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“Intellectual developmental disorders”: reflections on the international consensus document for redefining “mental retardation-intellectual disability” in ICD-11

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Abstract

Purpose—The debate as to whether intellectual disability (ID) should be conceptualized as a health condition or as a disability has intensified as the revision of World Health Organization’s (WHO’s) International Classification of Diseases (ICD) is being finalized. Defining ID as a health condition is central to retaining it in ICD, with significant implications for health policy and access to health services. The purpose of this paper is to include some reflections on the consensus document produced by the first WHO Working Group on the Classification of MR (WHO WG-MR) and on the process that was followed to realize it. The consensus report was the basis for the development of official recommendations sent to the WHO Advisory Group for ICD-11.

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The authors report no conflicts of interest. This paper is based on a consensus document by an international group of experts identified by the Section Intellectual Disability of the World Psychiatric Association (WPA-SPID), which includes Colleen Adnams, Leyla Akoury-Dirani, Marco Bertelli, Sab Bhaumik, Nick Bouras, Jacob Burack, Sally-Ann Cooper, Sherva Cooray, Shoumitro Deb, Satish Chandra Girimaji, Jim Harris, Judith Hollenweger, Gregorio Katz, Henry Kwok, Rex Jung, Ruth Luckasson, Rafael Martínez-Leal, Kerim Munir, Geoffrey Reed, Per-Anders Rydelius, Luis Salvador-Carulla, Rune Simeonsson, Peter Tyrer, Carolyn Walsh, and Germain Weber. Many of these contributors were members of the first World Health Organization (WHO) Working Group on the Classification of Intellectual Disabilities, reporting to the WHO International Advisory Group for the Revision of ICD-10 Mental and Behavioural Disorders, Department of Mental Health and Substance Abuse, WHO. Others were members of a broader Review Group that WHO established in this area. Unless specifically noted, statements in this paper represent the views of the authors and not the official views or policies of the International Advisory Group or of the WHO.

Design/methodology/approach—A mixed qualitative approach was followed in a series of meetings leading to the final consensus report submitted to the WHO Advisory group. These recommendations combined prior expert knowledge with available evidence; a nominal approach was followed throughout with face-to-face conferences.

Findings—The WG recommended a synonym set (“synset”) ontological approach to the conceptualisation of this health condition underlying a clinical rationale for its diagnosis. It proposed replacing MR with Intellectual Developmental Disorders (IDD) in ICD-11, defined as “a group of developmental conditions characterized by a significant impairment of cognitive functions, which are associated with limitations of learning, adaptive behaviour and skills”. The WG further advised that IDD be included under the parent category of neurodevelopmental disorders, that current distinctions (mild, moderate, severe and profound) be continued as severity qualifiers, and that problem behaviours removed from its core classification structure and instead described as associated features.

Originality/value—Within the ID/IDD synset two different names combine distinct aspects under a single construct that describes its clinical as well as social, educational and policy utilities. The single construct incorporates IDD as a clinical meta-syndrome, and ID as its functioning and disability counterpart. IDD and ID are not synonymous or mirror concepts as they have different scientific, social and policy applications. New diagnostic criteria for IDD should be based on a developmental approach, which accounts for the complex causal factors known to impact the acquisition of specific cognitive abilities and adaptive behaviours. The paper focuses on a new clinical framework for the diagnosis of IDD that also includes and complements the existing social, educational and policy components inherent in ID.

Keywords

Classification; Diagnosis; Intellectual disability; Health terminology; Intellectual developmental disorders; Mental retardation

Introduction

The conceptualisation and the positioning of Mental Retardation (MR) within the taxonomy of mental disorders have long been controversial. MR represents a cluster of syndromes that start early in life and are characterized by varying patterns of severe and persistent impairments in cognitive and personal functions, such as intelligence, learning, adaptive behaviour and skills. It is also recognized that these cluster of syndromes that constitute MR have complex array of neurodevelopmental underpinnings. The classification and diagnostic criteria are therefore crucial issues and have important implications for their prevalence, development of interventions, as well as measures of outcome, service provision and planning. The debate has gone on for more than two decades in parallel to the discussion on the concept of intelligence, human functioning and disability.

The diagnostic criteria for MR had remained unchanged since 1968 when the World Health Organization (WHO) sponsored a first international committee that provided the recommendation for its diagnosis in International Classification of Diseases (ICD), Eight Revision (ICD-8). In the current WHO classification systems, MR appears in the ICD and

Related Health Problems, Tenth Revision (ICD-10) within the grouping of psychiatric conditions, and at the same time, it can be coded as a disability in the WHO International Classification of Functioning (ICF) (as impairments in intellectual functions, code b117). In 2009, the WHO established the Working Group on the Classification of MR (WHO WG-MR) reporting to the WHO International Advisory Group for the Revision of ICD-10 Mental and Behavioural Disorders in the ICD Eleventh Revision (ICD-11), now slated for publication in 2017. Strikingly, the WG-MR was the first international and multidisciplinary expert group established by an international organization to debate the issue in 40 years.

The WHO WG-MR produced the first international consensus document incorporating views of representatives from the major professional as well as consumer organizations, including key members from the World Psychiatric Association (WPA), American Psychiatric Association (APA), American Association for Intellectual and Developmental Disabilities (AAIDD) and the International Association for the Scientific Study of Intellectual Disabilities (IASSID). The APA assigned a liaison from its organization as delegate to represent the views of the APA and harmonize with the ongoing Task Force on DSM, Fifth Edition (DSM-5) process and ICD-11. The WG-MR produced a position paper in 2011 (Salvador-Carulla *et al.*, 2011) with an extensive consensus report that formed the basis for the development of the official recommendations sent to the WHO Advisory Group on ICD-11 in 2012.

With the publication of the DSM5 in 2013, the APA replaced MR with the term Intellectual Disability (ID)/Intellectual Developmental Disorder (IDD) (ID/IDD) noting in the text that the diagnostic term ID (intellectual developmental disorder) with intellectual developmental disorder in parenthesis is the equivalent term for the proposed ICD-11 diagnoses of IDD and so listed for the purposes of harmonization. It is included under the meta-category neurodevelopmental disorders to make clear that the focus is on the disorder category and not the disability construct. The DSM Third Edition (DSM-III) in 1980, and the DSM Third Edition-Revised (DSM-III-R) in 1987, included MR under “developmental disorders” but nevertheless placed it under Axis II. This separation was with coding on Axis II was continued in DSM-IV and DSM-IV-R when MR was included under the Chapter “Disorders Usually First Diagnosed in Infancy, Childhood, or Adolescence”.

The term “Intellectual Developmental Disorder” (IDD), proposed by the WHO WG-MR, as noted was adopted and included in parenthesis in the DSM5 ID (IDD), underscoring the conceptualisation of ID as a brain based health condition and not as a disability. In contrast to the DSM5, the AAIDD defines ID as a “disability” and not as a “health condition” with a primary focus on the individual’s functioning, adaptive behaviour and support needs. This was consistent with the preceding MR classification by the AAMR (former name for AAIDD) in 1992, specifying “Patterns and Intensity of Supports Needed” (i.e. intermittent, limited, extensive and pervasive), not directly comparable with the codes for degrees of severity in DSM. Although the last AAIDD manual states that the interpretation of ID is consistent with the conceptual model proposed by the WHO (2001), the fidelity of the AAIDD approach to the ICF model has recently been revised by Luckasson and Schalock (2013), when they proposed a more comprehensive and complex functionality approach to ID including a recognition of the need to coding it in ICD.

Extreme positions in this debate might have very negative implications if a future scientific consensus cannot be reached. The conceptualisation of ID as a “disability” would be logically followed by its exclusion from ICD, and its classification solely under ICF coding, which in turn could imply, ensuing arguments for restriction in access to health-related services, and particularly mental health care. Furthermore, removing ID from the ICD could have a major impact on the visibility of the health conditions that constitute the cluster of syndromes that cause the health condition, in terms of both national and global health statistics as well as health policy (Salvador-Carulla and Saxena, 2009). Conversely, if ID is considered solely as a health condition, its definition as a “disability” could be replaced; this could have a very negative impact on the provision of social and educational services, as well as public perceptions, policy formulation and future legislation. This would be inconsistent with the already adopted positions by many governments and international organizations and advocacy groups.

This debate on classification of ID/IDD has overarching implications for the domain of health ontology and classification. In addition to clarification of disease and disability distinctions in ICD and ICF, respectively, it seems clear that an international consensus ought to be found so the health condition component (IDD), be appropriately coded within a classification of diseases, whilst its functional aspects (ID) be equally conceptualized under a classification of functioning and disability. The consensus, however, cannot be based on a non-scientific arrangement. The pragmatic use of IDD in ICF, as well as ID in ICF, as though they are synonymous would be a taxonomic anomaly that will undoubtedly reflect a major flaw in the conceptual robustness of the overall WHO Family of Classifications (WHO-FIC). The usage of DSM5 and placement of IDD in parenthesis also reflects such an underlying concern. Moreover, the USA has not adopted ICF. The two terms ID and IDD are not “mirror image” codes and their usage cannot be permissible in two WHO complementary classifications.

The current debate on ID/IDD has implications on the naming, definition, and classification of other neurodevelopmental disorders such as autistic spectrum disorders where a specified “with or without accompanying intellectual impairment” is required. Attention deficit hyperactivity disorder too sheds similar light on problems of modern taxonomy and nosology, as well as disease and disability distinctions.

Moreover, there are unsolved questions in the definition of ID/IDD as a health condition, including the age cut-off for onset, the operationalisation of intelligence quotient (IQ) reduction, and the association between cognitive impairments and behavioural skills.

This paper is undertaken to complement and update the information provided in the position paper (Salvador-Carulla *et al.*, 2011). We describe a qualitative method followed for developing the WHO consensus document on IDD. We review the main agreements reached with regards to the clinical diagnosis of IDD, including the conceptual framework, the key components for a clinical diagnosis of IDD, the differential diagnosis including Borderline Intellectual Functioning (BIF), and the key associated features.

Methods

A mixed qualitative approach was followed. It combined a purposive and rapid synthesis review of the available evidence with the prior expert knowledge particularly in areas without enough research or lack of prior consensus (Gibert *et al.*, 2010). A nominal approach was used in four face-to-face meetings following a preliminary international conference in Watford (UK) in 2009. Minutes were prepared and approved by the group after every meeting. In addition extensive electronic exchange and seven teleconferences were organized to generate a consensus report and the recommendation submitted by the WG to the WHO Advisory Group. Once the recommendation was submitted to WHO, experts from this group continued to cooperate together to refine several aspects of the position agreement. A further review of the relevance of the multiple neurocognitive impairment approach vs the IQ single cut-off was carried out in 2014 (Bertelli *et al.*, 2014).

This paper provides a review of the conceptual proposals agreed upon by the WG related to the parent (or “supra-ordinal”) category for IDD, as the name of the new entity, its definition, and its subtypes.

As a first step, the existing ICD-10 category of MR was discussed at a meeting of the WHO International Advisory Group held in Geneva in September 2009. This discussion focused on whether: MR should be retained in ICD-11; MR should be kept in the ICD-11 chapter on mental and behavioural disorders; the name should be changed in ICD-11; and a specific WG was necessary on this particular topic. Following agreement on these issues by the Advisory Group, three expert meetings were organized in the following months. The first was a preliminary working meeting aimed at identifying key issues and recommending to WHO a work plan for the development of the ICD-11 in relation to prior expert knowledge and existing evidence on “MR/intellectual and learning disabilities”.

Subsequently, a 12-member WG was appointed by WHO, consisting of experts from all six WHO global regions with a range of different backgrounds, including psychiatrists (child, adult and neuropsychiatry), psychologists, and education professionals from academia, clinical practice and research. An extended review group was also formed to give feedback on the products of the WG. The WG had two meetings in 2010 to reach a consensus on recommendations for the name, definition, subtypes and architecture of IDD classification for ICD-11. During the second meeting, these proposals were finalized and the general structure of the content model developed.

Adaptations of frame analysis and the nominal group technique were used to facilitate WG interactions and to achieve a consensus. Frame analysis is a broadly defined method of enumerating and defining ideas and themes within a larger topic that is particularly useful for defining new concepts (Goffman, 1986; Salvador-Carulla *et al.*, 2014). Of the four components of frame analysis, the WG particularly focused on “frame bridging” – collaboration among experts who are interested in a topic but do not commonly interact due to different training backgrounds or other reasons – and on “frame amplification”, or the clarification and elaboration of a conceptual framework for discussion (Snow *et al.*, 1986). Four members of the extended WG searched the relevant literature, arranged it according to

key topics and prepared a list of key areas and questions to be debated at the preliminary meeting. Material collected prior to every meeting was made available for review prior to the meeting by all participants.

The nominal group technique helps to deal with poorly structured domains and facilitates the integration of prior information and expert knowledge. Once ideas and related questions are formulated, their relevance to the central problem is discussed, and ideas are re-formulated and clustered into coherent groups. All members are encouraged to participate in the discussion following a sequential order and every round was followed by a final debate.

Questions discussed by the WG included: name and terminology; placement in the ICD-11; definition; diagnostic criteria; severity/extent (subtypes); associated features, mainly problem behaviours (PB), aetiology, course, and co-morbidities.

All sessions at the three meetings were coordinated by a single identified facilitator, tape-recorded and summarized by appointed rapporteurs to produce a final document, which was subsequently circulated and agreed on by all participants. Voting was used in the four WG meetings, but not in the preliminary meeting, as the aim of the preliminary meeting was to delineate topics and produce recommendations for consideration by the WG (Figure 1).

Results

All the members of the WHO WG-MR agreed on the conclusions included in the consensus report until the third meeting, with the exception of the role of PB in IDD and in functioning. During the last meeting in 2012, one member expressed a general disagreement with most of the achievements of the consensus.

Conceptual framework

Terminology

After a thorough debate the term identified by the WHO WG-MR was “IDD”. The group acknowledged that there are several terms currently used worldwide to name this entity (World Health Organization (WHO), 2007a, b, c), that some of these terms have diverse meanings or are only used in specific countries, and that most of them are used interchangeably in both singular and plural forms. In any case, the final approach followed current developments in health ontology, mainly the development of “shared ontologies” and the use of “synsets”. A synset is a set of synonyms and a glossary corresponding to a word sense (Wordnet, 2011). A “synset” of this category is very complex. It encompasses synonyms, semantically similar terms, and inappropriate terms (names that cannot be used as they are socially unacceptable or incorrect from a taxonomical point of view) (see Table I).

A polysemic-polynomial approach has been endorsed by the WHO WG-MR (Salvador-Carulla and Bertelli, 2008). This approach which is related to the use of “shared ontologies” in ICD-11, distinguishes between “Intellectual Developmental Disorder” (a clinical meta-syndrome coded at ICD) and “ID” (its functioning/disability counterpart coded at ICF). The group acknowledged that these terms have different scientific, social, and policy

applications, although they describe two different but related aspects of the same construct (IDD/ID). According to the group these two terms should be regarded as “semantically-similar” (Sanchez *et al.*, 2010) but they are not “mirror codes”.

Conceptualisation of IDD and placement in the ICD-11 as a developmental disorder

The WHO WG-MR agreed that the primary parent category for the condition formerly named MR should be that of “Neurodevelopmental Disorders”. A neurodevelopmental focus was maintained throughout the whole consensus process. The disorders included in this meta-structure are not chiefly united by onset in childhood and adolescence, as in previous classifications, but by aetiopathogenetic risk and clinical factors, such as genetic alteration, abnormal neural circuit development, dysfunctions in cognition, learning, communication, behaviour, early emergence, and the continuing course. Within-cluster co-morbidity also supports grouping these disorders together.

IDD encompasses therefore a broad grouping of heterogeneous developmental conditions which result from significant interference with the growth and maturation of the brain during its early developmental phases including the prenatal and perinatal periods, infancy, childhood, and extending into adolescence.

The group considered IDD as a group of “life-span neurodevelopmental conditions with onset in early life, and early diagnosis is important in view of the need for early intervention services”. There is wide variability in the developmental presentation of persons with IDD, as many conditions underlying IDD have unique developmental trajectories (Harris, 2006). Although many known prenatal and perinatal conditions associated with IDD can be diagnosed at birth or during the neonatal period, other causes can emerge during the postnatal period. Dynamic interaction between biological and environmental factors influences development over time. Early diagnosis of IDD is critical especially given that children with delayed or no clear diagnosis usually fare poorer than those with an identified diagnosis (Goldberg *et al.*, 1986).

In spite of a major focus on the brain substrate of cognitive impairment, the group underlined the fact that “cognitive development and brain architecture are influenced by environment and experience”, apart from biology (Fox *et al.*, 2010). It also stressed that acquisition of cognitive abilities follows a hierarchical sequence such that attainment of more complex abilities is built upon foundations that have been established earlier in development (Shonkoff and Phillips, 2000). The cognitive, language, social, and emotional competencies develop in an interdependent fashion and are driven by experience and environmental factors.

Furthermore, the group made a clear call for a better understanding of “critical periods” in brain development and its impact on IDD, as “adaptation continues throughout life, and cognitive abilities are developed sequentially during times when the individual is optimally receptive to environmental influences, times referred to as sensitive periods” (Knudsen, 2004; Knudsen *et al.*, 2006). Critical periods are a type of sensitive period, in which the effect of experience on the brain is actually essential for normal development. Although much work has been done to identify the timing of these periods, and there is evidence

showing different sensitive periods for areas including language, attachment, and indiscriminate behaviour in young children (Nelson *et al.*, 2009), more research in this area is required to enhance understanding of the dynamics of cognitive development, and how interventions may best be timed to improve developmental outcome.

This approach had implications for early detection. The group emphasized that “persons with IDD often first presents to clinicians because of parental or other caregiver concerns about their development, e.g., limitation in or age-appropriate acquisition of skills, difficult behaviours, or co-occurring medical concerns. Parents may suspect a delay in development when younger siblings surpass the index child in developmental skills. Many severe and profound cases of IDD often may present earlier with impairments and delays in multiple domains of development. Many children with mild to moderate severity of IDD may not present to the clinician until school-age, when problems in learning become more noticeable, when they have increasing difficulties in managing their school work, or when they have growing difficulties following complex household activities and meeting expectations (World Health Organization (WHO), 2008). Some individuals with IDD may remain undetected until much later during adolescence or early adulthood years”.

This developmental perspective was combined with a life course approach to IDD group. Therefore: “While every person with IDD has a unique developmental trajectory, the longitudinal course of IDD can be described in a number of general patterns distinct from that of typical development, particularly when considering the features of particular etiological conditions (Munir *et al.*, 2008; Szymanski, 1989). These groupings may include: non-progressive linear trajectory; progressive trajectory often associated with metabolic disorders in which the manifestations of the underlying process occur after birth and evolve over time into a progressively deteriorating course; acquired trajectory with normal course of development up to the point of an insult with subsequent modification of the course of development (e.g. traumatic brain injury or severe psychosocial deprivation); and environmental disorder with fluctuating developmental course, with periods of stress, and periods of nurturance, or positive intervention, or both.

These different developmental patterns have relevant implications for diagnosis. Therefore the group recognized that “at any given cross-sectional encounter point with convergence of the various possible trajectories it is difficult for a clinician to make an informed decision about the developmental course of IDD. Since many genetic, metabolic and environmental factors can lead to the development of IDD across the life-span, it is therefore especially vital to undertake full medical history, physical examination, and requisite work-up, for a principal cause of IDD” (Shevell *et al.*, 2003).

The introduction of a developmental longitudinal perspective therefore is likely to improve understanding of how different presentations of IDD, with or without a known aetiology, may change and evolve over time. In addition, this could also help differentiate how different genotypes of known conditions associated with IDD, may lead to specific behaviours and cognitive profiles across the life-span. For example, young children with Down syndrome show increasingly better receptive language skills in advance of expressive abilities (Abbeduto *et al.*, 2007; Chapman and Hesketh, 2000). Likewise, boys with fragile X

syndrome show more pronounced pattern of higher-level abilities in simultaneous processing than in sequential processing over the course of development (Hodapp *et al.*, 1991).

Diagnosis of IDD: critical components

Following this conceptual framework the group expanded on the critical aspects of the diagnosis of IDD.

Definition of IDD

IDD is a group of developmental conditions characterized by significant impairment of cognitive functions, which are associated with limitations of learning, adaptive behaviour and skills.

Fundamental to the definition of IDD are not only impairments in the rate and level of cognitive development such as knowledge, reasoning, and symbolic representation, but also the associated functioning. These limitations of functioning are related to difficulties in learning, adaptation and acquisition and use of language. These limitations of functioning can be classified in the ICF. The ICF version for children and youth ICF-CY (WHO, 2007a) should be used to classify limitations of functioning in children as well as adults because it is sensitive to disorders of development and permits coding of basic and elemental functions associated with IDD.

Given the complex nature of IDD, a large number of associated functional impairments can be identified with this condition. Impairments related to the neurodevelopmental disorder of cognitive functioning are essential to the definition of IDD.

Diagnostic criteria

In the context of ICD, intelligence is not considered a unitary characteristic but an umbrella term that includes cognitive functioning, learning, adaptive behaviour and skills, that is age-appropriate and meets the standards of culture-appropriate demands of daily life.

IDD can be envisioned as an early cognitive “meta-syndrome” analogous to the syndrome of dementia in later life (Salvador-Carulla and Bertelli, 2008) in that a variety of conditions lead to these two meta-syndromes, they involve extensive impairment of core cognitive functions required for daily living and the evaluation process involves a broad consideration of biological, personal, and environmental factors.

IDD is a life-span condition requiring consideration of developmental phases and life transitions. A developmental approach which accounts for the complex causal factors known to impact the acquisition of cognitive abilities and adaptive behaviours is necessary to understand IDD, and to provide the appropriate interventions and long-term care that assist an individual in reaching his or her developmental potential (Hodapp and Burack, 2006).

There is still no conceptual map or hierarchy of the cognitive functions and domains important in IDD (Bilder *et al.*, 2009). Different terms are often used for the same functions, and vice versa. However, a series of cognitive domains are significantly impaired in persons with IDD. These core cognitive domains mainly include perceptual reasoning, working

memory, processing speed, and verbal comprehension (Deary, 2001; Holdnack *et al.*, 2011), and these domains are significantly impaired in persons with IDD. Several authors have listed other functions but these four domains are consistently cited (Holdnack *et al.*, 2011).

In general, persons with IDD have marked impairment of these cognitive functions. These cognitive functions are necessary for the development of knowledge, reasoning, and symbolic representation of the level expected of one's age, peers, culture, and community environment. Nevertheless, very different patterns of cognitive impairments and atypical features appear for particular conditions of IDD (Simon, 2010). As an example, persons with Down syndrome usually manifest impairments in specific areas of language, long-term memory and motor performance, while showing relative strengths in visuo-spatial construction (Edgin *et al.*, 2010). In contrast persons with Williams Syndrome show deficits in attention, visuo-spatial construction, short-term memory and planning (Tiekstra *et al.*, 2009), while showing a distinctive pattern of auditory processing and relative strengths in auditory processing and concrete language (Thornton-Wells *et al.*, 2010).

The impairment of the cognitive functions in IDD is generally assessed using the IQ, a score derived from an intelligence test which should be used for diagnosis only with consideration of locally standardized norms. Usually an IQ score 70 or below is indicative but not sufficient to diagnose IDD. Other alternative clinical and cognitive assessments could be used including locally standardized developmental tests. Cognitive assessment should not be limited to standardized IQ but also include neuropsychological testing, for example executive functioning testing to determine an individual profile. Moreover, retesting during critical life periods is recommended, as distinct cognitive developmental trajectories have been identified in different conditions of IDD and because there may be cognitive losses related to ageing.

To be diagnosed with IDD, the identified cognitive impairments should be associated with difficulties in different domains of learning, including academic, social, and practical knowledge. The objectives of the elementary school curriculum will barely be met. Reading, writing and calculating are hard tasks to achieve and will need considerable effort and repetitions. In addition, becoming independent and self-directed/self-care in the daily life will require practice and long-term monitoring. For example, a person with IDD will need a regular and consistent coaching and support to learn to solve everyday life problems, such as dressing skills and how to appropriately interact with others. These difficulties also vary across levels of cognitive functioning and other aspects of the disability and environment, as well as across different etiological conditions. For example, persons with Down syndrome show more problems in motor learning (Holdnack *et al.*, 2011). The person should also manifest limitations in adaptive behaviour; that is, meeting the demands of daily life expected for one's age peers, as well as cultural and community environment. These difficulties include limitations in relevant skills for daily life. They have been previously categorized into conceptual, social, and practical skills (American Association on Intellectual and Developmental Disabilities (AAIDD), 2010).

Persons with IDD often have difficulties in managing their behaviour, emotions, and interpersonal relationships, and maintaining motivation in the learning process. Cognitive impairment and adaptive behaviour limitations in IDD:

- IDD is characterized by a marked impairment of core cognitive functions necessary for the development of knowledge, reasoning, and symbolic representation of the level expected of one's age peers, as well as cultural and community environment. Nevertheless, very different patterns of cognitive impairments appear for particular conditions of IDD.
- In general, persons with IDD have difficulties with verbal comprehension, perceptual reasoning, working memory and processing speed.
- The cognitive impairment in persons with IDD is associated with difficulties in different domains of learning, including academic and practical knowledge.
- Persons with IDD manifest significant limitations in adaptive behaviour; that is, meeting the demands of daily life expected for one's age peers, cultural, and community environment. These difficulties include limitations in relevant conceptual, social, and practical skills.
- Persons with IDD often have difficulties in managing their behaviour, emotions, and interpersonal relationships, and maintaining motivation in the learning process.
- IDD is a life-span condition requiring consideration of developmental phases and life transitions.

IDD is an early-onset syndrome generally manifesting in infancy, childhood, and extending into adolescence. Although IDD is usually a stable diagnosis, there can be significant variability in cognition and functioning across different clinical severity levels throughout the lifecycle (Nelson *et al.*, 2007). Therefore, IDD is considered a dynamic health condition, and it should be reassessed at key developmental phases, life transitions (e.g. at school entry age, puberty, early and later adulthood), other life events, and traumatic events. No specific temporal qualifier is necessary for the diagnosis (Martorell *et al.*, 2009).

Severity/extent (subtypes)—Though the degree of cognitive deficits and adaptive skills vary from person to person with IDD, in most instances it is possible to categorize the condition into one of the four following clinical severity levels: mild, moderate, severe and profound. Although the prevalence is highest for mild, followed by moderate subtypes, severe and profound subtypes show similar rates (Stromme and Valvatne, 1998; Heikura *et al.*, 2003). Specific prototype characteristics of the severity subtypes are noted in the Annex.

Major issues in differential diagnosis: BIF

A detailed list of differential diagnosis was analyzed in the consensus document. Within them, “Borderline Intellectual Functioning” (BIF) deserved special attention as this other health condition is critical to define IDD, and to understand the difference between a “nosological” approach and a “normality approach”.

A previous strict normality approach, in which the diagnosis of MR was made by measuring impairments in cognitive and adaptive functioning that were one or two standard deviations below the population mean for the person's age and cultural group, is no longer considered adequate in the current diagnostic approach to IDD (Bejab and La Veck, 1972; Scheerenberger, 1983) by the expert panel. The bell curve approach to IDD is regarded as insufficient, and it is questionable from a methodological point of view. IQ ratings may vary as a result of the specific test being used, the testing conditions, status of the person being tested, the individual's life course, and even the historical generation. For example, the "Flynn effect" describes a substantial increase in average IQ scores on intelligence tests which may be about three IQ points per decade (Colom *et al.*, 2005) although stagnation has been identified in Scandinavian countries after the 1990s (Flynn, 2009). These variations are especially relevant for the diagnosis of mild IDD and BIF, and highlight the need for periodic standardization of IQ tests. Nevertheless, BIF may not just be understood from a normality approach, as evidence is emerging that there are significant similarities between individuals with BIF and IDD; for instance, a significant proportion of persons with BIF show mild cognitive developmental impairments and higher rates of mental illness. This population group have similar patterns of service response to mental health disorders, and have similar communication and support needs (Emerson *et al.*, 2010). In IDD, IQ rating is not an isolated diagnostic criterion, but should be considered a proxy measure of the "marked impairment of core cognitive functions necessary for the development of knowledge, reasoning, and symbolic representation" which characterizes IDD. Standardized developmental tests provide a complementary information source for the diagnosis of IDD where traditional IQ tests cannot be used (e.g. for infants, young children or persons with severe or profound IDD). In addition, simple screening tests are already available in several LMIC countries. It may be expected that simpler and more specific testing of the cognitive functions related to IDD may be available in the future.

BIF is a complex clinical entity typically characterized by IQ ratings between approximately 71 and 84, with problems in adaptive behaviour and academic performance beginning in childhood and adolescence (Salvador-Carulla *et al.*, 2011). Impairments in cognitive functions such as executive functioning and concentration have been identified in this population group, in addition to difficulties coping with more demanding, complex or unfamiliar tasks, and behavioural problems due to frustration and difficulties adjusting to social norms (Artigas-Pallares *et al.*, 2007). The recent evidence on the clinical characteristics, support needs and health policy implications indicate that BIF is a health condition that may require specific health attention as well as attention from other areas such as education, work, and the justice system (Emerson *et al.*, 2010).

On a standardized test, a significant limitation of adaptive behaviour and/or health-related functioning may be demonstrated in having a limitation in one of the following three types of adaptive skills: conceptual, social, or practical. In considering the assessment of adaptive behaviour, it is critical to consider limitations in present functioning must be considered within the context of community environments typical of the individual's age peers, as well as cultural environment; no existing measure of adaptive behaviour completely measures all aspects of adaptive behaviour; assessment depends upon understanding an individual's typical behaviour that goes beyond what can be observed in a formal testing situation; and

an adaptive behaviour score should not be considered as a precise score, because test properties such as standard error of measurement apply. Problem behaviour is not a dimension of adaptive behaviour, even though it may influence the acquisition and performance of adaptive behaviour and thus may be important in the interpretation of adaptive behaviour scores.

Adaptive behaviour must be understood in the context of developmental life-span periods and within the context of the individual's own culture, both of which may influence opportunities, motivation, and performance of skills (Schalock *et al.*, 2010).

Associated features: PB, aetiology, course, and co-morbidities

Associate features which are not core features to IDD but are very important to understand this health condition were also discussed and agreed by the expert group.

PB

PB are common in adults with IDD, with a point prevalence of about 20 per cent, depending upon the exact definitions and criteria used (Cooper *et al.*, 2007a; Deb *et al.*, 2001; Emerson *et al.*, 2001; Lowe *et al.*, 2007). PB present across a range of personal and social situations, although may be more frequent and/or severe in particular settings. Whilst they relapse and remit, they are also pervasive, and impact on development and subsequent opportunities. The commonest types of PB for persons with IDD are aggression (physically aggressive behaviour, verbally aggressive behaviour/screaming, destructive behaviour/aggression to property), self-injurious behaviour, overly-demanding behaviour, oppositional defiant behaviour, and sexually inappropriate behaviour (Deb *et al.*, 2001; Emerson *et al.*, 2001; Lowe *et al.*, 2007; Jones *et al.*, 2008). Furthermore, these behaviours often co-exist (Cooper *et al.*, 2009; Kwok *et al.*, 2010). Additionally, PB previously recognized in other populations are also experienced by some children and adults with IDD, such as pica, feeding disorders, stereotypies, and oppositional defiant disorder.

PB have biological, psychological, social and developmental aetiologies (Cooper *et al.*, 2007a). In view of the multi-factorial aetiology, where possible, assessments should consider all these areas, so that appropriate treatments and supports can be designed.

PB can occur as symptoms of other psychiatric or physical disorders. This is not dissimilar to, but occurs more commonly than in the general population in whom, e.g. pre-existing traits can be accentuated by super-imposed psychiatric disorder, and heightened irritability or paranoid ideation can lead to verbal or physical aggression. In these situations, other symptoms of the psychiatric/physical disorder will also be present and a temporal relationship will exist with regards to onset. Assessment of PB therefore always requires a thorough assessment of other possible psychopathology or ill-health as part of the differential diagnosis. Hence a special focus is needed on assessment of PB in IDD as they are a major cause of misdiagnosis, inadequate treatment, and service use in this population group.

Although the WHO WG-MR agreed on removing PB as a core component of IDD, there was no agreement on how to code them in ICD, either as a category coding at the same level as that for other developmental disorders or as an additional coding.

Behavioural phenotypes

Certain genetic syndromes that cause IDD are associated with other mental disorders and physical disorders. The term “behavioural phenotype” has been coined for a characteristic pattern of motor, cognitive, linguistic and social abnormalities which is consistently associated with a biological disorder. Whilst these associations are common in particular genetic syndromes, they are not inevitable; also, the presentation is not static and changes over the life course. Presentations are also variable, depending upon the persons severity of IDD, and can be influenced by a number of environmental, developmental and therapeutic factors (O’Brien, 2006). For these reasons, the occurrence of particular behaviours that are characteristic of a known behavioural phenotype should be coded as distinct, additional mental disorders within the ICD classificatory system, as is the case for physical phenotypes – for example, additional physical disorders that include cardiac anomalies in people with Down syndrome. There are numerous examples of behavioural phenotypes, e.g. person-dependent social problems and attentional deficits in DiGeorge syndrome (velo-cardio facial syndrome) (Shprintzen *et al.*, 1992; Pulver *et al.*, 1994; Papolos *et al.*, 1996; Murphy and Owen, 1996), self-injurious behaviour in Lesch Nyhan Syndrome and hyperphagia or skin picking in Prader-Willi syndrome (Dykens, 2004; Beardsmore *et al.*, 1998; Soni *et al.*, 2007).

Aetiology

There are over a thousand genetic disorders that are associated with IDD (Hodapp and Burack, 2006). These conditions damage and interfere with the growth and maturation of the brain in during the prenatal, perinatal and postnatal periods. Prenatal conditions include genetic diseases such as chromosomal disorders (e.g. Down syndrome), and single gene disorders (e.g. inherited metabolic disorders such as phenylketonuria); adverse environmental influences such as severe malnutrition, deficiencies such as iodine or folic acid, and infectious diseases during pregnancy like rubella; and exposure to adverse materials and toxins such as pollutants, excessive radiation, substance use during early pregnancy (e.g. alcohol, leading to fetal alcohol syndrome), heavy metals, or harmful medications. Perinatal conditions include late pregnancy complications such as placental dysfunction, labour and delivery problems such as asphyxia, severe prematurity or birth trauma, and neonatal complications such as severe jaundice or septicaemia during the first four weeks of life. Postnatal conditions in infancy and childhood, extending into adolescence include traumatic brain injury, infectious encephalopathies, chronic lead exposure, severe and prolonged childhood undernutrition especially during the first two years of life (Victora *et al.*, 2008), and gross psychosocial under stimulation (World Health Organization Regional Office for South-East Asia, 2006; Maulik *et al.*, 2011; AAIDD, 2010; Nelson *et al.*, 2007). Coding of aetiology, when known, can be accomplished through diagnostic coding of the underlying disease. Such common codes may also be cross-listed under aetiology section of the clinical manual.

Course

IDD is a dynamic condition that continues throughout the life-span in most individuals. It is typically manifested during early childhood, may show high individual and condition-related variation in developmental trajectories (e.g. the development of sensory and cognitive functioning in adults with Down syndrome) and should be reassessed at key developmental phases, life transitions (e.g. at school entrance, transition from educational to occupational setting, and with changes in living arrangements), and after other life events and traumatic events. The experience of life events and trauma can be particularly difficult for a person with IDD who has limitations in adaptive functioning. While the timing and type of life transitions vary across societies, it is generally the case that individuals with IDD need additional support adapting to changes in routine, structure, educational or living arrangements. Traumatic life events (e.g. learning of the sudden death of a family member, being in a life-threatening accident, being physically or sexually assaulted) are important predictors of psychopathology in this population (Martorell *et al.*, 2009).

IDD is influenced by many factors including the maturation of the brain, gene-environment interactions, environment, culture, education, availability of supports and training opportunities as well as by the physical and mental health of the person (Beadle-Brown *et al.*, 2005). Most individuals with IDD continue to acquire skills and competencies, especially with optimal care, training, supports, education and opportunities for learning. Features across different age periods are summarized in Table II.

Other associated features and co-morbidities

The types of health conditions most commonly experience by persons with IDD differ from the most common types for the rest of the population, as do their leading causes of death (NHS Health Scotland, 2004), due to differing aetiologies and health-related behaviours. Persons with IDD have a very high level of co-morbid ill-health, and premature death (Tyrer *et al.*, 2007), and more so the more severe their IDD. There are multiple causes for this. Many causes of IDD are associated with physical and/or mental ill-health disorders, e.g. tuberous sclerosis with epilepsy and hypertension; Down syndrome with thyroid disorder, impacted cerumen, numerous visual anomalies, and dementia; congenital rubella and sensory impairments. Due to complex and multiple health needs, adults with IDD are also more likely to be prescribed multiple drugs. This polypharmacy can adversely affect health, due to side-effects and interactions. Adults with IDD are less likely to exercise (Finlayson *et al.*, 2009) and eat healthily (Melville *et al.*, 2007) than the general public. Persons with IDD do not always have the knowledge or understanding to make healthy choices, and are reliant on others for support. This is affected by carers' own background, training, motivation, and understanding. Often persons with IDD face difficulties accessing the health services they need, compounding these issues.

Clinical presentations are often atypical, with change in behaviour, loss of skills, or onset of PB sometimes being the first indicator that there is an underlying health problem. Adults with IDD typically have high levels of unrecognized physical and mental health conditions (Howells, 1986; Wilson and Haire, 1990; Beange *et al.*, 1995; Kapell *et al.*, 1998; Baxter *et al.*, 2006; Kerr *et al.*, 2005; Felce *et al.*, 2008) with ill-health presenting late, at more severe

stages of disease progression. Chronic disease monitoring is also less well addressed for persons with IDD (Chauhan *et al.*, 2010). Several factors contribute to this, such as the limited verbal communication skills of adults with IDD, limited self-awareness, impaired mobility, and PB. They are reliant on carers recognizing that they may have a problem and seeking out help. Sometimes, health conditions are misattributed by carers or health professionals as being part of the adult's IDD (diagnostic overshadowing), and hence are not addressed for this reason. It is therefore very important during contacts with persons with IDD to be aware of the high potential that they have a previously unrecognized health condition warranting investigation and intervention.

Associated psychiatric symptoms and psychiatric disorders

IDD is associated with a high rate of mental disorders; point prevalence is about 40 per cent (Corbett, 1979; Cooper *et al.*, 2007a; Bailey, 2007; Deb *et al.*, 2001) and annual incidence 8 per cent (Smiley *et al.*, 2007). Persons with IDD can experience the full range of mental disorders, and there is evidence that some occur more commonly than in the general population, such as schizophrenia (Turner, 1989; Cooper *et al.*, 2007a), bipolar affective disorder (Cooper *et al.*, 2007b), dementia (Cooper, 1997; Strydom *et al.*, 2007), autism (Fombonne, 2003), and attention deficit hyperactivity disorder (Cooper *et al.*, 2007a; Hastings *et al.*, 2005; Deb *et al.*, 2001). PB are also frequent in this population (Cooper *et al.*, 2007a; Deb *et al.*, 2001). A high index of suspicion is needed for mental disorders in this population, as they are frequently underdiagnosed or misdiagnosed, and mental disorders can markedly reduce a person's cognitive and adaptive functioning, and quality of life. Aetiology is often multi-factorial, including social, psychological and developmental factors as well as biology and genetics (Cooper *et al.*, 2007a; Smiley *et al.*, 2007). Hence it is important to be alert to the possible emergence of mental disorders at difficult life transitions, following life events, where there are known vulnerabilities, and in cases of IDD with known behavioural phenotypes.

Clinical presentations may vary from the typical pattern seen in persons without IDD, with the distortion of psychopathology increasing the more severe the person's IDD (Cooper *et al.*, 2003; RCP, 2001; Fletcher *et al.*, 2007). Additionally, sophisticated concepts such as guilt, recurrent thought of death or suicide, intrusive dread of gaining weight, etc. require attainment of a developmental level that will never be achieved by people with severe IDD, so will not be part of the clinical presentation of the disorder. Some other symptoms of mental disorders are difficult to elicit in persons with IDD, particularly where there is limited or no verbal communication skills, for example, that obsessional thoughts or images originate in the mind of the patient, or that the person recognizes that anxiety symptoms are excessive or unreasonable. Further considerations in assessment of mental disorder are the need to always distinguish long standing traits from new-onset symptoms, and to interpret finding within the context of the person's developmental level, e.g. there is nothing abnormal in a person with severe IDD having imaginary friends.

The diagnosis of personality disorder is made inconsistently in this population. This diagnosis should be considered when the difficulties in interpersonal functioning are considered to be independent of IDD (i.e. when there is a habitual negative interaction with

others because of personality attributes leading to distress and difficulty) then the diagnosis of personality disorder should be considered.

Discussion

To our knowledge the consensus report produced by the WHO WG/MR is the most extensive international effort undertaken to date to reach a consensus on a new name, positioning, definition, and diagnostic principles for MR. It has involved 30 experts from 13 countries, representing the different WHO global regions, and experts from both high income and developing countries.

One of the major changes agreed by the WHO WG-MR is the re-inclusion of IDD in the group neurodevelopmental disorders. In ICD-9 (WHO, 1977), MR was moved with the intention to provide greater visibility to the other disorders of this group and to align the system with the DSM. In introducing the multiaxial system in DSM-III (American Psychiatric Association (APA), 1980), the APA placed MR on Axis II (developmental disorders). Nevertheless, analogous meta-syndrome adult (presumably non-developmental) categories such as dementia were maintained on Axis I. It was supposed that such an approach would favour the better identification and recognition of co-occurring psychiatric disorders, coded on Axis I. Unfortunately, such a separation of MR from other developmental and mental disorders did not necessarily spur the anticipated progress of psychiatric knowledge, as well as promotion of further development of targeted services in most countries for individuals with ID/IDD, as reported by the WHO's Global Atlas on ID (WHO, 2007a). The APA Task Force on DSM5's representative who participated as non-voting member of the WHO consensus process, described plans for the creation of the neurodevelopmental meta-category (Chapter on neurodevelopmental disorders in DSM5). disorders. The incorporation of IDD under neurodevelopmental disorders parent category is likely to have significant implications, and hopefully will induce clinicians and researchers to re-analyze the hierarchy and the conceptual map of this meta-structure, as well as to better define boundaries between specific disorders and to consider differential diagnoses and co-morbidities more carefully.

The term IDD, as well as its definition, clearly identifies this condition as a health condition. This agreement is consistent with those of the 2008s position statement of the section of psychiatry of ID of the WPA (Salvador-Carulla and Bertelli, 2008), which recommended for ID to use more than one name and meaning for different audiences, purposes and classificatory contexts (Sanchez *et al.*, 2010). The WHO WG-MR distinguishes between the clinical meta-syndromes and their functioning/disability consequence, which have different scientific, social and policy applications. The first component should have a place in the ICD, while the second being included in the ICF. This approach is consistent with the one promoted by the WHO within the family of international classifications (see www.who.int/classifications/network/en/) and supports the public health mission of WHO and the provision of appropriate services and opportunities to people with IDD. However, it appears likely that the WHO has temporarily dropped the use of "IDD" in favour of "Disorders of Intellectual Development" (ICD-11 beta XXX). This new terminology proposed by WHO was in fact discussed during the WHO WG-MR review process, among others terms that

were proposed, and was not considered to be particularly preferable, as it was felt to “accentuate” the disordered nature of intellectual development per se rather than equally emphasize the intellectual and the neurodevelopmental nature of IDD, analogous to the approach in defining other neurodevelopmental disorders such as Autism Spectrum Disorders. This new WHO term also seems to target more on “intellectual development” rather than on the aetiological and preventative nature of neurodevelopmental disorders implicit in IDD.

The DSM-5 chose the term disability with disorder in parenthesis. Its use, although semantically similar to the term one used by the AAIDD (Schalock *et al.*, 2010), as well as other American professional and disability groups (American Psychological Association’s Division 33, Arc of US), the US Federal Government (President’s Committee for Persons with Intellectual Disabilities, Social Security Administration, Rosa’s Law PL 111-256), is not intended to reflect the disability construct. The APA did not provide any rationale for the choice to include an entity that is labelled as a disability in a classification of mental disorders other than to cite its ambiguous use in the scientific literature in reference to both disorder and disability. However, the DSM5 in essence did adopt a disorder construct and maintained the meaning of a health condition, aligned with the WHO WG-MR consensus document and the recommendations sent to the WHO Advisory Group for ICD-11. The term IDD was clearly expressed and clearly included in parenthesis to make clear that it is used in the same context as the ICD proposal of intellectual developmental disorder.

The need to define all the different cognitive conditions included in the meta-syndrome of IDD is expressed in the DSM5 by the removal of references to IQ scores from the body of definition, the highlighting of clinical judgment, operationalised re-definition of intelligence, as well as linking of cognitive and related adaptive functions though a focus on adaptive reasoning. In the explanatory text the DSM5 does require standardized individual intelligence testing but does not list require IQ ranges in defining severity categories. The convergence on the inadequacy of a precise age limit for onset of cognitive and adaptive impairments is also expressed in DSM5 by the removal of the age limit of 18 and its substitution with a generic reference to the developmental period.

The WHO WG-MR proposal of a more articulated model of cognitive impairment might have relevant implications not only for education and services provision, but also for the general cultural attitude towards IDD, by juxtaposing a new concept of dimensional cognitive characterization to that of normal/subnormal intelligence and by substituting the measurement of IQ with the assessment of specific cognitive functions (i.e. spatial attention switching in audition or working memory chunking in vision) and a contextualized description of consequent adaptive and learning difficulties (see Figure 2). The evaluation should aim to identify the cognitive dysfunctions that have the greatest negative impact in terms of behaviour, adjustment, autonomy, and above all quality of life, across the life-span.

This document exemplifies an integration of expert knowledge and empirical evidence in developing consensus related to clinical processes. The use of techniques that incorporate quantitative data analysis and formal prior expert knowledge had proven to be particularly

useful in order to transform existing evidence into beneficial information for taxonomy and classification in highly multifaceted domains with lack of research data.

Conclusions

The classification of IDD involves very complex and challenging conceptual issues. In recent years, considerable work has been done in the revision of the classification systems resulting in the APA's DSM5 and the WHO's ICD-11. In addition there has been significant contribution by the AAIDD and other American professional and disability groups. However, major conceptual differences exist between the social/educational perspective followed by AAIDD, and the clinical health condition approach taken by APA DSM and the WPA. It is important to note that intelligence testing was designed and developed with a goal being an implementation tool to determine needs and services in schools and selection in the military. This approach is at the center of the AAIDD's advocacy for normalization in using a normative approach to adaptive behaviour and focusing on the use of supports separately from the IQ. On the other hand the nosological approach to diagnosis seeks to understand the aetiology of neurodevelopmental disorders and seeks to apply neuroscience to the understanding of cognitive impairment and intelligence following a methodically focused discovery approach. The educational/social and the clinical approaches, although apparently antithetical, are legitimate and complementary and provide a content-rich framework to explain the different facets of one of the more complex constructs in the classification of health conditions.

In 2011 the WHO WG on the Classification of MR tried to encompass both perspectives and suggested to maintain the standardized measurement of general intelligence as a diagnostic criterion while it recognized that many different patterns of intellectual impairment in neurodevelopmental syndromes exist, and people diagnosed with them show very different results in subtest score measurement. To reach this final consensus on key questions regarding the new term, definition, and diagnostic principles, the WHO WG-MR used a mixed qualitative approach by combining available evidence with prior expert knowledge. This approach might be particularly suitable for the public health mission of the WHO and for the provision of appropriate services and opportunities to people with IDD.

The WHO WG-MR conceptualized "IDD" (IDD) as a metasyndromic health condition, which may be related to a variety of specific aetiologies. For the re-naming the WG endorsed a polysemic-polynomial approach with an aggregate meaning that distinguished between IDD (clinical meta-syndromes) and ID (their functioning/disability counterpart), with different scientific, social and policy applications.

IDD was thought to be included in a wider group of neurodevelopmental disorders, together with Autism Spectrum Disorders and Specific Developmental Disorders. ICD-10 diagnostic criteria were considered highly inconsistent with current scientific knowledge and clinical experience and were suggested to be conceptually clarified and changed. The WG agreed on a more articulated model of cognitive impairment, and proposed to complement the measurement of IQ with the assessment of specific cognitive functions and a contextualized description of consequent adaptive and learning difficulties. The WG also agreed on a more

operationalised and support-oriented criterion for adaptive functioning deficit and on the elimination of a precise age limit for onset of cognitive and adaptive impairments. This approach took into account the broader perspective of the WHO Family of Classifications (WHO-FIC) towards coding the health status in ICD, as well as functioning and environmental aspects in ICF, that allows for the classification of IDD in ICD and the additional use of ICF to code its functional aspects.

In any case the existing differences between the different approaches should be considered in the context of the philosophy of science, the separate history of the ID/IDD as a diagnostic construct, and the evolution of intelligence testing.

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Appendix

Table AI

Main characteristics of mild IDD

Persons with mild IDD are noticeably slower in learning to speak than others but usually achieve sufficient language abilities for everyday purposes, to hold conversations, and to engage in the clinical interview. However, such as relating complex experiences and complex discourses are limited. They may only show delays in cognitive and motor milestones

Difficulties are seen distinctly in academic skills and most have difficulties in the age-appropriate skills of comprehending, writing, and reading. The majority manage to acquire primary school education with support

Most achieve bowel and bladder control, and skills needed for independence in basic self-care activities (eating, washing, and dressing), as well as practical and domestic life. However, the development of these skills is considerably lower than the average person. As adults, most live a relatively independent life looking after themselves with appropriate support. They develop a capacity for open employment with initial support as long as it involves simple (unskilled/semi-skilled) tasks

In a socio-cultural context requiring little academic achievement, some degree of mild IDD may not be readily evident. However, activities that place greater demands on cognitive/adaptive abilities such as long-term partnership, child-rearing, fitting in with cultural traditions and expectations, dealing with increased demands on performance, or adaptation to stressors will make their difficulties more noticeable In general the behavioural, emotional, and social difficulties of persons with mild IDD, and the needs for treatment are more closely akin to those found in the general population. Evaluation of co-occurring psychiatric disorders may require some modification of diagnostic approaches to elicit psychopathology

The IQ is usually in the range 50–70

Table AII

Main characteristics of moderate IDD

As children they show impairments in multiple areas that include cognitive, motor, language and social domains

Persons with moderate IDD are typically slower in learning to speak, walk, in developing comprehension and learning simple tasks to look after themselves. However, they are able to become independent in basic self-care activities with adequate training by adolescence

School achievement is limited to the basic skills needed for reading, writing, and counting

The level of developed language in adulthood is variable: most of those affected can take part in simple conversations in domestic and social situations while some may develop better language skills in expressing more complex ideas such as giving directions to others

Discrepant profiles of abilities are common in this group, with some individuals achieving higher levels in visuo-spatial skills than in language-dependent skills

Usually, people with moderate IDD are fully mobile and physically active and the majority show evidence of social development in their ability to establish contact, to communicate with others, and, to engage in simple social activities

As adults, many can work productively if given extensive on-the-job support. They will rarely be able to live independently without considerable and regular support from others

In general the behavioural, emotional, and social difficulties of persons with moderate IDD are different to those shown by the general population, particularly for individuals with limited expressive language skills. Persons with moderate IDD show more associated medical problems than people with mild IDD, particularly epilepsy and mental ill-health, including problem behaviours

Co-occurring psychiatric disorders should be assessed using complementary diagnostic guidelines adapted for IDD

The IQ is usually in the range 35–49. It should be taken into account that several of the common IQ tests

Table AIII**Main characteristics of severe IDD**

As children individuals with severe IDD show gross impairments in attainment of all developmental milestones

They cannot speak beyond phrases or simple sentences. They often use signs or gestures to communicate

They may learn to read and write very simple words with intensive training

The person needs assistance with basic self-care activities such as toileting functions; however, by adulthood, they are likely to have mastered some of these with intensive training

Usually, people with severe IDD are mobile, but may be uncoordinated and lack adequate visuo-motor skills. The majority shows some evidence of social development and enjoy interaction

As an adult, most are able to perform very simple repetitive tasks household chores such as fetching, carrying and cleaning in closely supervised settings. They typically require considerable and regular support from others throughout the entire day

In general the behavioural, emotional, and social difficulties of persons with severe IDD differ largely from the general population

Persons with severe IDD may often have a psychiatric illness that goes undiagnosed due to limitations in verbal expression, and differences in presentation

Co-occurring psychiatric disorders must be assessed using complementary diagnostic guidelines adapted for IDD

These persons show more associated medical problems such as epilepsy, sensory impairment, gastro-intestinal disorders, and cerebral palsy than persons with mild or moderate IDD

The IQ is usually in the range 20–34. It should be taken into account that the accuracy of IQ measurements is decreased in this group

Profound IDD

In childhood, they show profound impairments in attainment of milestones in all areas of development; for instance, a two year-old-child with profound IDD is not able to sit without support, reach for objects, recognize close family members, or localize sounds

Persons with Profound IDD are unable to understand or comply with most except the simplest of verbal requests or instructions. They are capable of rudimentary forms of non-verbal communication and may develop vocabulary of a few words or phrases understood by caregivers

Persons with profound IDD are more likely to be immobile or severely restricted in mobility than persons at other ability levels

They are often incontinent, possess little or no ability to care for themselves, and so require intense assistance, for nearly all self-care activities, including attention during the night

In general the behavioural, emotional, and social difficulties of persons with profound IDD differ largely from other clinical severity levels of IDD. Psychiatric illness may go undiagnosed due to severe limitations in verbal expression and differences in presentation. Co-occurring psychiatric disorders must be assessed using complementary diagnostic guidelines adapted for IDD

They have more associated medical problems than persons with less severe IDD. Severe neurological or other locomotor disabilities affecting mobility are very common, as are epilepsy and visual/hearing impairments, and gastro-intestinal problems such as gastro-oesophageal reflux disorder and constipation

The IQ is usually under 20, but this measure is of limited value in this group

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Figure 1.
Model of IDD (WHO-IDD WG)

Figure 2.
Working group on IDD: procedure followed for developing recommendations for the international advisory group for the revision of ICD-11

Table I

Intellectual developmental disorders synset

Synonyms	Mental retardation
	Disorders of intellectual development
Semantically similar terms	Intellectual disability
	Global developmental delay (for Unspecified IDD) (Shevell <i>et al.</i> , 2003)
	Early developmental impairment (for Unspecified IDD) (Francoeur <i>et al.</i> , 2010)
Inappropriate terms	Learning disability/difficulty (ambiguous)
	Mental deficiency/subnormality (not acceptable)
	Mental handicap (ambiguous and not acceptable)
	Developmental disability (ambiguous and too broad)

Table II

IDD features across different age periods

Adolescence	During this stage, major transitions take place in physical growth and sexual maturation. Puberty and adolescence may cause increased social, emotional and behavioural difficulties for individuals with IDD. There may be changes in educational or care settings
Young adulthood	During young adulthood, individuals with mild or moderate IDD often have difficulties with personal relationships, managing finances and planning for the future. In most societies, this is also a time of significant life transition (e.g. moving to adult services, moving from living at home to a group home, moving from an educational to occupational setting or getting married), and this can put an individual with IDD at increased risk for problem behaviours and psychiatric illness. New psychiatric illnesses may have their onset in young adulthood. It is important to note that persons diagnosed with mild IDD in childhood may no longer meet criteria for IDD in later life, as their skills may be better suited to the new environmental demands or because of new brain maturation and learning. Even in this optimal group, individuals with a BIF diagnosis will require specific health attention as well as attention from other areas such as education, work, and the justice system
Adulthood	Many older adults with mild IDD may continue to work and live independently. There may be known condition-specific health risks that should be checked for in mid – adulthood (Esbensen, 2010). For example, older adults with Down syndrome have a higher prevalence of Alzheimer's disease and epilepsy. Family life transitions include bereavement of older family members and parents
Old age	Ageing is accelerated in many genetic conditions with IDD, for example, in Down syndrome. Dementia occurs at a higher prevalence in the population with IDD compared with the general population (Strydom, 2007). Early retirement is available for persons with IDD in countries with extensive social inclusion programmes. In addition to the onset of condition-specific health problems, age-related health conditions occur in old age (WHO, 2010; Haveman <i>et al.</i> , 2009)