

Chapter 4

Assessment of Psychiatric Disorders



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Background

In the last 20 years, the average life expectancy of people with intellectual and developmental disabilities (IDD) has increased considerably [1–3]. Since IDD represents a condition of high physical and mental vulnerability [4], the lengthening life expectancy has been associated with an increasing prevalence of physical health issues, mental health problems, and multi-morbidity related to aging, much higher than in the general population. In fact, in persons with IDD both the ageing process and the onset of mental disorders are reported to start earlier [5–7]. See Chap. 2.

This is particularly true for dementia, which appears to affect between 30% and 40% of persons with IDD and around 50–70% of those with Down syndrome (DS) older than 60 years [8]. In persons with non-DS IDD the average age of onset of dementia is around 10 years lower than the general population [9]. In DS, dementia in Alzheimer's disease affects at least one-third of individual in their forties [10, 11].

Psychiatric disorders are present in more than 40% of older adults with IDD, especially depression, anxiety, and psychotic disorders [5, 12]. In those with co-occurrence of autism spectrum disorder, the prevalence can further increase up to five times more than in people who present only IDD [13, 14], with a proportionate negative impact on individual functioning and quality of life as well as on family and the community [15].

Research indicates many different factors being associated with this high mental health vulnerability, including specific genetic syndromes, the extent of the central

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nervous system compromise (leading to associated neurological disorders and disabilities such as epilepsy, cerebral palsy, and sensorial impairment), life events, psychological factors (i.e. low skill for coping with stress, lack of environmental mastery), low level of health literacy, low compliance with hygiene rules, lifestyle and environmental conditions, health promotion/disease prevention practices, access to basic healthcare services, and reliance on other people for care.

The risk of psychiatric disorders in older people with IDD seems to be increased by exposure to more age-related life events than in younger adults. In particular, depressive and anxious symptoms were significantly heightened in individuals who reported more total and negative life events during the preceding year, also after controlling for a history of depression or anxiety disorder [1, 7]. Several surveys have reported positive correlations between lifetime exposure to life events and the development of psychiatric disorders, somatoform disorders and physical health issues, in persons with IDD [16–22].

However, in people with IDD mental disorders are difficult to diagnose, especially in old age. The first difficulties relate to clinical presentation, which is substantially different from that of the general population and which would, therefore, require the application of adapted diagnostic criteria as well as specific diagnostic procedures and tools, which are instead unknown to most clinicians. Differential diagnosis can be even more difficult to make, including that between dementia and some psychiatric disorders, such as depression or psychosis. Research findings on the co-occurrence of depression and dementia amongst older adults with DS are controversial precisely due to different interpretations of clinical presentations [23–25].

Peculiarities of Psychiatric Symptoms

In persons with IDD, the presentation of psychiatric disorders can considerably vary from that of the general population, especially for symptoms related to subjective experience in those with low or absent verbal communication skills, who may only be able to express themselves through changes in behaviour [4]. Symptoms can appear as both qualitative and quantitative changes of basic behaviours, the latter of which are indicated with the term “baseline exaggeration” [26–28]. Thus, symptomatology can be chaotic, intermittent, fluctuating, mixed, scarcely defined, extremely rigid, atypical, or masked [4, 29, 30]. Sometimes even the nuclear elements of some syndromes, such as suicidal ideation or delusion, may not be identifiable, especially if verbally mediated [31]. Some researchers have found that, concerning schizophrenia, the only first-rank symptom which can be detected with a good frequency through direct observation is auditory hallucination [32].

Scientists have proposed some explanatory models of this phenomenal complexity and difficulty to diagnose. One of the most common has been called “diagnostics overshadowing” [26], which consists in the tendency to attribute behavioural changes to the person’s disability rather than to one or more symptoms of an underlying psychiatric disorder and/or to dementia or another physical or mental problem.

Other explanatory models are the “intellectual distortion”, that is the presence of alterations of the level of cognitive, communicative, physical and social functioning [33], the “developmental inappropriateness”, that is the non-correspondence between the individual developmental level expected for chronological age and the level of effective individual development [34], and the “psychosocial masking”, that is the peculiarities of social, cultural and environmental experiences [26]. Another aspect that strongly characterizes psychiatric symptomatology in people with IDD is “neurovegetative vulnerability”: somatic symptoms, pain, changes in circadian rhythms, dystonias of the autonomic nervous system are frequently the main expression of many emotional dysfunctions [27].

Behavioural Equivalents

In people with IDD problem behaviours are common and represent one of the factors with the greatest negative impact on the implementation of rehabilitative interventions and on the quality of life of disabled persons and their families [35]. Some of the largest studies in this area found that about 25% of people with IDD have at least one problem behaviour, 5% show self-injury, and 10% physical outwardly directed aggression [36, 37]. When considered as a psychopathological category in persons with severe and profound IDD, problem behaviours account for 75% of all the diagnoses [6].

The term ‘problem behaviour’ refers to culturally abnormal behaviours of intensity, frequency, and duration such as to put at risk the safety of the person acting on them or others, or behaviour that severely limits or prevents access to the community’s ordinary services. It is often difficult to determine whether problem behaviours are an expression of physical pain, organic problems, drugs, psychological, environmental, socio-relational factors, or whether they represent psychiatric symptoms or even a combination of some of these aspects. The same behaviour can be interpreted in a quite different way by the various professionals, even within the same multidisciplinary team, with consequences that are sometimes relevant for the interventions.

Several studies have shown a relationship between problem behaviours and psychiatric disorders [38–43] particularly evident in persons with lower levels of functioning [39]. Some problem behaviours have been identified as symptoms, or groups of symptoms, specific to some psychiatric disorders, named “behavioral equivalents” [44]; they are characteristic for onset, development, maintenance, and extinction, especially compared to other possible symptoms of a psychiatric disorder [44–47].

Not all researchers support the idea that problem behaviours can be considered as “behavioral equivalents” of an underlying psychiatric disorder. More they are indicators of underlying emotional stress [48, 49]. In one of the most recent researches in this area, problem behaviours have been found to be associated more with dysregulation of wide psychological dimensions than with specific

psychopathologic areas [50]. For detection and interpretation of psychopathological symptoms and “behavioral equivalents”, dimensional symptomatologic models can have higher predictive validity than a categorical one. Further advantages can be given by a careful specification of problem behaviour and symptoms associated with them. For example, in persons with IDD and autistic features, self-oriented aggressive behaviours seem to occur more frequently during depressive episodes than during manic or hypomanic episodes. Within depressive episodes, verbal aggression seems to prevail in the clinical presentations characterised by the prominence of emotional symptoms (irritability, anxiety, and sadness). Physical aggression seems to be more common in individuals with more associated physical symptomatology (problems with energy, eating, sleeping, and stress) [51].

In persons with IDD, reference to behaviours as part of psychiatric psychopathology may represent relevant support for clinicians in the diagnostic processes and the treatment choice, especially during the old age, in which cognitive and communication skills often undergo further reduction.

Diagnostic Criteria

Research findings showed that the use of diagnostic criteria developed for the general population does not allow a correct diagnosis of psychiatric disorders in persons with IDD [45] and that adapted criteria should focus on observational-behavioural symptoms, especially in older people and in those with higher cognitive and communication impairment.

In the last 25 years, some adaptations have been produced, for both the Diagnostic and Statistical Manual of Mental Disorders (several versions) system and the International Classification of Diseases and Related Health Problems (several versions) by the Royal College of Psychiatrists (UK) and the National Association for Dual Diagnosis (USA). The former organisation has published an adaptation of the ICD-10; Diagnostic Criteria for Learning Disability (DC-LD) [52]. The latter organisation has adapted changes to the DSM-IV-TR and the DSM-5 and produced the Diagnostic Manual—Intellectual Disability (DM-ID) [53, 54]. As an example, the clinical guide of the DM-ID (version 2) states that depressed mood can be described by others in one or more of the following ways, that constitute a significant change from the baseline: sad or angry facial expression, crying, assaults, self-injurious behaviour, spitting, yelling, swearing disruptive or destructive behaviours [55].

Symptoms of mania or hypomania are more difficult to detect than depressive symptoms, and in some cases, the latter can mask the former, such as in the presence of irritability [56]. The differentiation can be supported by the identification of an increase of goal-directed activities, which are indicated by the clinical guide of the DM-ID (version 2) to present with engagement in activities in a “sped up manner”, rarely sitting down, being up and down from seat a lot, pacing, walking rapidly, appearing “driven”, racing around the room, and becoming very intrusive [57].

Another interesting example of behaviorally adapted diagnostic criteria can be provided in respect to visual, auditory, somatic, and multi-sensorial hallucinations, which are described in the DM-ID (version 2) and in some research reports in ways as the following: cover of eyes or ears, stare, nod, self-talk, make gestures as to remove something from the body, wear heavy, tight-fitting or multi-layered clothing, bandage ankles or wrists, move as if defending, fighting, or flirting [58, 59].

Although they need some improvements, these criteria have a valuable clinical utility, helping the clinician to better recognize and diagnose psychiatric disorders in persons with IDD and to better differentiate them from other psychiatric and neurological disorders [60].

Other Assessment Issues

In older people with IDD the assessment of psychiatric disorders should be appropriately modified and adjusted for the cognitive dysfunctions, language and communication limitations, sensory impairments, skill deficits, adaptations difficulties, and for physical disabilities often present. Another important issue is the reliability of the information source, including the persons with IDD, who may have poor verbal expression skills, tend to acquiescence and present cognitive distortion, that is difficulties in the ability for introspection, to define one's own experiences and to communicate the states of suffering or to recognize that they need help to identify the meaning of some changes in their psychic functioning [61].

Information from informants can be very helpful to the psychopathological assessment by adding historical information, clarity, and depth that might otherwise be missing. Conversely, some researchers have highlighted several limitations of the proxy perspective, which is often heterogeneous, contradictory, and with low inter-rater reliability. For example, family members are often struggling to answer questions about the presence of further disorder of mental functioning, and the indirect or atypical presentation of symptoms often limits the capability of identifying them for caregivers or other informants.

Several studies have shown that informant evaluation may vary depending on a number of factors, ranging from the cultural level and other personal characteristics to the specific nature of the relationship and involvement [62, 63]. Furthermore, the accuracy of the ratings depends strictly on the experience and insight of the informant, as well as on how well he/she knows the person being rated. The degree of agreement or disparity between ratings made through different informants is unclear [45]. The use of a comprehensive diagnostic system that includes the integration of different sources of information, such as the same individuals with IDD, carers, familiars, or other informants is highly recommended to measure any significant behavioural change that has occurred, to interpret it as symptom equivalent, and to increase the validity of the psychiatric assessment [45].

In addition to the sources of information, the setting of the evaluation also deserves special attention. The duration of the assessment should be short and

flexible, preferably in the context of the habitual life of the person with IDD, in order to reduce the impact of contextual variables on psychic and behavioural conditions. If this is not possible, a welcoming and silent place with limited sensory stimuli should be chosen. The evaluator should communicate in simple language, avoiding metaphors and idiomatic expressions. The question style should have many different answers and images and symbols are very useful for the communication between the evaluator and the person with IDD.

Assessment Tools

For all the above-mentioned issues, the application to people with IDD of the assessment tools developed for the general population also showed significant limits of validity and reliability. For example, screening tools and neuropsychological tests, such as the Mini-Mental State Examination [64, 65], determine a floor effect in most people with more severe disabilities and do not allow to identify a single cut-off threshold in mild and moderate forms [66]. Autoptic and neuroradiological studies have found changes in the central nervous system typical of Alzheimer's disease in almost all adults with DS over 45 years of age [67, 68], while the clinical manifestations do not seem so universal. The diagnostic difficulties could be precisely at the root of this discrepancy.

Researchers specialised in this field have worked partly on creating new tools, partly on translations and adaptations of questionnaires developed for the general population. In both cases, the tools have numerous limitations in applicability and effectiveness. The adjusted questionnaires also present sensitivity problems, due to the considerable atypical psychopathological symptoms of people with.

Currently, the most commonly used tools are general screening ones, which provide indications on all diagnoses compatible with the symptoms and behavioral equivalents detected in a person, but there are also tools which are specific for the psychopathological area. Instead, there are few standardized diagnostic interviews, which are long, expensive, and less useful for multidisciplinary collaboration. The tools must be chosen on the basis of the specific purpose, the characteristics of the person to be evaluated, the administration time, the training required, and the resource available [69].

Among standardized interviews addressed to people with good communication and introspective skills the Psychiatric Assessment Schedule for Adults with Developmental Disabilities (PAS-ADD) [70] was designed to provide all members of a care staff with a rapid psychopathological screening tool for adults with IDD and to collect information useful for setting the care plan. This instrument produces a diagnosis according to ICD-10. It showed reasonable reliability and validity [71, 72], and is also available in two other forms: the Mini PAS-ADD [73] and the PAS-ADD Checklist [74]. PAS-ADD boasts a high number of high-quality studies, is a reference tool in the United Kingdom and its use is also growing in other European countries [75–78]. However, some sensitivity problems have been identified, in

particular for disorders of the schizophrenic spectrum, reproducibility between different evaluators and consideration of anamnestic clinical information [4].

The Psychopathology Instrument for Mentally Retarded Adults (PIMRA) [79–81] is a structured interview, available both in a self- and other-report form, based on the DSM diagnostic criteria. It has been developed to screen for the presence of psychopathology in individuals with mild and moderate IDD. While showing increasingly frequent concordance difficulties between evaluators, PIMRA has proven to be particularly useful in the field of research, therapeutic planning, and evaluation of treatment outcomes. Furthermore, a revised version of the PIMRA has just been developed (PIMRA-II) and it seems to have good psychometric properties [82].

The Diagnostic Assessment for the Severely Handicapped (DASH-II) [83], which is the revised version of the DASH [84] is mainly based on the detection of key symptoms related to different syndromic groups, which can be defined by frequency, duration, and severity. The main limit of this scale is represented by the number and quality of the items, which are unbalanced for the various psychopathological dimensions, and present poor alignment with the criteria of the categories of the main diagnostic manuals [4].

The Psychopathology Checklists for Adults with Intellectual Disability (P-AID) is a battery of tools developed recently and allows to identify ten different psychiatric disorders and eight types of problematic behaviors according to DC-LD diagnoses. P-AID showed high values of internal consistency and inter-rater reliability, while sensitivity and specificity are still to be confirmed [85].

A comprehensive set of tools for all the different practical needs related to psychopathological assessment and monitoring, to be used by mental health professionals and the whole multidisciplinary team working with people with IDD is represented by the Systematic Psychopathological Assessment for persons with Intellectual and Developmental Disabilities (SPAIDD) [86]. It includes tools for every phase of the clinical intervention, such as general psychopathological screening, diagnostic categorical specification, dimensional diagnosis, and symptoms monitoring. The items of the SPAIDD system have been kept unchanged across all the battery tools, since they represent observable and behavioural aspects of cross-categorical symptoms. See Chap. 5 for further information.

In designing and implementing this system, authors tried to overcome also the other main limits of previous tools, such as the impossibility of being used for all the degrees of intellectual and communication impairment, the misalignment with the DSM or the ICD, the lack of some main symptoms or syndromes, the lack of chronological criteria, the long times of administration, and the scarce interdisciplinary usability [86]. The version for General screening (SPAIDD-G) is the first already validated and published tool of the SPAIDD system [4, 86]. It includes 56 items, which represent descriptions of the most frequent observable and behavioral aspects of all the symptoms that appear in the various DSM-5 diagnostic categories. These items were developed to be rated by a mental health professional through the information gathered by interviewing a family member of the person with IDD or another informant who has an awareness of changes in the behaviour of the people

for whom they care. The SPAIDD-G showed very good psychometric characteristics. To date, it is available only in Italian, although validations of the English, German and French translations are already underway.

Diagnosis and Differential Diagnosis of Dementia

Diagnosis of dementia is often difficult in people with preexisting cognitive deficits, and it is therefore important to make it by referring to valid specific diagnostic criteria and diagnostic tools. Important guidelines on this issue have been provided in the last decade by the British Psychological Society [87] and the National Task Group on Intellectual Disabilities and Dementia Practice [88], although there is currently no standard protocol for the assessment of dementia in persons with IDD.

Some of the early symptoms of dementia may be subtle or may present as an exacerbation of the existing behavioural traits or manifest differently in people with IDD than in the general population, as it has been described for psychiatric disorders. In most cases, these symptoms are noticed first by caregivers, as unusual changes in the individual's behaviour. Therefore, the use of informant-based instruments is recommended for screening or diagnostic purposes. Review of the literature [89, 90] in the last decade identified 79 instruments for direct evaluation and 35 for informant rating and provided detailed descriptions of selected neuropsychological assessments. Frequently used instruments in IDD research which have been thoroughly studied for their psychometric characteristics are; Dementia Scale for Down Syndrome (DSDS) [91], Dementia Questionnaire for people with Learning Disabilities (DLD) [92], and the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) [93].

The DSDS is an informant interview and includes 60 items organized into three categories, one for each of the stages in which the progress of the disorder is normally divided. The DSDS is designed for use with people with DS and standardized in a sample of adults the majority of whom had severe to profound disability, but may also be useful for people with IDD [94].

The DLD (formerly the Dementia Questionnaire for people with Mental Retardation—DMR) [95] has 50 items that have been divided up into the categories of 'short-term memory', 'long-term memory', 'spatial and temporal orientation' (making up the sum of cognitive scores; SCS), 'speech', 'practical skills', 'mood', 'activity and interest' and behavioural disturbance (making up the sum of social skills score; SOS). The cut-off scores for screening positive for dementia are different for individuals who have mild, moderate or severe disability respectively.

The DSQIID is another common screening tool, which is best used in clinical settings to deeply explore each item with a habitual caregiver in order to build up a valid clinical picture. It comprises 43 questions organised in three sections and should be used prospectively; however, certain items may show floor effect and total score may decrease as dementia progresses. The DSQIID has been included, with some adaptations, in the evaluation package of the National Task Group on

Intellectual Disabilities and Dementia Practices (NTG), called NTG-Early Detection Screen for Dementia (NTG-EDSD) [96], and recently translated and validated in various languages [97–100].

For people with good introspective and communication skills, there are also some adaptations of tools widely used in the general population, one of the most recent is the Cambridge Examination for Mental Disorders of Older People with Down's syndrome and Others with Intellectual Disabilities (CAMDEX-DS) [101]. The CAMDEX-DS consists of approximately 150 questions that ask about functional difficulties in different cognitive domains, whether such difficulties represent a deterioration in function, and the extent of the deterioration (slight or great) [102]. These tools can also support clinicians in the differential diagnosis between dementia and some psychiatric disorders, such as depression and psychosis, which are reported to be frequent co-morbid conditions and confounder of dementia in old persons with IDD [103, 104].

Most patients referred to as affected by depressive pseudo-dementia often show cognitive symptoms outside the range of dementia and a more rapid decline (over weeks to months rather than years). Depression does not significantly impair performance on cognitive testing if testers provide enough time and encouragement [105, 106]. Referral for complete neuropsychological testing can be helpful in clarifying the diagnosis in many cases. Treatment with antidepressants will significantly improve cognitive function in patients with pseudo-dementia, whereas persons with dementia may limit to partial improvements in overall functioning [107]. Recent research findings suggest that evaluation of facial recognition and left hippocampal volume may provide more reliable evidence for distinguishing pseudo-dementia from dementia in Alzheimer's disease [108].

Evidence on the relationship between psychosis and dementia in persons with IDD is lacking. Basing on data from the general population, psychotic symptoms are likely more common in prodromal and early dementia than previously indicated by factor analysis studies, although they are much more common in established dementia [109]. Psychotic features of dementia include hallucinations (usually visual), delusions, and delusional misidentifications. Delusional misidentification result from a combined decline in visual function and cognition and frequently manifest with strange suspicions or beliefs, such as family members being impostors, strangers living in one's own home, or with failure to recognize one's own reflection in a mirror. Hallucinations often do not upset patients and may be pleasant [110], contrary to what happens in the co-occurrence of schizophrenia, in which hallucinations usually have negative contents and determine severe problem behaviour. Symptoms of very late-onset schizophrenia show greater dissimilarity with psychotic symptoms associated with dementia with Lewy bodies than with those associated with Alzheimer's type, particularly more partition delusions and more auditory hallucinations of human voices. Processing speed and executive function seem to be comparably impaired among the three conditions. Patients with psychotic disorders at baseline such as schizophrenia seem to have a high risk of developing dementia over time, although finding are controversial [111].

Conclusion and Future Issues

Despite the evidence of need, adults and older people with IDD have a much lower rate of appropriate physical and mental healthcare (about a third of the population) and much larger difficulties in achieving services [112, 113]. The quality of mental health care is particularly poor and the rate of improvement is slow compared to that for the general population. Here the proportion of satisfied and unsatisfied care needs drops to a quarter of the norm [114, 115]. Already in early adulthood, the person with IDD and mental health problems is left without specialist reference. One of the main reasons for this serious situation seems to be the lack of experienced and properly trained specialists.

As mentioned in this chapter, people with IDD often have ways of communicating and displaying symptoms, especially symptoms of psychiatric disorders, which cannot be adequately understood by health professionals who have not received specific training. Graduate courses in medicine and specialization in psychiatry of national universities do not include the mental health issues of this population. The current attitude of neurologists, geriatricians, and psychiatrists towards IDD-specific problems is severely limited [116]. The need for training also concerns socio-health workers, educators, basic carers, family members, and other non-professional caregivers [117]. The current evaluation shortcomings are linked to even greater limits of socio-health systems and policies, determined in turn by long-lasting cultural issues, such as the prejudice that people with IDD cannot experience emotional suffering or the belief that rehabilitation interventions could have some efficacy only during childhood and adolescence. The idea that IDD is a life span condition is relatively recent. The first international document referring to it dates back to 2001 and the first European data on health problems are from 2008 [118].

The Edinburgh Principles, produced in 2001 by an international group of experts led by Wilkinson and Janicki [119], still represents a good reference for the care of the elderly person with IDD. Recently, the National Task Group (NTG) on IDD and Dementia Practices [120] and in the British Psychological Society and the Royal College of Psychiatrists [9] jointly produced consensus recommendations for the evaluation and management of dementia in adults with IDD. These recommendations follow a longitudinal perspective, forcing caregivers to take into account the individual's mental health history and to assess the symptomatological potential of behaviours, especially in terms of change from the baseline level. This perspective is not so obvious for providers of continuous assistance to people with IDD, often flattened into a sort of eternal present.

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