

# Un cas de démence fronto-temporale

l'Équipe de 2 nord

# Confidentiality

- ❖ As we are all health professionals, our codes de déontologie require that we respect patient confidentiality
- ❖ Please do not discuss today's case outside of this room; take especial care in elevators, the cafeteria, etc. where visitors and families may overhear

# History of FTD

- ❧ Described 1892 by Arnold Pick
  - ❧ frontotemporal lobar degeneration at autopsy
  - ❧ argentophilic intranuclear inclusions (Pick bodies) found in later cases
  - ❧ subsequently called “Pick’s disease”
- ❧ later found that most cases of frontal lobe degeneration did not have Pick bodies
- ❧ Pick bodies are also found in other neurodegenerative conditions

# Terminology

- ❧ Frontotemporal dementia (FTD)
  - ❧ behavioural presentation
  - ❧ overall disease
- ❧ Frontotemporal degeneration (FTD)
  - ❧ used for all pathological variants
  - ❧ may be used for the clinical disease

- ❧ Frontotemporal Lobar Degeneration (FTLD)
  - ❧ lobar added to distinguish the pathological designation from the behavioural presentation (FTD)
  - ❧ also used for the overall clinical disease
- ❧ Pick's Disease (PiD)
  - ❧ less used now for the overall clinical syndrome
  - ❧ histologically defined condition
- ❧ Pick Complex (FTD/Pick)
  - ❧ groups everything (clinical syndromes and pathological variants)

- ❧ Primary Progressive Aphasia (PPA)

- ❧ slowly progressive aphasia before anything else develops

- ❧ Semantic Dementia (SD)

- ❧ loss of meaning in multiple modalities, including visual

- ❧ difficulty with both comprehension and naming, especially nouns

- ❧ Corticobasal Degeneration Syndrome (CBDS)

- ❧ unilateral rigidity, immobility, apraxia, “alien hand”

- ❧ Progressive Supranuclear Palsy (PSP)

- ❧ vertical gaze palsy, slowness, falling, dysarthria

- ❧ overlaps with CBDS

# Frontotemporal dementia behavioral variant (FTD-bv)

- behavioural changes often begin with apathy, disinterest (may look like depression)
- disinhibition symptoms (may suggest mania, OCD, sociopathic PD)
- unable to plan or carry out complex tasks
- inattentive, impulsive, distractible



- ❧ childish behaviour
- ❧ rudeness
- ❧ inappropriate sexual remarks
- ❧ impatient, careless driving
- ❧ excessive spending
- ❧ hoarding of certain items
- ❧ inappropriate joking
- ❧ perseverative routines
- ❧ compulsive roaming
- ❧ insistence on certain foods
- ❧ excessive food intake
- ❧ neglect of personal hygiene
- ❧ disinterest in the immediate family
- ❧ pilfering, shoplifting, swearing, undressing in public, unexpected urinary or fecal incontinence may involve police

# Primary Progressive Aphasia (PPA)

- ❧ initially: word finding difficulty or anomia
- ❧ relatively preserved memory and nonverbal cognition (cf. AD)
- ❧ progresses to a non-fluent aphasia with increasing word finding difficulty, reduced speech output
- ❧ sometimes see grammatical errors and phonemic paraphasias
- ❧ may see stuttering, slow segmented speech, difficulty articulating
- ❧ progresses to mutism

### FRONTAL BEHAVIORAL INVENTORY (FBI)

Name: \_\_\_\_\_ Age: \_\_\_\_\_ Date: \_\_\_\_\_

Duration: \_\_\_\_\_ Caregiver: \_\_\_\_\_ Examiner: \_\_\_\_\_

Explain to the caregiver that you are looking for a change in behavior and personality. Ask the caregiver these questions in the absence of the patient. Elaborate if necessary. At the end of each question, ask about the extent of behavioral change, and then score it according to the following:

**0 = none    1 = mild, occasional    2 = moderate    3 = severe, most of the time**

1. Apathy: Has she/he lost interest in friends or daily activities? \_\_\_\_
2. Spontaneity: Does she/he start things on his/her own, or does she/he have to be asked? \_\_\_\_
3. Indifference, Emotional Flatness: Does she/he respond to occasions of joy or sadness much as ever, or has she/he lost emotional responsiveness? \_\_\_\_
4. Inflexibility: Can she/he change his/her mind with reason or does she/he appear stubborn or rigid in thinking lately? \_\_\_\_
5. Concreteness: Does she/he interpret what is being said appropriately or does she/he choose only the concrete meanings of what is being said? \_\_\_\_
6. Personal Neglect: Does she/he take as much care of his/her personal hygiene and appearance as usual? \_\_\_\_
7. Disorganization: Can she/he plan and organize complex activity or is she/he easily distractible, impersistent, or unable to complete a job? \_\_\_\_
8. Inattention: Does she/he pay attention to what is going on or does she/he seem to lose track or not follow at all? \_\_\_\_
9. Loss of Insight: Is she/he aware of any problems or changes, or does she/he seem unaware of them or deny them when discussed? \_\_\_\_
10. Logopenia: Is she/he as talkative as before or has the amount of speech significantly decreased? \_\_\_\_
11. Verbal Apraxia: Has she/he been talking clearly or has she/he been making errors in speech? Is there slurring or hesitation? \_\_\_\_
12. Perservation: Does she/he repeat or perseverate actions or remarks? \_\_\_\_
13. Irritability: Has she/he been irritable, short-tempered or is she/he reacting to stress or frustration as she/he always had? \_\_\_\_
14. Excessive Jocularly: Has she/he been making jokes excessively or offensively or at the wrong time? \_\_\_\_
15. Poor Judgment: Has she/he been using good judgment in decisions or in driving, or has she/he acted irresponsibly, neglectfully or in poor judgment? \_\_\_\_
16. Inappropriateness: Has she/he kept social rules or has she/he said or done things outside what are acceptable? Has she/he been rude or childish? \_\_\_\_
17. Impulsivity: Has she/he acted or spoken without thinking about consequences, on the spur of the moment? \_\_\_\_
18. Restlessness: Has she/he been restless or hyperactive, or is the activity level normal? \_\_\_\_
19. Aggression: Has she/he shown aggression, or shouted at anyone or hurt him or her physically? \_\_\_\_
20. Hyperorality: Has she/he been drinking more than usual, eating excessively anything in sight, or even putting objects in his/her mouth? \_\_\_\_
21. Hyper Sexuality: Has sexual behavior been unusual or excessive? \_\_\_\_
22. Utilization Behavior: Does she/he seem to need to touch, feel, examine, or pick up objects within reach and sight? \_\_\_\_
23. Incontinence: Has she/he wet or soiled his or herself? (Excluding physical illness, such as urinary infection or immobility). \_\_\_\_
24. Alien hand: Does she/he have any problem using a hand, and does it interfere with the other hand? (Excluding arthritis, trauma, paralysis, etc). \_\_\_\_

**Total Score** \_\_\_\_\_