in treatment to match evolving clinical needs.

Like the *DSM-5*, the *DM-ID-2* is based on phenomenological rather than neurobiological subtypes. Although not included in the neurodevelopmental disorders, oppositional defiant and conduct disorders represent the convergence of biologically based (e.g., relatedness to attention or impulse dyscontrol), socially "deviant" behaviors (callous unemotionality, irritability/overt defiance, violations of property or individual "rights"). For example, the relationship between ADHD, oppositional defiant, and conduct disorders represents a subset of externalizing behavioral disorders that requires a level of awareness for rule-governed social behaviors. They may also lie on a continuum of impulse dys-control, affective dysregulation, neuroticism, and deficits in conceptual, social, and practical skills. In this sense, conduct and oppositional defiant disorders have limited utility for individuals with severe and profound ID. It may be more helpful to describe and address underlying temperamental, psychophysiological, and behavioral response to threat or physical trauma, as well as genetic risk for ADHD or DMD. The goal is to include more than a descriptive diagnosis and focus instead on associated functional impairments.

Trauma- and Stressor-Related Disorders

Trauma- and stressor-related disorders include disorders in which exposure to a traumatic or stressful event is listed explicitly as a diagnostic criterion.

This is a new chapter within *DSM-5* and includes reactive attachment disorder (RAD), disinhibited social engagement disorder (DSED), (PTSD), acute stress disorder, and adjustment disorder. Within *DM-ID* these disorders were described in separate chapters, but in keeping with *DSM-5* they are brought together to reflect the increased understanding in the variation of expressing psychological distress when an individual is exposed to a traumatic or stressful event. The inclusion of reactive attachment disorders and disinhibited social engagement disorder, which develop early in life due to lack or absence of adequate caregiving, show a recognition in the importance of early experiences on the later development of an individual, including those with IDD.

Despite the frequency of pathogenic care and risk for neglect or abuse in people with IDD, it can be difficult to diagnose attachment disorders such as RAD and DSED in people with IDD. This is due to a variety of reasons including biological and genetic factors that influence an individual's ability to make attachments such as an ASD. In addition, there is a significant lack of research pertaining to both RAD and DSED in children and adults with IDD. There are instruments to assess the attachment behaviors of individuals with intellectual disabilities such as the Secure Base Safe Haven Observation List (De Schipper & Schuengel, 2010). The need to screen individuals for difficulties in attachment behaviors within adult care settings is becomingly increasingly recognized (Schuengel, De Schipper, Sterkenburg, & Kaf, 2013). Adults and children with ID can show signs and symptoms of disordered attachment even with a secure attachment pattern (Minnis, Fleming, & Cooper, 2010). Behaviors alone should not be used to diagnose RAD or DSED, but evidence of early life experiences of abuse, deprivation, and neglect should be sought. In individuals with borderline intellectual functioning or mild IDD referred for psychiatric consultation, the following prevalence figures have been reported with 42% exhibiting symptoms of overall disordered attachment, 16% showing symptoms of RAD, and 11% showing symptoms of both RAD and DSED. Individuals in this study were aged 5 to 11 and had borderline or mild IDD, with IQs ranging between 50 and 85 and a mean IQ of 71.7 (Giltaji, Sterkenburg, & Schuengel, 2013).

PTSD is a chronic disorder in response to trauma. With respect to ascertainment of PTSD in people with IDD, the research supports three important points: (1) people with IDD seem to be more vulnerable to the development of PTSD than members of the general population; (2) people with IDD are more often exposed to conditions known to contribute to the development of PTSD, such as interpersonal abuse and violence; and (3) for people with only mild IDD, the presentation of PTSD is similar to that seen in members of the general population (Wieland, Wardenaar, Dautovic, & Zitman, 2013). For people with more severe IDD, the presentation may be complicated by differing presentation of symptoms, with a lower

developmental functioning increasing the risk for developing PTSD (Mevissen & de Jongh, 2010).

Acute stress disorder is characterized by symptoms similar to those of PTSD that occurs immediately following exposure to one or more traumatic events. There is very little research on acute stress disorder presenting in those with IDD. Most of the evidence is from studies of PTSD, with no reference to the first month of presentation in these studies when an acute stress disorder would be present.

Adjustment disorders involve the development of clinically significant emotional or behavioral symptoms in response to an identifiable psychosocial stressor or stressors. The stressor may be a single event or events or circumstances that are recurrent or continuous. This definition incorporates an extremely valuable diagnostic concept, suggesting that environmental stressors, so common in the lives of persons with IDD, might be a critical source of psychopathology, which could otherwise be mistaken for other behavioral or mental health disorders. As simply stated in the *DSM-5*, "When bad things happen, most people get upset…the diagnosis should only be made when the magnitude of the distress…exceeds what would normally be expected…" Also noted in *DSM-5* is the fact that what is normally expected may vary in different cultures, so clinicians serving people with IDD must take into consideration that the world of a person with IDD is a culture (e.g., residential setting) within a broader culture (geographic region, ethnic community, etc.). In one study, Tsakanikos, Bouras, Costello, and Holt (2007) looked at a clinic sample of people with IDD and demonstrated that IDD is associated with a general increase in psychological vulnerability to life events, with adjustment disorder being more likely for people with ID exposed to *multiple* events but not as closely associated with *single* life events.

Neurocognitive Disorders

DSM-5 replaced the term "dementias" with a characterization of these neurodegenerative disorders as neurocognitive disorders. Previously known as Dementia, Delirium, Amnestic, and Other Cognitive Disorders in *DSM-IV-TR*, these disorders comprise delirium, and major and mild neurocognitive disorder (NCD) in the *DSM-5*. The term "dementia" may still be used where physicians and patients are accustomed to this, but "neurocognitive disorder" is preferred, especially for conditions affecting younger adults. NCD is also seen as broader and encompasses disorders previously included under Amnestic Disorders in *DSM-IV-TR*. The conceptualization of delirium as a disturbance in consciousness (specifically in attention and awareness), which develops over a short period of time and is due to a direct physiological consequence of another medical condition, has not changed significantly.

Major Neurocognitive Disorder (Major NCD)

The *DSM-5* has adopted a hierarchical approach to the diagnosis of NCDs, so that the criteria for major or mild NCD must be met before criteria for etiological subtypes such as Alzheimer's disease (AD), Lewy body disease, or vascular disease can be applied. The essential feature of a major neurocognitive disorder is the development of multiple cognitive deficits that are severe enough to cause impairment in daily functioning and represents a decline from a previous level of functioning. To meet the diagnostic criteria for a major NCD, individuals must present with significant cognitive decline in one or more domains (including complex attention, executive function, learning and memory, language, perceptual-motor, or social cognition). The *DSM-5* definition therefore differs from the *DSM-IV* and *ICD-10* definitions which require an impairment in memory *as well as* at least one other cognitive disturbance; the rationale being to ensure that the diagnosis of an NCD would apply to most dementia etiological subtypes, whereas previous definitions were based on the typical presentation of AD and therefore less valid in other subtypes (Ganguli et al., 2011). Most of what we know about the course of neurocognitive disorders in individuals with IDD comes from the study of individuals with Down syndrome (DS) and probable Alzheimer-type dementia. Although individuals with DS often present with memory decline and behavioral and other cognitive changes, such as deficits in executive functioning, are also prominent and may be the presenting symptoms in many cases (Strydom et al., 2010; Wiseman et al., 2015). As life expectancy of people with IDD extends into older age, dementia is an increasing cause of morbidity and mortality. To update and summarize current knowledge on dementia in older adults with IDD, the authors conducted a comprehensive review of the published literature from 1997–2008 with a specific focus on: (1) epidemiology of dementia in IDD in general as well as in specific genetic syndromes; (2) presentation; and (3) diagnostic criteria for dementia. The authors report that varied methodologies and inherent challenges in diagnosis yield a wide range of reported prevalence rates of dementia. Rates of dementia in the population with IDD without DS are comparable with or higher than the general population. Alzheimer's disease onset in DS appears earlier, and the prevalence increases from under 10% in the 40s to more than 30% in the 50s, with varying prevalence reported for those 60 and older. Incidence rates increase with age. Few studies of dementia in other genetic syndromes were identified. Presentation differs in the IDD population compared with the general population; symptoms of depression, sleep disturbances, delusions, and auditory hallucinations may also be apparent when individuals with IDD develop dementia (Strydom et al., 2010), particularly in adults with DS (Dekker et al., 2015).

Since the requirement for both memory and at least one other cognitive decline has been shown to affect the performance of dementia diagnoses in individuals with DS or IDD (Sheehan et al., 2014; Strydom et al., 2013), the new streamlined criteria for NCDs, therefore, have considerable face validity in this population. Nevertheless, several limitations in applying the criteria for NCD in individuals with IDD are apparent, the most important of which is the difficulty in objectively defining cognitive impairment and decline in a population with pre-morbid deficits. Individuals may have a wide range of baseline abilities across different domains, and there is considerable between-individual variation. Criteria therefore need to clearly state that a change from an individual's own baseline is required for a diagnosis of dementia; the new *DSM-5* NCD criteria do indeed require significant decline from a previous level of performance. This must be based on concern from an individual, knowledgeable informant, or clinician, as well as documented by standardized neuropsychological testing, or in its absence, another qualified clinical assessment.

Formal neuropsychological testing is difficult due to the limited range of tests suitable for the population with ID, especially for those with more severe intellectual disability. It is also difficult to "optimize" cognitive tests due to factors such as emotional states, sensory problems, and medical status. Questionnaire-based assessment of cognitive functioning reported by caregivers are often used instead, though these may not map well onto specific cognitive domains and the "context" may not be considered, e.g., whether a task is regularly ignored because the person is unwilling to do it rather than unable to do it. Reliability of informant reports of impairment or decline may also be an issue, particularly when information is obtained from different caregivers at different times. However, retrospective report has been found to be as good as prospective ratings (Jamieson-Craig, Scior, Chan, Fenton, & Strydom, 2010) and an adjustment to the *DSM-5* major NCD criterion for neuropsychological testing may therefore be to use informant-based questionnaires to demonstrate decline, such as the Dementia Questionnaire for Persons with Mental Retardation—DMR; (Evenhuis, 1996) or the CAMDEX-DS (Ball, Holland, Huppert, Treppner, & Dodd, 2016).

Mild Neurocognitive Disorder

DSM-5 introduced the term "mild neurocognitive disorder" to refer to a preclinical state of having symptoms akin to dementia which precedes significant functional impairment. Mild NCD can be distinguished from major NCD by the severity of the cognitive decline (modest vs. significant) and the impact the symptoms have on everyday function; for a diagnosis of mild NCD the cognitive deficits do not interfere with capacity for independence in everyday activities. However, in individuals with IDD this may be difficult to

apply, given their variable pre-morbid abilities and lifelong dependence on support. Indeed, a similar definition of mild cognitive impairment has been found to have poor predictive validity in individuals with IDD (Strydom et al., 2013) and could result in over-diagnosis. However, this diagnosis may be useful in individuals at high risk for dementia such as those with DS, particularly in research settings, and could help to diagnose dementia at an earlier stage as long as the individual's own baseline is used to define decline or level of independence in everyday activities.

Summary

The authors in this article have first explored the evolution of the *DSM* to the *DM-ID* nosology systems. Then a discussion about neurodevelopmental disorders follows. These disorders share three basic features—an age of onset during the developmental period, diverse etiologies, and a large number of overlapping symptoms that co-occur in what appear to be discrete syndromes. Clinicians will have to judge how best to modify inclusion, specifiers, and exclusion criteria to apply these diagnostic criteria to individuals with IDD. The authors address trauma- and stressor-related disorders, which include disorders in which exposure to a traumatic or stressful event is listed explicitly as a diagnostic criterion. Most of the recent research around trauma- and stressor-related disorders has focused on PTSD and there remains a need for further research to look at the impact of the early experiences of trauma on the attachment behaviors of people with IDD. The benefits of *DM-ID-2* will be to support how we recognize these disorders from a clinical perspective but also be a basis to support further research of these disorders in persons with IDD. Finally, the authors point out clinical issues as they pertain to neurocognitive disorders and their relationship to IDD. The *DSM-5* and the *DM-ID-2* include criteria for major neurocognitive disorder and mild neurocognitive disorder. While the criteria for major neurocognitive disorder have several characteristics which will help to diagnose dementia in individuals with ID, the validity of mild neurocognitive disorder should be established before it is widely applied.

ORCID

Robert J. Fletcher ⓘ http://orcid.org/0000-0002-4702-1939
Jarrett Barnhill ⓘ http://orcid.org/0000-0002-4702-1939
Jane McCarthy ⓘ http://orcid.org/0000-0002-4702-1939
Andre Strydom ⓘ http://orcid.org/0000-0002-2502-6704

References

Adam, D. (2013). Mental health: On the spectrum. *Nature*, 496(7446), 416–418. doi:10.1038/496416a

American Psychiatric Association. (2013). *Diagnostic and statistical manual of mental disorders (DSM-5;* 5th ed.). Washington, DC: Author.

Aupperle, R. L., Melrose, A. J., Stein, M. B., & Paulus, M. P. (2012). Executive function and PTSD: Disengaging from trauma. *Neuropharmacology*, 62(2), 686–694. doi:10.1016/j.neuropharm.2011.02.008

Ball, S. L., Holland, A. J., Huppert, F., Treppner, P., & Dodd, K. (2016). *The Cambridge examination for mental disorders of older people with Down's syndrome and others with intellectual disabilities (CAMDEX-DS)*. Cambridge, UK: Cambridge University Press.

Barnhill, J. (2003). Can the *DSM-IV-TR* be salvaged for individuals with severe developmental disabilities? *Mental Health Aspects of Developmental Disabilities*, 6(3), 85–98.

Barnhill, J. (2011). Obsessive-compulsive disorders or not: Differential diagnosis of repetitive behavior among individuals with intellectual and developmental disorder. In S. Selek (Ed), *Different views of anxiety disorders* (pp. 37–68). Rijek, Croatia: InTech.

Barnhill, J. (2012). Velocardiofacial syndrome and schizophrenia. *NADD Bulletin*, 15(1), 14–15.

Barnhill, J. (2014). Child development and intellectual disabilities: The challenge of hitting a moving target. In D. Baker, & E. Blumberg (Eds.), *Mental health and wellness supports for youth with IDD*. Kingston, NY: NADD Press.

Cochrane Library. (2001). *Issue 2 update software*. Oxford, UK: Author.

Cooper, S. A., Smiley, E., Morrison, J., Williamson, A., & Allan, I. (2007). Mental ill-health in adults with intellectual disabilities: Prevalence and associated factors. *British Journal of Psychiatry*, 190, 27–35. doi:10.1192/bjp.bp.106.022483

De Schipper, J. C., & Schuengel, C. (2010). Attachment behavior towards support staff in young people with intellectual disabilities: Associations with challenging behavior. *Journal of Intellectual Disability Research*, 54, 584–596. doi:10.1111/j.1365-2788.2010.01288.x

Dekker, A. D., Strydom, A., Coppus, A. M. W., Nizetic, D., Vermeiren, Y., Naudé, P. J. W., ... De Deyn, P. P. (2015). Behavioural and psychological symptoms of dementia in Down syndrome: Early indicators of clinical Alzheimer's disease? *Cortex*: *A Journal Devoted to the Study of the Nervous System and Behavior*, 73, 36–61. doi:10.1016/j.cortex.2015.07.032

Evenhuis, H. M. (1996). Further evaluation of the Dementia Questionnaire for Persons with Mental Retardation (DMR). *Journal of Intellectual Disability Research*, 40(4), 369–373. doi:10.1111/j.1365-2788.1996.tb00642.x

Fletcher, L., Loschen, E., Stavrakaki, C., & First, M. (Eds.). (2007). *Diagnostic manual–intellectual disability: A textbook of diagnosis of mental disorders in persons with intellectual disability*. Kingston, NY: NADD Press.

Fletcher, R. J., Havercamp, S. M., Ruedrich, S. L., Benson, B. A., Barnhill, L. J., Cooper, S. A., & Stavrakaki, C. (2009). Clinical usefulness of the diagnostic manual–intellectual disability for mental disorders in persons with intellectual disability: Results from a brief field survey. *The Journal of Clinical Psychiatry*, 70(7), 967–974. doi:10.4088/JCP.08m04429

Ganguli, M., Blacker, D., Blazer, D. G., Grant, I., Jeste, D. V., Paulsen, J. S., ... Sachdev, P. S. (2011). Classification of neurocognitive disorders in *DSM-5*: A work in progress. *The American Journal of Geriatric Psychiatry: Official Journal of the American Association for Geriatric Psychiatry*, 19(3), 205–210. doi:10.1097/JGP.0b013e3182051ab4

Gardner, W. I., Griffiths, D. M., & Hamlin, J. (2012). Biopsychosocial features influencing aggression: A multimodal assessment and therapy approach. In J. K. Luiselli (Ed.), *The handbook of high-risk challenging behaviors: Assessment and intervention* (pp. 83–102). Towson, MD: Brookes.

Giltaji, H. P., Sterkenburg, P. S., & Schuengel, C. (2015). Psychiatric diagnostic screening of social maladaptive behavior in children with mild intellectual disability: Differentiating disordered attachment and pervasive developmental disorder behavior. *Journal of Intellectual Disability Research*, 59, 138–149. doi: 10.1111/jir.12079.

Gresham, F. M., & Vellutino, F. R. (2010). What is the role of intelligence in the identification of specific learning disabilities? Issues and clarifications. *Learning Disabilities Research & Practice*, 25(4), 194–206. doi:10.1111/ldrp.2010.25.issue-4

Guilmarte, A., Dubourg, C., Mosca, A., Legallic, S., Goldenberg, A., Drouin-Garraud, V., & Champion, D. (2009). Recurrent rearrangements in synaptic and neurodevelopmental genes and shared biologic pathways in schizophrenia, autism, and mental retardation. *Archives of General Psychiatry*, 66, 947–956. doi:10.1001/archgenpsychiatry.2009.80

Jamieson-Craig, R., Scior, K., Chan, T., Fenton, C., & Strydom, A. (2010). Reliance on carer reports of early symptoms of dementia among adults with intellectual disabilities. *Journal of Policy and Practice in Intellectual Disabilities*, 7(1), 34–41. doi:10.1111/j.1741-1130.2010.00245.x

Mevissen, L., & de Jongh, A. (2010). PTSD and its treatment in people with intellectual disabilities: A review of the literature. *Clinical Psychology Review*, 30(3), 308–316. doi:10.1016/j.cpr.2009.12.005

Minnis, H., Fleming, G., & Cooper, S.-A. (2010). Reactive attachment disorder symptoms in adults with intellectual disabilities. *Journal of Applied Research in Intellectual Disabilities*, 23, 398–403. doi:10.1111/jar.2010.23.issue-4

National Core Indicators. (2016). *Chart generator 2013–14*. National Association of State Directors of Developmental Disabilities Services and Human Services Research Institute. Retrieved from http://www.nationalcoreindicators.org/charts/

Piek, J. B., Dawson, L., Smith, L. M., & Gasson, N. (2008). The role of early fine and gross motor development on later motor and cognitive ability. *Human Movement Science*, 27, 668–681. doi:10.1016/j.humov.2007.11.002

Reiss, S., Levitan, G. W., & Szysko, J. (1982). Emotional disturbance and mental retardation: Diagnostic overshadowing. *American Journal of Mental Deficiency*, 86(6), 567–574.

Schuengel, C., De Schipper, J. C., Sterkenburg, P. S., & Kaf, S. (2013). Attachment, intellectual disabilities and mental health: Research, assessment and intervention. *Journal of Applied Research in Intellectual Disabilities*, 26, 34–46. doi:10.1111/jar.12010

Sheehan, R., Sinai, A., Bass, N., Blatchford, P., Bohnen, I., Bonell, S., . . . Strydom, A. (2014). Dementia diagnostic criteria in Down syndrome. *International Journal of Geriatric Psychiatry*. doi:10.1002/gps.4228

Strydom, A., Chan, T., Fenton, C., Jamieson-Craig, R., Livingston, G., & Hassiotis, A. (2013). Validity of criteria for dementia in older people with intellectual disability. *The American Journal of Geriatric Psychiatry*, 21(3), 279–288. doi:10.1016/j.jagp.2012.11.017

Strydom, A., Shooshtari, S., Lee, L., Raykar, V., Torr, J., Tsiouris, J., . . . Maaskant, M. (2010). Dementia in older adults with intellectual disabilities—epidemiology, presentation, and diagnosis. *Journal of Policy and Practice in Intellectual Disabilities*, 7(2), 96–110. doi:10.1111/j.1741-1130.2010.00253.x

Szymanski, L., & King, B. (1999). *Summary of the practice parameters for the assessment and treatment of children, adolescents, and adults with mental retardation and comorbid mental disorders*. Washington, DC: AACAP Publications Department. Retrieved from http://www.aacap.org/clinical/parameters/summaries/mr.htm

Tsakanikos, E., Bouras, N., Costello, H., & Holt, G. (2007). Multiple exposure to life events and clinical psychopathology in adults with intellectual disability. *Social Psychiatry and Psychiatric Epidemiology*, 42(1), 24–28. doi:10.1007/s00127-006-0128-1

Wieland, J., Wardenaar, K. J., Dautovic, E., & Zitman, F. G. (2013). Characteristics of posttraumatic stress disorder in patients with an intellectual disability. *European Psychiatry, 28*(1), 1.

Wiseman, F. K., Al-Janabi, T., Hardy, J., Karmiloff-Smith, A., Nizetic, D., Tybulewicz, V. L. J., . . . Strydom, A. (2015). A genetic cause of Alzheimer disease: Mechanistic insights from Down syndrome. *Nature Reviews. Neuroscience, 16*(9), 564–574. doi:10.1038/nrn3983