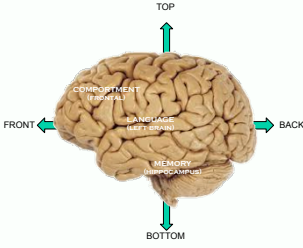


**INTRODUCTION TO
PRIMARY PROGRESSIVE APHASIA (PPA)
AND BEHAVIORAL VARIANT FRONTOTEMPORAL DEGENERATION (bvFTD)**

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BRAIN FACTS
3-4 LBS, 30 SQ. FT.
40,000,000,000 NEURONS, 10¹⁴ CONTACTS (SYNAPSES)



BRAIN INJURY CAN BE SUDDEN OR SLOWLY PROGRESSIVE OVER YEARS.
DISEASES THAT CAUSE PROGRESSIVE BRAIN INJURY (ATROPHY) ARE KNOWN AS NEURODEGENERATIONS.
THE LOCATION OF ATROPHY DETERMINES THE NATURE OF THE DEMENTIA- bvFTD, PPA, MEMORY LOSS.
TWO MAJOR NEURODEGENERATIVE DISEASES ARE ALZHEIMER'S DISEASE (AD) AND FRONTOTEMPORAL LOBAR DEGENERATIONS (FTLD).
THE TWO MAJOR TYPES OF FTLD ARE FTLD-TAU AND FTLD-TDP.

PRIMARY PROGRESSIVE APHASIA (PPA)

a) Impaired usage, retrieval, spelling, or comprehension of words (i.e., aphasia)
b) The language disorder initially arises as the *principal deficit*, (i.e., primary),
c) The cause is a neurodegenerative disease (i.e., progressive)

NEURODEGENERATION TYPES IN PPA:
(30% FTLD-TAU; 30% FTLD-TDP; 40% ALZHEIMER PATHOLOGY)

THE ONE COMMON DENOMINATOR IS THE LOCATION OF
NEURONAL LOSS IN THE LEFT HEMISPHERE LANGUAGE
NETWORK.

TYPES OF PPA AND RELATIONSHIP TO NEURODEGENERATION

<p>AGRAMMATIC (PPA-G) consistently low word output distorted grammar good comprehension</p>	<p>Mostly FTLD-Tau, a few AD.</p>
<p>SEMANTIC (PPA-S) good word output & grammar poor comprehension & naming</p>	<p>Mostly FTLD-TDP, a few AD.</p>
<p>LOGOPENIC (PPA-L) word-finding hesitations good grammar & comprehension</p>	<p>Mostly AD, a few FTLD.</p>

THERAPEUTIC CONSIDERATIONS- PPA

MEDICAL-

- Is it AD or not?-DETERMINED BY AMYLOID PET SCANS AND SPINAL FLUID ANALYSIS.
- If AD- Galantamina, Donepezil, Rivastigmine, Memantine.
- If not- Is in FTLD-Tau or FTLD-TDP- a goal for the future.
- In either case, refer to proper clinical trials.

NON-PHARMACOLOGICAL-

- Education- AFTD, IMPACT (ppaconnection.org).
- Life-enrichment programs & psychosocial interventions.
- Speech therapy- the internet program funded by AFTD.
- TMS?

BEHAVIORAL VARIANT FRONTOTEMPORAL DEMENTIA (bvFTD)

THE CLINICAL PICTURE INCLUDES THE NEW ONSET OF ONE OR MORE OF THE FOLLOWING FEATURES IN RELATIVE ISOLATION

- 1) Diminished motivation (apathy) alternating with loss of restraint (disinhibition, gullibility, indiscretions).
- 2) Impaired judgment, insight, problem solving, multitasking, flexibility.
- 3) Emotional blunting with erosion of empathy or concern for the feelings of others.

DEFINITE DIAGNOSIS REQUIRES THREE STEPS:

1. Documentation of the clinical picture by history or neuropsychological testing.
2. Documentation of progression.
3. Documentation of abnormality in the frontal lobes by MRI, SPECT or metabolic PET.

IF THE CLINICAL PICTURE LOOKS LIKE bvFTD BUT THERE IS NO DOCUMENTATION OF PROGRESSION OR FRONTAL LOBE ABNORMALITY ON IMAGING, MORE TENTATIVE TERMS SUCH AS "POSSIBLE" bvFTD OR bvFTD "PHENOCOPY" ARE MORE APPROPRIATE

NEURODEGENERATIONS IN bvFTD:
(40% FTLD-TAU; 40% FTLD-TDP; 20% AD)

THERAPEUTIC CONSIDERATIONS-bvFTD

THE MEDICAL-

- Is it AD or not?
- If AD- use AD drugs.
- If not- Is in FTLD-Tau or FTLD-TDP- a goal for the future.
- In either case, refer to proper clinical trials.

NON-PHARMACOLOGICAL-

- Education.
- Psychosocial interventions.
- Life-enrichment programs.
- Behavioral modification.
- TMS?

QUIZ

1. ARE PPA AND bvFTD MOST COMMONLY CAUSED BY FTLD? **YES**
2. ARE PPA AND bvFTD ALWAYS CAUSED BY FTLD? **NO**
3. IS IT POSSIBLE TO HAVE BOTH PPA AND ALZHEIMER'S DISEASE? **YES**
4. CAN bvFTD BE CAUSED BY ALZHEIMER'S DISEASE? **YES**