Researchers have shown that individuals with mental retardation or with developmental disabilities are at a greater risk of developing health problems and among others dementia than the general population. As the literature points out, dementia is characterized by progressive loss of cognitive functions, until the individual has lost all independency and ability in daily life. It is therefore necessary to carry out a systematic assessment throughout the developmental phases at the beginning of these signs. The purpose of this paper is to present the importance of systematic assessment of early symptoms of unspecified dementias in people with developmental disabilities. The methodology is based on the pre-dementia analysis of two study cases followed by a 2-year period. In conclusion, it can be argued that the diagnosis of dementia in people with developmental disabilities, in the early stages, has become difficult because of the lack of reliable and standardized criteria and diagnostic procedures and difficulties to investigate cognitive decline versus an already vulnerable developmental disability base. Therefore, in people with developmental disabilities, a diagnosis of dementia needs to be done based on changes in mental status from basic functioning. This helps a clinician to determine an accurate diagnosis in later years as hypothetically results from two case studies with later subcortical dementia. However, this endeavour remains to be discussed widely by mental health specialists, public health and cognitive neuroscience in order to determine whether this contribution provided actually has the power of explanation understandable or is understandable by the part of interest.

**Key words:** systematic assessment, early symptoms of unspecified dementias, people with developmental disabilities, case studies, pre-demental analysis

**INTRODUCTION**

*What we call the future is the shadow that our past casts before us.* Marcel Proust (1871-1922).

For most people, the image for people with developmental disabilities (PDD) is shaped by our personal experiences as we may have seen closely or in the community how they function in everyday life. In this regard, according to the DSM-V (2013), the PDD is characterized by constraints on general mental abilities and adaptive functioning that result from the course of a person's developmental life. These limitations are apparent in comparison with other people of the same age, gender and socio-cultural background. The PDD significantly demonstrate low intellectual performance in perceiving and processing information, learning quickly and efficiently, applying their knowledge and skills to developing problems, thinking creatively and responding in a fast and accurate way. Although these signs are reflected in their functioning in everyday life, PDD also present a comorbid state of mental health.

In the last decade, the professional knowledge associated especially with the abovementioned situations has increased significantly as they cause serious obstacles to their social integration. But what can these inferior or
comorbid states exacerbate or alter their normal and healthy functioning in their everyday lives, regardless of the limitation they have?

Moreover, recent years have been noted, a significant increase in the population with intellectual disabilities (World Health Organization, 2011, 2016). Over a billion people, about 15% of the world's population, have some form of disability. People with disabilities have largely been unrecognized as a population for public health attention, but recent efforts have made the poor health of this population visible (Krahnnet et al., 2015). In the last decade, the health status of individuals with disabilities has emerged as an explicit focus of public health attention, with consumers, policymakers, and researchers joining in defining and implementing an agenda in this area. (Wilber et al., 2002). With age development, these individuals are identified with changes that affect their cognitive, physical, emotional and adaptive functioning. (Kapell et al., 1998, Borson, 2010).

In this regard, contemporary literature in the field of medicine, clinical psychology (Guss, et al., 2014) and recently also cognitive neuroscience (Kennedy, 2013) provides advanced explanations based on authentic scientific studies where the core lies in determining the apparent decline in cognitive functions such as concentration, memory, and so on. Among others, researchers have shown that individuals suffering from mental retardation or being with developmental disabilities are at a greater risk of developing dementias than the general population. (Burns, 1992; Zigman, Silverman and Wisniewski, 1995, Cooper, 1997).

As the literature points out, dementia is a syndrome where progressive deterioration is very severe in cognitive abilities and interferes with ordinary social functioning or at work or at their personal level (APA, 1994). It is also characterized by progressive loss of cognitive functions, until the individual has lost all independency and ability in daily life. (Eckerström, 2017). In the revised Diagnostic and Statistical Manual of mental disorders (DSM-V,2013) the term dementia has been replaced with 'major neurocognitive disorder'.

Moreover, according to International Classification of Functioning, Disability and Health (WHO, 2007,2013) it also includes memory loss, personality changes, and lowers self-care skills. Initial symptoms often appear gradually. There may be a minimal memory loss, especially of recent events. The individual may experience difficulty finding the right words to use during casual conversations. Work experience may begin to worsen, and behavioral changes can become clear. As the disease progresses, memory loss becomes even worse emphasized. There may be special problems with language skills. Affected persons may have difficulty naming objects or keeping a logical conversation. They may have difficulty in understanding guidance and be misled about the time of day, where they are and with whom. People may begin to experience loss of self-care skills, including eating and using the toilet. Serious changes in personality can be made clear, and social behavior can be marked by mistrust and illusions. It is therefore necessary to carry out a systematic assessment throughout the developmental phases at the beginning of the emergence of these signs in order to determine a later diagnosis. The aim of this paper is to present the importance of systematic assessment of early symptoms of unspecified dementias in people with developmental disabilities.

**METHODOLOGY**

The methodology is based on the pre-dementia analysis of two case studies.

There is usually a period between appearance of first symptoms and a clinical diagnosis of dementia. Changes may occur even before the first symptoms. Routine clinical practice shows that the cognitive and functional changes of dementia are typically accompanied by changes in behavior and in personality, but these have not become core criteria when they have been considered to lack sufficient diagnostic specificity. (Chertkow,2013).

How can one professional refer to the specific criteria? Kendell's Criteria (1989) for validating clinical syndromes suggests six following steps:

1. Identification and description
2. Demonstration of boundaries or ‘points of rarity’ between related syndromes
3. Concurrent validity
4. Establish a distinct course or outcome
5. Establish a distinct treatment response
6. Association with more fundamental abnormality

Let us stop only at the first **Criterion 1**: Identification and description:

The first and most important question to be asked when assessing the memory impaired person with developmental disability is “Is this dementia?” Conditions mimicking dementia are considered under the term “pseudodementia” and often relate to affective disorders that represent treatable psychiatric pathology, such as anxiety or depression. (Cooper & Greene, 2005). In this regard, one clinician would say that it is difficult to identify the symptoms of dementia when it is the case of comorbid state but one may also say that it is possible. But it may not be the same for all because we may always find problems with criteria because of multiple definitions, heterogenous criteria eg subjective, objective, variations in content and...
amount of detail eg which tests to use for evaluation of cognitive performance. Referring to this, one always one should take into account the state of cognition and functional ability between normal aging, general cognition normal for age normal for age and cognitive complaints. Lastly, the criterion is then based on a pure clinician judgement. In order to make a good clinician judgement, it may be suitable to be based on the first criteria: identification that can be done following a pre-dementia analysis (psychological changes, cognitive changes and physiological changes) of the given clinical case.

Case Study 1

M. M is 17 years old, diagnosed with moderate mental retardation according to ICD-10. Since the age of 15, in addition to the symptoms that come as a cause of mental retardation, has begun to demonstrate significant negative changes that have been exacerbating the last eight months. The signs are like: staying confused at the table, forgetfulness in the application of eating ethics (in the use of knife, spoon, fork etc.), secretly trying to feed herself with her hands and wipe off her lips with a tablecloth, starting with eating in front of others, watching mildly and savagely the others, displaying psychopathic smile, leaving her head completely unwashed when doing bathing, with foaming shampoo in the hair, showing no interest in changing clothes in the morning, forgetting about carrying out personal hygiene. During the activation time at the Center of Occupational Therapy and Socialization, it has manifested mostly apathic behavior, and has also performed the tasks or instructions mechanically by being distracted and standing in a flaccid state. It does not show interest in doing the usual things. Performs mechanically only when asked but forgetting the main details. It does not present any intellectual ability to refuse for any reason or feelings of the mood (eg cleaning the environment, only taking the broom mechanically and moving only to one place). Occasionally she leaves the activation site as frightened, does not recall how many times a day she gets medicines, or confuses them. She is often shaken by talking and opening her eyes hard. She is not helpful and shows no interest in the feelings of others, does not use verbal support for others (such as encouragement, etc.). She displays symptoms of withdrawal, anxiety, mechanical engagement. She displays the task by repeatedly looking for things that come to mind. Displays forgetting signs of forgetting the work file, and then asks if it is possible going to take it. Forgets the instructions but does not require help again, asks the appraiser to do it for her. She is easily dismissed by the task, displaying a state of persistence and persistent confusion. It does not present any intellectual ability that it can perceive the recent changes that have occurred and have affected the interruption with school relations. She just mentions certain episodes that are left in the memory and occasionally obsessively seeks to realize what's left in the mind. She shows other signs like: increased heartbeats in a social or mental situation that require performing complex movements, sharp eyesight, breathing difficulties, skin changes and body temperature and anxiety. Also, she begins to bend her back and lower her head. Changes her voice, begins to speak normally and then the voice gets blocked into her lips and reveals only incomprehensible custom; the word is barely understood due to poor articulation and pronunciation as well.

Case Study 2

E. H is 32 years old, diagnosed with mild mental delay according to ICD-10. Since the age of 30, in addition to the symptoms that come as a cause of mental retardation has begun to demonstrate the apparent negative changes that have been exacerbating the last 5 months. The signs are like eating fast, not chewing food and eating uncritically, confusion in eating ethics, crying sharply, then showing mechanical smiles. She shows signs of hyperactivity and impulsivity during the work process, sweats too much and as a result often changes and significantly the body temperature. During the activation period, has manifested mostly apathic behavior, multiple physical complaints without being aware to show the motive. She also performs duties or instructions mechanically by being distracted and standing in a fuzzy state. In some cases, she leaves the given duty by focusing on things that are not relevant at the moment, forgets and does not turn back to begin again. It is often heard by talking (eg knees or lower leg). She shows symptoms of anxiety and mechanical co-operation. She interrupts the task by looking for things that come to mind for which it has created illusions in her mind. She shakes often talking (eg knees or lower jaw) showing symptoms of anxiety and mechanical co-operation. She interrupts the task by looking for things that come to mind for which it has created illusions in her mind. It shows signs of forgetfulness, such as forgetting the task left out, for example, if asked to get it, feels bad, usually justified, occasionally simply say "I forgot”. She soon forgets the instructions and confuses them too. It is easily distracted from the task by displaying a state of perseverance and constant confusion. She also fails to perceive the recent changes that have taken place that have affected her physical and mental health. It poses problems with receptive and expressive language skills (eg forgets important portions of the sentence when giving or receiving information). She shows other signs such as being aggressive, irritated, increased heart rate, severe breathing difficulties, body temperature changes, frequent urination, anxiety, and vibration.
Pre-dementia analysis

Instruments for determining dementia are often difficult to evaluate when it comes to assessing the diagnosis of individuals with developmental disabilities. (King et al., 2016). Tests are frequently not appropriate for individuals with intellectual disabilities as they often require abilities these individuals may find more difficult due to their pre-existing impairment. However, the two case studies presented have been evaluated over a 2-year period based mainly on observations in psychological, physiological and low-level cognitive impairment.

Psychological changes

Based on the symptoms demonstrated in the two study cases, it appears that the PDD have negative changes in the psychological state. Historically, many professionals believed that persons with mental retardation were incapable of developing emotional problems due to their lack of "the right power of the ego" (Reiss, 1994). From this, it is reflected that the atypical behaviors of the PDD have been simply as a result of the cognitive limitations of the individual. In 1982, the term "diagnostic overshadowing" (Reiss, 1994; Reiss, Levitan, and Szyszko, 1982) emerged, because mental retardation is claimed to "overwhelm" the symptoms of psychological distress.

Referring again to two clinical cases, apart from the developmental delay, they present a comorbid state of mental health which hinder the identification of the main pathology. Consequently, there is evidence from many experts that individuals with developmental delays are suspected to have a wide range of emotional and personality disorders (Davidson et al., 1994; Reiss, 1994). In both case studies there is noted an increase in emotional and behavioral difficulties, although there is little likelihood in the preliminary assessment to recognise them exactly regardless of the consciousness of comorbid psychiatric states.

In addition to the behavioral observations, based on the used neuropsychiatric inventory, in the Case I, there are delusions (some of which show the feeling of fear of abandonment), agitation/aggression (sadness, spatula with certain cases, agitated behavior in some cases), it is noted disfiguring/depressive states in Case I and Case II (often crying, often discouraged), anxiety in both cases (in the Case I, often is shown anxiety, for example during an activity that is being performed, avoid having to go somewhere else, do not know how to complain, is worried/obsessed when separated from loving figures or who are interested in her), while in Case II, it happens to be unconsciously disconnected and involved in other things without connection), the euphoric state is present in both cases (sometimes show abnormal humorous feelings, feel too good or too happy in childish form, laugh inappropriately) in Case I does not like to start a conversation, shows up the emotions, avoids the chores that she has preferred in the past, is largely less spontaneous and less active than usual, is less enthusiastic about things than before), in Case II, it is noted a disinhibited state (acts occasionally impulsively, approaches too much with others (having powers), it is noted noticeably in both cases irritability and emotional lability (the mood changes frequently, which causes delays in performing even a simple assignment, they are shown impatient.

Other aspects that are noticed to the Case I, are abnormal motions (walking without a goal, excessive nervous disturbances, buzzing movements and adverse changes in appetite and eating habits.) In these individuals, the factors that favor the fastest decline include the age and weight of psychotic symptoms (Wilkoszet et al., 2010), worsening in walking (Patti et al., 2010). In addition to anxiety and high levels of mood swings, researchers have noted an increase in the prevalence of age-related psychiatric disorders in mentally retarded persons. Such a condition is dementia.

Physiological changes

Changes in physiological conditions are noted based on the symptoms demonstrated in two case studies such as body temperature changes, eye pupil swelling, foot tremors or gut, etc. Referring to comorbid mental health conditions, researchers have hypothesized that these conditions may result from physiological changes, medical conditions, the use of pharmacological therapy, or changes in lifestyle (Moss et al., 1997).

From this it can be hypothesized that the PDD often experience a need for additional support due to physiological and psychological changes but based on the cases there is no evidence to be directly related to age, especially in Case I. This is likely to occur irrespective of age. What can be supplemented in description of Case II relates to slight eye impairment, decreased muscle tone and flexibility. Although these conditions are common to the PDD and the general population, these individuals are more likely to have a higher incidence of various illnesses.

Low cognitive functioning.

With age the changes come, not only in appearance, but also in certain mental functions or "cognitive functions" as provided in the two mentioned cases. It should be noted that "cognition" (Borson, 2010) refers to a wide range of invisible activities largely carried out by the human brain. Perception, thinking, knowledge, reasoning, remembering, analyzing, planning, attention, birth and synthesis of ideas, creation, judgment, being aware, having knowledge - all this and more are aspects of cognition.
Based on my psycho-clinical experience with the PDD, memory can be influenced by a variety of factors, sometimes because he / she has failed to receive the information correctly or because of the development delay there is difficulty in coding and storing the information for a long time. Since episodic memory deals with particular events and facts, while semantic memory is free from contextual facts, it is often one of the first cognitive functions that can be noticed and that can cause considerable distress to them and caring figures, or others that surround them. The deterioration in memory functioning is characteristic of dementia (Mitrushina et al., 1991) but may also indicate other malfunctions that are always needed in the assessment.

In both case studies, there is a growing difficulty in remembering and needing more time to even remember certain episodes of events. Sometimes, this reduction in speed to remember things and facts related to the simple things they need to perform in their daily lives becomes apparent and can accompany the onset of depression (Thompson, 2010) characteristic in both given cases. So, it is worrying because signs of memory deterioration are noted and continue to be so that it can show us the signs of a pre-dementia phase (we may call it like that), as it is associated with other failures in cognitive function (APA, 1994).

In the Case I, the tone of voice has dropped significantly, speaks with a drowning voice, and in the Case II, is forgotten important parts of the sentence by mentioning the deterioration of the pronunciation of the word as well.

Evaluating all cognitive functions, we also mention the ability to learn as reading or writing: in Case II, there is a difficulty in linking the letters together and forming the sentence as the writing becomes incomprehensible. For example, if we refer to the Mini-Mental State examination according to Foldstein (1975), the given case presents confusion for the month instead of the week or to determine the day of the week.

While instant memory results in having the ability to record the words after being told, but if referred to later memory does not tell it in the order. In the Case II, for example, it fails to copy the displayed geometric figure and indicates a lack of concentration. Naturally, we may wonder why their cognition has changed over time? Hedden & Gabrieli (2004) have evidenced an accurate review of neural changes related to brain aging by explaining two critical systems that serve cognitive processes.

The first component involves changes in the frontostriatal system, mainly related to executive skills and adaptation to a new environment and changes in the physical and mental ones, and in the latter the changes in the temporal media lobe and the two-way strata that link the hippocampus and the associated cortex. Based on this explanation, can we make an assessment from the point of view of "cognitive decline" or "pre-dementia indicators"?

Another explanation comes from the neurobiological perspective (Borson,2010) that implements it as a model of the "deficit" that comes as a cause of aging; namely a progressive decline in nerve tissue based on "toxic shocks" toward delicate macromolecules, oxidative stress, reduced bioremediation mechanisms, and sensitive disease processes that produce impairment in cognitive abilities. But what can we say about case studies: one at age 17 and the other at 32? Such explanations can help to assess the reorientation of local factors that delay or accelerate changes resulting from aging and consider different variations in individuals at the speed at which brain systems grow. It can be as well taken into account the individual differences with the mechanisms responsible for these differences which are not considered most of the time (Wilson et al., 2002).

**SUMMARY**

Based on psychological, physiological, and low functioning of cognitive functions that are systematically evaluated, a comparison can be hypothesized between a decrease in cognitive functions but also as a pre-dementia phase at which one can rely on a later stage on the advancement or severity of the above-mentioned changes. Bear in mind that classical symptoms should be neurologically based and require the presence of memory problems as well as one of the following: aphasia, apathy, agnosis and worries in executive functioning.

Obviously, this problem has dramatic effects on how well a person is able to take care of him / herself. Referring to this, we can argue that in both cases there is a noticeable decline in the ability to remember and execute certain actions.

According to DSM-V, (2013), there are twelve categories of dementia and based on changed conditions in the two given cases it can be hypothesized one that is related to signs of subcortical pre-dementia characterized by primary dysfunction in subcortical areas in the brain. These dementia states lead to motor dysfunction, speech impairment / maintaining a lecture, memory dysfunction, executive disorders and disturbances in mood and personality.

Furthermore, conceptualizing dementia in terms of social disability highlights the way in which symptoms such as memory problems—and the secondary effects of these, such as loss of confidence or negative reactions from others—affect the possibility of engaging in activities and participating in society. (Clare,2017)
Clinical assessment of pre-dementia

Changes in both the psychological and physical condition of the presented case studies are worrying given the frequency of occurrence of those signs:

- In order to evaluate the clinical signs, however, and not to conclude ahead of time with an unambiguous argument, it is necessary to carefully observe the data of the previous mental and physical status and to then make corresponding comparisons.
- It should also not be based solely on memory signs; this should be seen in relation to other functions and how often and how they affect the functioning of the PDD life.
- And in order to do this, all the first basic information combined with behavioral observations (even non-standardized), clinical interviews, neuropsychological instruments, medical assessments, advanced neurodevelopmental literature guidance are indispensable to detect changes only of cognition and memory, but also of non-perceptible changes that affect social and occupational functioning.
- Conducting a continuous assessment that evaluates any possible psychological, physiological and functional change of cognitive function helps the clinician to build a baseline having an accurate information of the case in which should be included the intellectual assessment, the assessment of adaptive and social functioning (which is a major diagnostic feature for dementia), psychopathological evaluation and mental status examination (adapted on the basis of the person's intelligence coefficient) but without ever avoiding behavioral observations. These latter are sensitive instruments at the end of the cognitive spectrum compared to other measuring instruments.
- This systematic assessment can be used to track changes in cognitive abilities and to compare individual performance over time. But it is always necessary to keep in mind that pre-dementia phases can be difficult to identify to these people because immediate cognitive impairment may be indistinguishable at the moment (Shultz et al., 1998).

CONCLUSION

The aim of the research work was to present the importance of systematic assessment of early symptoms of unspecified dementias to people with developmental disabilities:

- Based on the pre-dementia analysis it turns out that since the PDD may not exhibit traditional signs of dementia (e.g., cognitive impairment) or may be difficult to investigate (Menolascino and Potter, 1989), a systematic assessment is needed.
- This is because, for example, the PDD often demonstrate cognitive deficits and certain symptoms as follows: changes in personality, long periods of inactivity, apathy, attention deficit, and loss of speech, lack of orientation, hyperflexibility, stereotypical behavior, neurological abnormal signs at an early age. (Shooshtari et al., 2011)
- Based on the systematic assessment so far of determining a pre-demential condition we can reach hypothetical conclusion that the two cases coincide with the symptoms between the stage of a mild cognitive decline where social carers, mental health professionals and others noticed difficulties in the PDD compared to the previous situation (to find the right word, greater difficulty in performing tasks in social or work environment, forgetfulness of the material one have just read, loss of valuable objects, difficulty planning or being organized) and a stage of a moderate cognitive decline (related to frequent forgetting, impairment of the ability to perform mathematical actions, difficulties in performing complex tasks such as planning, being bored in challenging mental or social situations).
- Moreover, all of these mental health problems are associated with other health problems such as sensory impairments, arthritis, and history of infections, self-injurious behaviors, and depressive states.
- Among other things, importantly, it may be mentioned the emergence of inferential conditions as pre-psychotic distressing behavior.
- In conclusion, dementia affects the PDD in the same way as other persons in the general population. Therefore, it can be reasonably argued that the diagnosis of dementia in the PDD, mainly in the early stages, has become difficult because of the lack of reliable and standardized criteria and diagnostic procedures and difficulties to investigate cognitive decline versus an already damaged base of developmental disabilities (Aylward, Burt, & Thorpe, 1997).

SUGGESTIONS

- In order to carry out a systematic assessment of the early symptoms of unspecified dementias in the PDD, it is necessary to take into account that it requires patience, dedication to follow the case and professionalism in determining relevant changes. Also:
  - Cognitive deficits can not always determine the diagnosis of dementia because these injuries can be present throughout a person's life (Haveman et al., 1994).
  - Thus, at the PDD (Aylward, Burt & Thorpe, 1997), a final diagnosis of dementia should be made based on changes in their baseline status. This helps a clinician to determine an accurate diagnosis in later years as hypothetically results from two case studies with later subcortical dementia.
It is also necessary for a clinical psychologist to evaluate data based on the systematic functioning of multiple cognitive fields not only focusing on memory change and moreover combining the development of a clinical interview, physical examination, neuropsychological test or inventory for determining type of dementia.

Moreover, due to the development delay of these persons in certain areas, the use of self-reporting forms or self-measuring instruments should be avoided.

Therefore, a range of measuring instruments can be adapted or formulated for specific uses for the PDD. This can help establish durable methods to diagnose unspecified dementias to these individuals.

By identifying signs of memory decline and overall cognitive functioning as early as has advantages (Thompson, 2010) including health equation, planning and providing specialized care. It can help to reduce direct and indirect costs regardless of whether the underlying disease (PDD) causes dementia or whether (primary) dementia will cause the deficits observed in PDD.

It is then, possible to profit from rehabilitation-focused service. Rehabilitation involves working with people to achieve the goals that are important to them. It is based on individual formulations and not a one-size-fits-all approach (British Psychological Society, 2016) acknowledging that each individual has a unique set of experiences, values, motivations, strengths, and needs.

From my personal perspective, the focus is that by making a systemic assessment of early symptoms of unspecified dementias enables people with developmental disabilities dementia to participate in everyday life, and in their families and communities, in a way that is meaningful to them. Thus, helps to achieve the aim of rehabilitation based service and pharmacological treatment as a complement of service and as a coherent practical framework for supporting people with dementia and their families or caregivers.

Finally, systematic assessment can be a qualitative indicator as Stokes (1996) states, as recognizing challenging behavior in demented persons can ... represent the communication of a need that is not anchored in time, for example, be painless, to be loved, to be clean, to be sure, to "know".

However, this endeavor remains to be discussed more widely by mental health specialists, public health and cognitive neuroscience in order to determine whether this contribution provided actually has the power of comprehensible explanation or is understandable by the persons of interest. For this reason, logic of productivity takes place everywhere, but as theoretical chemist Roald Hoffmann says, sometimes acceptance is nothing more than an intuition.

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