

## Degenerative Neurologic Disorders

### Overview

By their definition, degenerative neurologic disorders get worse over time. Some progress quickly while others progress over decades. A major goal of intervention is to help the client achieve an optimal level of functioning in light of their neurological deficits.

**Common Nursing Dx**  
 Altered Thought Process  
 Memory Deficit  
 Visual-Perceptual Alteration  
 Impaired Physical Mobility  
 Incontinence  
 Self-Care deficit  
 Impaired Individual and Family Coping

### Dementias

Dementia refers to the loss of memory, reasoning, judgment and language to such an extent that it interferes with daily life. How the changes come about is a key factor in determining if the dementia is chronic or temporary. Pts with dementia experience impairment of cognitive activities and may undergo behavioral/personality changes as well (depending on the area of the brain affected). The causes and types of dementia are numerous. The most common form in people over 65 is Alzheimer's Disease (AD).

### Alzheimer's Disease

AD is a progressive degenerative disease of the cerebral cortex and hippocampus that presents as progressive memory loss. Recall that the cerebral cortex is the part of the brain that plays a key role in memory, attention, perceptual awareness, thought, language and consciousness. Researchers recognize two forms of AD: familial and sporadic. In the familial form genetics cause the disease (pretty rare). In sporadic AD (most common type) genes don't cause the disease but may influence the risk of developing the disease. Not much is known about the cause of the disease, but four factors are thought to play a role: neurochemical transmitters (such as deficiencies in acetylcholine), somatostatin, substance P, and norepi.

The characteristic microscopic findings of AD include "senile plaques", which are collections of degenerative presynaptic endings along with astrocytes and microglia. These plaques are more numerous in the cerebral cortex and hippocampus.

**A & P Review**  
 Astrocytes = support cells  
 Microglia = CNS immune defense  
 Hippocampus = short term memory

The brain tissue of Alzheimer's patients has three distinguishing features:

- 1) neurofibrillary tangles formed of cytoskeletal intermediate filaments
- 2) beta-amyloid plaques (deposits of protein-like substances)
- 3) granulovascular degeneration of neurons

The brain mass of an Alzheimer patient atrophies and the brain has narrowed gyri and widened sulci mainly in the frontal and parietal regions.

| Clinical Manifestations of Early-Stage AD | Clinical Manifestations of Mid-Stage AD | Clinical Manifestations of Late-Stage AD |
|---|---|--|
| Memory loss, especially recent            | Speech problems                         | Bedridden                                |
| Neglect of responsibilities               | Nonsense talkativeness                  | Spastic paralysis/contractures           |
| Carelessness                              | Complete disorientation                 |  |
| Job performance suffers                   | Restless/Wandering                      |  |
|   | Failure to recognize                    |  |
|   | Movement problems                       |  |

## Communicating with an Alzheimer's Patient

A big part of communicating is being an excellent listener. You must be a more active listener when communicating with your loved one because he may need assistance expressing his thoughts. Be sure to:

- **Show patience.** Your relative can sense when you're impatient or agitated, and this only increases her own frustration.
- **Provide reassurance.** If your loved one is having trouble communicating, tell him that it's fine and encourage him to keep trying to put his thoughts into words.
- **Focus on the positive.** Criticizing or correcting is nonproductive and may be harmful. Instead, focus on what your loved one has said and try to find meaning in her message.
- **Agree instead of argue.** If you don't agree with -- or are offended by -- a statement made by your loved one, just let it go.
- **Offer alternatives.** If your relative is really having difficulty finding the right words, it's OK to offer a guess as long as he appears to want some help.
- **Concentrate on feelings.** Although the content of a message may be hard to understand, it's often possible to uncover the feelings behind it by observing tone of voice, facial expressions, gestures, and body language.
- **Reduce distractions.** It's harder for people with Alzheimer's to communicate in chaotic environments, so try to talk in a quiet, calm place.
- **How to Communicate to Your Loved One**
- While being a good listener is vital to communication, it's also important to clearly convey your own ideas and information. Regardless of how difficult communication becomes, people at all [stages](#) of Alzheimer's benefit -- and have the right to -- frequent communication from those who care for them. When speaking to your loved one, try to:
- **Set yourself up for success.** Approaching your relative from the front will eliminate the possibility of startling her and getting off to a rocky start.
- **Be aware of tone and body language.** Speaking clearly in a relaxed tone of voice will put your loved one at ease. Friendly gestures will also foster positive interactions.
- **Keep it simple.** Difficult words or long sentences may overwhelm someone with Alzheimer's.
- **Wait for a response.** It might take longer for your loved one to respond, so be patient and give him time.
- **Be clear.** Avoid phrases that can be interpreted literally, such as "break a leg" or "chew the fat," which might be confusing.
- **Focus on the key word or idea.** Emphasize the most important word in your message either verbally or nonverbally (pointing).
- **Account for [hearing](#) or [vision](#) problems.** Make sure that your loved one is wearing a working hearing aid and/or clean glasses, if prescribed.

## Drug Treatment for Alzheimers

- Acetylcholinesterase inhibitors increase acetylcholine levels in synapse. Symptoms improve, but the disease does not:
  - Rivastigmine (EXELON) brain-selective
  - Donepezil (ARICEPT)
  - Galanthamine (RAZADYNE, REMINYL)
  - Tacrine (COGNEX) ~~too many SE~~

- Glutamine blocker is useful in later stages and allows combo therapy: Memantine
- Ginkgo biloba extract better than placebo. Vitamin E slows progress by 7 months. NSAID naproxen being studied. Estrogen not helpful.

As of 2007, drug therapy for Alzheimer's typically involves selection of one of these 3 cholinesterase inhibitors: donepezil, galantamine (made from the tulip bulb) or rivastigmine. Recently approval of a drug from another class, memantine, has opened up more possibility for combination drug therapy.

Other updates from 2007 include:

- *Immunization against amyloid* (amyloids are insoluble fibrous protein aggregates that can accumulate and play a role in neurodegenerative diseases such as AD.) In 2002 research on this was stopped due to brain inflammation in 6% of patients, but followup studies of those who developed antibodies shows significant improvement.
- *Flurizan* decreases production of amyloid.
- *Alzheimed* decreases plaque formation.

**Q.** Given that acetylcholine is the neurotransmitter in the PNS, and given that acetylcholinesterase breaks down acetylcholine, what side effects would an acetylcholinesterase inhibitor have?

**A:** Diarrhea and frequent urination (remember that the PNS is "rest and digest")

### Diagnosing AD

Sadly, AD can not be confirmed until the patient has died and an autopsy can be performed. However, tests are conducted to rule out other disorders:

- *PET scan* measures the metabolic activity of the cerebral cortex
- *CT scan* can show more brain atrophy than occurs in normal aging
- *MRI* evaluates the condition of the brain and rules out intracranial lesions
- *EEG* evaluates the brain's electrical activity and may show brain wave slowing late in the disease. It also helps to differentiate tumors, abscesses, and other intracranial lesions
- *CSF analysis* helps determine whether S&S stem from a chronic neurological infection
- *Cerebral blood flow studies* may detect abnormalities in blood flow to the brain

### Management of patient's with AD

Management of AD involves protecting the patient from injury, assisting with memory and providing respite for caregivers.

## Parkinson's Disease

It is estimated that PD affects 1 to 1.5 million people in the U.S. It most commonly strikes people over the age of 50, but can affect people even before age 40. Approximately 10 percent of PD cases are estimated to be young-onset...like Michael J. Fox. I was so sad when he got a girlfriend on Family Ties...I was kinda hoping he'd notice me ;-)

The etiology of this horrible disease is unknown, but one source I found says that some cases result from exposure to toxins that destroy cells in the substantia nigra of the brain...so stay away from manganese dust and carbon monoxide!

The disease is mainly caused by a deficiency of dopamine. Recall that dopamine is an important neurotransmitter in the brain and is essential for governing movement, balance and walking. By the time symptoms have developed, 80-90% of the dopamine producing cells have been lost.

### Symptoms of PD

- Muscle rigidity
- Tremor
- Slow movement (bradykinesia) or loss of movement (akinesia)
- Difficulty with balance or walking.

You can check out the symptoms at

[http://medweb.bham.ac.uk/http/depts/clin\\_neuro/teaching/tutorials/parkinsons/pdtremor.mov](http://medweb.bham.ac.uk/http/depts/clin_neuro/teaching/tutorials/parkinsons/pdtremor.mov)

[http://medweb.bham.ac.uk/http/depts/clin\\_neuro/teaching/tutorials/parkinsons/parkinsons2.html](http://medweb.bham.ac.uk/http/depts/clin_neuro/teaching/tutorials/parkinsons/parkinsons2.html)

### Associated Problems

60-90% of people with PD will develop some difficulty speaking (dysarthria). This is often characterized by weak, slow or uncoordinated speaking. Often, speech problems worsen over time. Speech therapists may help with this but there is limited data about which techniques are most successful.

50% develop swallowing problems (dysphagia). Food or fluids may spill from the mouth and the pt is more prone to choking on food or drink (and even on their own saliva). It is usually easier for this pt to swallow thickened fluids. A speech therapist performs a swallow eval.

A good strategy when feeding the Parkinsons' pt is to have them sit up and swallow twice with each bite. They should concentrate one eating and take small bites. Food should be pureed or chopped small.

More than 50% of Parkinson's pts have mild intellectual changes, and about 20% have more substantial cognitive impairment. The memory problems with PD are typically milder than with AD. The pt with PD may have difficulty concentrating, acquiring new information and recalling names.

### Prognosis

There is no cure for Parkinsons' disease, but symptoms can be managed

#### **Other symptoms of PD**

Depression  
Constipation  
Weight loss  
Sleep disturbances  
"Masked" facial expression  
Forced eyelid closure  
Drooling  
UTIs  
Excessive sweating  
Sexual dysfunction  
High-pitched monotone voice  
Fatigue

with medications levodopa & carbidopa are most common) or surgical procedures where part of the brain is destroyed or stimulated. Transplantation of fetal dopamine producing cells is promising, but ethically controversial.

### Treatments

- Selegiline (ELDEPRYL) is an MAO inhibitor that may be started at the time of diagnosis. It blocks the breakdown of dopamine.
- As the disease progresses beyond minor symptoms, additional drug treatment may be indicated.
  - Dopamine cannot cross the blood-brain barrier, so we give the precursor L-DOPA (levodopa). Levodopa is rapidly metabolized so only a small fraction of the drug is available to the CNS, so we give an inhibitor to the metabolism (carbidopa).
  - Sinemet, a combination of levodopa and carbidopa, is usually the drug most doctors use to treat Parkinson's disease patients. While side effects including large uncontrollable movements called "dyskinesias" and confusion may develop after about 8 years, drug therapy for Parkinson's typically provides relief for about 10-15 years or more. Keep the drug dose low and timing regular to minimize the "on-off phenomenon" where the pt responds to the meds ok and then at some point before the next dose the dopamine level seems to drop below a threshold and the patient develops sudden bradykinesia.
  - Others: entacapone, anticholinergic, amantadine
- Thalamotomy is a surgical procedure that surgically destroys cells in the thalamus to correct a disabling tremor in the hands or arms. It is used for pts with few other symptoms. Risks are considered low and immediate improvement is seen in 80-90 percent of patients after the operation. Full recovery takes about 6 weeks.
- Pallidotomy surgically destroys certain cells in the globus pallidus (part of the brain that controls movement). The operation may help correct problems in slow movement, tremor and imbalance. It is performed with the patient conscious. Generally, the most dramatic result is the decrease in dyskinesia. Post-surgical recuperation is similar to that of thalamotomy.
- Stimulator implantation involves the implantation of an electrode with the tip in the target site. It is connected to a wire running beneath the skin that leads to a stimulator placed in the chest wall. When electrical current is activated it modifies the function of the target site. The stimulator can be adjusted externally using a programmer with an electromagnetic head.
  - Follow this link to see a video or two of deep brain stimulation: <http://www.georgetownuniversityhospital.org/body.cfm?id=1236>
- Fetal tissue implantation is an experimental technique used to restore the brain's ability to produce dopamine. It is rife with controversy and we may see genetically engineered cells for this type of surgery in the future. To date, long-term studies are not available on the efficacy of this procedure. Preliminary findings indicate that the surgery can dramatically decrease the need for meds, although the improvement may not begin until 6 months after the surgery and may not peak for 12 to 24 months.
- Physical therapist can help develop and monitor a home exercise program. A good exercise routine should include strengthening and flexing all limbs, stretching legs and

- feet, walking, facial and breathing exercises, and specific exercises to gain better control in swallowing.
- Occupational therapist can help the person accomplish everyday activities.
  - Speech therapist can help the person with PD improve voice volume, quality and articulation.
    - Therapeutic exercises including verbalizations and tongue movements
    - Where speech is severely impaired, a machine or computer-generated voice
    - Families must learn new strategies to help the person communicate.
    - The inability to articulate can be frustrating...offer reassurance and support.

### **Multiple Sclerosis**

MS is an autoimmune process in which immune cells mistake myelin as foreign and attack it. This results in the progressive demyelination of the white matter of the brain and spinal cord, leading to widespread neurologic dysfunction. The structures usually involved are the optic and oculomotor nerves and the spinal cord tracts. Depending on where nerve damage occurs, MS can affect vision, sensation, coordination, movement, and bladder & bowel control.

Approximately 350,000 people in the US have MS, and 50% of them will need a walking aid within 15 years of diagnosis. The prognosis of the disease varies. It may progress rapidly causing death or disability by early adulthood, but about 70% of people with MS lead active, productive lives with prolonged remissions.

### Who gets MS?

MS is more common in women and generally first occurs in people in their 20s and 30s. Evidence shows that people of northern European descent, especially Scandinavian) may be genetically predisposed to MS, and many of these people have settled in the northern U.S. The disease does appear to run in families, with as many as 20% of people with MS having at least one affected relative. Compared to the general population, first-degree relatives (children, siblings) of people with MS have a 20-40-fold increase in risk of eventually having MS. However, it's not just one gene that passes down the disease...multiple genes likely contribute. My best friend has MS, but right now she is doing just fine...thank goodness!

### Causes

The exact cause of the disease is unknown, but there are several theories. It may be caused by a slow-acting viral infection, an autoimmune response of the nervous system or an allergic response. Exposure to common bacteria or viruses (such as the human herpes virus 6) may trigger the disease in some people with a genetic tendency. This may explain why in identical twins, only one twin develops MS 70% of the time. Because MS is twice as common in women as in men, it's been suggested that hormonal factors predispose women to the disease. This theory has not been confirmed, but one study showed that symptoms of MS were reduced during the second and third trimesters when estrogen levels are high.

Other possible causes include trauma, anoxia, toxins, nutritional deficiencies, vascular lesions and anorexia nervosa, all of which may help destroy axons and the myelin sheath.

MS does not affect the PNS.

### Types of MS

**Benign:** In 10 percent to 15 percent of patients, symptoms are mild to moderate, don't worsen and don't lead to permanent disability.

**Relapsing-remitting:** About 85% of people with MS begin with this form, and more than half have this form of the disease at any one time. It is characterized by one or two flare-ups every 1 to 3 years, followed by periods of remission. Such flare-ups, which may include any of the symptoms of MS alone or in combination, typically appear suddenly, last a few weeks or months, and then gradually disappear. Symptoms may worsen with each recurrence.

**Primary progressive:** From the onset of symptoms, neurological function deteriorates without periods of remission. About 10% of patients begin with this disease pattern.

**Secondary progressive:** Usually after years of having relapsing-remitting MS, at least half will enter a stage of continuous deterioration. Relapses may still occur.

**Progressive relapsing:** This is primary progressive MS with the addition of sudden episodes of new symptoms or worsened existing ones. This form is quite rare.

### Treatments for MS

- Steroids
- Immunomodulating agents
- Betaseron, Avonex, Rebif (interferons) and Copaxone (synthetic)
- Immunosuppressive drugs such as Mitoxantrone
- Caution pt that immune boosting herbs may worsen disease

Treatments for MS aim at shortening exacerbations and relieving neurologic deficits. Corticotropin, prednisone or dexamethasone is used to reduce edema of the myelin sheath during exacerbations, relieving symptoms and hastening remission. These drugs do not prevent future exacerbations.

Currently (as of 2009) the preferred treatment during an acute attack is a short course of methylprednisone (with or without a short prednisone taper). Interferons may also be given to decrease the frequency of relapses.

During acute exacerbations, supportive measures include bed rest, massage, prevention of fatigue and pressure ulcers, bowel and bladder training, treatment of bladder infections with Abx, physical therapy and counseling.

### **Myasthenia Gravis**

Myasthenia gravis (MG) is a rare autoimmune disease that produces sporadic, progressive weakness and abnormal fatigue of voluntary skeletal muscle. It usually affects muscles in the face, lips, tongue, neck and throat...but can affect any muscle group including bulbar muscles, ocular muscles, and respiratory muscles. Muscle fibers may eventually degenerate and weakness becomes irreversible. When this happens to the respiratory muscles it can be life-threatening.

The cause of MG is unknown, but it commonly accompanies autoimmune disorders and disorders of the thymus. For some reason that no one has yet figured out, the patient's blood cells and thymus gland produce antibodies that block, destroy or weaken the neuroreceptors, causing a failure in transmission of nerve impulses at the neuromuscular junction. During normal

neuromuscular transmission, acetylcholine is released to diffuse across the synapse and receptor sites in the motor end plate react and depolarize the muscle fiber. The depolarization spreads throughout the muscle fiber, causing contraction. In MG, antibodies attach to the acetylcholine receptor sites making them insensitive to acetylcholine, thereby blocking neuromuscular transmission.

### Treatment

- Cholinesterase Inhibitors: cause considerable improvement in some patients, but little to none in others. Strength rarely returns to normal. Most common ChE inhibitors are Pyridostigmine bromide (MESTINON) and neostigmine bromide (PROSTIGMIN).
  - SE of Mestinon: muscarinic- abd. cramping and skeletal muscle fasciculations, weakness from excessive depolarization
- Thymectomy is recommended for most patients with MG. The maximal favorable response generally occurs 2-5 years after surgery. Response is unpredictable.
- Corticosteroids.
  - Marked improvement or complete relief of symptoms occur in more than 75% of pts treated with prednisone with the other 25% showing some improvement. About one-third of the pts become weaker within 7-10 days of starting pred, but this only lasts for up to 6 days. The major disadvantage of this therapy are the side effects.
  - 30% have remission, 45% marked improvement (???)
- Immunosuppressant Drugs (decrease Ab to ACh receptor)
  - Azathioprine reverses symptoms in most pts, but the effect is delayed 4-8 months
  - Cyclosporine inhibits predominantly T-lymphocyte-dependent immune responses and is sometimes beneficial in treating MG. Most pts with MG improve 1-2 months after starting the drug, and improvement is maintained as long as the therapeutic doses are given. SE include renal toxicity and HTN.
  - Cyclophosphamide has been used IV and PO. More than half of the pts become asymptomatic after one year. SE are common.
- Plasma Exchange is used as a short-term intervention for pts with sudden worsening of MG symptoms, to rapidly improve strength before surgery, and as a chronic intermittent treatment for pts who are refractory to all other treatments.
  - Plasmapheresis improves for 1-2 months.
- Intravenous Immune Globulin (IVIG). Several groups have reported favorable responses to high-dose (2 grams/kg infused over 2-5 days) IVIG. Possible mechanisms of action include down-regulation of antibodies directed against AChR and the introduction of anti-idiotypic antibodies. Improvement occurs in 50-100% of patients, usually beginning within 1 week and lasting for several weeks or months. \$\$\$\$

Age of onset:  
women ages 20-30  
Men ages 70-80

### The Big Picture

These are only a few of the many neurologic diseases, but there are common themes.

- Safety
- Are pulmonary muscles weak? (think atelectasis and respiratory distress)

Most pts with MG have hyperplasia. Get CT or MRI to evaluate all pts.

- Are the esophageal muscles weak? (think aspiration)
- Is the brain dysfunction affecting judgment or control of movement (think injury, falls)
- Is innervation to bowel/bladder compromised? (think constipation, retention)
- Are skeletal muscles affected? (think immobility)