Appreciating the Common Changes, Symptoms and Challenges of Atypical Dementias:
What if it is not one of the usual suspects?

Dementia

Four Truths About All Dementias:
- At least two parts of the brain are dying
- It keeps changing and getting worse – progressive
- It is not curable or reversible – chronic
- It results in death – terminal

<table>
<thead>
<tr>
<th>Alzheimer's</th>
<th>Lewy Body</th>
<th>Vascular</th>
<th>Frontotemporal</th>
</tr>
</thead>
<tbody>
<tr>
<td>New skills lost first</td>
<td>Movement problems – walks/coordination</td>
<td>Severe changes in behavior</td>
<td>Non-verbal</td>
</tr>
<tr>
<td>Memory problems – short term</td>
<td>Delusional thinking</td>
<td>Sensory changes</td>
<td>Non-verbal</td>
</tr>
<tr>
<td>Severe problems with eating</td>
<td>Gastrointestinal symptoms</td>
<td>Mental changes</td>
<td>Non-verbal</td>
</tr>
<tr>
<td>Incontinence</td>
<td>Other symptoms</td>
<td>Social withdrawal</td>
<td>Non-verbal</td>
</tr>
<tr>
<td>Mood swings</td>
<td>Mental confusion</td>
<td>Loss of muscle tone</td>
<td>Non-verbal</td>
</tr>
<tr>
<td>Agitation</td>
<td>Sleep disturbed</td>
<td>Movements slow or stiff</td>
<td>Non-verbal</td>
</tr>
<tr>
<td>Confusions</td>
<td>Paranoia</td>
<td>Loss of speech</td>
<td>Non-verbal</td>
</tr>
<tr>
<td>Agitation</td>
<td>Hallucinations</td>
<td>Loss of speech</td>
<td>Non-verbal</td>
</tr>
<tr>
<td>Personality changes</td>
<td>Changes in sleep</td>
<td>Memory changes</td>
<td>Non-verbal</td>
</tr>
<tr>
<td>Severe depression</td>
<td>Loss of function</td>
<td>Loss of speech</td>
<td>Non-verbal</td>
</tr>
</tbody>
</table>

- Family history
- Psychiatric – bipolar disorder, mania
- Memory loss
- Vision loss
- Sleep or eating problems
- Speech changes
- Immediate and long-term memory impairment
- Comorbidities
- Seizures

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What if it is not one of the usual suspects?
What does **Atypical Mean to Us, Under the Umbrella?**

- Because it is not what we would usually notice, we might very well:
  - Miss it
  - Think it is something else
  - Think it is just *part of the person*
  - Believe there is nothing we can do about it
  - Stay in *seek mode* a long time, without figuring out our *response mode*

What are some of the **Atypical Dementias?**

- CTE – Chronic Traumatic Encephalopathy
- PCA – Posterior Cortical Atrophy
- Huntington's Chorea and other genetic syndromes
- Alcohol-related dementia
- PSP: Progressive Supranuclear Palsy
- Creutzfeldt-Jakob disease
- Battens disease
- Multi-System Atrophy

Some of the Other **Atypical Dementias:**

- Drugs/toxin exposure: heavy metals, pesticides
- White matter diseases: MS
- Mass effects: tumors, NPH
- Depression and other psychological conditions
- Infections that cross the blood-brain barrier: C-J, HIV/Aids, Lyme?
- Or, could be a mixture of two or more types
Chronic Traumatic Encephalopathy:
- Caused by repeated head injuries or concussions, but doesn’t happen to all
- Symptoms:
  - Frontal lobe issues
  - Temporal lobe issues
  - Sometimes rapid progression into Alzheimers-type patterns
  - Sometimes rapid progression into FvFTD patterns
  - Sometimes rapid progression into temporal lobe changes
  - Emotional episodes of extreme distress and inability to cope or appreciate others’ perspectives – not seeing them as valuable or reasonable

List of Common Possible Symptoms of CTE:
- Early signs: headaches, attention-concentration problems, depression and suicidal ideations, but no changes in the structures of the brain
- Episodes of extreme impulsivity in actions, words, decisions
- Episodes of paranoia or mis-interpretation of visual or auditory info
- Challenges with hyper- and hypo-attention and ability to focus attention on what is important in sustaining relationships or work situations
- Episodes of more acute confusion, frustration, and inability to appreciate any other perspective
- Difficulty with tremors, coordination, and skilled task performance that only occurs occasionally at first, especially when under stress
- Episodes of getting lost or disoriented to place or situation, missing key elements
- Emotional lability with extremes: anger, sadness, anxiety, apathy, and fears of being trapped or abandoned

A Few CTE Resources that Might be Helpful:
Progressive Supranuclear Palsy (PSP):

- A subcortical dementia first highlighted in modern times in the 70s (Steele, Richardson-Olszewski)
- Typically happens to people over age 60: marked slowing and weakening
- Large numbers of tau pathologies spreading from the deep brain and cerebellum to the pre-frontal, frontal, temporal, parietal lobe regions.
- Challenges with movement, balance, coordination, overall slowing of all functioning, multiple falls due to lack of ability to react to gravity’s demands during normal movements and actions
- Major changes in visual gaze abilities: slowed tracking and limited gaze, inability to track up and down (“doll eyes”)
- Issues with slurred and slowed speech with pauses and difficulty with breath and voice control
- Problems with swallowing, drinking, or chewing and pneumonia
- Problems as PSP progresses with sleep, light sensitivity, apathy, depression, anxiety, emotional lability, and limited reasoning and impulse control
- Can be mis-identified as Parkinsons

A Few PSP Resources that Might be Helpful:


Alcohol-Related Brain Damage or Dementia:

May be called Wernicke’s and Korsakoffs syndrome

- Possibly caused by or tightly related to neurotoxicity and/or Vitamin B1 and thiamine deficiency
- Common Symptoms:
  - Decreased ability to learn ‘new’
  - Decreased interest in valued activities, people, life
  - Impaired judgment and decision making
  - Emotional lability or apathy
  - Problems with balance and coordination
  - Problems with social control and behaviors
  - Problems with initiation and termination
A Few Alcohol-Related Brain Damage Resources that Might be Helpful:

- https://www.alzheimers.org.uk/about-dementia/types-dementia/alcohol-related-brain-damage
- https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3580328/
- https://vertavahealth.com/alcohol/dementia/
- https://www.verywellmind.com/alcohol-dementia-62980

Huntingtons Disease (Chorea):

- Genetically induced dementia, symptoms appear between 30-40 years of age; if younger when the disease begins it is referred to as Juvenile Huntingtons
- Average length of life is 15-20 years after first symptoms
- A neurodegenerative condition that has diffuse and pervasive impact throughout the brain, progressing and ultimately causing extreme challenges for the person to interact with the world or people around them in meaningful ways.
- Movement challenges: writhing, tremors, slow and abnormal eye movements, impaired gait and balance, difficulty with speech and swallowing
- Cognitive challenges: disorganization, not able to focus, perseveration, anosognosia, slowed processing, difficulty learning new, lacking impulse control for speech, action, or visual regard, limited decision-making skills
- Emotional challenges: depression, social withdrawal, apathy, irritability, insomnia, fatigue, thoughts of ending life, isolation, loss of energy, OCD, mania, bipolar disease
- Lack of appetite and weight loss as symptoms escalate along with pneumonia

A Few Huntingtons Resources that Might be Helpful:

- https://www.mayoclinic.org/diseases-conditions/huntingtons-disease/symptoms-causes/syc-20356117
- https://medlineplus.gov/genetics/condition/huntington-disease/
- https://rarediseases.org/rare-diseases/huntingtons-disease/
Multi-System Atrophy:
- Previously called Shy-Drager syndrome – either a Parkinsonian type shift or a cerebellar shift affecting younger people 40-60 years of age (MSA-P or MSA-C)
- A neurodegenerative condition that has a primary early feature of problems in basic homeostasis dysfunction
- Blood pressure: both hypotension upon standing and hypertension when supine – can be life threatening or resolve on its own
- Sweating issues with cold extremities – Sighing or gasping
- Fainting episodes – Sexual dysfunction
- Episodes of fixed off-center postures (Pisa)
- Respiratory patterns – Bladder-bowel control
- Sleep problems – waking dreams – Blood sugar control
- Cardiovascular issues – Motor control losses
- Emotional control issues
- Typically swallowing problems and severely impaired mobility are primary challenges combined with loss of communication

A Few MSA Resources that Might be Helpful:
- [https://www.multiplesystematrophy.org/about-msa/](https://www.multiplesystematrophy.org/about-msa/)
- [https://my.clevelandclinic.org/health/diseases/17250-multiple-system-atrophy](https://my.clevelandclinic.org/health/diseases/17250-multiple-system-atrophy)

Post Cortical Atrophy:
- Primary initial symptom is declining visual abilities – age of onset is typically 50-65 years of age – Bensons disease
- It can have more alpha-synuclein malformations, tau pathologies, or beta-amyloid abnormalities – LBD, Alzheimer, or cortico-basal patterns can emerge
- Visual spatial and object recognition abnormalities
- Apraxia: motor planning issues
- Trouble recognizing faces
- Visual perceptual changes
- Loss of wayfinding skills, environmental disorientation
- Reading and writing skill decline
- Eye-hand coordination changes
- Inability to use numbers effectively
- Challenges with getting eye muscles to coordinate
A Few PCA Resources that Might be Helpful:
- [https://www.raredementiasupport.org/posterior-cortical-atrophy/](https://www.raredementiasupport.org/posterior-cortical-atrophy/)
- [https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3740271/](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3740271/)
- [https://www.orpha.net/data/patho/Pro/en/PosteriorCorticalAtrophy-FRenPro10748.pdf](https://www.orpha.net/data/patho/Pro/en/PosteriorCorticalAtrophy-FRenPro10748.pdf)

Why Bother Getting a Good/Complete Diagnosis?
- Future plans
- Progression and prognosis
- Finances
- Health
- Being in control while staying connected in relationships
- Skillful use and careful management of medications can make a difference in quality of life

Building Care Partner Skills and Knowledge:
- Understand dementia and its progression
- Know how symptoms affect behavior
- Describe needs connected to behavior
- Optimize interaction skills
How Can We Become Better Care Partners?

Be willing to try something new
Be willing to learn something different
Be willing to see it through another’s eyes
Be willing to fail and try again