

## Review Article

## Clinical Practice after Diagnosis of Different Dementias

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## Abstract

As the estimated 5.4 million people in the U.S. with Alzheimer's disease or a related dementia (ADRD) approach a predicted 16 million by 2050, careful diagnosis, treatments, and family care management become more critical than ever. While health providers educate patients and family caregivers about diagnosis and treatments for progressive dementia, important strategies that address the family's upcoming lifestyle changes in their lifestyle, job or home routines, driving safety, financial and legal matters, and linkage to important community and internet resources may not be covered. Not only are health and behavior changes of progressive dementia important to know for long-term care planning and management, but also understanding how different types of dementia impact different abilities over time is critical for timing and prioritizing steps for care planning and management over the long term. The varied decline and challenges of different progressive dementias mandates a strong role for the health provider and health team to guide the family over the long-term of clinical care. This article overviews briefly major categories of dementia (Mild Cognitive Impairment, Alzheimer's disease, vascular dementia, frontotemporal dementias, and Parkinson disease with dementia as well as parkinsonisms such as Lewy body dementia, progressive supranuclear palsy, and corticobasal degeneration) and provides a tool, a *Checklist on Family Matters*, for the clinician to offer the patient and family caregiver(s) to plan for the long-term care management.

**Keywords:** Diagnosis; Alzheimer's disease; Dementias; Mild cognitive impairment; Parkinson disease

## Introduction and Background

The estimated 5.4 million people in the U.S. with Alzheimer's disease or a related dementia (ADRD) receive more than 17.5 billion hours of care from about 15.4 million unpaid family and significant others; the unpaid care represents \$216 billion [1]. By 2050 the numbers of people with ADRD are expected to reach 16 million; the current estimated \$203 billion costs of direct care are predicted to reach \$1.2 trillion in 2050 [1]. These anticipated increases of a dementia-tsunami amplify the *silver-tsunami* [2] threatening the U.S. socio-economic-healthcare infrastructure.

As used here, *progressive dementia* is the ongoing decline of a person's memory and other mental abilities to the point of interfering with interactions and accomplishing daily tasks on the job or at home. Progressive dementias most often diagnosed as Alzheimer's disease (AD) though there are more than 100 different progressive dementias including vascular dementias, frontotemporal dementias, and Parkinson disease with dementia as well as parkinsonisms (such as Lewy body dementia, progressive supranuclear palsy, and corticobasal degeneration).

## The Problem

Early medical evaluation, diagnosis, intervention, treatments, guiding families about planning ahead for long-term care management are critical to reverse, stop, slow down, or manage treatable or progressive dementias [3]. After the comprehensive medical evaluation when the patient and family/significant other(s) learn of the clinical diagnosis of ADRD, they often feel overwhelmed, not only by the diagnosis, but also by their simultaneous anticipation

of the impending encumbrances of future decline. Foreseeing the long-term challenges of the disease, the family may experience a *family-tsunami*, feeling at a complete loss about the next steps to take, especially for less commonly known progressive dementias, such as Lewy body dementia, frontotemporal dementias, or a parkinsonism. While physicians may recommend treatments, they typically do not cover important strategies to address upcoming related lifestyle changes in job or household routines, driving safety, or financial and legal matters. Often the patient or family is aware of changes from Alzheimer's disease, but naive about the possible health and behavior changes of other progressive dementias or how to prepare for future changes in their lives. The health provider should educate the patient, when appropriate, and the family caregiver(s) about how the specific diagnosed dementia will impact different abilities and offer challenges over time, important to the family's timing and prioritizing steps for long-term care planning and adaptations.

Addressing lifestyle issues early after the diagnosis allows the family time to learn about long-term care planning and strategies, communication with members of the extended family, anticipating stress-reactions from future lifestyle adjustments, and developing back-up plans to minimize the fallout from unexpected health or family crises. In families where the person with the diagnosis is the matriarch or patriarch carrying most of the responsibility for the family finances, legal matters, running a family-owned business, or making household decisions, the other family members may be unaware of the numerous tasks which should be gradually transferred to others. It may take time to identify which family members can help, are available, have good communication and care skills, and

which ones may not be as available, skilled, overwhelmed with health or personal problems, not reliable, or not trustworthy.

The health team (physician, nurse, social worker, therapists, etc.) should provide education about the diagnosed dementia, recommended medical treatments and care plan, and local as well as e-resources. The varied decline in different progressive dementias, [4] however, mandates expertise from the health providers. The following information briefly describes a clinical template for education and guidance for some different types of dementia: Mild Cognitive Impairment (which may not decline further to become a progressive dementia), Alzheimer's disease, vascular dementia, frontotemporal dementias, and Parkinson disease with dementia as well as parkinsonisms (that is, Lewy body dementia, progressive supranuclear palsy, and corticobasal degeneration). An outline, *Checklist on Family Matters*, (see Figure 1.) keys on points for the health team to overview as they guide the patient and family caregiver (or significant other) to plan ahead.

### Clinical Template: After the Diagnosis

#### Education, Treatment, Prioritizing, and Planning Ahead First, Education

A health care professional with special training and experience in progressive dementia should take time to explain the diagnosis resulting from the comprehensive medical evaluation and include three points:

- Highlight of the areas of brain function that remain strong
- Description of current and anticipated decline
- Compensatory strategies and available community and internet resources

In addition to answering questions from the patient and family, a health expert or health team should provide handouts, educational references, internet links, and referrals to local community resources that provide information, services, and support.

#### Second, Medical Treatment and Healthy Lifestyle Recommendations

While the physician-patient-family caregiver discussion should cover the many treatments such as cholinergics, serotonergics, and neuroprotective medicines to improve or slow decline of memory and other cognitive functions, it is important to treat and stabilize existing chronic health conditions such as hormone imbalance or cardiovascular problems, and stop anticholinergics such as antihistamines. The health team should encourage the patient and family to pursue a lifestyle that benefits brain health such as:

- Regular physical exercise such as walking, bicycling, jogging, or resistance training (30-40 minutes at a time for 5 days a week) [5-8]
- Daily brain exercise, such as crossword and jigsaw puzzles, word games, reading, arts and crafts, playing a musical instrument; discussing topics, news, or books with friends, etc [9].

- Good nutrition such as the Mediterranean diet; lots of fresh fruits and vegetables[9]
- Hydration –8 -10 glasses of water a day unless physician restricts fluids[9]
- Quality sleep[9]
- Reduced (negative) stress[9]
- No chemical abuse[9]
- Meaningful activities[9]
- Positive relationships[9]
- Following medical and dental care recommendations

### Third, Prioritizing

The health team should encourage the patient and family/significant other caregivers to think about priorities, values and interests. The patient should prioritize a Wish List and form plans to pursue wishes to: travel, visit family, join a new club, take a class, learn a new skill or refresh an old one, hold a reunion, etc. Independent or interdependent functioning to fulfill the Wishes List depend upon the patient's level of health, cognitive and physical functioning, and resources. The person with greater cognitive decline will be wise to partner with one or more loved ones or a trustworthy person to pursue the items on the Wish List.

### Fourth, Planning Ahead (see Figure 1. CHECKLIST ON FAMILY MATTERS)

Soon after a diagnosis of a progressive dementia, because financial, legal, and other family matters may require detailed discussions and decisions, the health team should share a guide such as the *Checklist on Family Matters* to allow enough time for the patient and family to deliberate and take action steps. The patient should identify preferences for diet, daily routine, and activities for current as well as future reference when abilities change, i.e., when personal assistance is needed in-home or at a community facility and later end-of-life care. These preferences should be discussed, written down, and shared with the health provider and, as appropriate, trusted loved ones.

The points on the *Checklist on Family Matters* may take a few weeks or months to consider. After pondering on the points of the *Checklist on Family Matters*, convening a family meeting to review the points may clarify to relatives how to become involved with caregiving over time, such as helping with home repairs, cleaning tasks, hands-on care, and volunteering for back-up plans, such as for an unexpected family illness, caregiver hospitalization, or harsh-weather emergencies.

[Please insert Figure 1. CHECKLIST ON FAMILY MATTERS here.]

### Guidance for different dementias

The following sections on Mild Cognitive Impairment, Alzheimer's Disease, Vascular Dementia, Frontotemporal Lobar Degeneration, and Parkinson Disease with Dementia as well as Parkinsonisms (i.e., Lewy Body Dementia, Progressive Supranuclear Palsy, and Corticobasal Degeneration) suggest different approaches to guide the care planning steps.

## When the Diagnosis is Mild Cognitive Impairment

**Mild Cognitive Impairment (MCI)** refers to a mild decline in one or two cognitive functions such as short-term memory, recall of words, or other thinking function such as planning and decision-making. However, the decline typically does not interfere in the ability to manage daily routines, job or household responsibilities, relationships, or driving skills, and thus, is considered to not be dementia. MCI function may stay at the same level over time, but each year 10-15% of people with MCI who are 65 or older convert to early Alzheimer's disease or a related dementia [10-11].

Most people with a diagnosis of MCI are self-sufficient and likely will benefit from:

- **Education:** From the physician about MCI and memory tips may include reminders such as computer or paper calendars and iPod/iPad, clock or wristwatch alarms. Memory exercises include repeating details to oneself and aloud, visualizing the information in an emotional or funny setting, creating a silly sentence out of the details or items to remember, and chunking 2 or 3 similar items together such as grouping fruits versus grouping bath supplies when shopping.
- **Treatment:** Should remedy fixable conditions such as hormone or vitamin imbalances, infections, insomnia, etc. The clinician should discontinue anticholinergics and underscore the adoption of a healthy lifestyle incorporating regular physical exercise, daily brain exercise, good nutrition, etc.
- **Prioritizing:** Values and interests should involve creating a Wish List (Things I Want to Do within the Next 5-10 Years) and forming action plans for the top wishes.
- **Planning Ahead:** The clinician should encourage the person with MCI to think through preferences and organize "personal business matters" (i.e., finances, legal documents, back-up plans for unexpected crises, etc.) with the aid of the *Checklist on Family Matters* (see Figure 1), then to identify a trusted family member, significant other, elder law attorney, and/or an accountant to take into confidence regarding preferences, plans, and decisions.

To assist with leadership responsibilities, such as on the job, in a community group, or with the extended family, the person with MCI should appoint, identify, or elect a partner to train in the role of CoChair, assistant CEO, or future family matriarch/patriarch in order to share responsibilities with that "mentee" or for succession training as the next leader.

## When the Diagnosis is Alzheimer's Disease

**Dementia of the Alzheimer's Type (AD)** refers to the person who has a decline in two or more cognitive functions such as short-term memory, recall of words, keeping up with bills or appointments, and problems making decisions [12,13]. The decline interferes in the person's ability to manage relationships and daily routine tasks on the job or at home. Typically, the first decline occurs in short-term

memory (forgetting appointments, details of an event, or location of items such as keys, eyeglasses, shoes, etc.). The second area of decline is usually in the recall of names or other specific words. The third area of decline may involve visuospatial skills (getting lost while driving), and/or apraxia (becoming more clumsy with learned hand skills such as when repairing or using appliances). Typically in early AD, mobility, such as gait and using stairs, is fine until much later in the course of decline.

The progressive decline of AD may be characterized as three stages: 1) the Forgetful Stage, 2) the Confusion Stage, and 3) the Severe Stage. As people with a diagnosis of AD progress from early stages to the severe stage of decline, they slowly shift from being self-sufficient to dependent. Meanwhile the health provider's management of care adapts to the ever-increasing need for respite and stress reduction of the family caregiver.

While most people with a diagnosis of **Early AD** or the **Forgetful Stage of AD** can function independently, the clinician should encourage the patient to bring at least one other trustworthy person (with normal memory and cognition) to the medical evaluation to offer a different perspective to supplement the patient's self-report of functioning and to receive the care plan updates. The patient and caregiver should benefit from:

- **Education:** From the physician about the condition (strengths versus areas of decline) and coping strategies such as:
  1. For memory problems, reminders and memory exercises mentioned for MCI (above) are useful.
  2. For difficulty recalling words (anomia), listing names on a small handy piece of paper to review should help. A few days before attending a social function, reviewing a short list of the names of attendees should help the recall of names at the function. Repeating the name of an object, focusing on the object's appearance, qualities, and setting should help to store and recall it.
  3. For visuospatial difficulty, studying a map beforehand, keeping a simple map handy; traveling during daylight, reduced traffic times, and going the most familiar way should help one avoid getting lost.
  4. For difficulty with skilled hand movements (apraxia) fewer steps for a task and simpler tools should help.
- **Treatment:** Should address fixable conditions such as hormone deficiencies; if appropriate, the physician will start cholinergic medicine. Emphasis to the patient should cover the importance of carefully following medical recommendations, such as medicines taken appropriately including cholinergics, serotonergics, neuroprotective medicines, and a healthy lifestyle, for example regular physical exercise, daily brain exercises, good nutrition, etc.

Physician concerns about the patient's driving safety during the clinical evaluation or from reports of fender-benders or near-

misses merit a referral for a comprehensive driving evaluation (a test that takes about 3 hours and evaluates visual fields and acuity, cognitive functions, movement, and actual driving skills). Some people stop driving at the first sign of being an at-risk driver or when a family member requests for them to stop driving. The physician may recommend that the person stop driving until s/he passes a comprehensive driving evaluation.

- **Prioritizing:** Values and interests should occur with the primary caregiver (family member or significant other). The Wish List should have a short timeline (Things I Want to Do within the Next 2-3 Years) and involve the family caregiver to fulfill the top, few wishes as soon as possible.
- **Planning Ahead:** The clinician should encourage the patient along with the family/significant other caregiver to develop future plans using the *Checklist on Family Matters* as a guide, and then schedule an appointment with an elder law attorney or accountant to set-up the documentation that specifies preferences for current and long-term decisions regarding financial or legal matters, preferences for later decline and end-of-life care, and back-up plans for unexpected events, such as future health crises.

The patient in the **Confusion Stage of AD**, when reminders and available assistance are important 24/7, the patient should attend any clinical appointment along with a family/significant other caregiver.

- **Education:** The clinician should educate the family/significant other caregiver about the condition (strengths versus areas of decline), and coping strategies such as simple discussions (one or two brief points at a time), repeating simple directions, and following usual routines for any tasks. Also, before and at an appointment or social function, it may help the patient to review a short reminder list identifying the function, activity, scheduled time, place, and names of people attending. A few seconds before greeting someone, the caregiver should whisper the name to the patient.
- The clinic should stress the importance of encouragement and positive interactions. Typically people at this stage are physically agile unless there is a health condition such as joint problems that impede movement; thus, regular physical exercise such as walking for 30-40 minutes 5 days a week and doing household or yard chores are important.
- **Treatment:** If appropriate, should involve the physician starting a cholinergic medicine and then a neuroprotective medicine. Emphasis to the patient and family caregiver should cover the importance of adhering carefully to medical recommendations and a healthy lifestyle.
  - **Prioritizing:** Values and interests should involve the patient with the caregiver undertaking the primary responsibility for decisions and activities. The Wish List

should identify “Things I Want to Do in the next 1-2 Weeks’. Caregiver attention to the activities the patient enjoys and can do.

- **Planning ahead:** The clinician should emphasize that burden of planning ahead falls on the family caregiver and the need for assistance from relatives, community volunteers, or paid caregivers. Complex decisions involving financial, legal matters, family, or home-setting matters should include input from the patient when possible but is primarily the responsibility of the caregiver. To receive input from the patient, questions should be simple, merit a very simple one or two word response, and should occur when the patient is sharpest cognitively such as after breakfast.

In the **Severe Stage of AD**, the family caregiver’s memory and other cognitive functions serve 24/7 as the memory and cognition of the patient. The patient may have a 30 second long memory and be able to do a one-step command, or not.

- **Education:** The clinician should help the family caregiver to obtain as much information as possible about the current level of health, cognition, and care strategies, especially hands-on care strategies for basic daily needs such as feeding, bathing, dressing, toileting, and mobility. Hired help to assist with the personal care of the patient or household duties is essential to reduce caregiver burden and to provide respite for the family caregiver.
- The clinician may remind the family caregiver that the cognitive capacity of the patient typically functions best with one or two brief points or words, repetition, simple one-step directions, following the routine way and schedule of tasks, daily exercise. The clinician should remind the family caregiver to keep interactions positive with a pleasant, soothing voice; smiles, and a gentle touch.
- **Treatment:** Full directions to the family caregiver should cover medicines, physical therapy, occupational therapy, speech therapy, etc. General basic health issues should include good nutrition, hydration, frequent appointments for dental care, skin care and massage, simple leisure activities such as going for walks with a companion, enjoying a pet, time outdoors, or listening to music.
- **Prioritizing:** The schedule and activities should balance the daily quality of life for *both* the patient and the family caregiver.
- **Planning Ahead:** The clinician should encourage the family caregiver to carry out the patient’s preferences. The family caregiver should seek advice from experts about finding assistance such as in-home help for personal care or caregiver respite or to use a skilled care, residential facility. Planning ahead and honoring the preferences and priorities of the patient are the responsibilities of the caregiver(s) who must remember what has been expressed previously if the written preferences are not available. If

the patient's preferences have not been documented nor communicated clearly to other family members, family disagreements about care at the severe stage or the end-of-life time may lead to family discord and fractured relationships.

### When the Diagnosis is Vascular Dementia (or Cardiovascular Dementia)

**Cardiovascular dementia** is considered the second most commonly occurring dementia in the U.S. (Lewy body dementia, which some researchers consider to be the second most common progressive dementia, is discussed later in the section on parkinsonisms.) In early decline the health provider, patient, and family caregiver should optimize and stabilize cardiac or blood circulation conditions, such as hypertension, high LDL or triglycerides, arteriosclerosis, atherosclerosis, diabetes, or strokes [14].

- **Education:** The clinician should educate the patient and family caregiver about the condition (strengths versus deficits) and the possibility in some cases of slow, steady improvements in function. Personality changes in the patient as well as an unawareness of deficits such as memory loss may lead to family conflicts, thus appropriate coping strategies are important for the clinician to teach the caregiver.
- **Treatment:** The clinician should emphasize that the first step after the evaluation is to adhere to careful treatment of any cardiovascular conditions. Cholinergics may boost memory and other cognitive functions. The physician should refer the patient to therapists, for example: a physical therapist to increase mobility, design an appropriate exercise schedule for the patient, and recommend assistive devices such as a cane or walker; a speech therapist for speech and swallowing difficulties; and an occupational therapist for access and safety issues, especially in the home setting. The physician should guide other elements of a daily healthy lifestyle such as good diet, careful dental hygiene, increasing or decreasing calories or water intake, decreasing stress, etc.
- **Prioritizing:** Values, interests, and health goals, depending upon the patient's level of functioning should involve a collaboration of the patient, the family/significant other caregiver, and the health provider team including therapists. Wish List priorities, depending upon the level of dysfunction and goals of rehabilitation, should be set for the short-term, i.e., one day, one week, one month, etc.
- **Planning Ahead:** The clinician should help the family caregiver stay in tune to the preserved physical and cognitive abilities and partner with the patient to identify preferences. Planning and making complex decisions for financial, legal matters, transportation, and other matters take time so an early start can accommodate changes more easily as function improves or worsens.

### When the Diagnosis is Frontotemporal Dementia [15-17]

**Frontotemporal Dementia, also called Frontotemporal Lobar Degeneration (FTD)** involves slow decline in the frontal and/or temporal lobes appearing in people 40 – 60 years of age. There may be limited insight about personal and cognitive changes, increased or decreased sleep. The three categories of FTD are: 1) Behavioral Changes, 2) Language Changes, and 3) Movement Changes.

*Early Behavioral Changes* may involve: unsocial behavior such as rude or embarrassing remarks, inappropriate conduct such as anger outbursts in public; lack of emotional expression such as a neutral response that seems uncaring to someone else's grief or pain; a problem with boundaries or limits such as hugging strangers, overeating, or insatiable cravings for sweets; or difficulty with choices.

*Early Language Changes (including speech, comprehension, writing, or reading)* may involve paucity of speech (nonfluency) or being unable to: recall words, use correct words, understand speech or written words, read, or correctly pronounce words.

*Early Movement Changes* may involve being unable to move one's limbs correctly to walk, move facial muscles fully to express feelings, or do skilled movements such as use a key, phone, or zipper.

- **Education:** The clinician should educate the patient and family about the different strengths versus areas of decline. In addition the discussion should include strategies to keep the patient engaged in a routine involving a variety of activities such as keeping a weekly or daily calendar posted on a bulletin board or iPod.

For *behavioral difficulties* supervision may be necessary to set diet boundaries (keeping sweets out of the home) and limit inappropriate interactions with strangers. Distractions that may work include calling the patient by name, making eye contact, and using strong, pleasant, positive words to do something else, such as look at a favorite photo, shop for a snack, go for a walk, groom a pet, etc.

*Language difficulties* may benefit from simpler, briefer statements, repeating phrases, using specific names of people and items (not pronouns such as her, him, it, them, or this/those) and allowing extra time for responses. In short, apply **KISSSSS (keep it sweet, same, simple, short; safe)**. Pointing or using body language while talking or using pictures may help conversations.

For *movement difficulties* an occupational therapist can help to safety-proof the home/yard setting and vehicle. A physical therapist can offer help for balance, walking difficulties, and an exercise routine. A speech therapist can offer tips for swallowing difficulties.

- **Treatment:** The physician can prescribe cholinergic and serotonergic medicines to benefit function. If the patient is still driving, based on the clinical evaluation, the physician should revoke driving privileges or refer the patient for a comprehensive driver evaluation. The physician should prescribe a physical therapy evaluation for recommendations regarding movement difficulties such as for gait, using a chair and stairs, daily exercise,

and getting into and out of a vehicle. An occupational therapist can offer ways to improve safety in the home, yard, and other settings.

- **Prioritizing:** Values, interests, and health goals should involve a collaboration of the patient, the primary caregiver (family member or significant other), and the health provider. The patient should keep as active as long as possible in their hobbies (or simpler versions of their hobbies) and relationships.
- **Planning Ahead:** The clinician can recommend early adaptations to improve the home setting for ease of use and safety before injury to the patient or caregiver occurs, for example adding hand rails and ramps, and removing sharp objects, such as knives and scissors. The clinician can help the patient and family caregiver to discuss preferences and decisions regarding current and long-term care adjustments. The caregiver with guidance from a member of the health team such as a social worker or other professional with expertise in elder and dementia issues along with a guide such as the *Checklist on Family Matters* should plan complex decisions for financial, legal matters, home-setting matters, and adaptations for the ongoing decline.

### **When the diagnosis is Parkinson disease with dementia or a Parkinsonism such as Lewy body dementia, progressive supranuclear palsy, or corticobasal ganglionic degeneration**

**Parkinson Disease with Dementia:** Primarily a movement disorder, Parkinson disease features any combination of slowed movement and thinking, balance and gait difficulties, cogwheel rigidity (in muscles), resting tremor, tiny handwriting, decreased facial expression (masked facies), and depression; dopaminergic medicines provide significant benefit as do antidepressants, where indicated. Up to 80% of people with Parkinson disease will develop memory and cognitive disorders and they benefit from cholinergic medications [4].

**Parkinsonisms:** Many people with movement difficulties which resemble Parkinson disease have a parkinsonism, often called a “cousin of Parkinson disease”. These atypical disorders, for reasons not clearly understood, may benefit somewhat or not at all from dopaminergic medicines. In the following brief discussions of three parkinsonisms, Lewy Body Dementia, Progressive Supranuclear Palsy, and Corticobasal Ganglionic Degeneration, the discussion of Lewy Body Dementia is lengthier because it is considered the 2<sup>nd</sup> or 3<sup>rd</sup> most commonly occurring [4].

**When the Diagnosis is Lewy Body Dementia [4,18,19] :** Recent scientists have insisted that **Lewy body dementia (LBD)** is the second most commonly occurring slowly progressive dementia. The onset of LBD symptoms occurs in people in their 40s or 50s. Typically, the early symptoms of LBD include visual hallucinations, movement problems, periods of on-off physical or thinking functions, and sleep problems, explained further below:

*Early symptoms:*

1. Visual hallucinations (less often auditory hallucinations)
2. Movement difficulties such as stiffness, slowness, or clumsiness when moving such as when walking; difficulty sitting in or rising from a chair, problems with balance; sometimes better movement and sometimes worse (or freezing up of) movement; lack of facial expression (masked facies)
3. On-off mental function
4. REM-sleep (Rapid Eye Movement) disorder which may include kicking, arm swinging, and yelling as if acting out a dream. The patient may be unaware of the activity, often reported by the sleep-partner.

#### **Later symptoms:**

1. Personality changes, i.e., social withdrawal, more irritable, more outgoing or withdrawn, or socially inappropriate
2. Less interested in leisure activities
3. Unwilling to do routine tasks
4. Decline in personal hygiene
5. Frequent stumbling or falling

#### **Much later:**

1. Decline in short-term memory, word recall, and other cognitive functions
2. More irritable, restless, and/or withdrawn
3. Frequent falls

### **When the Diagnosis is Progressive Supranuclear Palsy (PSP) [4,18,20,21]**

An atypical progressive dementia, the parkinsonism, **Progressive Supranuclear Palsy (PSP)** usually appears in people above 50 years of age. [21] PSP prevalence has been estimated as 1 to 6 or more per 100,000 people [18,21]. The early key features of PSP are balance problems, gait difficulties, midline rigidity, and “doll’s eyes” (vertical palsy - difficulty with upgaze) [20,21]. Much later there is decline in memory, language, and other cognition [20,21].

**When the Diagnosis is Corticobasal Ganglionic Degeneration (CBG):** [22]. Also an atypical progressive dementia, the parkinsonism, **Corticobasal Ganglionic Degeneration (CBG)** occurs in 4 to 7 or more people per 100,000 [4,23]. The key feature of CBG is the asymmetry of movement difficulties, usually beginning with increased clumsiness or failure to control the fingers to perform skilled movements (apraxia) in one hand. The problem in one hand expands to the whole arm before affecting the same-side leg or the other hand. In some cases the hand seems to have its own will (alien hand) and respond to external stimulus rather than obey the will of its owner. Later swallowing and speech difficulties occur. Much later there may be decline in memory and other cognitive functions [22,24-27].

**After the Diagnosis of Parkinson disease with Dementia or a Parkinsonism such as Lewy Body Dementia, Progressive Supranuclear Palsy or Corticobasal Ganglionic Degeneration**

**Education:** Should involve the clinician explaining to the patient and family caregiver/significant other the typical picture of initial slow decline in movement and then later decline in memory and other cognitive functions. It is important for the patient to function as fully and as long as possible. Early on, physical therapy for balance, gait training, finger-hand exercise, and regular physical exercise are key. The patient and family need to compensate for the motor slowing by increasing their patience, allowing more time for physical activities, and allowing more time for the patient to think and respond during interactions. Later when swallowing becomes a problem, speech therapists can offer meal tips such as avoiding dry foods (i.e., crackers, plain meat, chunks of fresh vegetables or fruits, etc.) and including wet, smooth foods such as stews, custard, and thickened beverages. Suggestions for future decline include patient-caregiver teamwork to help with personal care and other activities, memory tips, and maintaining familiar routines and ways to do tasks.

The physician may do a one-time, brief trial of a Parkinson medicine (a dopaminergic medicine) to see if there is improved movement and thinking. A clear improvement in movement and thinking function may indicate a diagnosis of Parkinson disease (with or without memory or other cognitive problems). Partial improvement may indicate a parkinsonism. No improvement from the dopaminergic medicine strengthens the suspicion of parkinsonism.

For Lewy body dementia education should include early coping strategies to address hallucinations, insomnia, and on/off times, for example:

1. People with Lewy body dementia usually realize and accept that their visual hallucinations are not real when others report not seeing them. Sometimes changes in the setting (removing dark or intimidating furniture or covering mirrors and other reflective surfaces) or a distraction will help.
2. Early on, help from a physical therapist should address difficulties with balance and gait and design of an exercise routine; an occupational therapist should recommend ways to assist movement struggles and making the home and yard settings safe.
3. Ways for family caregivers to ensure that the patient is paying attention and “tuning in” include: addressing the patient by name, making eye contact, repeating comments, speaking in a strong, pleasant voice, and asking for a simple answer to check alertness and comprehension.

Sleep changes may be problematic and may improve with sleep hygiene such as limiting caffeine later in the day, daytime exercise, relaxed pre-bedtime activities and massage, routine bedtime and awakening times, and no daytime naps.

For PSP, the education should address problems with upgaze (vertical gaze palsy, often called doll’s eyes) and teaching the patient and caregiver the importance of compensating for the decrease in visual fields by

increasing horizontal and vertical neck-turning for more careful looking when walking; an occupational therapist can offer suggestions regarding decreasing fall and injury risks in home and other settings.

**Treatment:** The physician can prescribe cholinergic and serotonergic medicines to benefit function. The physician should emphasize the importance of avoiding any antipsychotic medicine, especially to treat hallucinations or sleep problems associated with Lewy body dementia to steer clear of paradoxical effects or worsening of dysfunctions. The physician should refer the patient for a physical therapy evaluation for early symptoms of movement and balance difficulties evident in a slower, stiffer, tipsy gait; physical therapy to aid movement, an exercise plan, decrease fall risks, and especially to identify appliances such as a simple cane, a quad-cane and later a cane with a sling seat or a walker. Other useful referrals in an occupational therapy to evaluate the home setting, and a sleep study to evaluate sleep architecture. The physician should recommend that the patient stop driving until passing a comprehensive driving test, which is more in-depth than a driving test at a Department of Motor Vehicles.

A trial of a dopaminergic may help mobility; treating depression will help physical and cognitive functioning and contribute significantly to the quality of daily life.

**Prioritizing:** Values, interests, and health goals should be a collaboration of the patient, the primary caregiver (family member or significant other), and the health provider. The patient should consider a Wish List of priorities to undertake in the 6 -12 months and another Wish List for later that accommodates ongoing decline in mobility and safe activities for a person with balance and walking problems or using a cane or walker. The patient should continue their hobbies, relationships, safe physical activities, and daily brain exercise for as long as possible.

**Planning Ahead:** The clinician should work with the family and offer a guide such as the *Checklist on Family Matters* to motivate the family to begin to plan for future health changes. The patient should indicate preferences to help them remain as active in tasks, personal care, hobbies and relationships for as long as possible. The patient and family should consider care preferences of the patient as decline occurs and should write down decisions for future reference.

In the case of Lewy body dementia the clinician care plan should work with the family to deal first with visual hallucinations and mobility changes, next sleep changes, and later the memory and other cognitive changes, as they begin to surface.

## Conclusion and Summary

After the clinical evaluation indicates a progressive dementia, the physician (and health team) should educate the patient and family about the condition, offer clear instructions about treatments, and additional information such as educational handouts, references, and community as well as internet resources to provide assistance and support to the family over the long-term management of the health condition. Healthy lifestyle changes such as good nutrition, drinking plenty of water every day, regular physical exercise, daily brain exercise, and reducing stress may improve cognition or slow further decline.

Treating and stabilizing cardiovascular conditions may slow down the decline of vascular dementias. Recommending, as appropriate, cholinergics, neuroprotective medications, and antidepressants for progressive dementias such as Alzheimer's disease, frontotemporal degeneration and the parkinsonisms, should optimize function, delay decline in routine tasks and personal care, and enhance the quality of life for the patient and loved ones. Different progressive dementias require different care plan timing strategies, for example:

- **Mild Cognitive Impairment:** first address decline in memory or word recall.
- **Alzheimer's disease:** first address decline in short-term memory and word recall, later other cognitive difficulties and much later, movement difficulties.
- **Vascular dementias:** first address cardiovascular issues, next rehabilitation to restore or improve stroke-related dysfunctions, and address therapy, physical exercise and brain exercise toward specific cognitive and behavioral changes.
- **Frontotemporal Dementia:** depending upon the category:

For *behavioral difficulties*: first supervise and set boundaries, limit inappropriate interactions with strangers; use distractions; set up team approach to activities; structure daily and weekly schedules. Later, address decline in memory and language skills. Much later, address decline in movement.

For *language difficulties*: first approach with **KISSSSS (keep it sweet, same, simple, short; safe** for language (speech, writing, reading, comprehension). Later, address memory and other cognitive functions which decline. Much later, address decline in movement.

For *movement difficulties*: first refer to a physical therapist and then occupational therapy for balance and gait problems and apraxia and home/yard safety evaluations. Much later, address decline in memory and other cognitive dysfunctions.

- **Parkinson Disease with Dementia:** first address movement difficulties with referrals to physical and occupational therapists; also address decline in memory, other cognitive deficits, and, as appropriate, depression.
- **Parkinsonismssuch as Lewy Body Dementia, Progressive Supranuclear Palsy, or Corticobasal Ganglionic Degeneration:** first address movement difficulties with physical therapy, occupational therapy, and as appropriate, speech and swallowing therapy; also address depression and insomnia (avoid anti-psychotics for Lewy body dementia), and much later address general cognitive deficits such as decline in short-term memory, executive function, and language. **Note: avoid antipsychotics for visual hallucinations and sleep disorder problems related to in Lewy body dementia.**

The physician and other members of the health team have

important clinical roles when addressing progressive dementias, in particular after the diagnosis. Different progressive dementias vary greatly in onset and decline of symptoms; thus, the clinician has a critical role to guide the patient and family/significant other caregiver to learn about the diagnosed health condition and steps to take with guidance from a tool such as the *Checklist on Family Matters* in planning for adaptation over the long-term care management.

### Figure 1: Checklist on family matters(2014edition)

**Money & finances** Organize money matters early and with trusted people.

- Cash flow, safe, bank & other money accounts (Location? Keys or combination? Co-signer?)
- Checking, savings, money market, fund beater, IRA, CD
- Safety deposit box; safe (Keys or combination to open?Co-signer?)
- Social security (direct deposit) or pension (if under age 66, social security disability):
- Check, mailed home or direct deposit
- Change the representative payee?

**LEGAL MATTERS** Contact attorney or financial expert about:

- Durable power of attorney (DPA) for:** Finances, Health, Springing DPA (give date to start)
- Living will; health surrogate; full code; prehospital DNR (Do Not Resuscitate)
- Trust funds
- Legal changes, update when: move to another state, stay in different states more than a few weeks each year, change in caregiver(s)
- Guardianship (**Plan ahead to avoid the need for guardianship.**)

**FAMILY BUSINESS** Someone must handle:

- Regular bills:  Utilities  Rent/mortgage  Car repair  Yard care  Pet care  Insurance
- Back up plans.** If something happens (hurricane, fire, etc.), what is the family backup plan? Who helps? List items to toss into a box or suitcase for an emergency evacuation: important papers, medicines, clothing, incontinence items, water, food, first aid kit, pet food, etc.
- Enroll in local emergency shelter with driver license/ID two emergency contacts
- Wallet-size page: brief Health Record: allergies, major illness, surgery, medicines, etc.; ICE (In Case of Emergency) number on cell phone with health information & contacts
- Medicine (Dose? Where is medicine located in home? Who renews & picks up refills?)



Help with grocery shopping, bills, cooking, cleaning, laundry, house repairs, yard, travel

Real estate (Location of property and deeds? Co-owners?)

**INSURANCE** Know policy schedule, location and agent. CHECK WAIVERS& CO-PAYS!!

House;property  Life Insurance  General Medical  Dental  Veterans medical

Medicare  Long-term care  Disability  Medicaid  Car  Theft  Fire  Flood

**SAFETY:** Safety-proof the home. Plan ahead for crises: remove firearms; lock up or remove sharp knives, razors, scissors, sharp tools and appliances with cutting edges, etc.

**Drivers must be safe!** Plan ahead for other transportation.

Identity bracelet/necklace for missing persons [Silver Alert (FL)]; locks on doors, fenced yard, recent photo on file, Medic-Alert/Safe Return Program; patient-caregiver bracelet set available; Life Line; ICE (In Case of Emergency) medical&contact info (1-877-564-6423)

ABUSE Registry (abuse,neglect,threat/exploitation)for the state, toll free hotline

National Suicide Prevention Lifeline 24/7, 1-800-273-8255; www.suicidepreventionlifeline.org/gethelp.aspx

**IN EVENT OF DEATH** Know & honor person's preferences.

Summary of assets  Funeral & memorial service details  People to notify

Autopsy for diagnosis  Cemetery lot (Deed? Location?)

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