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Wisdom for Your Life.

Neurodegenerative Dementias and the Multidisciplinary Approach to Patient Care

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Overview

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- Overview of Alzheimer's disease and other age-related dementias
- Diagnostic approach to dementia
- Multidisciplinary approach to dementia care
- The benefits of multidisciplinary care

Causes of Dementia

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- Alzheimer's Disease
- Vascular Dementia
- Dementia with Lewy Bodies
- Frontotemporal Dementia
- Chronic Traumatic Encephalopathy
- Other (Metabolic, Autoimmune, Infectious)

Alzheimer's in Numbers

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ALZHEIMER'S DISEASE IS THE
6TH LEADING CAUSE
OF DEATH IN THE UNITED STATES

5.8 MILLION Americans are living with Alzheimer's

BY 2050, this number is projected to rise to nearly **14 MILLION**

 # NUMBER OF DEATHS FROM ALZHEIMER'S DISEASE (2017)

3,522

4th *highest Alzheimer's death rate in America*

244% *increase in Alzheimer's deaths since 2000*

The only top 10 cause of death that cannot be prevented, effectively treated or cured

Caregiver Burden

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MORE THAN 16 MILLION AMERICANS provide unpaid care for people with Alzheimer's or other dementias

These caregivers provided an estimated **18.5 BILLION HOURS** valued at nearly **$234 BILLION**

- Twice as likely to report financial, emotional and physical difficulties compared to non-AD caregivers
- 30-40% suffer from clinical depression
- Risk of depression is 2x higher

Clinical Presentation: Typical

- Impairment of recent episodic memory is most common early symptom.
- Working memory and semantic memory initially preserved
- Non-amnestic symptoms are frequent and may precede memory deficits (visuospatial, language, apraxia, dysexecutive, behavioral)
- Neuropsychiatric symptoms include apathy, anxiety, irritability and depression
- Hallucinations, delusions and disinhibition occur later, but can also happen sooner in behavioral variant

Clinical Presentation: Atypical

- **Frontal variant**:
  - Early personality change out of proportion to cognitive impairment
  - Irritability, impulsivity and disinhibiton
- **Posterior cortical atrophy**:
  - Visuospatial and visuo-perceptual impairments
  - Bálint's syndrome (simultagnosia, oculomotor apraxia, optic ataxia)
  - Gerstmann's Syndrome (agraphia, acalculia, finger agnosia, left-right disorientation)
  - Deficits in working memory
- **Logopenic variant of primary progressive aphasia**:
  - Confrontation anomia and impaired repetition with preserved grammar and no speech apraxia
- **Corticobasal syndrome**:
  - Apraxia, parkinsonism, visuospatial deficits

Age-related cognitive change

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DECLINE WITH AGE	
FREE RECALL (NO-CUE)	Remembering items on a shopping list
SOURCE OF MEMORY	Recalling where or in what circumstances a fact was learned
PROSPECTIVE MEMORY	Remembering to take a medication before going to bed
PROCESSING SPEED	Time to complete tasks, reaction times
ATTENTION	Divided selective, and sustained attention
EXECUTIVE FUNCTION	Abstraction, mental flexibility, concept formation decline after age 70. Response inhibition.
CONSTRUCTIONAL	Constructional abilities and learning new tasks can decline

Age-related cognitive change

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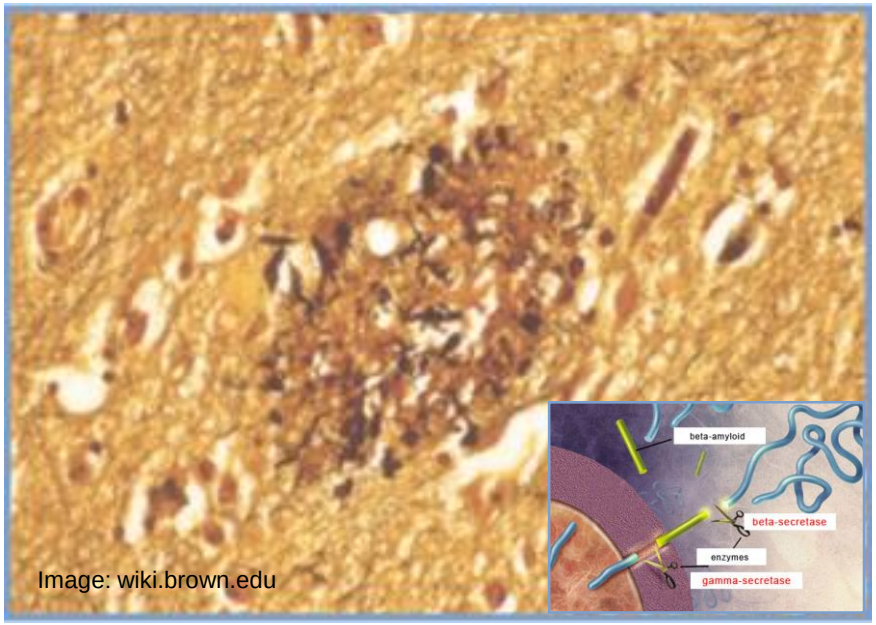
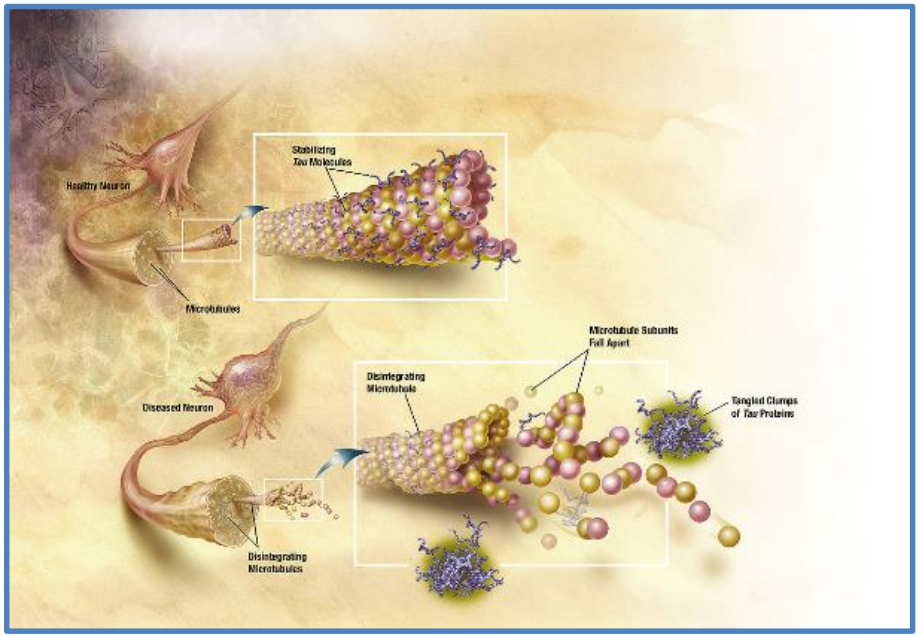
STABLE WITH AGE	
RECOGNITION MEMORY	Retrieving memory when given a cue (e.g. recalling details of a story when asked yes/no questions)
TEMPORAL ORDER	Recalling the sequence of events
PROCEDURAL MEMORY	How to tie a shoe lace, ride a bike
LANGUAGE	Overall intact with aging. Vocabulary may improve. Some decline in confrontational naming and word search. Sporadic word finding difficulty.
VISUOSPATIAL	Navigation, orientation, depth perception tend to remain Intact

Pathology: Amyloid Plaques



Amyloid Plaques

- Amyloid is a naturally occurring protein
- In it's abnormal form, it has tendency to aggregate forming plaques



Tau Tangles

- Normal tau protein plays crucial role in neuronal structure and function
- In AD and several other dementias, Tau changes its configuration, forms tangles, cause cell dysfunction and eventually cell death
-

Pathology: Anatomical Distribution

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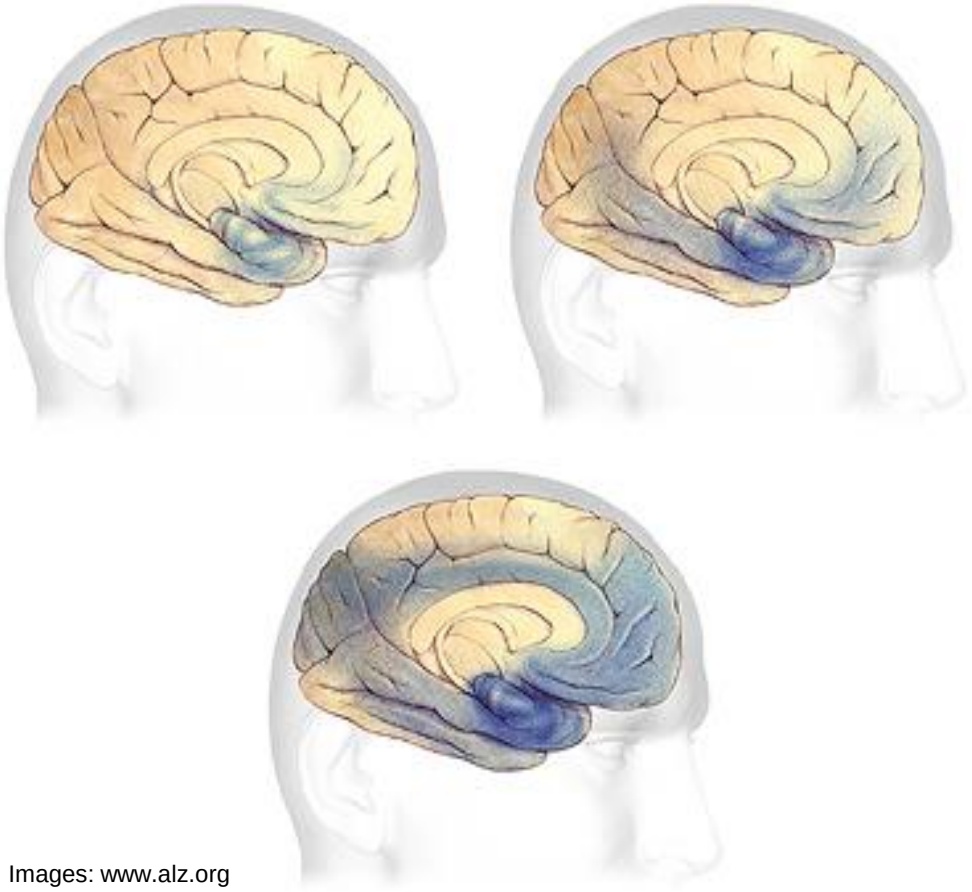
- Not all brain regions are affected equally or at the same time
- Some areas are more vulnerable
- Hallmark changes are first seen in temporal lobes
- Other brain regions may be affected first
- Spreads in a predictable pattern



Images: www.alz.org

Pathology: Genetics

- APOE4 is a variant of a gene that has been established as the most common genetic **risk factor** for sporadic Alzheimer's of late onset (usually after age 65)

- Presence of one or two copies of this gene increases the risk of Alzheimer's but it is also a poor predictor of who will or will not get the disease

- Familial, autosomal dominant, early onset forms of the disease (e.g. Presenilin 1 mutation) are very rare and account for less than 2% of cases

Treatment Strategies

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- There are currently no approved medications that can cure, slow down or revert Alzheimer's
- Approved medications are intended to treat symptoms and may provide temporary improvement
  - **Donepezil** (Aricept), rivastigmine (Exelon)
  - **Memantine** (Namenda)
- Non-pharmacological interventions can improve quality of life and may slow down progression (diet, exercise, social interaction, and caregiver support)
- Experimental drugs target know mechanism of disease through different approaches

Preventive Measures

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- Healthy lifestyle may slow cognitive decline and may reduce risk of developing dementia
- Study from **Lancet** showed evidence that a number of dementias (up to 1/3) may be preventable and that the risk can be significantly reduced by risk factor modification at different stages in life:
  - Early life - Level of education
  - Middle life - Hypertension, hearing loss and obesity
  - Late life - smoking cessation, treating depression, increased physical activity, social interaction, diabetes

Some Recommendations

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- Participate in intellectually engaging activities and maintain social interactions
- Routine physical activity, especially exercise that improves cardiovascular health
- Maintain a heart-healthy diet
- Maintain healthy sleep habits and treat sleep conditions such as sleep apnea
- Minimize alcohol use and do not smoke

Frontotemporal Lobar Degeneration

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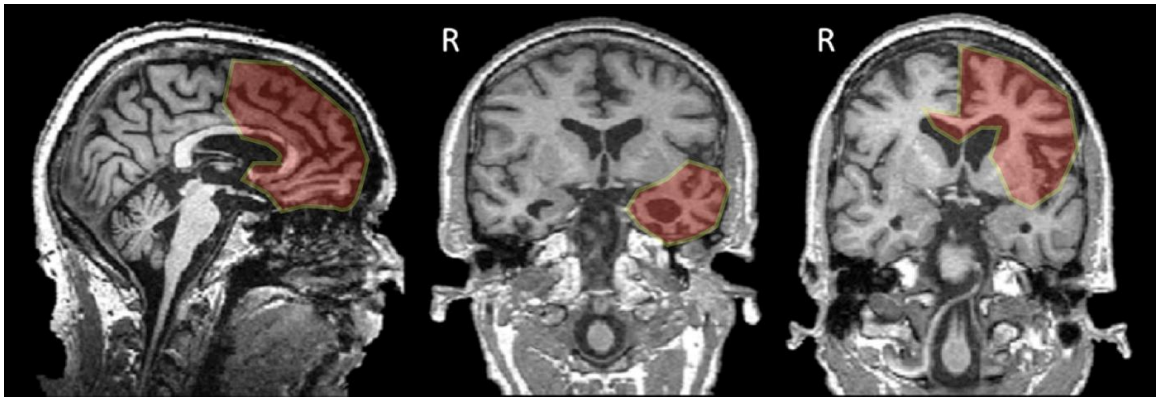
- Diverse group of syndromes
- Characterized by focal degeneration in the frontal and anterior temporal lobe



- Typically presents with behavioral symptoms, language impairments, or both
- Patients may also have motor symptoms and may develop other neurodegenerative diseases such as ALS
- In contrast with Alzheimer's, there are multiple types of pathological types, with different abnormal proteins

Frontotemporal Lobar Degeneration

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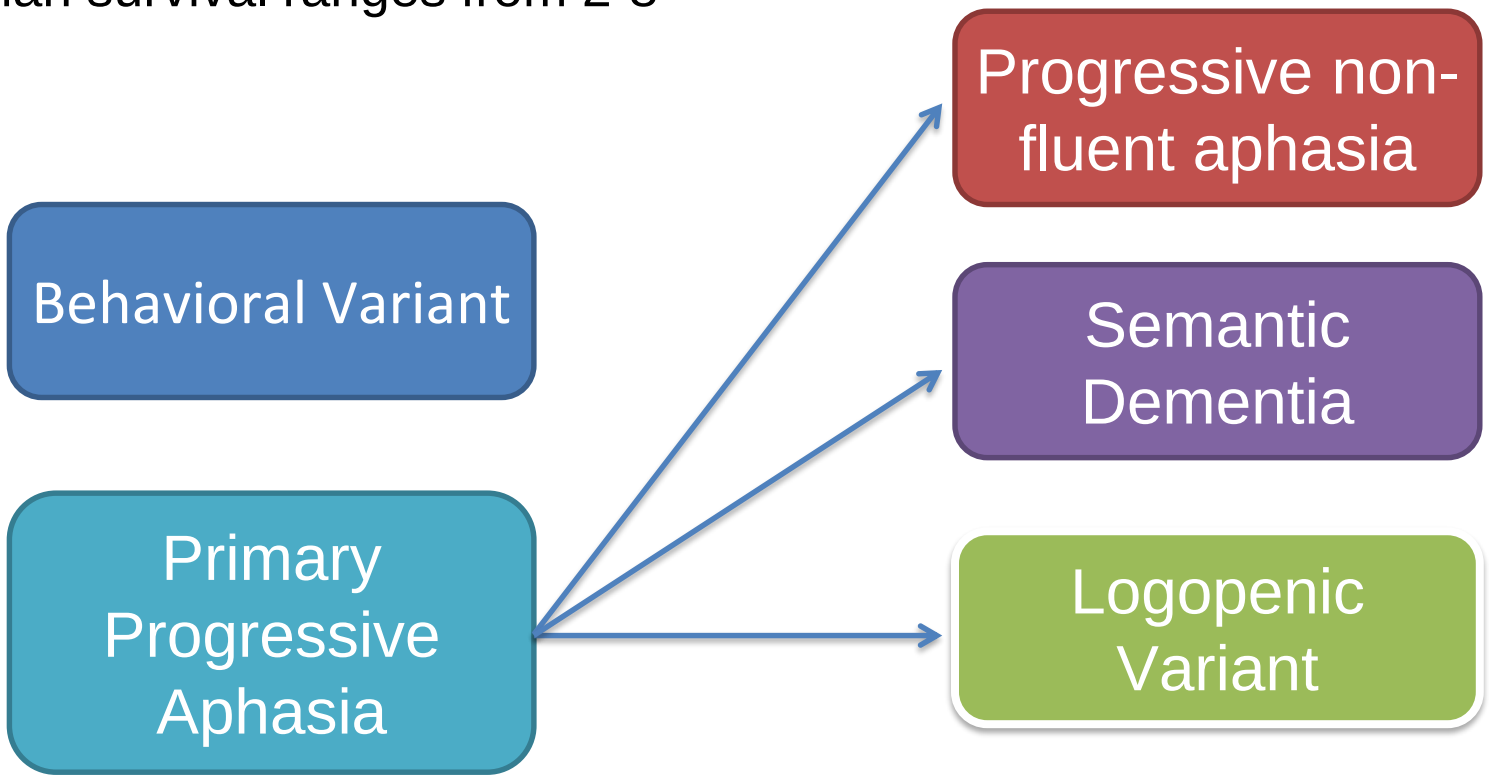
- Third most common cause of neurodegenerative dementia after AD and DLB
- Prevalence close to AD 60-70
- Age of onset **45-65**
- Median survival ranges from 2-8



Behavioral Variant

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- Insidious onset of changes in social decorum and personal regulation including:
  - ✓ Apathy
  - ✓ Overeating
  - ✓ Emotional blunting
  - ✓ Loss of empathy
  - ✓ Personality changes: Coldness and Submissiveness
  - ✓ Repetitive motor behaviors, ritualistic behaviors
  - ✓ Impairment of judgment and insight
  - ✓ Inappropriate behaviors and disinhibition
- Deficits in executive control as reflected by difficulties performing tasks such as:
  - ✓ Organization
  - ✓ Planning
  - ✓ Multitasking
  - ✓ Disengaging from specific activities
  - ✓ Generating ideas
- Behavioral symptoms are **very** common in other dementias. Behavioral and personality changes **do-not** equal FTD

Primary Progressive Aphasia

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Group of clinical syndromes with diverse pathology

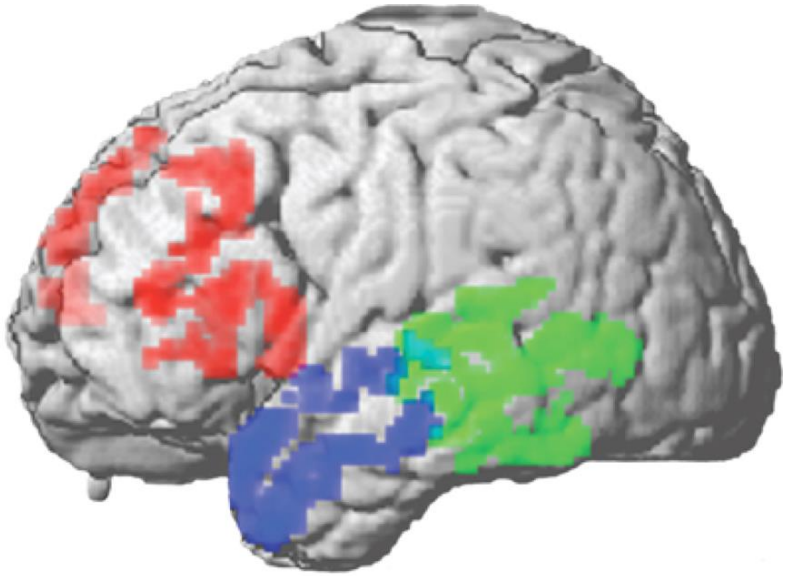
Most prominent clinical feature is difficulty with language

These deficits are the principal cause of impaired function

Distinct brain regions affected in each variant

Logopenic variant tends to be a language variant of Alzheimer's

- Non-fluent
- Semantic
- Logopenic



Non-Fluent Aphasia

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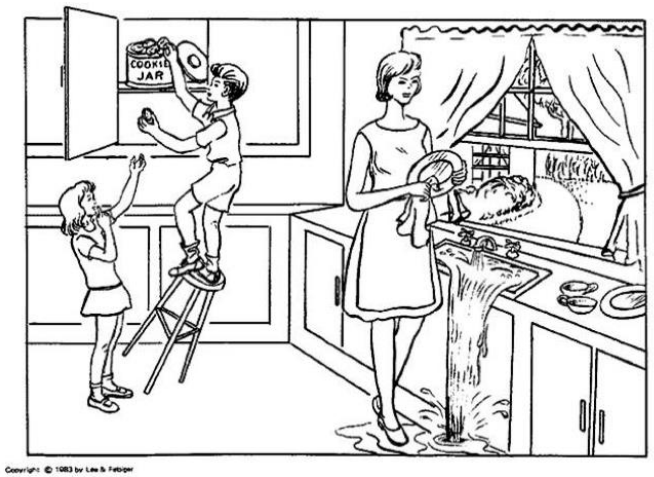
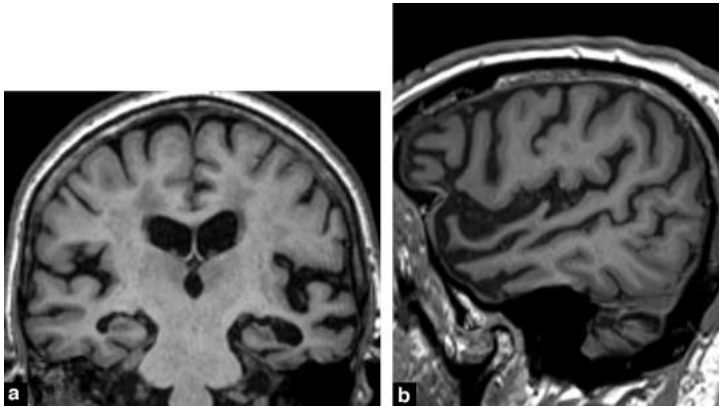
- Patients speak in simple phrases, with grammatical errors (e. g. errors in tense, use of prepositions)



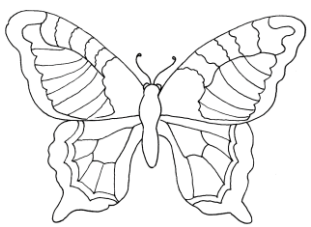
- Effortful speech: Slow, labored speech production
- Mispronunciation of words and errors in sequencing of syllables

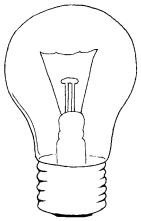
  “aminal” for “animal”
  “Sable” for “Table”

- Phrases are short, generally less than 4 words
- Inferior frontal and left antero-superior temporal atrophy

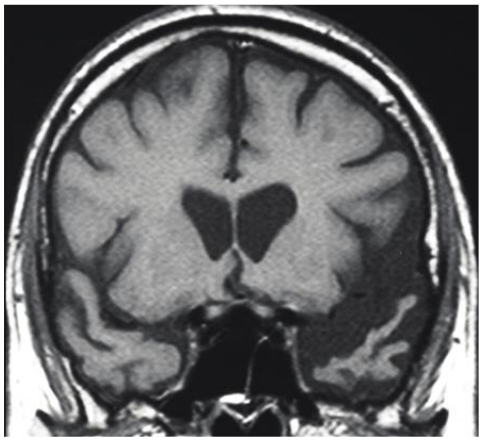


Semantic Dementia





- Difficulty naming objects and comprehension of single words with fluent speech and preserved grammar



- Patients often repeat the word and ask what it means
- May have difficulty interpreting facial expressions of emotion and recognizing familiar faces
- Right side: Prosopagnosia, some degree of anomia, mild loss of object knowledge. Often present behavioral symptoms similar to bvFTD
- Left side: Fluent aphasia beginning with profound anomia, later progressing to globally impaired knowledge of objects (what they do, where they are found, etc)

Treatment of FTD Syndromes

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- Management of inappropriate or aggressive behavior with non-pharmacological measures when possible
- Discussion of tolerance for disruptive but non-dangerous behavior
- Speech therapy for language variants
- Some types of antidepressants may help with some behaviors
- Atypical antipsychotics have risks but may be necessary
- No evidence to support use of Alzheimer's medications and in fact they may worsen symptoms and cognitive function

Dementia With Lewy Bodies (DLB)

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- Third most common type of adult onset dementia after AD and vascular
- Difficulties with attention, executive function and visual-spatial function
- Difficulties with memory that tend to improve with cuing
- Frequent hallucinations
- Rapid fluctuations in cognitive function (minutes or hours)
- REM behavior disorder
- Parkinsonism
- Can respond favorably to cholinesterase inhibitors (e.g. donepezil)

Diagnostic Approach to Dementia

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Diagnostic Approach to Dementia

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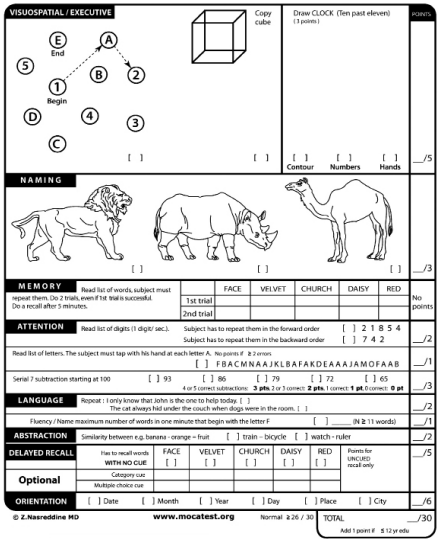
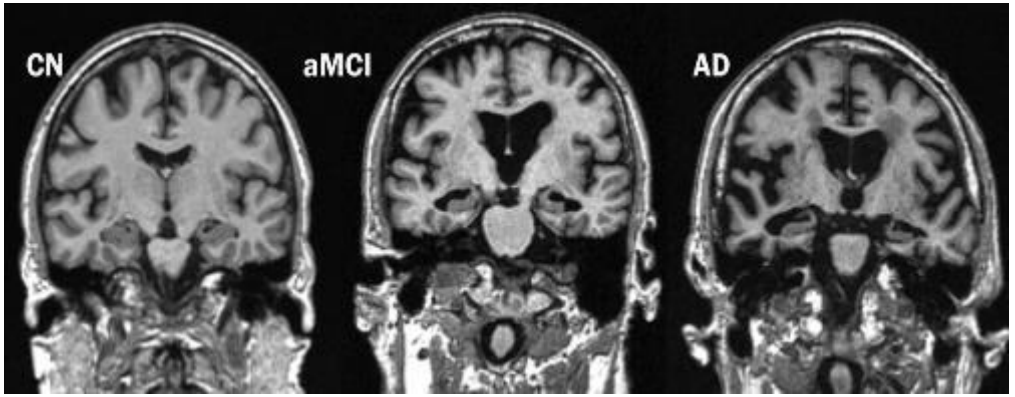
- Clinical history is most important
- Neurologic Exam
- Cognitive testing
  - Screening tests
  - Comprehensive neuropsychological testing
- Brain Imaging (MRI or CT scan)
- Spinal fluid markers or PET scans in complex cases (not routinely done)





Biomarkers

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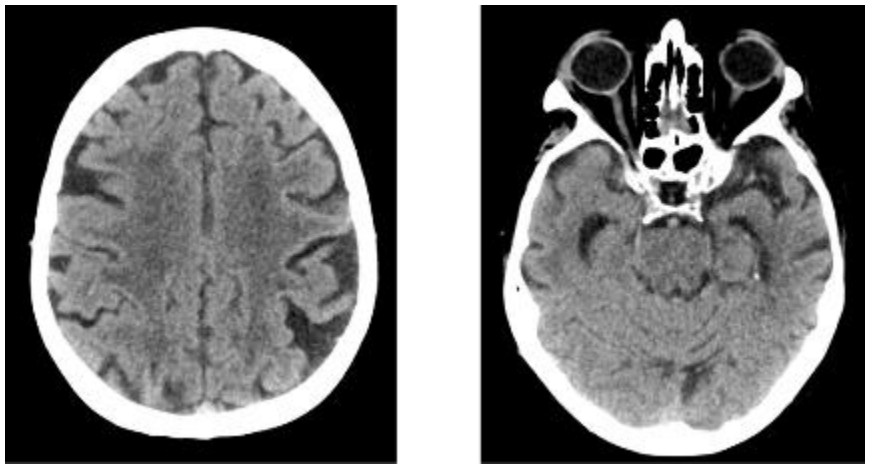
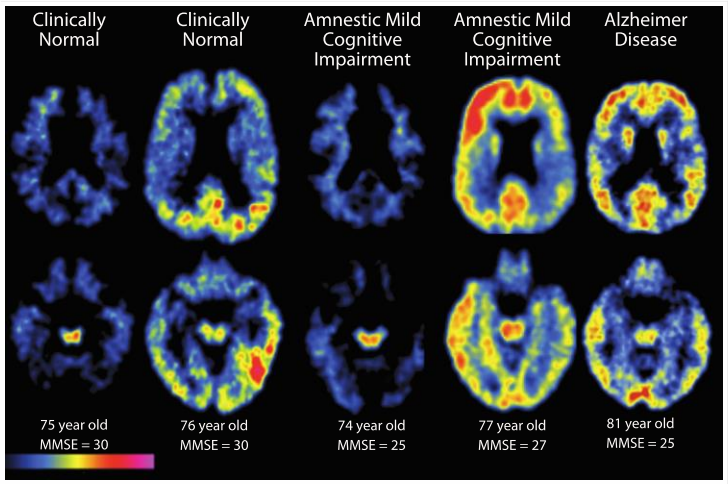
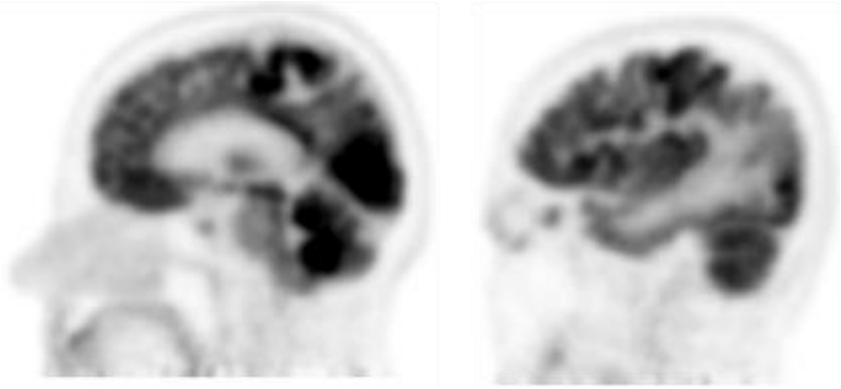
Structural: CT and MRI



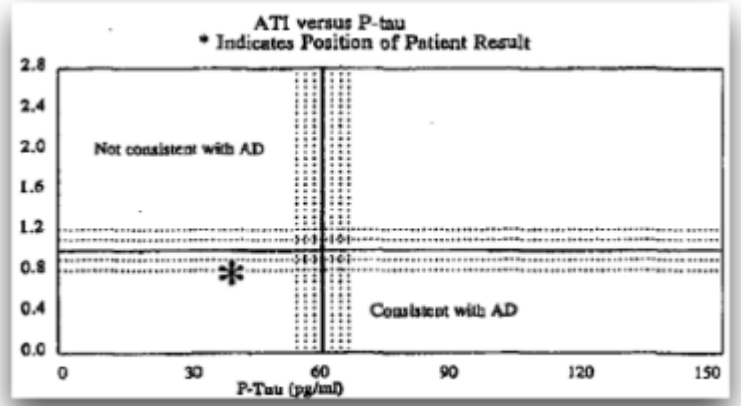
Amyloid PET



Functional: FDG-PET



CSF Aß and Tau



Multidisciplinary Care Model

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Multidisciplinary Team

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Behavioral Neurologists
Roberto Fernandez, MD, MPH, PhD
Bruce R. LeForce, MD
Mary Widmeyer, MD
Lauren McCollum, MD

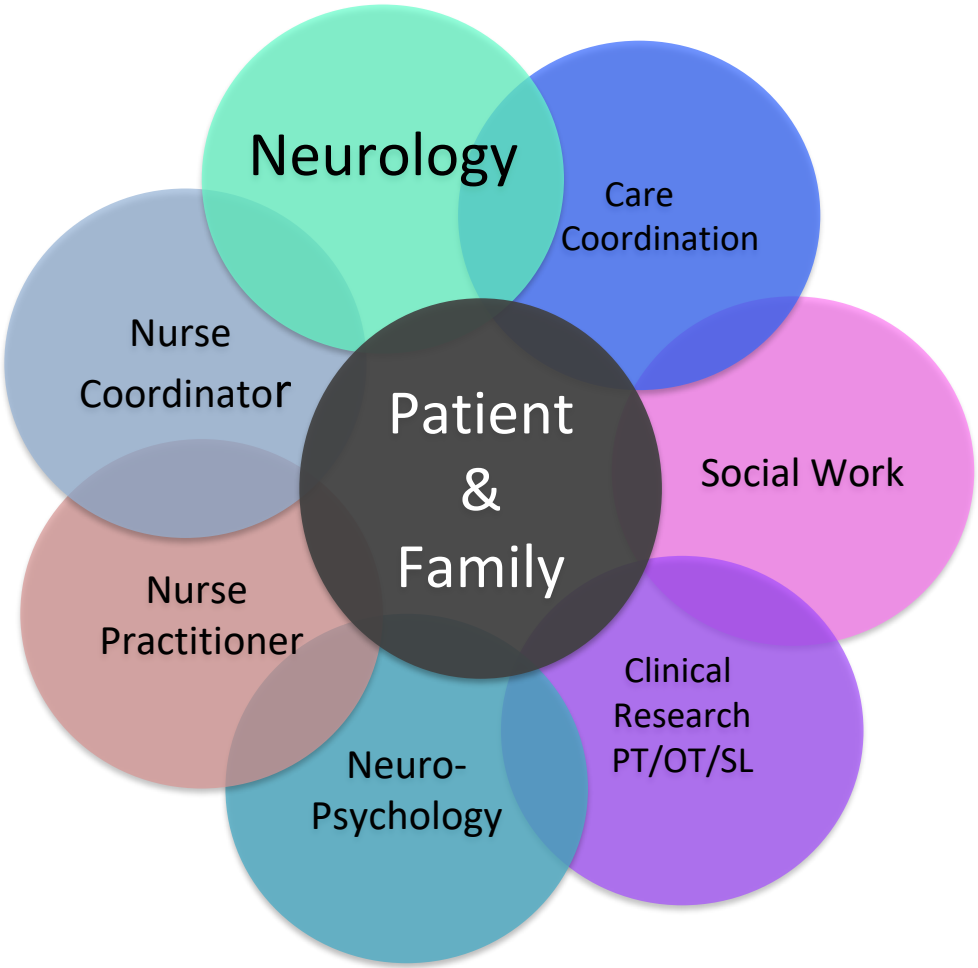
Clinical Neuropsychologists
Malcolm D. Spica, PhD
Nichole K. Miller, Psy. D

Nurse Practitioner
Heather Massengill, NP

Nurse Coordinator
Jan Alexander, RN

Social Worker
Sallie W. Gentry, LCSW, CCM
Charlotte Sorensen, MSW

Speech-Language Pathologist
Mandie Oslund, MS, CF-CSP



Medical Assistants
Elaine Leonard
Megan Pierce

Cognitive Testers
Sydney Michelson
Taylor Leonard

Clinical Care

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- **Comprehensive extended visits**
- **Individualized care focused on patient and caregivers**
- **Standardized cognitive testing performed at each visit**
- **Multidisciplinary team involvement**
- **Comprehensive neuropsychological testing**
- **Specialized cutting edge diagnostic techniques**
- **Care coordination and caregiver support**
- **Clinical and basic science research**

Social Work

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- Caregiver support
- Educational programs
- Support Groups
- Transportation
- Housing
- Power of Attorney/Living wills
- Driving Concerns
- Community resources
- Placement
- Durable medical equipment
- Therapy, PT, OT, Speech
- Patient letters
- In-home care
- Hospice/Palliative care
- Capacity questions
- Elder abuse
- Patient assistance programs

Nurse Coordinator

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- Visit with all new patients to review plan, offer visit summary and any additional individualized teaching and educational materials as indicated by provider
- Meets with follow-up patients as needed when new interventions or changes in management are implemented
- Maintain information of ongoing clinical trials and other research studies and discuss with patients and families who are interested in possible participation
- Follow up telephone calls to families and patients with information and support as needed regarding test results, caregiver support and questions, and medications
- Coordination of communication between physician and patient, families, health care team

Neuropsychology

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Neuropsychological evaluations:

- Designed to identify the extent & severity of a person's cognitive and behavioral impairments
- Help determine a person's areas of cognitive strength/weakness
- Help assess patients capacity for decision making
- Use standardized tests to evaluate cognitive abilities such as:
  - Attention
  - Memory
  - Language
  - Processing speed
  - Visuospatial function
  - Planning and Organization

- Not all patients are candidates for full testing. Indication and extent of testing is determined by behavioral neurologist at time of referral

Initial Visit

- Time for provider to meet separately with caregivers and with patient
- Brief standardized cognitive screeners (MoCA and Cognivue)
- Administration of multiple diagnostic instruments for assessment of depression, anxiety, caregiver strain and ADLs
- Caregiver meeting with Social Work
- Visit with Nurse Coordinator to review plan and education
- Patient and family should plan for a 3 hour visit
- Diagnostic work-up: May include brain imaging, full neuropsychological testing, blood work and advanced diagnostics in very specific cases (e.g. CSF biomarkers, PET imaging)

Follow-up Visits

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Diagnostic Follow-up

- Review of work-up results
- Discussion of diagnosis
- Addressing treatment and plan of care with provider and Social Work
- Meeting with nurse coordinator as needed

Routine Follow-up

- Usually every 6 months
- May alternate with MLP (patients will see neurologist at least once a year)
- Repeat brief neuropsychological testing at 6 month intervals
- Meeting with nurse coordinator and/or Social Worker as needed

Savvy Caregiver Program

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- Program intended to train caregivers in the basic knowledge, skills and attitudes needed to handle the challenges of caring for family members with dementia
- 12 hours of training, divided in 2 hour sessions over 6 weeks
- A total of 20 caregivers have been trained
- Respite care provided for patients
- High satisfaction and impact according to surveys
- We will continue to offer this program several times a year

Benefits of Multidisciplinary Care

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- Timely and accurate diagnosis
- Personalized treatment and plan of care
- Optimized treatment tailored to condition and stage of disease
- Access to educational resources
- Access to support resources
- Opportunities for participation in clinical trials and research studies
- Helps facilitate transitions through the course of disease and end of life

- Improve patient outcomes
- Decrease hospitalizations and delays in institutionalization
- Increase patient satisfaction
- Decreases unnecessary health care
- Improves patient and caregiver quality of life.
- Reduces caregiver burden
- Increases independence

2nd Annual Symposium

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Symposium on Alzheimer's and Dementia

Save the Date!
May 7-8, 2020

Downtown Knoxville

Tyler Summitt

Ronald Petersen
MD, PhD

Nicole Dawson
PT, PhD

Thank You

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“Put the team before yourself”

From Pat Summitt’s Definite Dozen