

**Social Security Administration
Compassionate Allowance Outreach Hearing on
Early-Onset Alzheimer's and Related Dementias
July 29, 2009, Chicago, Illinois**

Primary Progressive Aphasia And Other Atypical Young-Onset Dementias

Marsel Mesulam, MD
Center Director

Northwestern Alzheimer's Disease Center
Northwestern University Feinberg School of Medicine
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Website: <http://www.brain.northwestern.edu>

ALZHEIMER'S DISEASE, DEMENTIA, AND OLD AGE

Alzheimer's disease (AD) is the most widely recognized form of dementia. It is most frequently seen in the elderly and typically causes profound memory impairments.

Many of us therefore develop the impression that AD and dementia are the same thing, that dementia can only occur in the elderly and that a diagnosis of dementia cannot be made in the absence of memory loss.

The major message that my colleagues and I would like to bring to this hearing is the need to critically re-examine these assumptions.

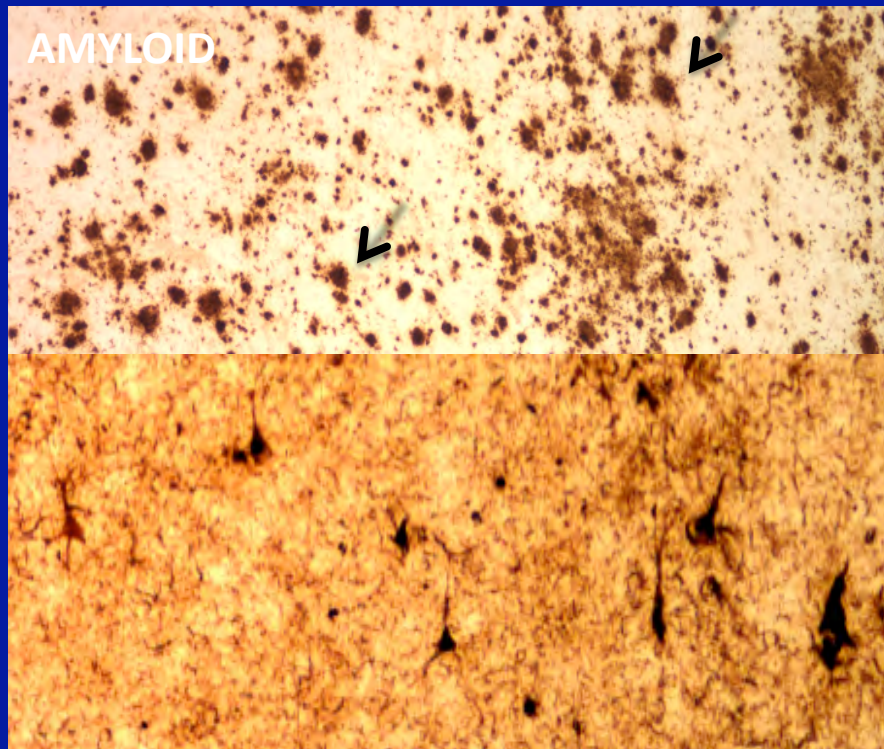
REVISING OUR CONCEPTS OF DEMENTIA AND ALZHEIMER'S DISEASE (AD)

1. AD CAN ARISE IN THE YOUNG. THE YOUNGEST PATIENT I SAW WAS 24.
2. DEMENTIA CAN BE CAUSED BY DISEASES OTHER THAN AD. THE MOST FREQUENT YOUNG-ONSET, NON-AD DEMENTIA IS KNOWN AS FRONTOTEMPORAL LOBAR DEGENERATION (FTLD).
3. FTLD AND UNUSUAL FORMS OF AD CAN CAUSE DISABILITY BY IMPAIRING LANGUAGE, SPATIAL ORIENTATION OR BEHAVIOR RATHER THAN MEMORY. IN SOME OF THESE PATIENTS A DIAGNOSIS OF DEMENTIA IS MADE ALTHOUGH MEMORY IS NORMAL.

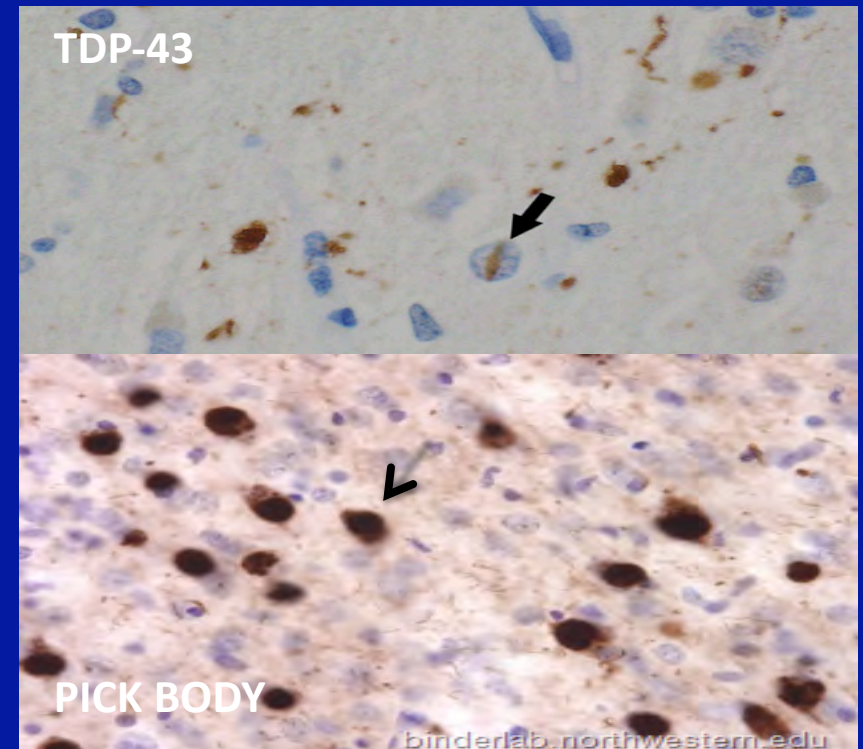
FRONTOTEMPORAL LOBAR DEGENERATION (FTLD) -A MAJOR CAUSE OF NON-AD, YOUNG ONSET, ATYPICAL DEMENTIA-

- ALZHEIMER'S DISEASE HAS AMYLOID IN THE BRAIN, FTLD DOES NOT.
- HEREDITARY AD IS LINKED TO CHR 1, 14, 21. FTLD IS LINKED TO CHR 3, 9, 17.
- TYPICAL ALZHEIMER'S LEADS TO AN AMNESTIC DEMENTIA, FTLD USUALLY SPARES MEMORY.
- FTLD CAN CAUSE TWO DIFFERENT CLINICAL SYNDROMES OF DEMENTIA- bvFTD AND PPA.

ALZHEIMER



FRONTOTEMPORAL



THE TWO DEMENTIAS OF FRONTOTEMPORAL LOBAR DEGENERATION (FTLD)

bvFTD (aka FTD) causes a gradual loss of judgment, insight and comportment to the point where the person cannot make sound decisions and engages in inappropriate impulsive behaviors.

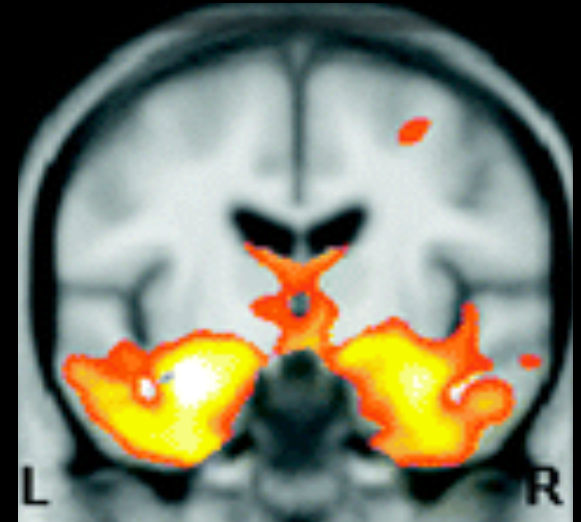
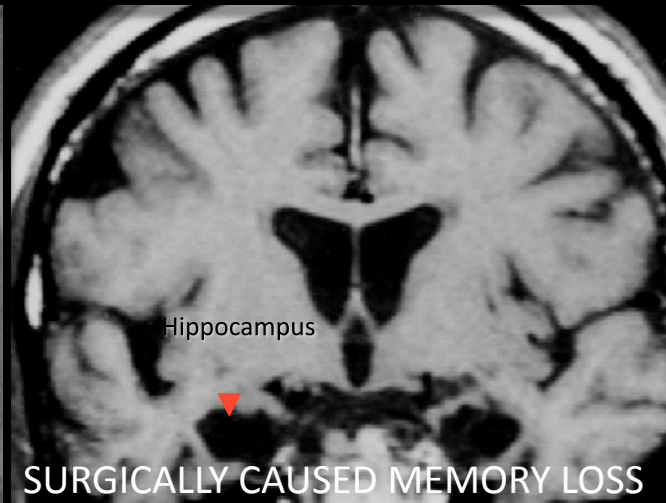
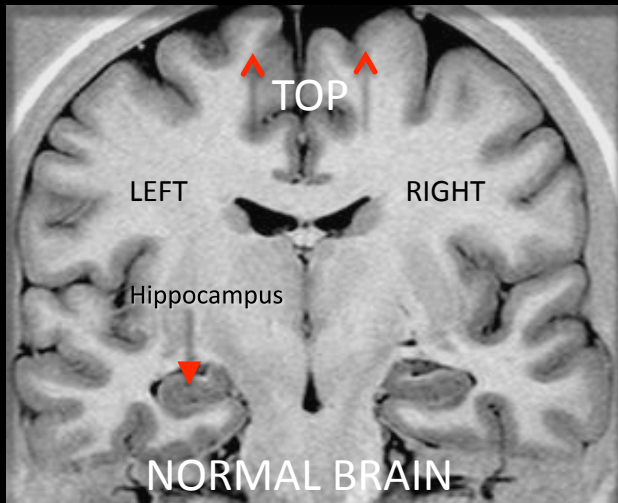
PPA results in a gradual loss of word usage and comprehension to the point where the person loses the ability to communicate thoughts or understand what others are saying.
The problem is not in articulating or hearing speech but in the brain's ability to use and interpret words.

**BOTH bvFTD AND PPA USUALLY START BEFORE THE AGE OF 60 AND
CAN SPARE MEMORY FOR MANY YEARS**

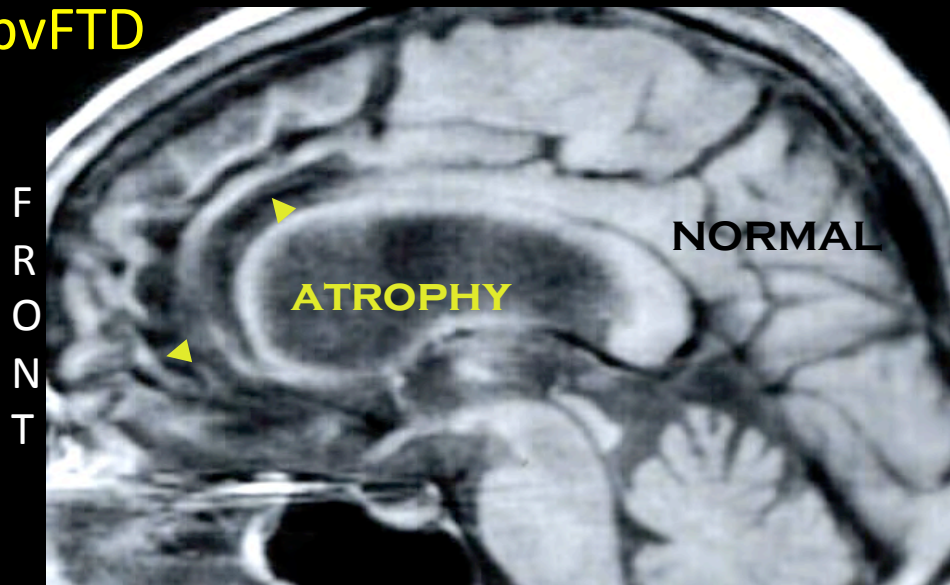
**bvFTD HAS BEEN INCLUDED IN THE COMPASSIONATE ALLOWANCES LIST
BUT PPA HAS NOT**

WHY ARE THERE SO MANY DEMENTIAS?

-different parts of the brain have different job descriptions
and each neurodegenerative disease has a different preferred target of destruction-



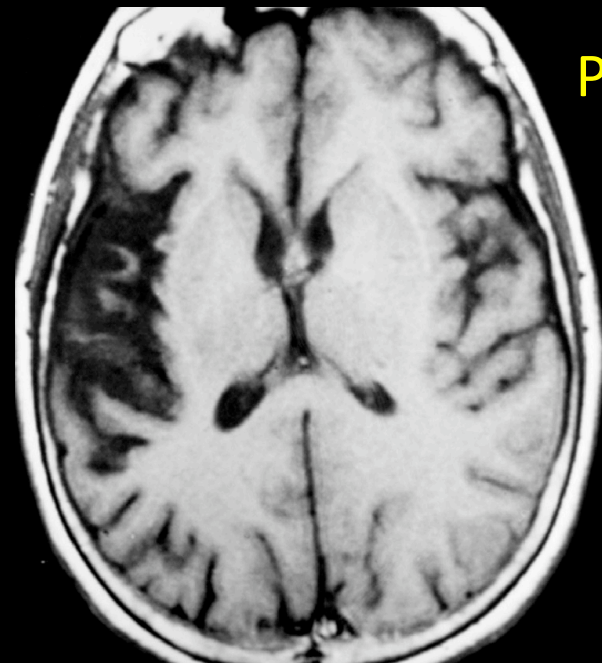
bvFTD



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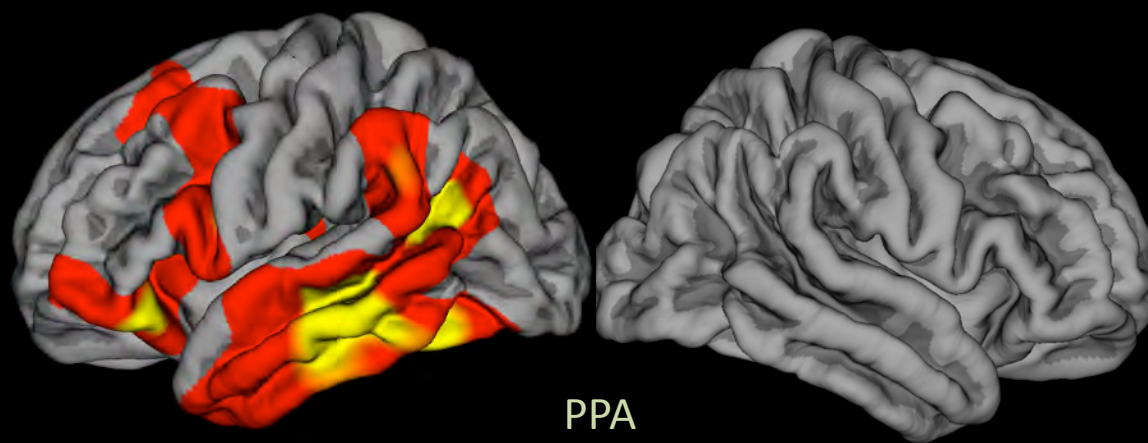
PPA



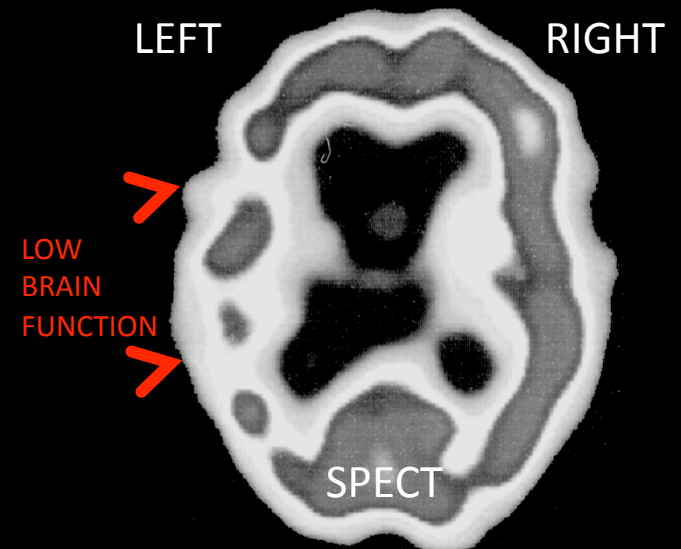
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DIAGNOSTIC CRITERIA FOR PRIMARY PROGRESSIVE APHASIA (PPA)

1. DOCUMENTED HISTORY OF DISABLING AND PROGRESSIVE IMPAIRMENT IN-
 - word-finding
 - word usage
 - ordering words into meaningful sentences
 - word comprehension and reading
 - writing and spelling
2. OBJECTIVE NEUROPSYCHOLOGICAL EVIDENCE OF LANGUAGE IMPAIRMENT AND RELATIVE SPARING OF OTHER COGNITIVE ABILITIES.
3. IMAGING EVIDENCE OF PROGRESSIVE BRAIN DAMAGE IN LANGUAGE AREAS



PPA
COMPUTERIZED MAPPING OF ATROPHY



SUMMARY FACTS ABOUT PRIMARY PROGRESSIVE APHASIA (PPA)

- PPA IS A LANGUAGE-BASED DEMENTIA**
- IT IS DIAGNOSED BY OBJECTIVE CRITERIA**
- IT INVARIABLY LEADS TO TOTAL DISABILITY**
- IT TYPICALLY ARISES BEFORE THE AGE OF 60**
- IT IS CAUSED BY FTLD IN 70% OF PATIENTS AND BY AD IN 30%**