

Intellectual Disabilities and Dementia

Learning to Recognize and Support
Individuals with Cognitive Decline





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- ▶ Doctorate in Clinical Neuropsychology
- ▶ Post-Doctoral Fellowship in Neuropsychology with Intellectual Disability
- ▶ Published on the topic of dementia and intellectual disability
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Goals of this Presentation

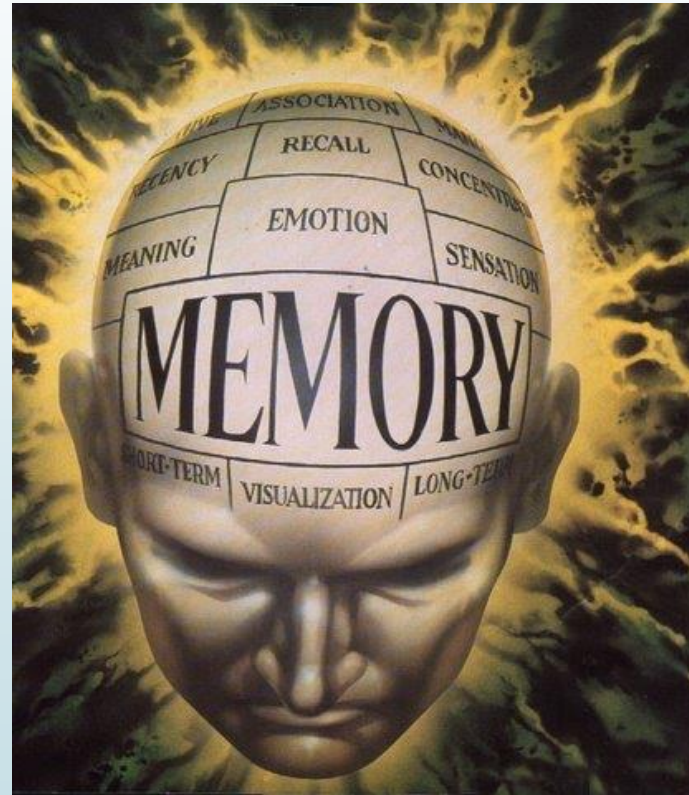
- ▶ 1. Attendees will learn different variations of dementia including course and treatment
- ▶ 2. Attendees will learn how to recognize signs of dementia
- ▶ 3. Attendees will learn how to treat, modify environments of, and preserve abilities for adults with comorbid intellectual disabilities and dementia



Common Misconceptions about Dementia

- ▶ “It’s a symptom of normal aging”
 - ▶ Truth: As the disease gradually worsens, it takes away someone's ability to think, eat, talk, etc.
- ▶ “It is a old person disease”
 - ▶ Truth: About 5% of people with the disease get symptoms in their 30s, 40s, or 50s
- ▶ “It won’t kill you”
 - ▶ Truth: It's the sixth leading cause of death in the U.S. Most people live 8 to 10 years after they're diagnosed.
- ▶ “There are treatments that stop the disease”
 - ▶ Truth: While certain treatments can help against Alzheimer's symptoms, there's no current way to stop or slow the disease itself
- ▶ “It’s caused by aluminum, flu shots, silver fillings, or aspartame”
 - ▶ Truth: There's no scientific evidence to support these claims

But First.....What exactly is
Memory?





Definition



- ▶ “Memory is a cortical network, an array of connective links formed by experience between neurons of the neocortex...the function of cortical neurons in memory derives exclusively from their being part of such networks.” ---Joaquin Fuster
- ▶ “The power or process of reproducing or recalling what has been learned and retained especially through associative mechanisms; A particular act of recall or recollection.” ---Webster’s Dictionary



Stages of Memory

- ▶ Encoding
 - ▶ Registering information after initial exposure
 - ▶ Consolidation-required for transfer to storage; implies learning the information. That “I got it” feelings
- ▶ Storage
 - ▶ Maintaining information over time
 - ▶ “Forgetting”
- ▶ Retrieval
 - ▶ Accessing information
 - ▶ Recall and Recognition

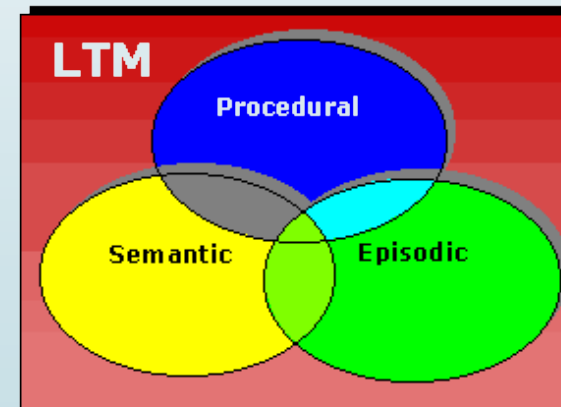
Short Term Memory

- ▶ AKA Immediate Memory
- ▶ 7 +/- bits of information
- ▶ Transferred to long term memory after rehearsed and organized
 - ▶ Rehearsal-repetitive mental process that strengthens duration of memory trace



Long Term Memory

- ▶ Types of Memory
 - ▶ Explicit/Declarative (intentional)
 - ▶ Episodic-memories; purposeful encoding (recent and remote)
 - ▶ Semantic-meaning; general knowledge
 - ▶ Implicit/Non-declarative (incidental)
 - ▶ Sensory memory, classical conditioning
 - ▶ Procedural memory
 - ▶ Vague recollections



What about “Executive Dysfunction?”

- ▶ Initiation
- ▶ Shifting behavior (perseveration/rigidity)
- ▶ Problems stopping (impulse control/disinhibition)
- ▶ Self-awareness (hygiene, overreacting, socially inappropriate)
- ▶ Concrete





Alzheimer's Disease

- ▶ DSM-5 Diagnostic Criteria
 - ▶ Course: gradual and progressive
 - ▶ Clear evidence of decline in memory and learning and at least one other domain of functioning (language, visuospatial functioning, etc.) and adaptive functioning
 - ▶ Evidence of genetic mutation or family history
 - ▶ Genetic testing
 - ▶ Neuropsychological screens
 - ▶ Cerebrospinal fluid (beta amyloid)
 - ▶ Neuroimaging
 - ▶ Neurological examination
 - ▶ Autopsy- only definitive way to determine diagnosis (neurofibrillary tangles)

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Facts about Alzheimer's Disease

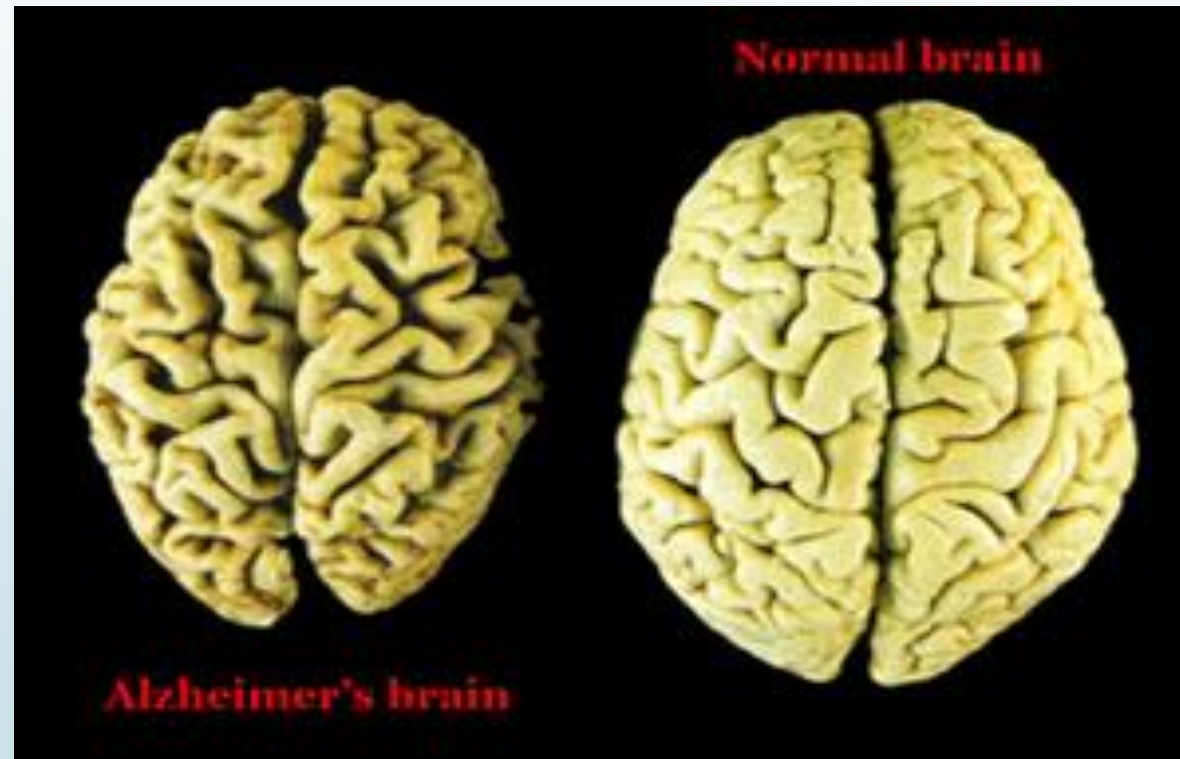
- ▶ 12% of adults age 65+ have Alzheimer's disease (about 5 million). Predicted number by 2025 is 34% of adults 65+ (about 16 million)
- ▶ 1/3 senior citizens die annually from dementia
- ▶ There has been an 86% increase in Alzheimer's deaths since 2000
- ▶ It's the 6th leading cause of death in Louisiana
- ▶ About twice as common in females
- ▶ More prevalent in minority populations



Alzheimer's Disease

- ▶ 60-80% of cases of dementia. $\frac{1}{2}$ of those are pure Alzheimer's while the other half have mixed pathology
- ▶ Common complaints include: difficulty remembering recent conversations, names or events, apathy and depression, impaired communication, disorientation, confusion, poor judgment, behavior change, trouble swallowing and walking

Alzheimer's Disease





Vascular Dementia

- ▶ DSM-5 Criteria:

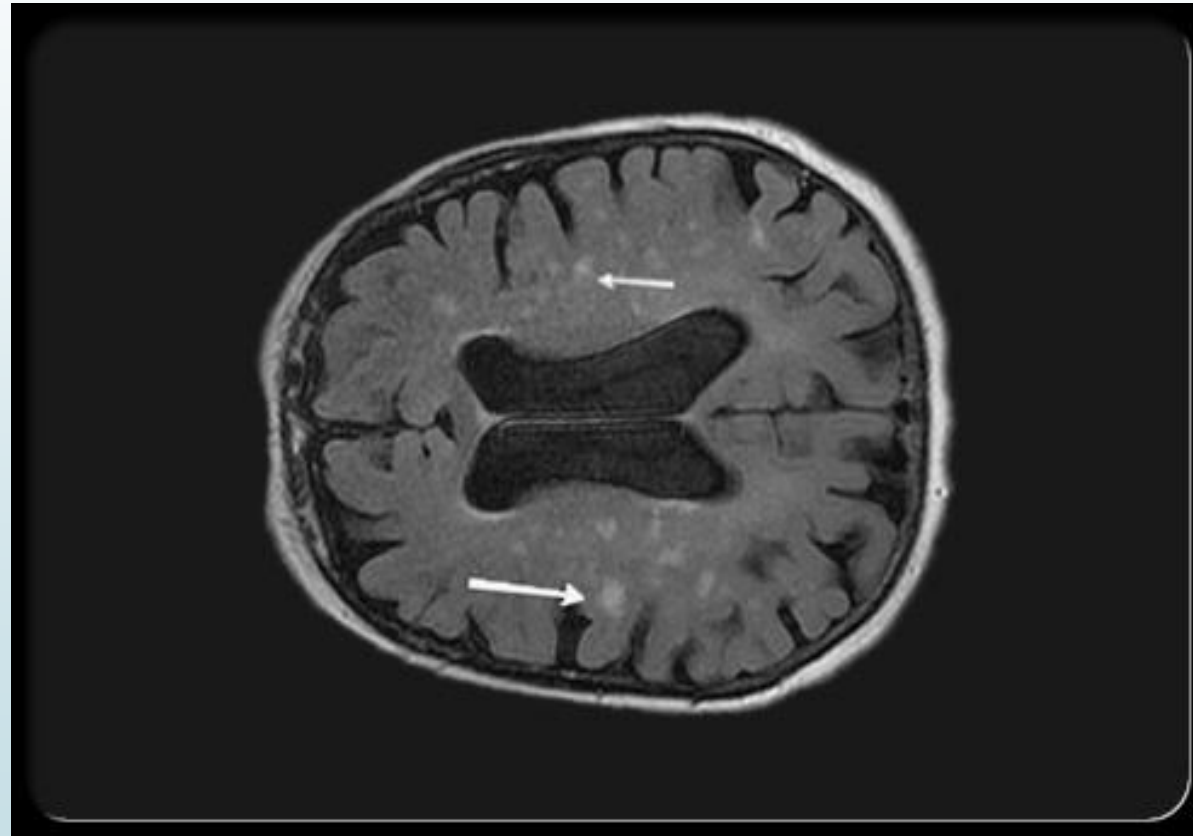
- ▶ Course: Occurs typically following a stroke, TIA or due to vascular changes in the brain over time. Stepwise progression
- ▶ Decline is evident following a cerebrovascular event
- ▶ Evidence in decline in complex attention/processing speed and frontal-executive functioning



Vascular Dementia

- ▶ AKA multi-infarct or post-stroke dementia
- ▶ 10% of cases of dementia but 50% having pathologic evidence of vascular decline (infarcts).
- ▶ Characterized by impaired judgement, poor ability to make decisions, poor planning and organization. Memory decline occurs secondary.
- ▶ Causes: blockage, stroke, brain bleed. Insults to the brain of this nature are cumulative.

Vascular Dementia





Frontotemporal Dementia

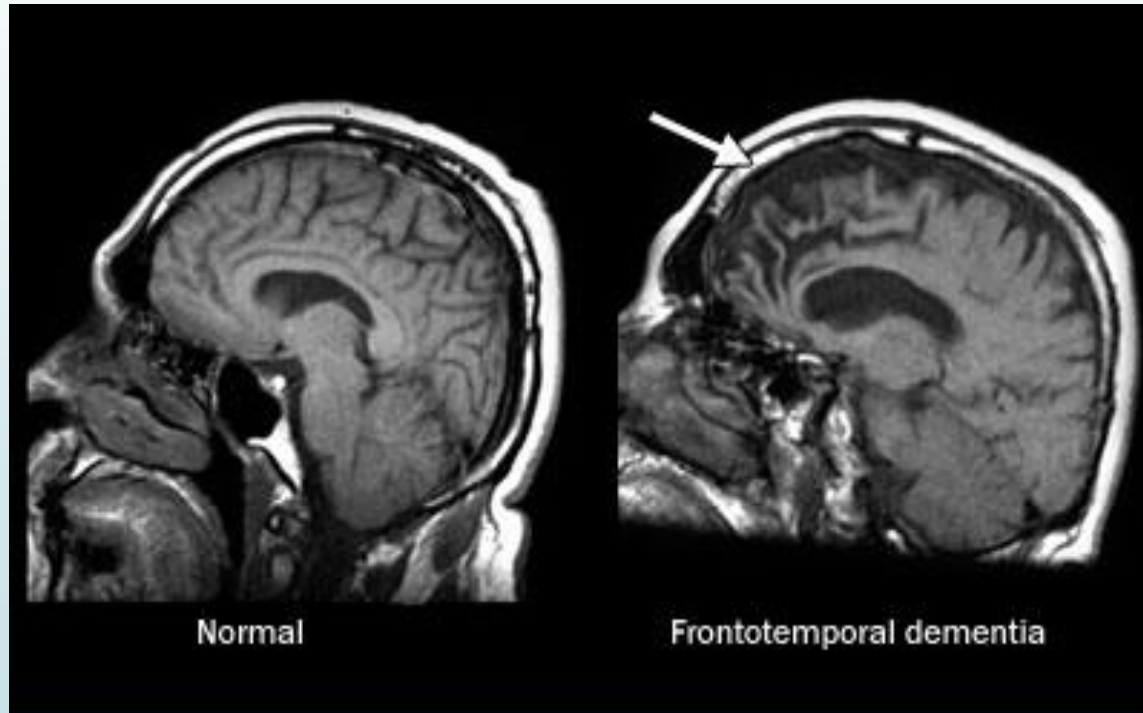
- ▶ DSM-5 Criteria
 - ▶ Course: gradual and progressive
 - ▶ Typically memory is intact but declines later
 - ▶ Behavioral Variant
 - ▶ Disinhibition
 - ▶ Apathy/inertia
 - ▶ Loss of empathy
 - ▶ Perseverative; ritualistic
 - ▶ Hyperorality or dietary changes
 - ▶ Language Variant (Primary Progressive Aphasia)
 - ▶ Prominent decline in language production, word finding, object naming, grammar, or comprehension



Frontotemporal Dementia

- ▶ Typical early symptoms include marked changes in personality and behavior and difficulty with producing or comprehending language. Memory impairment comes later
- ▶ Cause: nerve cells in the front (frontal lobe) and side regions (temporal lobes) of the brain atrophy.

Frontotemporal Dementia



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Lewy Body Dementia

- ▶ DSM-5 Criteria
 - ▶ Course: gradual progression
 - ▶ Core features
 - ▶ Good days and bad days
 - ▶ Visual hallucinations
 - ▶ Parkinsonism
 - ▶ Cognitive decline
 - ▶ Suggestive features
 - ▶ Sleep behavior disorder
 - ▶ Severe neuroleptic sensitivity



Other Types of Dementia

- ▶ Parkinson's dementia
- ▶ Huntington's dementia
- ▶ Dementia due to Traumatic Brain Injury (TBI)
- ▶ Dementia due to substance abuse (Korsakoff's syndrome)
- ▶ Dementia due to Prion disease (e.g. "mad cow" disease)
- ▶ Unspecified Neurocognitive Disorder



Mild Cognitive Impairment

- ▶ Pre-dementia phase
- ▶ Mild changes typically in memory but can be present first in language, executive functioning, or visuospatial abilities
- ▶ Still independent in Activities of Daily Living (ADLs)
- ▶ Typically aware of decline



Dementia in Intellectual Disability

- ▶ Alzheimer's is the most common type of dementia with Lewy Body being the second most common type in ID
- ▶ Approximately 13% of individuals with ID will develop dementia under the age of 60
- ▶ Up to 18% over the age of 60 with ID will develop dementia
- ▶ Almost all individuals with Down's syndrome will develop dementia
- ▶ Once diagnosed with dementia, may live anywhere from 3-10 years on average.

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Determining if it's Dementia

➤ Rule Outs

- Vitamin B12 deficiency
- Vitamin D deficiency
- Hypothyroidism
- Medication changes
- Sleep schedule
- Depression
- Urinary tract infection or other infection causing fever
- Result of a fall
- Sudden or recent changes in schedule

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Determining if it's Dementia

- ▶ Indicators it could be dementia
 - ▶ Slowly progresses or occurs following a neurological incident
 - ▶ Does not improve over time once other variables are accounted for
 - ▶ Sleep-wake schedule changes
 - ▶ Confusion/disorientation that does not change
 - ▶ Wanders and seems to get lost
 - ▶ Major changes in personality
 - ▶ Toileting changes (incontinence)
 - ▶ Decline in daily living skills
 - ▶ Lack of energy
 - ▶ Difficulty ambulating
 - ▶ Being uncooperative



What should you do if you think it's dementia?

- Schedule appointment with Primary Care Physician or if they already have an established neurologist, schedule appointment with them
- Attend appointment with them if possible
- Schedule a neuropsychological assessment



How do we assess dementia with intellectual disability?

► Complications

- Low premorbid functioning makes it difficult to measure decline
- Sometimes difficulty responding on standardized tests
- Normative data mostly doesn't account for ID
- Fallacy that all individuals with ID have dementia



Indirect Assessment

- ▶ **Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID)**
 - ▶ 53 items and requires 15 minutes to administer
 - ▶ Require a reliable informant
 - ▶ Assesses speech, daily living skills, and needed accommodations
 - ▶ Rate each question
 - ▶ Used qualitatively
 - ▶ Pros-takes into account premorbid ability
 - ▶ Con-normed primarily on individuals with Down's syndrome
 - ▶ Good sensitivity and specificity (e.g. rules out those with dementia and identifies those with dementia)
 - ▶ <http://www.birmingham.ac.uk/documents/colleges/psych/ld/lddementiascreeningquestionnaire.pdf>



Indirect Assessment

- ▶ **Dementia Questionnaire for Mentally Retarded Persons (DMR)**
 - ▶ 50 items and 8 subscales
 - ▶ Used for adults with and without Down's syndrome
 - ▶ High degree of false positives (38.5%)
 - ▶ May underdiagnose vascular dementia



Indirect Assessment

- ▶ **Short Informant Questionnaire on Cognitive Decline in the Elderly (IQCODE)**
 - ▶ 16 items; easy to administer and used qualitatively
 - ▶ Shortcomings: requires informant to know individual 10 years prior; does not account for comorbid symptoms; inconsistent findings in research
 - ▶ <http://m.patient.media/pdf/9309.pdf>

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Indirect Assessment

- ▶ **Early Signs of Dementia Checklist (ESDC)**

- ▶ 37 total items

- ▶ Shortcomings: little research on the measure; does not account for premorbid functioning; questions can be misleading

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Direct Assessment

- ▶ **Test for Severe Impairment (TSI)**
 - ▶ 24 items assessing memory, knowledge, and language
 - ▶ Good reliability and validity



Direct Assessment

- ▶ **Severe Impairment Battery (SIB)**

- ▶ Short form and long form
- ▶ Six major subscales: attention, orientation, language, memory, visuo-spatial ability, and construction.
- ▶ Can identify severe dementia
- ▶ Good reliability and validity
- ▶ Developed for individuals without ID



Assessment Take Home notes

- ▶ 1. It's important to have a reliable collateral informant, preferably someone who has known and worked with the individual being assessed for years
- ▶ 2. It's important to get a good assessment of adaptive functioning (social, daily living, communication skills) and compare it to previous estimates
- ▶ 3. It's important to attempt some measure of cognitive abilities (language, memory, executive functioning, etc.)
- ▶ 4. Remember that a diagnosis of dementia often requires both decline in cognition and adaptive abilities.



Treatment Options

- Pharmacology

- There is little evidence that medication is effective in slowing progressive neurocognitive decline in individuals with ID
- Among the common medications, Namenda has been the most efficacious
- Because depression and dementia are often comorbid, treatment of depression has helped with clients who have a pseudodementia presentation.
 - SSRIs-increased energy, interest in activities, independence in eating, positive mood, better sleep, and decreased behavioral outbursts



Treatment Options

- ▶ Behavioral Interventions
 - ▶ Limited due to difficulty learning and retaining newly acquired information
 - ▶ There is some support for differential reinforcement of other behavior involving reinforcing agents contingent on the absence of challenging behaviors
 - ▶ Engage the individual and their family and/or caregivers in advance care planning (and prepare advance directives)
 - ▶ Establish a daily regime that provides for purposeful engagement based on individual needs and preferences, yet is organized so as not to cause anxiety and confusion.
 - ▶ Provide ongoing clinical supports to address behavioral and psychological symptoms associated with dementia.
 - ▶ Small group activities with interactions
 - ▶ Reminiscence
 - ▶ Skill maintenance



Treatment Options

- Environmental Modification
 - Separate spaces for occupational and recreational activities
 - Relaxation areas and activities
 - Use of adequate signage
 - Welcoming wall colors
 - Daylight or soft light
 - Contextual cues indicative of season
 - Orientation board
 - More decline seen in institutional settings rather than personalized
 - Use of a person-centered approach
 - Safety measures



Treatment Options

- Staff Interventions

- Most staff members are unaware of symptoms of dementia and as such, struggle with empathy and understanding of challenging behavior
- When they need increased assistance with ADLs, staff often report they were not prepared and struggled with burnout
- Need to continuously assess staff skill sets and doing continuing education regarding decline and what to expect.
- Shorter shifts for staff working with this population
- Training programs typically include: caring for people with ID, caring for people with dementia, culture competency, palliative care, and nutrition.



Treatment Options

- Pearlin Model

- Primary stressors: changes in cognition and their relationships, behavioral and personality changes, restlessness/wandering, sleep disturbance, incontinence, being uncooperative, and caregiving tasks themselves.
- Secondary stressors: support from family members and other staff, limited time for the caregiver to take a break, poor comprehension of job expectations, and staffing ratio issues.
- Proposes need for adaptive coping mechanisms, social support, and training
- 4 domains of caregiver stress when working with dementia
 - 1. Staff demographics
 - 2. Duration of caregiving
 - 3. Progression of dementia
 - 4. Participation in day services planning

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Effects of Caregiver Stress

- Depression
- Anxiety
- Physical ailments
- Absenteeism
- Burnout
- Turnover



Case Example 1

- “Greg” is a 65-year-old male with Down’s syndrome. Growing up, Greg was always known for his eccentric fashion sense. He would often wear multiple layers of clothing, a ring on every finger, and several hats at once. Greg has always loved to color, go on walks, eat, and interact with staff. Spider man is his absolute favorite character and he collects Spiderman items. Around the age of 60, Greg’s staff starting noticing he was becoming confused. He started walking to his old home (where he hadn’t lived in several years) and asking for staff members who no longer worked at his facility. Greg started losing his interest in fashion and coloring (but not Spiderman!) and was noted to become quickly agitated and aggressive. While Greg had a history of challenging behaviors, they never escalated so quickly. He also started sleeping a lot during the day and staying awake all night. One day, he was observed yelling and swatting at images that were not present and yelling for them to “go away.” He began spending much of his day inconsolably crying. He started having toileting accidents and refusing to wear pants. His gait became unsteady and he sustained two falls with head injury. He began coughing during meals and forgot how to use his utensils properly.



Assessment and Treatment

- ▶ Greg's adaptive behavior scores (measured with the Vineland) showed substantial decline. He was given the TSI and ESDC which also indicated decline.
- ▶ His medical records were examined for possible contributing factors
- ▶ A neurology consult was requested. He was started on medication.
- ▶ A medical psychology consult was requested. He was started on an antidepressant
- ▶ Speech therapy was consulted for a swallow study
- ▶ Physical therapy was consulted for his gait
- ▶ Occupational therapy was consulted for mealtime modification and safety measures for in the bedroom
- ▶ His behavior plan, goals, and training objectives were altered to reflect his neurocognitive decline
- ▶ Movies and music from his childhood and early adult years were played at his home



Outcome

- ▶ Greg's mood changed substantially. He regained elements of his personality, was more alert and engaged, and appeared less confused. He was happier and more aware of his surroundings. He was no longer hallucinating and sleeping better. He became confused in the evenings but the time at which he became confused was later in the day (previously around 2:00pm)
- ▶ Unfortunately, despite efforts from all treating parties, Greg reached a plateau and began to decline again. This time, interventions were unsuccessful. He spent much of his day sleeping. An enteral feeding tube was been placed. He was constantly confused and had to be moved to a medical home. He was no longer able to walk or care for himself. When awake, he was often seen crying and is difficult to console. He has since passed away.

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Case Example 2

- ▶ Lance was a 62-year-old Caucasian male with Down's syndrome. Lance always had a liking for the ladies. He enjoyed walking around, eating, dancing, and listening to music. Although nonverbal, Lance was always good at expressing himself with gestures and vocalizations. Around the age of 60, Lance's gait started to decline and he became incontinent. It was also evident that he was attending to images that were not present. He began engaging in Pica by attempting to eat his clothing. He also began damaging his clothing by pulling at and tearing them. He wanted to chew on everything. He ended needing a wheelchair for ambulation.



Assessment and Treatment

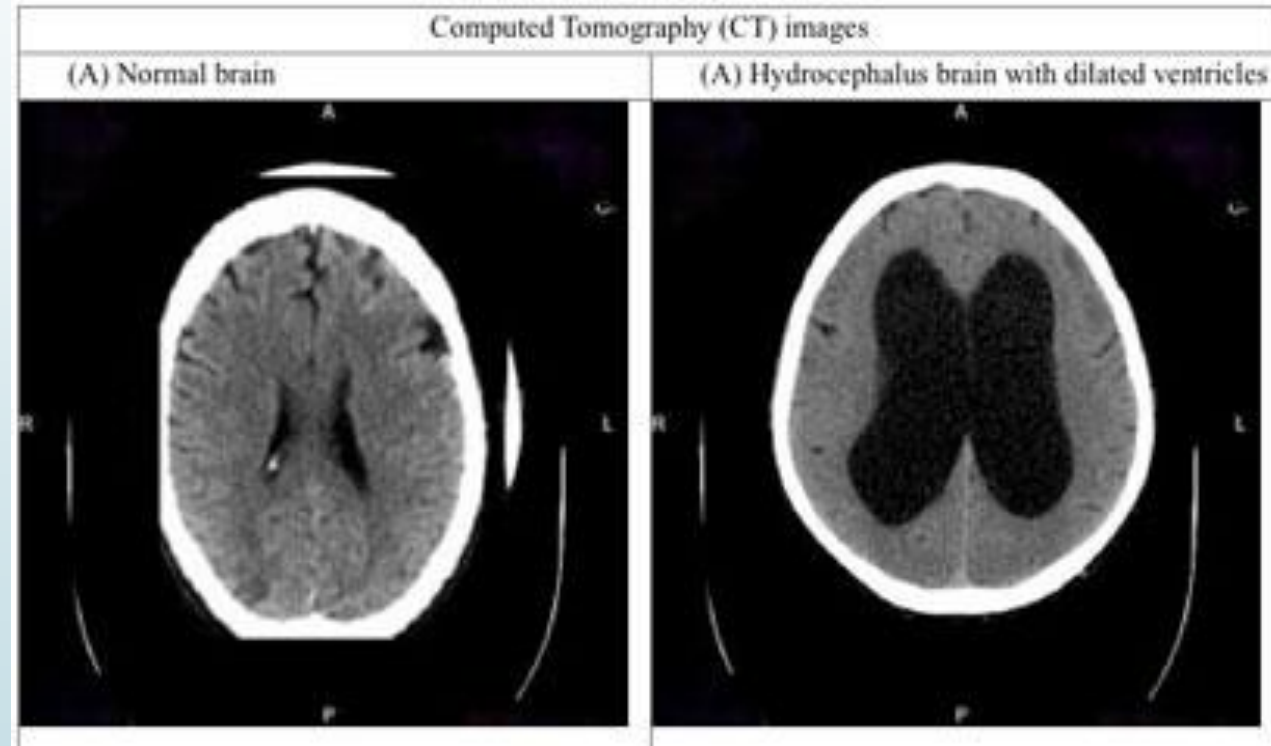
- ▶ A neurological consult was requested. Subsequent neuroimaging revealed severe cortical atrophy and hydrocephalus. The decision was made not to prescribe medication
- ▶ A neuropsychological assessment revealed global impairment as did his adaptive assessment
- ▶ He was treated with psychotropic medication for mood instability, which seemed to work temporarily
- ▶ OT/PT/ST were consulted. He was provided with adaptive equipment to help with meals and safety in the shower and in bed.
- ▶ Psychology implemented overalls so that he could not remove and eat his clothing.

Outcome

- ▶ Despite intervention, Lance continued to decline. He required 1:1 support for safety. He became increasingly irritable until his energy level decreased and he began to spend much of his time sleeping. He has since passed away.



Hydrocephalus



Questions or Comments?

