

**WHEN TRADITIONAL CARE FALLS SHORT:  
CARING FOR PEOPLE WITH ATYPICAL PRESENTATIONS OF CORTICAL DEMENTIA**

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**Introduction**

About four to six million Americans suffer from dementing illness. While the majority will be diagnosed with Alzheimer's disease and/or vascular dementia, diagnostic specificity for dementing illness may vary with a number of factors including perception and training of the diagnostician, age and location of the patient, symptom presentation, secondary symptoms, presence of comorbidities, sophistication of the family, and probably the third party payer. Additionally, there are concerns that the current emphasis on health care cost containment will reduce measures assuring diagnostic specificity to a simple dichotomy of determining which forms of dementia are reversible.

Once the diagnosis of an irreversible dementing illness is established, responsibility for planning and providing daily care is assumed by family and allied health professionals. Over the past 10-15 years, enormous strides have been made in both the understanding of pathophysiology of chronic dementing illnesses and the development of research-based techniques for providing care to victims of Alzheimer's disease and related disorders (ADRD) and their families. Yet, while neuroscientists have identified multiple variants of dementing illnesses, care improvements still focus primarily on typical presentations of Alzheimer's disease, assuming people with dementia have similar symptoms and therefore, similar care needs. Care plans, whether in special care units, assisted living programs, day care, or in-home care utilize a set of known needs and traditional beliefs about the caring for persons with Alzheimer's disease. Thus, care programming for people with dementia has become essentially a "one size fits all" strategy. Individualization is usually accomplished by identifying the person's culture, past interests, and preferences, and integrating them into the generic Alzheimer's care plan or with specific treatments for problematic secondary symptoms such as wandering or agitation.

This approach may be used successfully with most patients with dementia of the Alzheimer's type, however it often falls short with patients presenting with atypical presentations, rare, or multifactorial dementias -- perhaps as many as 20% of the patient population with irreversible dementias. While atypical cortical degenerative syndromes are described in the medical literature, there is little in the allied health literature aimed at caregiving professions. In fact, to date there is no research to date on specific needs of people with atypical cortical decline -- probably due to the relatively small number of subjects available for study in any given area and the lack of diagnostic specificity previously discussed.

The lack of information on atypical dementing syndromes in the caregiving literature is a paradox, as caregiving professions need this information for appropriate day-to-day decision-making. The purpose of this article is to describe characteristics of several common atypical syndromes of cortical degeneration and to suggest how those presentations might be accommodated in care provision. However, research is needed to validate the efficacy of the interventions suggested.

**Cortical Degenerative Syndromes**

Identifying atypical presentations, whether histologically linked to Alzheimer's disease or not, can only help practitioners and caregivers to understand observed behaviors, distinguish between primary and secondary symptoms, and plan care accordingly. For purposes of this article, several categories of atypical cortical dementias are discussed including progressive aphasia, perceptual-motor syndromes, frontal degenerative syndromes, and bitemporal syndromes. Because of its initial presentation with symptoms of

cortical decline, Diffuse Lewy Body Syndrome is often mistakenly diagnosed as Alzheimer's disease. Therefore, this syndrome will also be addressed.

Due to the volume and complexity of atypical cortical degenerative syndromes, a care table and glossary will be used at the end of the article for comparison and easier use by caregiving professionals. The format for the table uses core syndromes, clinical presentation, associated problems, and cortical structures involved (Caselli, 1995). Care issues have been added to assist caregiving professionals and to stimulate ideas for intervention research.

### **Care Issues with Atypical Dementias**

Care associated with dementias of the Alzheimer's type currently focuses on developing routines of meaningful activities that fall within the patient's range of abilities, and preventing excess disability (Baum, 1993; Hall, 1994). Excess disability is defined as reversible functional loss and secondary behavioral symptoms triggered by one or more of the following: 1) fatigue; 2) change in routine, environment, or caregiver; 3) affective responses to perceived losses; 4) inappropriate or excessive auditory and visual stimuli; 5) internal or external demands to achieve that exceed functional capacity; 6) not using previously learned skills (disuse); and 7) physical stressors including illness; medication reactions, pain or discomfort, or changes causing delirium (Baum, 1993; Hall, 1994). Care planning includes reducing environmental stimuli to a comfortable level; providing opportunities for adequate rest during the day; providing a calm, consistent, structured sequence of activities; providing appropriate levels of sensory input; developing meaningful programs of socialization and activities based on past interests; limiting demands to achieve to those the patient can realistically manage; and maintaining an optimal state of physical well-being that is free from pain (Baum, 1993; Hall & Buckwalter, 1987)

Care of the person with an atypical cortical degenerative syndrome may be modified to accommodate subtle differences in disease presentation or an entirely new strategy may have to be developed--especially early in the disease. Caregiving professionals need to understand both the functions of the areas of the cortex affected and how these areas influence symptom presentation in order to make decisions about care. Five types of atypical presentations and implications for planning care will be addressed, however, readers must remember that none of the implications/interventions set forth have been empirically validated.

#### **Progressive Aphasia**

Five types of progressive aphasia have been identified: non-fluent; fluent; anomic; mixed; and aphasia associated with motor neuron disease (Caselli, 1995; Caselli, Windebank, Petersen, et al, 1993). Each type differs in the following areas: patient understanding; type of language deficit; ability to comprehend written materials; write; and name things. Careful assessments by a neurologist and ongoing speech pathology consultations can assist caregivers with defining exact losses and identifying appropriate compensatory interventions.

Non-fluent aphasia is characterized by halting, effortful, labored speech production, whether expressing original ideas or repeating. The patient may still be able to read and visual memory is generally preserved. Patients with non-fluent aphasia may, therefore be safe to live alone. The non-fluent aphasic may be able to use language boards, lists, and written materials. They can generally respond to pictures and signs.

The person with fluent aphasia can produce language, but it is flawed. Comprehension may be impaired and the patient often makes paraphasic errors (e.g., calling a couch a "frouch"). The person is unable to recognize and describe things, but they can repeat what is said to them. Patients with fluent aphasia generally have severe language deficits with preserved cognition for about two years of an eight year disease course (Caselli, 1995).

Anomia is a pure localized inability to name. Probably the two most troubling progressive aphasias are the mixed type, where the patient demonstrates impairments in all aspects of language, and the aphasia associated with motor neuron disease.

Motor neuron disease usually advances rapidly and is associated with dysarthria and dysphagia. If diagnosed early the patient may be trained in use of alternative communication devices, such as a light writer. Swallowing function and management of secretions must be closely monitored with the increasing

danger of aspiration. The patient and family need to be counseled regarding advanced directives for alternatives to oral feeding.

Patients with progressive aphasias generally may be able to maintain daily function longer than people with global cognitive decline, although they will need assistance with emergency planning and communication aids to live independently. Moreover, because their level of comprehension may be high, patients with progressive aphasia should be evaluated for depression and, if present, treated in order to prevent excess disability and maintain optimum function.

### Perceptual-Motor Syndromes

Three perceptual-motor syndromes have been described (Caselli, 1995). The first two are visual syndromes. Asimultanagnosia limits the patient's ability to perceive a common scene as a whole. While able to understand bits and pieces of their environment, these patients are unable to comprehend "the big picture." The second, visual agnosia, is a failure to recognize objects and, more commonly, people. The visual perceptual deficits have been well documented by Sacks (1984) who described a case of a music professor who mistook his wife's hand for a hat.

When working with patients with asimultanagnosia, the caregiver must provide continuous verbal input, describing actions and the environment. Patients often learn to rely heavily on other sensory modalities including hearing, touch, and smell to function in their environment (Sacks, 1985). Caregivers would want to provide distinct identifying odors and tactile textures to areas of the environment to enable recognition. However, extraneous environmental stimuli, such as large crowds and high noise levels, could potentially add to the person's disability by taxing remaining senses. Sacks (1985) suggests having carers wear a single identifying object, such as a brightly colored hat or flower, to assist the patient with caregiver recognition.

Patients with visual agnosia do not recognize day-to-day objects. They must be supervised and assisted when using objects, such as feeding utensils inappropriately. It is not uncommon to see people with visual agnosia put ketchup on their cereal instead of milk or request help to find something while appearing to gaze directly at the object.

The two visual syndromes may confuse direct care providers. While the patient is able to "see," vision is often not functionally useful. Having the patient describe what he/she is seeing helps train direct caregivers. For example, a farmer explained that he saw tractors and trees piled atop of one another. While he recognized the information he was receiving was impaired, he was unable to develop compensatory strategies to enhance function. Ongoing work with occupational therapists and neuropsychological testing can help care-providers to modify the environment to meet the patient's needs. Services for people who are visually impaired may be helpful for these families. Caregivers must also remain alert for signs and symptoms of auditory and additional sensory deficits that may accompany this syndrome.

The third group of perceptual-motor syndromes is progressive motor syndromes. Patients develop inexorably progressive hemiparesis, hemipastically, and disabling apraxias. One of the more "common" of these syndromes is corticobasal ganglionic degeneration (CBGD). Over time people with CBGD become increasingly disabled with spasticity and loss of motor planning for purposeful movement. They can develop cramping and contractures. Visual loss may result from impaired eye movement. Speech and swallowing difficulties are common problems. Patients with CBGD have impairments in performance on neuropsychological testing however, insight into their condition remains and may produce depression or acute anxiety.

With severe apraxias and hemiparesis, occupational and physical therapies can be utilized to maintain function. Occupational therapists assist by assessing levels of function using task analyses and training caregivers to assist patients with tasks by breaking them into progressively simpler components the patient should be able to perform. Physical therapists assist families by demonstrating passive and active range of motion exercises, positioning, measures to manage spasticity and maximize function in affected extremities. Patients also report increased comfort with massage therapy.

Scrupulous skin care is required to maintain skin integrity and prevent breakdown over bony prominences, in contracted extremities, and in areas exposed to secretions or excreta. Botox injections have been used to release contracted hands, allowing for hygiene when there is danger of skin breakdown.

Accommodations for loss of coordinated eye movements can be accomplished by providing care/activities slowly and simplifying the environment. The slowed purposeful movement accompanied by verbal description can help patients gather more “perceptual information” as they try to focus on activities. Caregivers should try to stand directly in front of person and place items within the field of vision. Consistent calm caregivers provide reassurance and continuity when the patient is aware of continuing loss of control over their environment. In the final stages of motor syndromes, the patient may become immobile and mute. Difficulties with swallowing, coughing, and excretory functions occur in late stage disease. As with any degenerative condition, it is important to determine patient care preferences using an advanced directive prior to the onset of aphasia. Patients must be observed for aspiration pneumonia, constipation, urinary retention, and skin breakdown. Bowel and bladder regimens, feeding techniques, range of motion exercises, skin care, and care for immobility must be taught to caregivers.

At the end of the disease, the patient becomes unable to chew or swallow. Rapid weight loss and weight loss ensues. Providing high calorie, protein-rich, vitamin-enriched soft foods such as custards can help to maintain weight and nutritional status. A multiple vitamin syrup may also be given. Swallowing studies can determine optimal food consistencies to prevent aspiration. Use of an enteral feeding system may be selected to minimize the danger of aspiration pneumonia. Hospice care is an excellent alternative for care during terminal stages as personnel are trained to manage pain, secretions, fear, and help with family support.

#### Progressive Frontal Lobe Syndromes(FLS)

Patients suffering from Pick’s disease and other frontal lobe syndromes often provide some of the greatest caregiving challenges encountered. In very early disease, people developing FLS often appear quite normal and rational yet caregivers note changes in behavior patterns and judgment. This produces conflict and crises among family members (Caselli, 1995; Salmon, Degueldre, Franco, & Franck, 1996).

While some medical literature describes patients with FLS in passive terms, in a care setting such as the home, day care center, or nursing home, these patients often have little insight into their limitations, make poor choices, and can be disruptive, posing risks to themselves and others (Barber, Snowdon, & Crauford, 1995; Caselli, 1995). Many are disinhibited and act on impulse seemingly without reason. Patients with damage to prefrontal areas may verbalize risks and alternatives, yet consistently make poor decisions (Damasio, 1995). The greatest concern with FLS is maintaining safety both of patients and those around them.

The patient with FLS may refuse to stop driving, make unusually large purchases, use power tools inappropriately, have social outbursts, or eat non-food items such as glass ornaments. Their lack of insight and disinhibition may produce inconsistent behavior despite making verbally appropriate statements, constantly catching caregivers off guard.

A structured environment whereby the daily schedule, the level of noise, extraneous stimuli, and potential hazards can be controlled is one way to decrease the need for one to one supervision for patients with FLS. Yet, the FLS patient may not benefit from the structured program in the same way as the person with dementia of an Alzheimer’s type, meaning outcomes of appearing calm, interacting with others appropriately, sleeping through the night, or participating in activities will not be reached. A structured environment will, however, decrease the need for patient judgment, eliminate many potential safety hazards, enhance the likelihood of bathing and meeting basic needs. Moreover, structured settings often offer specialized training to employees about FLS, whereby the patient will receive unconditional positive regard.

#### Bitemporal Syndromes

Bitemporal degeneration is characterized by severe progressive amnesia (Caselli, 1995). Patients differ from those with Alzheimer’s disease in that they lack other areas of cognitive impairment. In a case described by Sacks (1985), a man who appeared to converse and reason in a perfectly normal manner totally forgot the encounter after the neurologist left the room. Sacks describes the man as doomed to repeat the same encounter over and over. Moreover, because the amnesia was so complete, it was impossible to convince the patient of his deficit.

The patient did not comprehend the passage of time, so he remained “frozen” at age 23. He could not believe that ages of family members increased, thereby arguing over their claims to be his relatives. Because of the severity of the amnesic syndrome, the man could never live independently.

Nursing care of people with amnesic syndrome includes making sure they wear identification as they appear and sound normal. In early disease, the patient may benefit from concrete memory aids such as calendars (marked off by others), schedules, and clocks. Later, careful supervision is required as the patient may become lost in all but the most overlearned environments. Direction and supervision would be required for completion of basic daily activities as the patient would not know if tasks had not been completed. Moreover, the patient’s lack of insight into deficits challenges caregivers to evaluate all statements made by the patient -- especially answers to questions requiring any memory at all such as “Did you have lunch?”

An environment offering twenty-four hour supervision would be required to assure safety and provide the necessary structure. Family and caregiving staff would require extensive ongoing training as the patient appears and sounds perfectly lucid. Moreover, the patient might be vulnerable to dependent adult abuse as caregivers might perceive his/her actions to be purposeful.

### Diffuse Lewy Body Variant (DLBV)

Although its relationship to Alzheimer’s disease is the subject of debate, DLBV is thought to be the second most common dementia (Kalra, Bergeron, & Lang, 1996). Often misdiagnosed, DLBV is mistaken for Alzheimer’s disease with Parkinsonian features, Alzheimer’s disease with psychosis, or Parkinson’s disease with Alzheimer’s disease (Hansen & Galasko, 1992). The early course of the disease closely resembles Alzheimer’s disease, but with progression the character of the symptoms change to include Parkinsonian features which can include bradykinesia, rigidity, or tremor. Psychotic symptoms appear in about half of cases (Robles, Rodriguez, Aldrey, et al, 1995). The early psychotic symptoms may include paranoid delusions, visual and auditory hallucinations, and illusions accompanied by dementia (Kalra, Bergeron, & Lang, 1996; McKeith, Perry, Fairbairn, et al, 1992). These may escalate until the patient becomes quite violent and/or may lose touch with reality altogether. Studies of DLBV are small and often behavioral findings are conflicting, but there have been no differences between Alzheimer’s disease and DLBV reported for severity of cognitive decline, or duration of illness (Forstl, Burns, Luthert, et al, 1993).

Using medications to control Parkinsonian features and/or to manage psychotic features is difficult due to competing side effects and increased potential for sensitivity to adverse effects of antipsychotics. Anti-Parkinson’s medications are strongly anticholinergic, producing increased confusion and potentiating psychosis, while many antipsychotics produce dopamine blockade worsening symptoms of Parkinson’s disease. Moreover, increased susceptibility to neuroleptic malignant syndrome has been noted in people with DLBV and one study reported a 50% reduction in survival time when neuroleptic medications were used (Kalra, Bergeron, and Lang, 1996; Kosaka, 1993). For the patient with DLBV, medications should be used as a last resort. When seeking medications, primary consideration must be given to maintaining safety of both patients and those around them. This may mean focusing and providing dopamine only to patients who are immobile, or antipsychotics only to those who are violent due to psychotic features. There are, however, some anecdotal reports of use of acetylcholinesterase inhibitors (Donepezil) in patients with DLBV, suggesting improved movement and cognition with no increase in psychotic symptoms.

Nursing care of the patients with DLBV consists first, of encouraging families to seek appropriate neurological diagnosis and follow-up. Patients with DLBV additionally may be followed by a psychiatrist. These patients require a multidisciplinary approach and, often, several medical subspecialties working in concert.

Much of the care focuses on helping the family to understand the disease process, providing for basic physical care, and safety. Patients with DLBV often behave far differently than those with Alzheimer’s disease or vascular dementias. Families are often in conflict about behavioral presentations as the caregiver may become quite fearful of the patient. Moreover, relatives living long distances from the patient may express disbelief at caregiver reports of psychotic or aggressive behaviors.

People with DLBV are at high risk for falls and injury to themselves from decreased mobility during simple tasks including eating and drinking. Exercise and range of motion activities may help maintain mobility. A quiet structured environment with few misleading stimuli may help to minimize hallucinations

and combative behaviors. Avoidance of fatigue, caffeine can assist with stability of mood. Yet, despite best intentions, these patients may become psychotic.

Management of early disease includes careful assessment of psychotic behaviors including descriptions of presenting symptoms; numbers, duration, and timing of episodes; antecedents (triggers) to episodes; and measures that provide relief. Safety hazards must be removed and documented. Even though the patient may be a resident of a long term care setting, careful documentation must be achieved for this patient to both plan care and measure outcomes. A primary care approach to staffing using psychiatric consultation, ongoing consultation from psychiatric nurse specialists, and supervision by licensed nurses is desirable. Supervisory personnel must evaluate direct caregiving staff for fatigue and burnout that might potentiate employee injury or patient abuse. If the person with DLBV becomes unmanageably violent, placement in a long term psychiatric institution where staff are specially trained in management of assault may be required.

### **Summary/Conclusions**

Atypical presentations of cortical dementias pose enormous challenges for allied health professionals and caregivers. Presented with patients whose behavioral symptoms do not fall within the expected range of Alzheimer's disease and related vascular dementias, professionals are expected to develop programs of care to meet the patient's individual needs. Understanding that atypical presentations may represent asymmetric cortical degeneration or DLBV, the caregiving professional can request a comprehensive diagnostic assessment and develop specific care strategies based on pathophysiology. Unfortunately, there are relatively few of each of the above patients in a given setting thereby limiting the potential for testing interventions in randomized controlled trials. The limited generalizeability of findings would be consistent with the conflicting results from studies of disease incidence and presentation as demonstrated by descriptive studies of DLBV.

In order to overcome this, researchers must utilize innovative designs to validate interventions for people with atypical presentations of dementia. This might include case study research, insisting subjects meet rigorous diagnostic standards for inclusion in large samples, which are then controlled for these subjects, multi-site intervention studies, and replicating single case or intervention studies using small samples.

In the meantime, nurses must be sensitive that varying presentations of dementia are due not only to personality, lifelong preferences, and cultural groupings. We must begin to incorporate developments in biological research and include pathophysiology when planning and evaluating care. One size does not fit all in caring for people with cognitive impairment.

## References

- Barber, R., Snowden, J.S., & Crauford, D. (1995). Frontotemporal dementia and Alzheimer's disease: retrospective differentiation using information from informants. Journal of Neurology, Neurosurgery, & Psychiatry. 59(1), 61-70.
- Baum, C. (1993). The effects of occupation on behaviors of persons with senile dementia of the Alzheimer's type and their carers. Doctoral dissertation. St. Louis: Graduate School of Arts and Sciences, Washington University.
- Caselli, R.J. (1995). Focal and asymmetric cortical degenerative syndromes. The Neurologist. 1(1), 1-19.
- Caselli, R.J., Jack, C.R., Petersen, R.C., Wahner, H.W., & Yanagihara, T. (1992). Asymmetric cortical degenerative syndromes: Clinical and radiologic correlations. Neurology. 42: 1462-8.
- Caselli, R.J., Windebank, A., Petersen, R., Komori, T., Parisi, J., Okazaka, H., Kokmen, E., Iverson, R., Dinapoli, R., Graff-Radford, N., & Stein, S. (1993). Rapidly progressive aphasic dementia and motor neuron disease. Annals of Neurology. 33 (2), 200-207.
- Damasio, A. (1995). Descartes' Error. New York: Grosset/Putman.
- Forstl, H., Burns, A., Luthert, P., Cairns, N., & Levy, R. (1993). The Lewy-body variant of Alzheimer's disease. Clinical and pathological findings. British Journal of Psychiatry. 162: 385-92.
- Hall, G.R. (1994). Managing people with Alzheimer's disease using the Progressively Lowered Stress Threshold conceptual model. Nursing Clinics of North America. 29(1), 129-142.
- Hall, G.R., & Buckwalter, K.C., (1987). A conceptual model for planning and evaluating care of the client with Alzheimer's disease. Archives of Psychiatric Nursing. 1(6), 399-406.
- Hansen, L., & Galasko, D. (1992). Lewy body disease. Current Opinion in Neurology and Neurosurgery. 5(6), 889-94.
- Harrington, C.R., Perry, R. H., Perry, E.K., Hurt, J., McKeith, I.G., Roth, M., & Wischik, C.M. (1994). Senile dementia of Lewy body type and Alzheimer type are biochemically distinct in terms of paired helical filaments and hyperphosphorylated tau protein. Dementia. 5(5), 215-28.
- Ishii, K., Mori, E., Kitagaki, H., Sakamoto, S., Yamaji, S., Imamura, T., Ikejiri, Y., & Kono, M. (1996). The clinical utility of visual evaluation of scintigraphic perfusion patterns for Alzheimer's disease using I-123 IMP SPECHT. Clinical Nuclear Medicine. 21(2), 106-10..
- Kalra, S., Bergeron, C., & Lang, A. E. (1996). Lewy body disease and dementia: A review. Archives of Internal Medicine. 156(5): 487-93.
- Kosaka, K. (1993). Dementia and neuropathology in Lewy body disease. Advances in Neurology. 60: 456-63.
- McKeith, I.G., Perry, R.H., Fairbairn, A.F., Jabeen, S., Perry, E.K., (1992). Operational criteria for senile dementia of Lewy body type (SDLT). Psychological Medicine. 22(4), 911-22.
- McShane, R., Gedling, K., Reading, M., McDonald, B., Esiri, M., & Hope, T. (1995). Prospective study of relations between cortical Lewy bodies, poor eyesight, and hallucinations in Alzheimer's disease. Journal of Neurology, Neurosurgery, & Psychiatry. 59(2), 185-8.
- Robles, A., Rodriguez, R.M., Aldrey, J.M., Vadillo, J., Suarez-Gil, P., Lema, M., & Noya, M. (1995). Clinical diagnosis of dementia associated with cortical Lewy bodies (abstract). Revista de Neurologia. 23(119), 62.
- Sacks, O. (1985). The Man Who Mistook his Wife for a Hat and Other Clinical Tales. New York: Harper Perennial.
- Salmon, E., Degueldre, G., Franco, G., & Franck, G. (1996). Frontal lobe dementia presenting as personality disorder. Acta Neurologica Belgica. 96(2), 130-4.
- Whitehouse, P., Tabaton, M., & Lanska, D., (1991). Pathological and chemical correlates of dementia. In F. Corkin (Ed.), Handbook of Neuropsychology and Aging (Vol. 5, pp. 29-37). New York. Elsevier.



instead of naming object, Semantic paraphasia are common and phonemic paraphasia occur as well.

Able to repeat sentences.

Relatively severe language deficits compared to other areas of cognitive function.

constructional apraxia, and apraxic agraphia.

(Caselli, 1995)

point to objects or requested items.

If patient still at home, Lifeline and emergency planning as patient may not convey information to emergency persons.

| <b>Syndromes</b> | <b>Sub-categories within Syndrome</b> | <b>Core Syndrome</b>  | <b>Associated Problems</b>   | <b>Associated anatomic Structures</b>  | <b>Implications for Care</b>   |
|------------------|---------------------------------------|---|--|--|--|
|                  | Anomic aphasia                        | Pure localized anomia. Able to read, comprehend well, repeat, and write. Speech lacks semantic precision. Generally unaccompanied by dementia | May be accompanied by frontal lobe signs<br>May develop impaired verbal memory with disease progression. | Left anterior temporal lobe may be associated with Pick's disease<br><br>(Caselli, 1995) | Written cues including lists and signs may be effective in prompting behaviors. Because of relationships to frontal lobe dementias, risks to safety from disinhibition must be assessed. Staff should wear identification.<br><br>Educate staff on extent of deficit |
|                  | Mixed                                 | Impairment of all aspects of language. Nonfluent with impaired comprehension, naming, repetition, writing, and reading                        |  | Temporal and perisylvian cortices<br><br>(Caselli, 1995)                                 | Speech pathology consultation periodically to determine extent of receptive and expressive aphasia and level of cognitive comprehension.   |
|                  | Association with motor neuron         | Progressive aphasia and dysarthria which can be   | Mild limb symptoms.  |  | Encourage to speak slowly, provide   |

disease

an early sign of motor neuron disease that often progresses to anarthria and death within two years

Some evidence of global dementia but less severe than aphasia

(Caselli, 1995)

additional time for response. Periodic swallowing evaluations to minimize potential for aspiration. Early assessment for alternative communication devices. Advanced directives regarding feeding.

| <b>Syndromes</b> | <b>Sub-categories within Syndrome</b> | <b>Core Syndrome</b>  | <b>Associated Problems</b>  | <b>Associated anatomic Structures</b>  | <b>Implications for Care</b>   |
|------------------|---------------------------------------|---|---|--|--|
| Perceptual-motor | Asimultanagnosia                      | Complex visual disorder in which patients can not integrate the numerous components of an ordinarily complex scene into a coherent whole. | Ocular apraxia - inability to voluntarily direct their gaze to a target of visual interest; and optic ataxia, the inability to benefit from visual guidance in reaching for an object.<br>Balint's Syndrome:<br>asimultanagnosia + ocular apraxia + optic ataxia<br>May also exhibit alexia, acalculia, right-left disorientation, and mild deficits of language and memory | Parieto-occipital<br>Bilateral dysfunction of dorsal cortical visual pathways in occipitoparietal association cortices.<br><br>(Caselli, 1995) | Limit visual input and extraneous stimuli: housing and feeding people in smaller areas; remove clutter; avoid potentially misleading visual stimuli such as TV..<br>Have staff provide verbal input whenever entering a room and offer simple explanations of basic procedures.<br>Have each staff wear a single item to provide a distinguishing characteristic such as a brightly colored hat or a flower (Sacks, 1984). |
|                  | Visual agnosia                        | Progressive prosopagnosia (the inability to recognize faces)  | May develop frontal degeneration with motor   | Parietotemporal<br>bilateral dysfunction of  | Have familiar people identify themselves entering a room.<br>Provide much verbal   |





preserved  
consciousness.

neglect. Immobility  
and contracture care.  
Swallowing function  
studies and  
nutritional support.  
Continuing to have  
patient participate in  
decision-making  
Advanced directives.

| <b>Syndromes</b>                   | <b>Sub-categories within Syndrome</b>   | <b>Core Syndrome</b>   | <b>Associated Problems</b>   | <b>Associated anatomic Structures</b>   | <b>Implications for Care</b>  |
|------------------------------------|---|--|--|---|---|
| Progressive frontal lobe syndromes | Neuropsychiatric syndromes: (Pick's disease, frontal lobe dementias, frontal lobe degeneration, or dementia of the frontal lobe type) | Abulia: functional errors of omissions: failing to pursue former interests, or initiate daily activities. May perseverate or fixate on specific activities such as watching TV, sorting, or eating. May appear depressed due to lack of interest and emotional lability, yet no overt signs of unhappiness. Inertia may prevent from meeting basic human needs such as hunger. Memory and language are often impaired, but patient may perform normally on mental status tests (exasperated spouse complains of problems no one else recognizes). Answer questions quickly, briefly, with "I don't know." May not argue when family asserts they | May develop impaired motor control with spasticity and/or impaired language. May also affect temporal lobes (Pick's disease). Syndrome is usually bilateral but often affects one side more predominantly. If dominant side may have aphasia -- especially anomia. | Frontal lobe: bilateral damage, but worse on one side<br>Pick's disease: frontal-temporal lobes | Structured environment cueing necessary activities. Anticipate hunger, thirst, and other physiologic needs. Continuous assessment of safety: ongoing evaluation of risks posed by decreased judgment, lack of insight, impaired executive function, and disinhibition. Teach caregivers implications of disinhibition and need to be constantly aware of safety needs.<br><br>Be aware of eating habits – especially non-food items. Assess impact of poor hygiene and antisocial behavior such as smearing |

are demented, yet generally lack insight into symptoms. May have disinhibited disruptive symptoms. unusual speech patterns such as echolalia patterns. Loss of executive function.

(Caselli, 1995)

feces in later disease. Caregiver support is especially important in early FLS. Early symptoms are vague and not reflected in neuropsych testing

| <b>Syndromes</b> | <b>Sub-categories within Syndrome</b>             | <b>Core Syndrome</b>   | <b>Associated Problems</b>  | <b>Associated anatomic Structures</b>                  | <b>Implications for Care</b>   |
|------------------|---|--|---|--|--|
|                  | Progressive spasticity: primary lateral sclerosis | Slowly progressive spasticity which is generally asymmetrical. Usually starts in lower limbs, but occasionally presents in arms or with dysarthria. Emotional lability without cognitive impairment. Severe spastic dysarthria is all patients progressing to anarthria. | Urinary incontinence (50%) and saccadic breakdown of smooth pursuit eye movements | Atrophy of the precentral gyrus<br><br>(Caselli, 1995) | Assess lability versus depression. Passive and active range of motion to prevent contractures. Periodic swallowing studies to minimize potential for aspiration<br>Prompted voiding program<br>Slowly paced activities to compensate for decreased visual processing<br>Patient may benefit from anti-convulsant or antispasmodic medications. |
| Bitemporal       | Progressive amnesia                               | Progressive severe amnesia without other cognitive impairment<br><br>(Caselli, 1995)   | May also be subtle psychotic features   |  | Use of written and visual memory cues, including lists, clocks, calendars, pocket reminder cards<br>“Medic-Alert   |

Bracelet”

| <b>Syndromes</b>          | <b>Sub-categories within Syndrome</b> | <b>Core Syndrome</b>  | <b>Associated Problems</b>   | <b>Associated anatomic Structures</b>                    | <b>Implications for Care</b>   |
|---------------------------|---------------------------------------|---|--|--|--|
| Diffuse Lewy Body Variant | Neurobehavioral syndrome              | Day to day fluctuations in mental state.<br>Psychosis: auditory and visual hallucinations, delusions, and paranoid ideation's with poor response to neuroleptic medications.<br>Generalized dementia followed by Parkinsonism of the rigid bradykinetic type. | May co-exist with Alzheimer's disease.<br>Severity of hallucinations positively correlated with poor eyesight.<br>(McShane, et al, 1995) | Neocortex, Cingulate cortex<br>(Harrington, et al, 1994) | Patients often present with florid psychosis. Psychosis, dementia, and movement disorders may respond well to acetylcholinesterase inhibitors.<br>Low stimulus environment free from large groups, noise, distractions, and potentially misleading stimuli.<br>Techniques to prevent injury either to patient or those around patient. Have patient hold objects during cares. Careful medical management of Parkinsonian features versus psychosis. In some cases, avoidance of anti-Parkinsonian medications. Careful monitoring of effects of neuroleptic |

medications.  
Maximizing visual acuity by having patient wear their glasses. One to one activities and cares, whenever possible. Evaluate speech content for clues to discern meaning. Family education and enhanced support. Liaison with long term care and psychiatric services to provide adequate support in non-psychiatric settings. Staff inservices to explain behavior.

## **Glossary of Terms**

Abulia - Functional errors of omission: failing to perform activities to meet basic human needs; inability to make decisions; lack of will or willpower.

Acalculia - inability to do simple arithmetical calculations

Anomia - inability to recall or recognize names of objects

Aphasia - loss of power of expression by speech, writing, or signs and/or loss of comprehension of spoken language or written language due to brain injury or pathology

Apraxia - loss of ability to carry out familiar, purposeful movements in the absence of paralysis or other motor or sensory impairments, especially the inability to make proper use of an object

Apraxic agraphia - inability to express oneself in writing due to apraxia

Asimultanagnosia - Inability to visually integrate the components of ordinarily complex scene into a coherent whole

Aural comprehension - understanding of stimuli perceived by the ear

Constructional praxis - inability to copy simple drawings or reproduce patterns of blocks or matchstick constructions

Dysarthria - imperfect articulation of speech due to muscular weakness resulting from damage to the central or peripheral nervous system

Echolalia - stereotyped repetition of another person's words or phrases

Executive function - ability to set a goal, make decisions, and implement appropriate activities towards meeting that goal.

Ocular apraxia - inability to voluntarily direct their gaze to a target of visual interest

Optic ataxia - the inability to benefit from visual guidance in reaching for an object

Paraphasia - speech defect characterized by disorderly arrangement of spoken words

Phonemic - speech sounds that are the basic units of speech (i.e. "leviator" instead of "elevator;" or "grontologs" instead of "gerontology")

Praxis - the performance of an action; "doing"

Proposagnosia - inability to recognize familiar faces

Prosody - the variations in stress, pitch, and rhythms of speech that convey meanings

Prosopagnosia - inability to recognize faces

Semantic paraphasia - substituting a similar word for an object, i.e., “staple” for “paper clip” (Caselli, 1995, p. 3)

Semantic precision - use of words appropriate or significant to the meaning of the intended communication. i.e. substituting “machine” for “automobile”.

Verbal memory - ability to remember speech

Visual memory - Ability to remember what is seen