DIAGNOSIS, CARE AND FOLLOW UP OF DEMENTIA IN PERSONS WITH INTELLECTUAL DISABILITIES AND AUTISM

CONTENTS

1. APPLICATION

This protocol applies to all organizational units of the Disabled Department that are involved in the care of persons with intellectual disabilities (ID) and dementia.

Target groups: medical doctors, coordinators, nurses and educators.

2. OBJECTIVES

The objectives of the present protocol are:

- creating a multi-professional team specialized in the care of aging persons with ID.

- establishing a framework of supports for persons with ID and autism affected by dementia, in keeping with Quality of Life (QOL) principles and models.

3. ABBREVIATIONS AND ACRONYMS

AADS	Assessment For Adults with Developmental Disabilities
AFAST	Alzheimer Functional Assessment Tool
BPSD	Behavioral and Psychological Symptoms of Dementia
DMR	Dementia questionnaire for people with mental retardation
DMS-IV	Diagnostic and Statistical Manual of Mental Disorders, 4th Edition
DS	Down Syndrome
ID	Intellectual disabilities
QOL	Quality of Life
SPAID-G	Strumento Psichiatrico per l'Adulto Intellettivamente Disabile (checklist for detecting psychiatric disorders in adults with intellectual disability)

4. INTRODUCTION

Aging adults with ID are at an increased risk of developing neurodegenerative diseases like dementia. While in individuals with typical development, the prevailing symptoms are cognitive deterioration and attendant performance loss, adults with ID frequently show an overall worsening functioning accompanied by the loss of adaptive skills.

Assessing persons with ID suspected of cognitive decline is extremely complex due to the intertwining of multiple individual variables (biological, psychological or functioning-related) as well as environmental factors that can determine a significant deterioration in the subject's behavior. Such difficulties increase if the basic personal functioning level is very low, as is the case in persons with severe ID.

The assessment and diagnosis of dementia in persons with ID starts off with an evaluation of the subject's highest functioning level, followed by subsequent assessments carried out with simple, validated instruments, used internationally, that can clearly highlight the areas of skill deterioration. The next step is the identification of the subject's needs and supports (through the Support Intensity Scale) that will inform the implementation of the social, environmental, psychoeducative, and medical interventions necessary to maintain the best autonomy and well-being level possible, in line with Shalock's QOL model.

5. TARGET GROUPS

The target groups are represented by persons with DS over 30 years of age as well as persons with ID and autism over 50 years of age.

6. DIAGNOSIS

A number of different pathologies can determine a loss of personal functioning. Differential diagnosis is therefore a priority. The diagnostic process typically involves the identification and possible exclusion of secondary, treatable causes of functional decline (e.g. metabolic dysfunctions, the use of psychotropic medications, thyroid, liver, and kidney pathologies, psychiatric conditions like depression), followed by targeted diagnostic measures, namely:

- Accurate case history: to identify attested dementia risk factors (e.g. brain trauma, depression, use of substances like alcohol, family history, genetic epsilon-4 positivity).
- Blood chemistry tests: complete blood count, esr, electrolytes, azotemia, creatininaemia, glycemia, hyperuricaemia, liver function test, electroforesis, serum proteins, thyroid function tests (TSH, FT4), B12, folic acid, syphilis serology test, HBV and HCV test, urine test; drug dosage.
- Check-up: including neurological screening to exclude sight and hearing impairment.
- Instrumental tests: neuroimaging, EEG, (other tests if necessary).
- Assessment procedures: to identify signs of reduction or loss of previous abilities, even when significantly compromised. In particular:

- orientation (spatial, temporal, personal)
- language
- recognition
- praxias
- balance and walking disorders
- disruption of sleep cycle
- eating disorders
- mood disorders
- changes in interactional patterns
- extinction or onset of conduct disorder

- loss of basic abilities (dining, dressing, toileting and personal care, sphyncter control).

6.1 Assessment tools: DMR, AFAST, SPAID-G, AADS, Vineland Scale.

DMR (Dementia Questionnaire for Mentally Retarded Persons): a screening tool for dementia in persons with ID of all types and levels that investigates key dementia dimensions and symptoms (cognition, BPSD) through observations of the subject's everyday life by direct caregivers with a good familiarity of the subject (limitations: floor effect in persons with severe ID; unsuitable for persons with sensory deficits).

AFAST (Alzheimer Functional Assessment Tool): a tool that ranks the subject's ability to autonomously perform everyday activities. Its items, except "Environmental Awareness", broadly overlap with those contained in the Activities of Daily Living test but allow a much greater accuracy and leveling, hence greater detail when it comes to devising assistance and supports programs, as well as individual procedural memory stimulation programs.

SPAID-G (Psychiatric Instrument for the Intellectually Disabled Adult): a tool that allows the preliminary identification of diagnostic areas with a high psychopathologic relevance. It covers the subject's entire life-span. Its items are designed to obtain data on symptoms through the observation of behaviors, the only data-gathering modality that suits all ID types. These items define the behavioral indicators of all the appearing symptoms, with different aggregation, in the different diagnostic categories contained in the DSM-IV, i.e. eating disorders, psychotic disorders, depressive disorders, anxiety disorders, delirium, dementia, substance-correlated disorders, personality disorders (cluster A [odd], cluster B [dramatic], cluster C [anxious]), impulse-control disorders, dissociative disorders, factitious disorders, sexual and gender identity disorders.

AADS (Assessment for Adults with Developmental Disabilities): a tool that assesses behavioral excesses (11 items) and deficits (17 items) associated with dementia in persons with ID, each item rated on a seven-point Likert scale.

VINELAND SCALE (Assessment of Adaptive Behavior): a tools that measures social competence, self-help skills, and adaptive behavior from infancy to adulthood. Personal and social skills are evaluated in the following areas: communication (listening, speaking, writing), daily living skills (general self-help, eating, dressing), socialization (interpersonal relationships, play and leisure, and coping skills), and motor skills (fine and gross, including locomotion). It consists of a 117-item semi-structured interview with a parent or other primary caregiver. It is used in diagnostic assessment and planning for therapy and/or individualized instruction.

6.2 ASSESSMENT SCHEDULE

Baseline: for **all** residents, to define the best possible functioning (DMR).

Annual assessment: persons with DS over the age of 30 and persons with ID and autism with risk factors (record is kept of all names).

Biennial assessment : for the entire population with ID and autism over the age of 50.

Diagnostic suspicion: see section 6. An intervention is activated (see section 8) when the diagnosis for possible or probable dementia is confirmed.

7. PROJECT MANAGER

The project manager will be chosen among the members of the team that, within the research group on psychogeriatrics, is specialized in the care of persons with ID and autism affected by dementia. The project manager's duties are as follows:

a. Making sure that the present guidelines are closely adhered to.

b. Monitoring the accomplishment of the life project, identifying work schedule and responsibilities.

c. Checking the project's results.

d. Identifying a reference figure in the team who, in turn, will define deadlines and modalities for the initial assessment, the intervention, and follow-up assessments.

8. INTERVENTION

8.1 GOOD PRACTICES FOR THE DEFINITION OF INTERVENTIONS FOR PERSONS WITH DEMENTIA

In what follows, we detail the clinical, relational and organizational guidelines that are to be applied whenever persons with ID and autism diagnosed with dementia are the target of a multidisciplinary intervention. Dementia, associated with a particularly fragile condition like that of ID and autism, makes these individuals extremely vulnerable to the changes occurring both in their environment and in their own bodies. It is thus essential to monitor not only the modifying functioning of the subjects but also to keep under control the environmental and relational variables surrounding them.

The multidisciplinary team is therefore expected to closely observe the subject's overall functioning through detailed and repeat assessments, and also to identify short-term rehabilitative and existential interventions. All interventions will have to address the following aspects:

A. ***Communication.*** Dementia in persons with ID can significantly alter their communicative skills (expressive and receptive). It is thus important to refer to the subject's past abilities and to reassess them periodically. It must be stressed that a form of communication organized around clear exchanges and tailored to the subject's competence (verbal, sign-based, iconic, etc.), allowing for appropriate decoding and recoding intervals, can enhance interpersonal relations and the subject's sense of content understanding as well as self-confidence. The subject's communicative behavior can also change in relation to his/her overall well-being, not only from day to day but also over one single day. For this reason, communicative style and pace must be adjusted to the objective conditions of the subject at each moment in time.

B. ***Relational stability***. Just because the person with dementia can have moments of lesser clear-headedness throughout the day, it is necessary to plan a continuous and well-thought out form of support. Professionals are the subjects who provide the primary support to persons with ID associated with dementia. For this reason, it is essential that their presence

be consistent. This consistency is beneficial to both the disabled person, who will not experience the relational stress determined by the need to get to know and connect with ever-changing staff, and the staff-members themselves, who can use their deep knowledge of the subject to implement the best intervention with minimal effort.

C. ***Environmental stability***. For the reasons discussed above, all living settings need to be stable and recognizable, and all interventions must be tailored to the specific features of such environments.

Life project. The definition of the life project and the intervention program is crucial for the person with ID diagnosed with dementia. This project must be very flexible and precise, that is it should allow periodic reevaluation and a clear detailing of the objectives, type, frequency and duration of the different supports. The aspects to be considered are as follows:

1. ***Preferences assessment***. Information on the subject's preferences and values—obtained directly from the subject or inferred from his/her history—is fundamental to a correct identification of his/her existential objectives and expectations, and hence to an accurate planning of appropriate supports.
2. ***Functioning assessment***. The assessment of the subject's overall functioning highlights the adaptive behavior areas that most need support.
3. ***Definition of the supports program***. Dementia leads to a slow and progressive worsening of the subject's functioning and health. For this reason it is necessary to plan repeat assessments of these two dimensions and to define clinical, personal, and functional objectives tailored to the subject's health conditions (supports program). For each objective, detailed information on the type, frequency and duration of the corresponding support(s) must be provided to the benefit of both the members of staff and the disabled person.
4. ***Objectives operationalization***. In the intervention program, objectives must be clearly operationalized, avoiding any ambiguity about the expected results (accomplishment criteria).
5. ***Objectives ranking***. Since the disabled person's priorities are likely to frequently change over time, the objectives derived from the different assessments need to be updated and ranked continuously, in agreement with the subject and his/her caregivers, with a view to

guaranteeing the best possible level of physical and personal well-being. To this end, a good balance must be obtained between clinical (health-related) and personal (existential) objectives. These, in turn, must be articulated in the 8 QOL domains.

6. ***Project manager.*** The project manager is chosen among the members of the specialized team dealing with dementia in persons with ID. His/her duties are:
    a. Making sure that the present guidelines are closely adhered to.
    b. Monitoring the implementation of the life project, identifying work schedule and responsibilities.
    c. Checking the project's results.
7. ***Six-monthly revision.*** Because of the changing nature of the disabled person's needs and functioning, both the initial assessment and the intervention program must be revised frequently, AT LEAST every six months.
8. **End-of-life care**: Upon diagnosis for dementia, the life expectancy of persons with ID is approximately eight-ten years, especially if associated with comorbidity, as is often the case in these subjects. Over this time span, the needs of personal and medical assistance increase steadily and become massive towards the final stages of the disease. The team in charge of the disabled person together with the latter's caregivers will define the best end-of-life care, also in keeping with the wishes previously expressed by the subject.
9. ***Disabled person and caregivers involvement.*** The life project should be drawn up involving the disabled person and all his/her relational environment (family, community friends, staff).

8.2 WORK TOOLS

The above guidelines will be implemented correctly only if the available work tools and resources are used appropriately. These are:

- Multiprofessional team
- Behavioral methodology
- Evidence-based medical and nursing interventions

8.3 TIMING

- Diagnosis: 60 days (necessary time span to obtain test results for differential diagnosis)

- Support program: 30 days

- Assessment and program revision: every 6 months

9. REVISION

The present document is revised every two years.

10. DISTRIBUTION

These guidelines will be made accessible via intranet (under Internal regulations) and circulated via email by the Hygienic and Sanitary Functions Executive Unit to the following addressees:

- General direction
- Director and MDs of the Disabled Department
- Area reference person and social worker of the Disabled Department

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