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Cognitive dysfunction and psychopathology: a cohort study of adults with intellectual developmental disorder

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Abstract

Background Cognitive impairment of intellectual developmental disorders (IDD) is determined by several different combinations of specific cognitive alterations. People with IDD present a rate of mental health problems that is up to 4 times higher than that of the general population. Despite this, the relationship between specific cognitive dysfunctions and co-occurring mental disorders has not been adequately studied. The aim of the present paper is to investigate the association between specific cognitive dysfunctions and specific psychiatric symptoms and syndromes in people with IDD.

Methods One hundred and twenty adults with mild to moderate IDD living in residential facilities underwent a clinical and instrumental assessment for specific cognitive and psychopathological features.

Results Participants with IDD and ASD have significantly lower scores compared to those without respect to who has not the diagnosis on the Processing Speed Index (PSI) and Perceptual Reasoning Index (PRI) on the WAIS-IV and higher time scores on the TMT A. Moreover, there is a significant association between years of hospitalisation and TMT B and TMT B A time scores; the longer a participant with IDD was hospitalised, the worse their performance on the TMT. Although not statistically significant, many psychopathological clusters showed substantial cognitive profiles.

Conclusions Although further research is needed, neuropsychological and IQ tests scores seem to be differently associated to various psychopathological conditions co-occurring with IDD, and with ASD especially. Cognitive assessment seems to support

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diagnosis and treatment of psychopathological co-occurrences in persons with IDD, also in consideration of indirect implications including a better knowledge of the patient's characteristics beyond IQ deficit.

Keywords ASD, Cognitive functions, Cognitive/neuropsychological batteries, Diagnosis, IDD, Psychopathology

Introduction

Intellectual disability (ID), renamed intellectual developmental disorder (IDD) in the DSM-5-TR (American Psychiatric Association 2022) and disorder of intellectual development (DID) in the ICD-11, is defined as a 'disorder with onset during the developmental period that includes both intellectual and adaptive functioning deficits in conceptual, social, and practical domains' (APA 2022).

Once IDD is diagnosed, clinicians should specify the level of severity (mild, moderate, severe and profound) based on clinical and test evaluations of intellectual and adaptive functioning rather than on intelligence quotient (IQ) scores alone (APA 2022). ICD-11 provides clinicians with behavioural indicators for additional guidance when interpreting levels of severity for IDD and when specific disorders co-occur (Lemay *et al.* 2022).

Evidence from neuropsychology, genetics, neuroimaging and functional anatomy increasingly shows a high variability in different cognitive abilities in both individuals with IDD and the general population, which challenges the usefulness of the concept of overall intelligence and the use of IQ (Bertelli *et al.* 2018). This is particularly relevant at this point of time, given that the ICD-11 conceptualisation of IDD includes a wide range of syndromes as well as non-syndromic conditions, which have only early cognitive impairments in common (Bertelli *et al.* 2018).

The case for using specific cognitive functions in assessing IDD is also supported by recent research on cognitive and behavioural phenotypes (Bertelli *et al.* 2018).

Epidemiological studies report a high prevalence of psychiatric co-morbidities in persons with IDD. Cooper *et al.* (2007) identified a point prevalence of mental ill-health based on clinical diagnoses of 40.9% (including challenging behaviours) or 28.3% (excluding challenging behaviours); prevalence rates based on classification manuals are lower: 35.2% (Diagnostic Criteria for Learning Disability, DC-LD), 16.6% (ICD-10-DCR) and 15.7% (DSM-IV-TR). More recently, Mazza *et al.* (2019) found a prevalence rate of 33.6%.

Causes determining the high psychiatric vulnerability of persons with IDD tend to consist of a complex combination of biological, psychological and socio-environmental factors (Luckasson *et al.* 2002; Bertelli *et al.* 2015b; Santambrogio *et al.* 2021a), but it is possible that the presence of cognitive alterations constitutes an element of vulnerability to developing psychopathological conditions (Day and Dosen 2002; Bertelli *et al.* 2018). In the light of the latest neuropsychological and neurobiological evidence of the multi-component structure of intelligence (Pascual-Leone *et al.* 1999; Naglieri and Das 2002; Johnson *et al.* 2008) and of the variability of cognitive profiles in IDDs (Friedman *et al.* 2006; Willner *et al.* 2010; Bertelli *et al.* 2018), it becomes necessary to investigate a possible relationship between mental health problems and specific cognitive deficits rather than a global intellectual capacity.

There is much evidence for this link in the general population (Millan *et al.* 2012). For example, the schizophrenic syndromes often have deficits in perception, attention, memory and problem-solving. Besides, in the mood disorders, there are frequent problems with focused and sustained attention, working and verbal memory, abstract reasoning, visuospatial and visuomotor skills, verbal fluency and executive deficits. Moreover, unipolar depression is associated with a lack of attention, language and visual memory. Finally, the obsessive–compulsive disorder is characterised by deficits in visuospatial and visuoconstructive skills, coding and recall, sustained attention, non-verbal memory, implicit memory and spatial memory (Trivedi 2006).

To the best of our knowledge, no studies have been conducted addressing the correlation between dysfunctional cognitive patterns and the presence of specific psychiatric symptoms in people with IDD.

A few studies have marginally identified a generic association between executive functions and co-occurrent psychiatric disorders. Woodcock and colleagues found a measure of switch cost in

Prader–Willi syndrome to be positively correlated to scores on preference for routine and predictability, which are related to repetitive questions and temper outbursts. In persons with fragile X syndrome, the same measure has been found to be related to excessive displays of anxiety, which involve stereotypical movement and repetitive self-injurious behaviour (Woodcock *et al.* 2009a, 2009b; Woodcock *et al.* 2010; Woodcock *et al.* 2011).

Demily *et al.* (2014) found that the 22q11.2 deletion syndrome, which is characterised by difficulties in sustaining attention, executive function, memory and visualspatial perception, not only presents a high rate of bipolar disorder and major depression, especially in adolescence and young adulthood, but also anxiety disorders and schizophrenia in adulthood (Gothelf *et al.* 2008; Santambrogio *et al.* 2020). Zarchi *et al.* (2014) compared the neuropsychiatric and neurocognitive phenotypes of this syndrome with those of Williams syndrome, which also has a lower Performance-IQ than Verbal-IQ but shows more severe impairment of visuospatial functions. They found that anxiety, mood and disruptive disorders have an equally high prevalence in both syndromes, but the 22q11.2 deletion syndrome presents a higher rate of psychotic disorders and the Williams syndrome a higher rate of specific phobia.

Some researchers have specifically addressed the association between ASD, co-occurring with IDD, and cognitive issues. ASD has been found not to be associated with general low IQ but with abnormalities in information processing, which in turn have pervasive effects on the overall functioning of the individual (Scheuffgen *et al.* 2000; Anderson 2008). Persons with ASD also show superior performance (in relation to their general mental age) on the Embedded Figures Test (Jolliffe and Baron-Cohen 1997).

The block design test was found to be quite useful in differentiating ASD and somatic symptom disorder (SSD) in persons with IDD. In fact, those with IDD alone or in association with ASD or SSD are reported to score significantly lower than those with only ASD or SSD. People with ASD show a weaker performance than those with SSD, also when their condition is associated with IDD. Although the data are not statistically significant, persons with ASD are reported to score better than those with SSD on digit span, picture completion, object assembly and digit symbol subtests. Statistically, persons with SSD perform significantly better than those with ASD on the digit symbol, which expresses visualspatial, visual-motor integrative and working memory skills. Those with IDD alone or in association with ASD or SSD also have significantly lower scores on Total IQ, as well as on Verbal and Performance indexes. The Total IQ score tends to be higher in ASD than in SSD, but the trend reverses when these conditions are associated with IDD. The Verbal and Performance scores are lower in persons with SSD than in those with ASD, also when the condition is associated with IDD (Piva Merli 2015; Bertelli *et al.* 2015a).

The objective of this study is to improve understanding of the association between general and specific cognitive functions and co-occurrent psychopathologies in persons with IDD.

Methods

The study

The present study is a cross-sectional cohort study including a convenient sample of 120 persons (90 male, 30 female) with IDD recruited from two Italian residential facilities to undergo an assessment battery of neuropsychological and psychopathological tests.

The study sample had a mean age of 57 years (range 22–83 years). The majority of the participants had severe IDD (59%), which impacted the proportion able to perform cognitive and neuropsychological tests. The mean length of stay was 36.44 years (range 1–76 years), and 52% of the sample had no formal education. Further details of the demographic and clinical characteristics of the sample are reported in Table 1.

Ethics

The study was devised adhering to the guidelines of the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) Statement (von Elm *et al.* 2008). The research project also complied with the principles of the Declaration of Helsinki regarding medical research in humans as well as local research ethical requirements and was approved by the University of Milano-Bicocca Ethics Committee (protocol number 575 – January 2021).

Consent was obtained according to the severity of the participants' IDD and their cognitive capacity:

Table 1 Socio-demographic and clinical characteristics

Variables		*N*	%
Gender	M	90	75%
	F	30	25%
Facility	IDD congregate setting	101	84%
	Psychiatric facility	19	16%
Education	Missing data	16	13%
	No formal education	62	52%
	Special schools for IDD	8	7%
	Elementary school	6	5%
	Middle school	23	19%
	High school	5	4%
Levels of IDD severity	Borderline	3	2%
	Mild–moderate	32	27%
	Moderate–severe	5	4%
	Severe	71	59%
	Profound	0	0%
	Not applicable	9	8%
Physical co-morbidities	None	13	11%
	At least one	107	89%
	of which: 1 co-morbidity	*34*	*32%*
	of which: 2 co-morbidities	*30*	*28%*
	of which: 3 co-morbidities	*25*	*23%*
	of which: 4 co-morbidities	*12*	*11%*
	of which: 5 co-morbidities	*6*	*6%*
Specific physical co-morbidities NB % with respect to *N* = 120, but a patient can be in more than one class	Epilepsy	48	40%
	Neurologic disease	8	7%
	Skeletomotor disease	34	28%
	Cardiovascular disease	30	25%
	Gastroenteric disease	21	18%
	Ocular disease	20	17%
	Endocrine disease	20	17%
	Urologic disease	19	16%
	Otorhinolaryngoiatric disease	8	7%
	Lung disease	8	7%
	Liver disease	8	7%
	Dermatologic disease	7	6%
	Obesity	6	5%
	Haematological disease	5	4%
	Cancer	3	3%
	Gynaecologic disease	2	2%
Psychiatric co-morbidities *from clinical records*	None	58	48%
	At least one	62	52%
	of which: 1 psychiatric co-morbidity	*53*	*50%*
	of which: 2 psychiatric co-morbidities	*7*	*7%*
	of which: 3 psychiatric co-morbidities	*2*	*2%*
Specific psychiatric co-morbidities NB % with respect to *N* = 120, but a patient can be in more than one class	Psychosis	40	33%
	Autism	13	11%
	Personality disorders	11	9%
	Anxiety	3	3%
	Obsessive–compulsive	2	2%
	Mood swings	2	2%
	Substance abuse	1	1%
Level of medication (BDZ, AP)	Missing data	4	3%

Table 1. *(Continued)*

Variables		*N*	%
	None	32	27%
	1	50	42%
	>1	34	28%
Psychopharmacological class of medication	Benzodiazepines	62	52%
NB % with respect to *N* = 120, but a patient can be in more than one class	Antipsychotics	71	59%
	Antidepressant	14	12%
	Anticholinergic	12	10%
	Antiepileptic	64	53%

- Persons lacking capacity with legal guardians: A specific consent form was e-mailed to the legal guardian with an explanation of the study procedures couched in plain language. The PI was available to answer any questions about the study.
- Persons with mild to moderate IDD deemed to have capacity: Informed consent was obtained directly.
- Persons with moderate to profound IDD lacking capacity: Consent was obtained from the caregivers who accepted to respond to the questionnaires.

Sample

The samples were obtained as follows:

- The congregate setting: 135 consent forms were sent to the legal guardians, of whom 32 (24%) refused consent. The clinical team of the present project discussed the cases of the remaining 103 participants, of whom 31 were considered eligible both for cognitive and neuropsychological evaluation and professional caregiver interviews and 72, due to their level of cognitive deficit or sensory impairments, were considered eligible only for professional caregiver interviews.
- The psychiatric residential facility: of 80 persons with various psychiatric diagnoses, 32 were diagnosed with mild IDD and were considered suitable for inclusion in the study. Of these, eight refused consent, three were excluded as being in an acute symptomatic phase, and two withdrew participation after consenting, leaving 19 people to participate in the study.

Of the total number of persons eligible for cognitive/neuropsychological evaluations (50), some withdrew due to clinical reasons (reactions of anxiety, impulsivity, anger, discontinuation of attention and concentration during the tasks, boredom, fatigue). Statistical analyses between diagnostic orientations from the SPAIDD-G (Systematic Psychopathological Assessment for persons with Intellectual and Developmental Disabilities – General screening) and STA-DI (Scala di valutazione dei Tratti Autistici nelle persone con Disabilità Intellettiva) and cognitive/neuropsychological tasks [WAIS-IV; Leiter-3; Trail Making Test (TMT); Stroop test; Tower Of London (TOL)] have been carried out in subsamples of, respectively, 29 (WAIS-IV), 14 (Leiter-3), 25 (TMT), 36 (Stroop test) and 36 (TOL) persons. Withdrawal rates were 14% (WAIS-IV plus Leiter-3), 50% (TMT), 28% (Stroop test) and 28% (TOL).

Inter-rater reliability

Eight raters (one psychiatrist and seven psychologists) were trained in the administration of the tests prior to starting the study, and a process of inter-rater reliability was performed for WAIS-IV (alpha = 1), Leiter-3 (alpha = 0.999), TMT (alpha = 1), Stroop test (alpha = 0.990), TOL (alpha = 0.999), SPAIDD-G (k_m = 0.76) and STA-DI (alpha = 0.997).

Materials

Leiter-3 was used with persons with moderate IDD and non-verbal persons, while WAIS-IV was used with the less impaired. Only the core subtests were

used in order to minimise the burden on the participants.

IQ tests

- WAIS-IV

The Wechsler Adult Intelligence Scale – Fourth Edition (WAIS-IV, 2008; It. ed. 2013) has 15 subtests: 10 core and 5 supplemental. The subtests are grouped in four main indexes, evaluating the following specific cognitive domains: Verbal Comprehension, Visuo-perceptual Reasoning, Working Memory and Processing Speed. The Total IQ score was still calculated (Lang 2022). The factorial structure of the WAIS-IV (in the US and Italian editions) allows the results to be read according to both the four-factor model (Wechsler 2008; Orsini and Pezzuti 2013, 2015) and the five-factor model or CHC model (Weiss *et al.* 2013; Pezzuti *et al.* 2018) for all age groups (16–90 years).

- Leiter-3

The Leiter International Performance Scale – Third Edition (Leiter-3, Roid, Miller, Pomplun, Koch, 2013; It. ed., 2016) mainly evaluates non-verbal intelligence. It can be administered without the use of oral language, including instructions, and requires no verbal responses. It does have verbal subtests, but if these are not used, it only measures non-verbal intelligence. Leiter-3 uses the three-stratum theory, the CHC psychometric model (Schneider and McGrew 2012; Wilhoit 2017). It has been calibrated for the Italian population (583 subjects; Cornoldi *et al.* 2016).

Neuropsychological tests

The three neuropsychological tests (Trail Making Test, Stroop Colour and Word Test, Tower of London) employed in this study were selected for their practicality and their easy-to-follow explanations, which make them ideal for use with persons with cognitive impairments.

- *The Trail Making Test* (TMT) (Reitan 1958; Lezak 1995; It. ed., Giovagnoli *et al.* 1996; Siciliano *et al.* 2019) measures visual-motor tracking and mental flexibility. It is administered in two parts (Part A and B), which involve visual scanning, number recognition, numeric sequencing and motor speed, the ability to manage multiple stimuli and shift attention (Lezak *et al.* 2004). The time difference between Part B and Part A reflects cognitive activity and shifting ability (Corrigan and Hinkeldey 1987).
- The *Stroop Colour and Word Test* (SCWT) (Stroop 1935; It. ed. Brugnolo *et al.* 2015) is based on the Stroop effect (i.e. the delay in reaction time between congruent and incongruent stimuli). The test is used to identify the attentional profile of persons with brain pathologies, exploring different aspects of attention (Brugnolo *et al.* 2015). It requires the participant to name the colour in which the word is printed that may or may not be the colour indicated by the word (i.e. the word green printed in red). Mismatches between the meaning of the word and the printed colour cause longer response times and mistakes. The present version consists of three tables with 100 stimuli each, arranged in 10 rows and 10 columns of (1) colour words printed in black ink, (2) coloured squares and (3) colour words printed with incongruent ink (Brugnolo *et al.* 2015).
- *Tower of London* (TOL) (Shallice 1982) was developed to investigate problem-solving abilities and standardised for the Italian population in 2017 (Boccia *et al.* 2017). The Simplified London Tower Test (SLTT; Allamanno *et al.* 1987) is designed for people with dementia (Spinnler 1985) and tests executive functions and problem-solving skills. The SLTT requires the problem solver to achieve the goal in the fewest moves possible by breaking it down into subgoals (Newell and Simon 1972). The task measures planning abilities with minimal verbal and perceptive load. Shallice (1982) claimed that this test can investigate the integrity of the SAS (Supervisory Attentional System) (Norman and Shallice 1980), which foresees the consequences of actions taken and assesses their adequacy for the purpose (Allamanno *et al.* 1987).

Tests of psychopathology

- *SPAIDD-G* (Systematic Psychopathological Assessment for persons with Intellectual and

Developmental Disabilities – General screening; Bertelli *et al.* 2012; Bertelli 2019) was developed as a screening instrument to assist clinicians in assessing the potential presence of psychiatric co-morbidities in adults with IDD. It includes 56 items describing the most frequent symptoms of common diagnostic categories (es. ICD-10 or DSM-5), adapted to how those symptoms appear in persons suffering from IDD. While it may be self-administered, it is usually done through a 'proxy' (a family member or a professional caregiver) who is in a position to observe eventual changes in the person's behaviour. The proxy is instructed to mark every item 'present' or 'absent' (dichotomous score). Although it is not devised to offer a complete diagnosis, SPAIDD-G is a sensitive and specific screening tool that can provide diagnostic orientation to be integrated with clinical observation.

Five psychopathology indicators have been created to group the 18 SPAIDD-G diagnostic orientations. The first four indicators have been derived from the Hi-TOP model (Kotov *et al.* 2017), and a fifth has been added for 'autism'. A person can have more than one indicator.

- *Somatoform disorders*, which include somatic symptoms disorders and side effects of drugs.
- *Internalising disorders*, which include anxious personality disorder, anxiety disorders, obsessive compulsive disorder, feeding and eating disorders, major depressive disorder and sexuality disorders.
- *Thought disorders*, which include manic/hypomanic episode (bipolar disorder), psychotic disorders, odd personality disorders, identity disorders, delirium and dementia.
- *Externalising disorders*, which include substance-related disorders, dramatic personality disorders and impulse-control disorder.
- Autism spectrum disorder.
- *STA-DI* (Scala di valutazione dei Tratti Autistici nelle persone con Disabilità Intellettiva; Kraijer *et al.* 2006) is a quick and valid screening tool structured in two parts for the evaluation of autistic traits in persons with IDD aged between 2 and 55 years. The first part is a clinical evaluation with three possible diagnostic orientations: N (the absence of disorder), D (doubt as to the presence of an autism spectrum disorder), PDD (presence of an autism spectrum disorder). The second part consists in evaluation of a 12-item scale: (1) social interaction with adults or caregivers; (2) social interaction with peers; (3) language and word – absent; (4) language and word – present but altered in content; (5) language and word – present but with voice alterations; (6) obsessive interests; (7) motor stereotypes; (8) stereotyped body manipulation; (9) dependence on fixed models; (10) self-injurious behaviour; (11) highly unpredictable behaviour; and (12) unusual, excessive and unmotivated panic and anxiety. Every item requires a dichotomic answer (present/absent) regarding these symptoms/behaviours over the previous 2–6 months. At the end of the report, the results are calculated to obtain four raw scores and the weighted scores that produce the final STA-DI score (0–19) and STADI classification: N: 0–6; D: 7–9; PDD: 10–19.

Statistical analysis

We performed descriptive statistics for socio-demographic and clinical variables, such as gender, facility, education, levels of IDD severity, physical co-morbidities, specific physical co-morbidities, autism co-morbidity, psychiatric co-morbidities from clinical records, specific psychiatric co-morbidity from clinical records, level of medication (BDZ, AP) and psychopharmacological class of medication.

The main goal was to analyse how psychopathological variables from the SPAIDD-G and autism from the STA-DI relate to cognitive (WAIS-IV, Leiter-3) and neuropsychological tests (TMT, STROOP test, TOL).

First of all, we checked the main assumptions for performing parametric analyses: normality and homogeneity of variances. Since they were not fully met and the sample size was small, we performed non-parametric tests to obtain reliable and generalisable results.

We carried out a Mann–Whitney *U* test to explore to what degree cognitive/neuropsychological scores differ if a specified psychopathological diagnosis is present or absent. Each diagnosis was considered

separately in relation with cognitive/neuropsychological test scores as the diagnoses are not mutually exclusive; in fact, almost all the participants in our study (79%) have more than one diagnosis.

The Spearman rank-order test was used to analyse the relationship between the cognitive/neuropsychological scores and years of hospitalisation, education and psychotropic medication load.

Missing data have been removed, and the significance level has been fixed at $P < 0.05$. All statistical analyses have been performed with R, version 4.0.4.

Results

Sample socio-demographic and clinical data

One hundred and seven (89%) participants had physical co-morbidities: 34 (32%) one additional disorder, 30 (28%) two disorders and 25 (23%) three additional disorders ranging from epilepsy through skeletomotor to cardiovascular, ocular, endocrine and other disorders. According to the medical records, 62 (58%) persons had psychiatric co-morbidities: psychoses (40; 33%), autism (13; 11%), mood disorders (2; 2%), anxiety (3; 3%) and obsessive–compulsive disorders (2; 2%). They have been prescribed psychotropic drugs, either benzodiazepines or antipsychotics, that affect cognitive performance [participants with one prescription: 50 (42%), with more than one: 34 (28%)]. The socio-demographic and clinical characteristics are summarised in Table 1.

Psychopathological assessment

Cluster A (Odd) Personality Disorders was the most common diagnostic orientation (68%), with Cluster C (Anxious) Personality Disorders being 49%. Other disorders in the shared neurodevelopmental spectrum: ASD 61% and psychosis 38%. Detailed results from the SPAIDD-G assessment are reported in Table 2.

Among groups of psychopathological indicators (Table 3), thought disorder was prevalent ($N = 91$; 76%), followed by ASD ($N = 73$; 61%) and SSD ($N = 70$; 58%). Externalising disorders ($N = 51$; 43%) were the least common, preceded by internalising disorders ($N = 66$; 55%).

Diagnostic orientations of ASD from STA-DI were found in 35 participants (29%), while 39 (33%) were doubtful and 46 (38%) were negative.

Table 2 Clinical diagnoses from psychological assessment (SPAIDD-G).

Variables		*N*	%
Psychiatric diagnostic orientations: co-morbidities	≤1	26	22%
	2	20	16%
	≥3	74	62%
Psychiatric diagnostic orientations N.B. % with respect to N=120, but a patient can be in more than one class	Nutrition/feeding	16	13%
	Psychotic	46	38%
	Mood-depression	46	38%
	Mood-mania	36	30%
	Anxiety	43	36%
	Side effects of drug use	60	50%
	Delirium	57	48%
	Dementia	52	43%
	Substance-related	39	33%
	Odd personality	81	68%
	Dramatic personality	39	33%
	Anxious personality	59	49%
	Impulse control	47	39%
	Autism spectrum	74	61%
	Dissociative identity	46	38%
	Somatic symptoms	36	30%
	Sexuality	9	8%
	Obsessive compulsive	45	8%

Table 3 Psychopathology indicators.

Variables		*N*	%
Psychopathological disorder from SPAIDD-G N.B. % with respect to N=120, but a patient can be in more than one class	Somatoform	70	58%
	Internalising	66	55%
	Thought	91	76%
	Externalising	51	43%
	Neurodevelopmental	73	61%
ASD diagnosis from STA-DI	Autism	35	29%
	Doubtful autism	39	33%
	No autism	46	38%

Autism diagnosis

Three different prevalence data regarding autism emerged from the present sample, depending on the method of assessment used:

- The clinical records showed 13 persons out of 120 (11%) to have been diagnosed with autism.
- The SPAIDD-G screening evaluation showed 73 persons out of 120 (61%) to have a diagnostic orientation to autism.
- The STA-DI screening evaluation showed 35 persons (29%) with a diagnostic orientation to autism. We used the STA-DI autism evaluation as the indicator of autism (Table 3), as it is more sensitive and specific to autism than SPAIDD-G.

Cognitive and neuropsychological assessment (WAIS-IV, TMT, Stroop, TOL)

Twenty-nine participants with a mean IQ of 49.96 (SD = 9.06) were tested with WAIS-IV, and 14 with a mean non-verbal IQ of 52.36 (SD = 10.46) were tested with Leiter-3. Neuropsychological assessment was performed on a larger number of persons (TMT-A: 32; TMT-B: 25; TMT B-A: 25; Stroop: 36; TOL: 36) (Table 4).

Participants with IDD and ASD had significantly lower scores on the Processing Speed Index (PSI) (estimated difference = 5.99, 95% CI = [0, 21.99]; $P = 0.0332$) and Perceptual Reasoning Index (PRI) (estimated difference = 10, 95% CI = [2, 18.99]; $P = 0.0258$) on the WAIS-IV test, compared to those without ASD. They also had higher time scores on the TMT-A test (estimated difference (s) = −97.99, 95% CI = [−377, −27.99]; $P = 0.0226$). It should be noted that the difference in location is estimated as the score of the non-psychopathological group minus the score of the psychopathological group (Table 5). Moreover, we showed a significant association between years of hospitalisation and TMT B (rho = 0.5; $P = 0.01438$) and TMT B-A (rho = 0.5; $P = 0.009$) time scores: the longer a participant with IDD was hospitalised, the worse was their performance on the TMT.

Table 4 Cognitive and neuropsychological assessments

Cognitive/neuropsychological test scores	*N*	Mean (SD)
WAIS – Verbal Comprehension	29	65.45 (7.6)
WAIS – Perceptual Reasoning	29	59.55 (9.66)
WAIS – Working Memory	29	61.45 (9.35)
WAIS – Processing Speed	28	58.03 (10.69)
WAIS – IQ	29	49.96 (9.06)
Leiter – Non-verbal IQ	14	52.36 (10.46)
Leiter – Memory	14	40.29 (19.36)
Leiter – Processing Speed	14	38.79 (16.56)
TMT – A (s)	32	130.31 (112.35)
TMT – B (s)	25	313.72 (156.64)
TMT – B-A (s)	25	218,48 (123.44)
STROOP – time (s)	36	47.25 (42.07)
TOL – total score	36	13.08 (5.13)

Discussion

General considerations

This study seeks to contribute to the sparse body of psychiatric literature concerning the investigation of cognitive functioning in specific categories of mental disorders in persons with IDD.

A key message from the review of cognitive dysfunction in psychiatry published by Millan *et al.* (2012) was that in addition to cognitive skills frequently being compromised in persons with mental disorders, deficits in specific domains are common to several conditions (Harrison 2019).

Cognition, as Millan *et al.* state, is often neglected in the study of mental disorders because changes in *emotions* are traditionally considered the core of mental illnesses.

Several studies in the past (Mansdorf 1977; McLaren and Richards 1986) aimed at testing and classifying IDD severity levels in 'institutionalised mentally retarded adults'. These authors showed that administering cognitive tests in large IDD/psychiatric institutions could be possible. Also, in the light of their results, the present study aimed at continuing and building on the research-oriented approach in a clinical context with persons affected by severe levels of IDD initiated in 2021 (Santambrogio *et al.* 2021b). To the best of the authors' knowledge, this is the first study conducted with a wide core of cognitive/neuropsychological assessments in IDD/psychiatric institutions, with the objective of improving comprehension of the impact that specific cognitive functioning of persons with IDD may have

Table 5 Estimated differences in location with their confidence intervals and *P*-values of Mann–Whitney *U* test in cognitive/neuropsychological scores in patients with and without psychopathological diagnosis

Cognitive/neuropsychological test scores	Somatoform *N* = 70 (58%)	Internalising *N* = 66 (55%)	Thought *N* = 91 (76%)	Externalising *N* = 51 (43%)	Autism *N* = 35 (29%)
WAIS – Verbal Comprehension	−1.99 [−8; 4]; *P* = 0.5995	4.99 [−3.99; 9]; *P* = 0.2345	−1.99 [−9.99; 3.99]; *P* = 0.5484	−3.99 [−11; 2]; *P* = 0.2423	−5.99 [−18; 10]; *P* = 0.3872
WAIS – Perceptual Reasoning	−1.99 [−8.99; 4.99]; *P* = 0.5395	4.78 [−3.08; 8]; *P* = 0.2339	0.000075 [−7; 6]; *P* = 0.9266	−0.000053 [−6.99; 6]; *P* = 0.9473	10 [2; 18.99]; *P* = 0.0258*
WAIS – Working Memory	1.99 [−6; 8.99]; *P* = 0.6929	−0.000079 [−8.99; 6]; *P* = 0.7913	0.000013 [−8; 8.99]; *P* = 0.9448	−2.99 [−10.99; 5]; *P* = 0.5815	6.44 [−3; 18]; *P* = 0.1828
WAIS – Processing Speed	−0.000061 [−5.99; 5.99]; *P* = 0.9812	0.000038 [−4.99; 5.99]; *P* = 0.7772	0.000032 [−4–99; 7.99]; *P* = 0.7311	−0.000041 [−7.99; 3]; *P* = 0.5366	5.99 [0; 21.99]; *P* = 0.0332*
WAIS – IQ	−1.99 [−8; 4]; *P* = 0.5995	−0.00067 [−6; 5]; *P* = 0.9825	0.4713 [−8; 6.99]; *P* = 0.9084	−2 [−8.99; 3.99]; *P* = 0.3792	4 [−3.99; 14.99]; *P* = 0.1614
Leiter – Non-verbal IQ	9.99 [−5; 27]; *P* = 0.2109	10.74 [−0.99; 26.99]; *P* = 0.0758	−4.99 [−26; 17]; *P* = 0.9009	6.84 [−5; 20]; *P* = 0.2704	5.58 [−16; 26]; *P* = 0.521
Leiter – Non-verbal Memory	11.99 [−9; 50.99]; *P* = 0.2555	6 [−9; 44.99]; *P* = 0.2098	−0.000018 [−24; 39]; *P* = 0.6979	0 [−23.99; 23.99]; *P* = 0.9463	24 [−3.06; 63]; *P* = 0.1531
Leiter – Processing Speed	0.000018 [−0.000046; 43.9]; *P* = 0.4646	0.000082 [−0.000034; 4.39]; *P* = 0.5611	−0.000045 [−5; 44]; *P* = 0.9999	−0.000021 [−5; 4.99]; *P* = 0.7618	5 [−0.000046; 49]; *P* = 0.1983
STROOP – time (s)	−2.25 [−20.25; 11.75]; *P* = 0.7623	−14.99 [−39.99; 1.99]; *P* = 0.07717	−7.49 [−30.25; 7.25]; *P* = 0.3492	0.47 [−14.25; 16.75]; *P* = 0.9619	1.03 [−100.99; 30.25]; *P* = 0.8601
ToL – total	−0.44 [−3.42; 2.5]; *P* = 0.6558	0.81 [−1.24; 4.12]; *P* = 0.4448	−0.15 [−2.67; 3.45]; *P* = 0.8876	−1.23 [−4.57; 0.94]; *P* = 0.1972	2.06 [−2.36; 10.71]; *P* = 0.3367
TMT – A (s)	−7.99 [−59.99; 37]; *P* = 0.806	−16.99 [−90; 30.99]; *P* = 0.5582	−34 [−92; 13.99]; *P* = 0.1665	14.99 [−32; 62.99]; *P* = 0.3878	−97.99 [−377; −27.99]; *P* = 0.0226*
TMT – B (s)	29.79 [−68.99; 198.99]; *P* = 0.7633	55.43 [−53.99; 210]; *P* = 0.2502	−43.9 [−193; 114]; *P* = 0.3501	34.56 [−63; 186]; *P* = 0.579	−184.59 [436.99; 145.99]; *P* = 0.1463
TMT – B A (s)	23.87 [−102; 146.99]; *P* = 0.848	70.12 [−31.99; 173.99]; *P* = 0.218	−38.27 [−159.99; 78]; *P* = 0.4117	32.61 [−59; 155]; *P* = 0.5603	−16.25 [−185; 197]; *P* = 0.8806

Sample size; difference [95%CI] and *P*-value of Mann–Whitney *U* test.
*$P < 0.05$.

on the onset and presentation of specific co-occurrent psychopathologies.

Considerations on the analyses on psychopathological and cognitive domains

There is substantial co-occurrence or clinical overlap between ASD and IDD (Stahlberg *et al.* 2004; Morgan *et al.* 2008), and it has been suggested that the two syndromes are part of the same group of neurodevelopmental disorders (Owen *et al.* 2011; Owen, 2012). Aetiologically, the two syndromes are considered as resulting from the overlapping of genetic and environmental factors that have important effects on brain development; this is proven to cause subsequent impairments in cognitive functioning. Autism is thought to be underpinned by abnormalities in brain circuitry related to social cognition and communication, while in IDD cognitive impairments are related to logical-deductive skills (Totsika *et al.* 2010; Bertelli *et al.* 2015a, 2015b; Keller *et al.* 2021).

Differences in cognitive profiles throughout the same neurodevelopmental continuum have been studied also between autism spectrum disorder and schizophrenia considered to be part of the same neurodevelopmental spectrum (Owen *et al.* 2011; Owen, 2012). Participants with ASD demonstrated significantly better performance than participants with schizophrenia for visuospatial perception, reasoning and problem solving, as well as visual attention and organisation. Participants with ASD also demonstrated better performance than those with schizophrenia for working memory and language and generally comparable performances on processing speed and verbal comprehension (Kuo and Eack 2020).

The intellectual functioning of people with ASD without IDD has not been widely studied, but recent findings highlight the relevance of WISC-V assessment for children with ASD without IDD to individualise intervention, especially remediation (Audras-Torrent *et al.* 2021). Moreover, Cooper *et al.* (2022) stated that anxiety-focused cognition in autism may drive insistence on sameness behaviours and that the relationship between repetitive cognition and behaviour is complex and warrants further investigation.

Previous researchers have stressed the importance of considering the association between ASD and IDD, hypothesising either shared genetic/organic load or different genetic/organic pathways underlying the two disorders (Morton and Frith 1995).

Variability between and within phenotypes is also present when ASD co-occurs with IDD. Of particular interest is the finding that low IQ scores in people affected by ASD are not necessarily associated with impairment of processing speed, but rather with abnormalities in information processing modules, which in turn have pervasive effects on the person's overall functioning (Anderson 2008). While processing speed may underpin individual differences in general intelligence (IQ), fast processing speed is not a sufficient condition for high intelligence in peers without IDD (Anderson 2008). In other words, low general ability at the cognitive level (determined by processing speed) is not part of the causal chain necessary to explain the low IQ scores of children with ASD (Anderson 2008).

In the present study, however, our analyses highlighted that when co-morbid with IDD, ASD has a worsening effect on the cognitive functions on the WAIS-IV Indexes: Processing Speed (PSI), Perceptual Reasoning (PRI). It has been postulated that aetiopathological factors of autism, shared by ASD and IDD, provide pointers as to why the participants with IDD and ASD in the present study had a lower performance on two of the four WAIS-IV Indexes; it is possible that these participants may have suffered from a common genetic/organic disease affecting cognitive functioning, combined with environmental factors that worsened its impact (Cicchetti 1984; Sroufe and Rutter 1984; Cicchetti and Cohen 2006). This hypothesis has been confirmed in that participants with IDD and ASD have higher time scores on the TMT-A, meaning that executive functions are also affected in the co-morbid neurodevelopmental condition.

Our finding that participants with IDD and ASD have lower scores on the Perceptual Reasoning (PRI) and the Processing Speed (PSI) Indexes and higher time scores on the TMT-A is in line with other literature that executive functioning (EF) consists of separated but related cognitive processes, such as inhibition, shifting and updating (Miyake *et al.* 2000; Maricle and Avirett 2012), while other studies (Menghini *et al.* 2010; Costanzo *et al.* 2013) use a

broader definition which includes attention. Persons with IDD are proved to perform statistically significantly lower on the EF tasks than those without IDD (Spaniol and Danielsson 2022).

Long-term institutionalisation may affect performance on the TMT-B and TMT B-A tests, increasing the time necessary to complete the task (rho = 0.48; $P < 0.05$). Indeed, the Spearman correlations on this subject were confirmed by the stepwise model: the longer a participant with IDD was hospitalised, the worse their performance on the TMT (many of our participants with IDD have been living in a congregate setting since their early childhood). Institutionalisation has been descripted in sociology as a deep and important phenomenon influencing the personal history of the subject (Foucault 1972): family bonds become more remote or even absent (in case of abandonment), the institution becomes 'a new family' for the person and professional caregivers their reference point. Their experience of environment, people and care settings are not equivalent to those of people who live on the 'outside'. It would be interesting to explore the impact of institutionalisation on the cognition of people with IDD in greater depth.

Considerations about the descriptive data

The literature shows that severe IDD samples tend to be under-diagnosed for mental disorders due to diagnostic overshadowing. When assessing psychiatric co-morbidities of persons with severe IDD through standardised screening interviews with professional caregivers, it should be noted that is often difficult to define if the symptom/behaviour is related to the mental disorder or to the original IDD condition (e.g. stereotyped and obsessive behaviours). This could explain why the clinical diagnoses on the medical records report less prevalent mental disorders.

In the present study, diagnoses of psychosis on the clinical records and from screening interviews were quite similar (40 vs. 46), but this similarity was not confirmed for ASD, with 13 participants having a clinical ASD diagnosis while the SPAIDD-G revealed 74 cases, very similar to the findings from the STA-DI: 35 ASD plus 39 possible ASD diagnoses (doubt). Misdiagnosis of ASD is much debated in the literature (Lord *et al.* 2018; Keller and Bari 2019). Given the levels of severity in the current sample, it could be assumed that the clinicians attributed the IDD condition with all the manifestations specific to ASD and low-functioning ASD, such as deficits in social–emotional reciprocity, non-verbal communicative behaviours used for social interaction, in developing, maintaining and understanding relationships; a restricted pattern of behaviour and interests; stereotyped motor movements; insistence on sameness; and hyper/hypo-reactivity to sensory inputs (APA 2022).

It must be kept in mind that SPAIDD-G is a screening tool and cannot replace clinical opinion in formulating definitive diagnoses. We selected it for the present study due to its broad sensitivity in identifying different symptom clusters that can be attributed to several diagnostic categories, with the goal of detecting statistical trends with other clinical factors. This clarification should be taken into consideration when interpreting all the epidemiological information from the current study obtained from the SPAIDD-G.

The relatively low prevalence of mental disorders noted on the medical records is not congruent with the descriptive data regarding psychotropic drug prescriptions (52% benzodiazepines, 59% antipsychotics, 53% antiepileptics). It is possible that rather than overmedication, mental ill-health has been under-ascertained and occurrences of challenging behaviour did not reflect the extent to which it may be the manifestation of an underlying mental disorder (Painter *et al.* 2018; Westlake *et al.* 2021). Further research-to-practice work should be done to study each person's psychopathological profile as evaluated with the SPAIDD-G, comparing their medications and assessing whether they are appropriate to the diagnoses (Sheehan and Hassiotis 2017). This could support the clinician in understanding if the present psychotropic medication load is correct and balanced with the correct diagnoses and psychiatric symptomatology underlying challenging behaviours or, if unbalanced and challenging-behaviour-oriented, it should be re-assessed.

These considerations could support a more extensive use of psychopathological screening tools such as SPAIDD-G (general battery) and specific tools such as SPAIDD-ASD (screening for autism), SPAIDD-P (screening for psychoses) and

SPAIDD-M (screening for mood disorders) in clinical routine. Clinical activities have times and urgencies that are often different from those of research, but an effort should be made to assess psychopathology in the severe IDD population with screening questionnaires for the caregivers or family members, given the difficulties in collecting symptoms, thoughts and emotions directly from the persons themselves.

Limitations

The main limitation of this study concerns the generalisability of results, as it is based on a small convenience sample without randomisation, which may have introduced a selection bias. A significant proportion of the sample had severe IDD (59%), and this has affected the ability of the participants to perform cognitive and neuropsychological tests. The recruited sample of persons with mild to moderate IDD, suitable for the overall cognitive/neuropsychological tests, numbered 54 persons. There were a number of withdrawals from the cognitive/neuropsychological tasks, that is, 2 (WAIS-IV), 4 (Leiter-3), 10 (TMT), 8 (Stroop Test) and 8 (TOL), due to clinical reasons (reactions of anxiety, impulsivity, anger, discontinuation of attention and concentration at the tasks, boredom, fatigue). Data available from cognitive/neuropsychological test scores were from 29 (WAIS-IV), 14 (Leiter-3), 31 (TMT), 36 (Stroop test) and 34 (TOL) participants. Regardless of these factors of bias, some statistically significant results have been highlighted. The results obtained can be verified in the future if a larger sample is used, and tested with computerised testing-check, which could be a simpler way of administering cognitive tasks (Sinai *et al.* 2016).

Another considerable limitation is that specific symptoms and psychopathological dimensions have not been considered in depth. Many studies indicate that cognitive dysfunction cuts across disorders and should therefore be considered a transdiagnostic dimension (McTeague *et al.* 2016; East-Richard *et al.* 2020). Future research should take a transdiagnostic approach, focusing on symptom-specific interactions with cognitive domains rather than investigating cognitive functioning within diagnostic categories.

Conclusions

Clinical implications

The present study assessed a sample of persons with IDD for psychopathological and cognitive features, with the objective of exploring a research area that as yet has not been sufficiently studied. Further research is definitely needed to study the impact of ASD and other mental disorders on the cognitive dimensions in persons with IDD.

This study points to the utility of using neuropsychological and IQ tests to improve diagnosis and construct better treatment paths for persons with IDD. Indeed, the person with IDD should not only be the object of an evaluation, but they should also be involved in the process and asked to engage in a process of self-knowledge and understanding. Neuropsychological tools can also provide clinicians with the means of better understanding the people in their care. The study also indicates that reinforcing multidisciplinary teams with clinical psychologists will also improve the care of persons with IDD. The result of this evoluted clinical process is an improvement in the therapeutic alliance.

Furthermore, the transdiagnostic nature of cognitive dysfunction and its heavy impact on daily functioning make it an important target for treatment (Millan *et al.* 2012), so treating cognitive dysfunction in association with psychopathology could lead to better outcomes for patients.

If this format is to have greater statistical power, it must be repeated in both comparable and dissimilar circumstances and also include comparison groups, such as peers without IDD.

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Conflict of interest

No conflicts of interest have been declared.

Data availability statement

Data available on request from the authors. The data that support the findings of this study are available from the corresponding author upon reasonable request.

References

Allamanno N., Della Sala S., Laiacona M., Pasetti C. & Spinnler H. (1987) Problem solving ability in aging and dementia: normative data on a non-verbal test. *Italian Journal of Neurological Sciences* **8**, 111–20.

American Psychiatric Association (2022) *Diagnostic and Statistical Manual of Mental Disorders*, 5th edn, text revision; DSM-5-TR. American Psychiatric Association, Washington.

Anderson M. (2008) What can autism and dyslexia tell us about intelligence? *Quarterly Journal of Experimental Psychology* **61**, 116–28.

Audras-Torrent L., Miniarikova E., Couty F., Dellapiazza F., Berard M., Michelon C. *et al.* (2021) WISC-V profiles and their correlates in children with autism spectrum disorder without intellectual developmental disorder: report from the ELENA cohort. *Autism Research* **14**, 997–1006.

Bertelli M. O. (2019) *SPAIDD-G Systematic Psychopathological Assessment for Persons with Intellectual and Developmental Disabilities – General Screening. Manuale*. Giunti Psychometrics, Firenze.

Bertelli M. O., Cooper S. A. & Salvador-Carulla L. (2018) Intelligence and specific cognitive functions in intellectual disability: implications for assessment and classification. *Current Opinion in Psychiatry* **31**, 88–95.

Bertelli M. O., Merli M. P., Bradley E., Keller R., Varrucciu N., Del Furia C. *et al.* (2015a) The diagnostic boundary between autism spectrum disorder, intellectual developmental disorder and schizophrenia spectrum disorders. *Adv Mental Health Intell Disabil* **9**, 243–64.

Bertelli M. O., Rossi M., Scuticchio D. & Bianco A. (2015b) Diagnosing psychiatric disorders in people with intellectual disabilities: issues and achievements. *Advances in Mental Health and Intellectual Disabilities* **9**, 230–42.

Bertelli M. O., Scuticchio D., Ferrandi A., Lassi S., Mango F., Ciavatta C. *et al.* (2012) Reliability and validity of the SPAID-G checklist for detecting psychiatric disorders in adults with intellectual disability. *Research in Developmental Disabilities* **33**, 382–90.

Boccia M., Marin D., D'Antuono G., Ciurli P., Incoccia C., Antonucci G. *et al.* (2017) The Tower of London (ToL) in Italy: standardization of the ToL test in an Italian population. *Neurol Science* **38**, 1263–70.

Brugnolo A., de Carli F., Accardo J., Amore M., Bosia L. E., Bruzzaniti C. *et al.* (2015) An updated Italian normative dataset for the Stroop color word test (SCWT). *Neurological Sciences* **37**, 365–72.

Cicchetti D. (1984) The emergence of developmental psychopathology. *Child Development* **55**, 1–7.

Cicchetti D. & Cohen D. J. (eds) (2006) *Developmental Psychopathology, Volume 1: Theory and Method*, vol. **1**. John Wiley & Sons.

Cooper K., Russell A., Calley S., Chen H., Kramer J. & Verplanken B. (2022) Cognitive processes in autism: repetitive thinking in autistic versus non-autistic adults. *Autism* **26**, 849–58.

Cooper S. A., Smiley E., Morrison J., Williamson A. & Allan L. (2007) Mental ill-health in adults with intellectual disabilities: prevalence and associated factors. *The British Journal of Psychiatry* **190**, 27–35.

Cornoldi C., Giofrè D. & Belacchi C. (2016) *Leiter-3 Leiter International Performance Scale*, Third edn. Standardizzazione italiana, Giunti Psychometrics, Firenze.

Corrigan J. D. & Hinkeldey N. S. (1987) Relationships between parts A and B of the trail making test. *Journal of Clinical Psychology* **43**, 402–9.

Costanzo F., Varuzza C., Menghini D., Addona F., Gianesini T. & Vicari S. (2013) Executive functions in intellectual disabilities: a comparison between Williams syndrome and Down syndrome. *Research in Developmental Disabilities* **34**, 1770–80.

Day K. & Dosen A. (2002) Recent developments in mental health and mental retardation. *Advances in psychiatry*, 91–7.

Demily C., Rossi M., Schneider M., Edery P., Leleu A., d'Amato T. *et al.* (2014) Neurocognitive and psychiatric management of the 22q11. 2 deletion syndrome. *L'encephale* **41**, 266–73.

East-Richard C., R-Mercier A., Nadeau D. & Cellard C. (2020) Transdiagnostic neurocognitive deficits in psychiatry: a review of meta-analyses. *Canadian Psychology/Psychologie Canadienne* **61**, 190–214.

Foucault M. (1972) *Histoire de la folie à l'âge classique*. Gallimard, Paris, France.

Friedman L., Glover G. H. & Fbirn Consortium (2006) Reducing interscanner variability of activation in a multicenter fMRI study: controlling for signal-to-fluctuation-noise-ratio (SFNR) differences. *NeuroImage* **33**, 471–81.

Giovagnoli A. R., Del Pesce M., Mascheroni S., Simoncelli M., Laiacona M. & Capitani E. (1996) Trail making test: normative values from 287 normal adult controls. *The Italian Journal of Neurological Sciences* **17**, 305–9.

Gothelf D., Schaer M. & Eliez S. (2008) Genes, brain development and psychiatric phenotypes in velo-cardio-facial syndrome. *Developmental Disabilities Research Reviews* **14**, 59–68.

Harrison J. E. (2019) Screening and measurement of cognitive impairment in psychiatry. *CNS Spectrums* **24**, 144–53.

Johnson W., Jung R., Colom R. & Haier R. (2008) Cognitive abilities independent of IQ correlate with regional brain structure. *Intelligence* **36**, 18–28.

Jolliffe T. & Baron-Cohen S. (1997) Are people with autism and Asperger syndrome faster than normal on the embedded figures test? *Journal of Child Psychology and Psychiatry* **38**, 527–34.

Keller R. & Bari S. (2019) Psychosis and ASD. In: *Psychopathology in Adolescents and Adults with Autism Spectrum Disorders* (ed. R. Keller), pp. 51–65. Springer, Cham.

Keller R., Costa T., Imperiale D., Bianco A., Rondini E., Hassiotis A. *et al.* (2021) Stereotypies in the autism spectrum disorder: can we rely on an ethological model? *Brain Sciences* **11**, 762.

Kotov R., Krueger R. F., Watson D., Achenbach T. M., Althoff R. R., Bagby R. M. *et al.* (2017) The hierarchical taxonomy of psychopathology (HiTOP): a dimensional alternative to traditional nosologies. *Journal of Abnormal Psychology* **126**, 454–77.

Kraijer D. W., Lassi S. & la Malfa G. (2006) *STA-DI: Scala di valutazione dei tratti autistici nelle persone con disabilità intellettiva*. Erickson.

Kuo S. S. & Eack S. M. (2020) Meta-analysis of cognitive performance in neurodevelopmental disorders during adulthood: Comparisons between autism spectrum disorder and schizophrenia on the Wechsler Adult Intelligence Scales. *Frontiers in Psychiatry* **11**, 187.

Lang M. (ed.) (2022) *I test che lo psicologo deve conoscere*. Cortina Editore, Milano.

Lemay K. R., Kogan C. S., Rebello T. J., Keeley J. W., Bhargava R., Sharan P. *et al.* (2022) An international field study of the ICD-11 behavioral indicators for disorders of intellectual development. *Journal of Intellectual Disability Research* **66**, 376–91.

Lezak M. D. (1995) Executive functions and motor performance. *Neuropsychological Assessment*, 650–85.

Lezak M. D., Howieson D. B., Loring D. W. & Fischer J. S. (2004) *Neuropsychological Assessment*. Oxford University Press, New York, New York (NY).

Lord C., Elsabbagh M., Baird G. & Veenstra-Vanderweele J. (2018) Autism spectrum disorder. *The Lancet* **392**, 508–20.

Luckasson R., Borthwick-Duffy S., Buntinx W. H., Coulter D. L., Craig E. M. P., Reeve A. *et al.* (2002) *Mental Retardation: Definition, Classification, and Systems of Supports*. American Association on Mental Retardation.

Mansdorf I. J. (1977) Learning concepts through modeling: using different instructional procedures with institutionalized mentally retarded adults. *American Journal of Mental Deficiency* **82**, 287–91.

Maricle D. E. & Avirett E. (2012) The role of cognitive and intelligence tests in the assessment of executive functions. In: *Contemporary Intellectual assessment: Theories, Tests, and Issues* (eds D. P. Flanagan & P. L. Harrison), pp. 820–38. The Guilford Press.

Mazza M. G., Rossetti A., Crespi G. & Clerici M. (2019) Prevalence of co-occurring psychiatric disorders in adults and adolescents with intellectual disability: A systematic review and meta-analysis. *Journal of Applied Research in Intellectual Disabilities* **33**, 126–38.

McLaren K. P. & Richards H. C. (1986) Adaptive behavior scale cognitive triad: discrimination and classification of institutionalized mentally retarded adults. *American Journal of Mental Deficiency* **21**, 304–7.

McTeague L. M., Goodkind M. S. & Etkin A. (2016) Transdiagnostic impairment of cognitive control in mental illness. *Journal of Psychiatric Research* **83**, 37–46.

Menghini D., Finzi A., Benassi M., Bolzani R., Facoetti A., Giovagnoli S. *et al.* (2010) Different underlying neurocognitive deficits in developmental dyslexia: a comparative study. *Neuropsychologia* **48**, 863–72.

Millan M. J., Agid Y., Brüne M., Bullmore E. T., Carter C. S., Clayton N. S. *et al.* (2012) Cognitive dysfunction in psychiatric disorders: characteristics, causes and the quest for improved therapy. *Nature Reviews Drug Discovery* **11**, 141–68.

Miyake A., Friedman N. P., Emerson M. J., Witzki A. H., Howerter A. & Wager T. D. (2000) The unity and diversity of executive functions and their contributions to complex "frontal lobe" tasks: a latent variable analysis. *Cognitive Psychology* **41**, 49–100.

Morgan V. A., Leonard H., Bourke J. & Jablensky A. (2008) Intellectual disability co-occurring with schizophrenia and other psychiatric illness: population-based study. *The British Journal of Psychiatry* **193**, 364–72.

Morton J. & Frith U. (1995) Causal modeling: a structural approach to developmental psychopathology. In: *Developmental Psychopathology, Vol. 1. Theory and Methods* (eds D. Cicchetti & D. J. Cohen), pp. 357–90. John Wiley & Sons.

Naglieri J. A. & Das J. P. (2002) Practical implications of general intelligence and PASS cognitive processes. In: *The General Factor of Intelligence: How General Is It*, pp. 855–84. Psychology Press.

Newell A. & Simon H. A. (1972) *Human problem-solving*. Prentice-Hall, Englewood Cliffs, New Jersey (NJ).

Norman, D. A., & Shallice, T. (1980). Attention to action: willed and automatic control of behavior. Technical Report No. 8006.

Orsini A. & Pezzuti L. (2013) *WAIS-IV. Contributo alla taratura italiana (16–69 anni)*. Giunti OS, Firenze.

Orsini A. & Pezzuti L. (2015) *WAIS-IV. Contributo alla taratura italiana (70–90 anni)*. *In WAIS-IV. Contributo alla*

taratura italiana (70–90 anni), vol. **1**, pp. 1–70. Giunti OS, Firenze.

Owen M. J. (2012) Intellectual disability and major psychiatric disorders: a continuum of neurodevelopmental causality. *The British Journal of Psychiatry* **200**, 268–9.

Owen M. J., O'Donovan M. C., Thapar A. & Craddock N. (2011) Neurodevelopmental hypothesis of schizophrenia. *The British Journal of Psychiatry* **198**, 173–5.

Painter J., Hastings R. P., Ingham B., Trevithick L. & Roy A. (2018) Associations between mental health problems and challenging behavior in adults with intellectual disabilities: a test of the behavioral equivalents hypothesis. *Journal of Mental Health Research in Intellectual Disability* **11**, 1–16.

Pascual-Leone A., Bartres-Faz D. & Keenan J. P. (1999) Transcranial magnetic stimulation: studying the brain--behaviour relationship by induction of 'virtual lesions'. *Philosophical Transactions of the Royal Society of London. Series B: Biological Sciences* **354**, 1229–38.

Pezzuti L., Lang M., Rossetti S. & Michelotti C. (2018) CHC model according to Weiss. *Journal of Individual Differences* **39**, 53–9.

Piva Merli, M. (2015). Advances in the psychiatric diagnosis for intellectual and relational developmental disorders. Tesi di Dottorato. Università degli Studi di Firenze.

Reitan R. M. (1958) Validity of the Trail Making Test as an indicator of organic brain damage. *Perceptual and Motor Skills* **8**, 271–6.

Santambrogio J., Colmegna F., Di Brita C., Merra A., Bertelli M. O. & Clerici M. (2020) Psychiatric and psychological support for an adolescent woman with chromosome 22 deletion syndrome and intellectual disability: a good outcome. *Rivista di Psichiatria* **55**, 106–11.

Santambrogio J., Masi G. & Bertelli M. O. (2021a) Attention deficit hyperactivity disorder in adults with intellectual disability: report 230 of the Royal College of Psychiatrists. *Journal of Mental Health Research in Intellectual Disabilities* **14**, 344–8.

Santambrogio J., Russo M., Terrevazzi S., Peschi G., Clerici M. & Bertelli M. O. (2021b) Mitigating the impact of SARS-CoV-2 on residential facilities for persons with intellectual disability and/or autism spectrum disorder: two experiences from the Italian red zone. *Advances in Mental Health and Intellectual Disabilities* **15**, 132–40.

Scheuffgen K., Happeè F., Anderson M. & Frith U. (2000) High "intelligence," low "IQ"? Speed of processing and measured IQ in children with autism. *Development and Psychopathology* **12**, 83–90.

Schneider W. J. & McGrew K. S. (2012) The Cattell-Horn-Carroll model of intelligence. In: *Contemporary Intellectual Assessment. Theories, Tests, and Issues* (eds D. P. Flanagan & P. L. Harrison), 3rd edn, pp. 99–144. Guildford Press, New York.

Shallice T. (1982) Specific impairment of planning. *Philosophical Transactions of the Royal Society of London. B, Biological Sciences* **298**, 199–209.

Sheehan R. & Hassiotis A. (2017) Reduction or discontinuation of antipsychotics for challenging behaviour in adults with intellectual disability: a systematic review. *The Lancet Psychiatry* **4**, 238–56.

Siciliano M., Chiorri C., Battini V., Sant'Elia V., Altieri M., Trojano L. *et al.* (2019) Regression-based normative data and equivalent scores for trail making test (TMT): an updated Italian normative study. *Neurological Sciences* **40**, 469–77.

Sinai A., Hassiotis A., Rantell K. & Strydom A. (2016) Assessing specific cognitive deficits associated with dementia in older adults with Down syndrome: use and validity of the Arizona cognitive test battery (ACTB). *PLoS One* **11**, e0153917.

Spaniol M. & Danielsson H. (2022) A meta-analysis of the executive function components inhibition, shifting, and attention in intellectual disabilities. *Journal of Intellectual Disability Research* **66**, 9–31.

Spinnler H. (1985) *Il decadimento demenziale: inquadramento neurologico e neuropsicologico*. Il Pensiero Scientifico, Rome, Italy.

Sroufe A. L. & Rutter M. (1984) The domain of developmental psychopathology. *Child Development* **55**, 17–29.

Stahlberg O., Soderstrom H., Rastam M. & Gillberg C. (2004) Bipolar disorder, schizophrenia, and other psychotic disorders in adults with childhood onset AD/HD and/or autism spectrum disorders. *Journal of Neural Transmission* **111**, 891–902.

Stroop J. R. (1935) Studies of interference in serial verbal reactions. *Journal of Experimental Psychology* **18**, 643–62.

Totsika V., Felce D., Kerr M. & Hastings R. P. (2010) Behavior problems, psychiatric symptoms, and quality of life for older adults with intellectual disability with and without autism. *Journal of Autism and Developmental Disorders* **40**, 1171–8.

Trivedi J. K. (2006) Cognitive deficits in psychiatric disorders. *Indian Journal of Psychiatry* **48**, 10–20.

von Elm E., Altman D. G., Egger M., Pocock S. J., Gotzsche P. C., Vandenbroucke J. P. & STROBE Initiative. (2008) The strengthening the reporting of observational studies in epidemiology (STROBE) statement: guidelines for reporting observational studies. *Journal of Clinical Epidemiology* **61**, 344–9.

Wechsler D. (2008) *WAIS-IV. Wechsler Adult Intelligence Scale*, Fourth edn. Ed it. Giunti O.S., Firenze 2013.

Weiss L. G., Keith T. Z., Zhu J. & Chen H. (2013) WAIS-IV and clinical validation of the four-and five-factor interpretative approaches. *Journal of Psychoeducational Assessment* **31**, 94–113.

Westlake F., Hassiotis A., Unwin G. & Totsika V. (2021) The role of behaviour problems in screening for mental ill-health in adults with intellectual disability. *The European Journal of Psychiatry* **35**, 122–5.

Wilhoit B. (2017) Best practices in cross-battery assessment of nonverbal cognitive ability. In: *Handbook of Nonverbal Assessment* (ed. R. S. McCallum), 2nd edn, pp. 59–76. Springer International Publishing, Cham, Switzerland.

Willner P., Bailey R., Parry R. & Dymond S. (2010) Evaluation of executive functioning in people with intellectual disabilities. *Journal of Intellectual Disability Research* **54**, 366–79.

Woodcock K., Oliver C. & Humphreys G. (2009a) Associations between repetitive questioning, resistance to change, temper outbursts and anxiety in Prader–Willi and Fragile-X syndromes. *Journal of Intellectual Disability Research* **53**, 265–78.

Woodcock K., Oliver C. & Humphreys G. (2011) The relationship between specific cognitive impairment and behaviour in Prader-Willi syndrome. *Journal of Intellectual Disability Research* **55**, 152–71.

Woodcock K. A., Humphreys G. W., Oliver C. & Hansen P. C. (2010) Neural correlates of task-switching in paternal 15q11-q13 deletion Prader-Willi syndrome. *Brain Research* **1363**, 128–42.

Woodcock K. A., Oliver C. & Humphreys G. W. (2009b) Task-switching deficits and repetitive behaviour in genetic neurodevelopmental disorders: Data from children with Prader–Willi syndrome chromosome 15 q11–q13 deletion and boys with Fragile X syndrome. *Cognitive Neuropsychology* **26**, 172–94.

Zarchi O., Diamond A., Weinberger R., Abbott D., Carmel M., Frisch A. *et al.* (2014) A comparative study of the neuropsychiatric and neurocognitive phenotype in two microdeletion syndromes: velocardiofacial (22q11.2 deletion) and Williams (7q11.23 deletion) syndromes. *European Psychiatry* **29**, 203–10.

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