



Your Questions Answered

LEWY BODY DEMENTIA

Q What is the difference between Lewy body dementia and other dementias, such as Alzheimer's disease?



DR. BRADLEY BOEVE RESPONDS:

A Lewy body dementia (LBD) is characterized by dementia plus two or more of the following features: parkinsonism; recurrent fully formed visual hallucinations; fluctuations in cognition and/or arousal; and REM sleep behavior disorder (RBD).

The parkinsonian features include less facial animation, a stooped posture and shuffling gait, and slowness of movement. Tremor, an unintentional to-and-fro movement, may also be present. The hallucinations can involve vivid visions of people or animals, often with the same imagery/perceptions from day to day. Cognitive problems are usually notable in judgment, problem solving, complex decision-making, and multitasking, as well as in visuospatial functions (required for activities such as driving). Memory may or may not be impaired.

Sleep disorders may cause problems with cognition and arousal as well as excessive daytime sleepiness. RBD refers to the tendency for people to "act out their dreams." During normal REM sleep, our brains are active but most of the muscles in our bodies are still. But in RBD, the muscles are abnormally active, and injuries such as bruising, pulled hair, and even fractured bones can occur. The dreams often have a nightmare quality, with the person being chased or attacked.

These features contrast with other common dementia syndromes. In Alzheimer's disease, forgetting the details of recent events and upcoming appointments is the prominent early feature. Parkinsonism and hallucinations occur late in the disease if at all, and RBD is extremely rare. Frontotemporal dementia is manifested by marked changes in personality and behavior and/or language, whereas memory and visuospatial functioning tend to be minimally affected. Parkinsonism and visual hallucinations are uncommon in frontotemporal dementia, and RBD is also rare.

Bradley F. Boeve, M.D., is associate professor of neurology at the Mayo Clinic in Rochester, MN.

MIGRAINE AND FACIAL PAIN

Q I've noticed that right before I get a migraine, my jaw and sometimes other parts of my face starts to feel sore or painful. Is this normal?



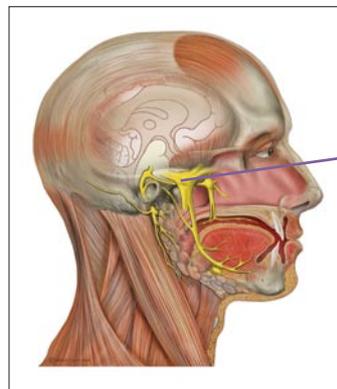
DR. NINAN T. MATHEW RESPONDS:

A It is not uncommon for some people with migraine to have facial pain, either at the onset or during the headache. This pain, abnormal sensation, or soreness in the jaw, facial skin, or the scalp is known as allodynia.

The pain is caused by changes in the central nervous system when a migraine occurs. During a migraine, the blood vessels that connect to the trigeminal nerve, which is responsible for facial sensation, become inflamed and dilated. The trigeminal nerve becomes sensitized and causes allodynia.

These symptoms are sometimes an early sign that a migraine is approaching and that you should therefore take your medication. The longer you wait, the less effective the medications are.

Ninan T. Mathew, M.D., is director of the Houston Headache Clinic in Houston, TX.



TRIGEMINAL NERVE

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DO YOU HAVE A QUESTION TO ASK THE EXPERTS?
Send it to neurologynow@lwwny.com

PROGRESSIVE SUPRANUCLEAR PALSY

Q My husband was just diagnosed with progressive supranuclear palsy. Is there any treatment for this disease?



**DR. LARRY
GOLBE
RESPONDS:**

A Progressive supranuclear palsy is a rare brain disorder that causes serious and permanent problems with control of gait and balance. People with this disease may also experience changes in their mood and behavior (such as depression and apathy), as well as difficulty with organizing mental tasks and dealing with abstractions. These problems slowly worsen over time. Eventually, most people also develop problems with swallowing and eye movement.

The drugs levodopa-carbidopa (Sinemet) or amantadine (Symmetrel) may provide modest and temporary relief for the limb stiffness that interferes with movement. However, dosages of amantadine beyond 100 mg twice a day may cause confusion, and if there is no benefit within a week, you should stop taking it. The levodopa-carbidopa should be titrated up (meaning the doctor will increase the dosage level until it produces an effect) to at least 1,200 mg per day before concluding that it is ineffective.

Donepezil (Aricept) or other “memory drugs” intended for Alzheimer’s disease may help with dementia symptoms. Depression may be treated with antidepressants, sleepiness with modafinil (Provigil), and insomnia with a sleeping medication such as zolpidem (Ambien)—however, it usually is not a good idea to take modafinil and zolpidem together because they have opposite effects. The urinary urgency may respond to tolterodine (Detrol) and oxybutynin (Ditropan), though these drugs often worsen the constipation that comes with the disease.

A swallowing evaluation, during which a doctor will usually take an x-ray of the person swallowing, should be performed as soon as he or she starts to cough on liquids. The disinhibition that is often part of this disease is a result of degeneration of the brain’s frontal lobe and may cause people to overload the mouth or to suddenly stand and run while eating.

While physical therapy helps little with balance, it can help doctors determine the best devices to aid gait and can be also be used to prescribe a safe exercise regimen to improve joint flexibility.

Lawrence I. Golbe, M.D., is professor of neurology at the University of Medicine and Dentistry of New Jersey-Robert Wood Johnson Medical School. He is also director of Research and Chair of the Scientific Advisory Board of CurePSP, the Society for Progressive Supranuclear Palsy.

DYSAUTONOMIA

Q I was told I have dysautonomia. What causes this, and what treatments are available for it?



**DR. PAOLA
SANDRONI
RESPONDS:**

A Dysautonomia refers to any disorder of the autonomic nervous system. The autonomic nervous system regulates bodily functions that are not under voluntary control, such as blood pressure and bladder function. When the autonomic nervous system is dysregulated—that is, not working properly—you will experience a number of symptoms. Common complaints include dizziness, light headedness, bladder dysfunction, constipation, sexual dysfunction, and inability to sweat.

There are many different causes of dysautonomia. One cause is autonomic neuropathy, which refers to damage of the peripheral nervous system that disrupts information flow from the brain and spinal cord to every other part of the body. Central autonomic disorders such as multiple system atrophy can also cause dysautonomia; these disorders are characterized by severe dysautonomia and Parkinson’s-like symptoms or incoordination.

Dysautonomia can also be hereditary. Some forms are fairly limited, but there is also a rare, devastating form affecting only the Jewish population.

To see what form of dysautonomia you have and how to treat it, a doctor will perform tests. These can include blood testing or autonomic testing, which involves monitoring your blood pressure, blood flow, heart rate, skin temperature, and sweating to see whether your autonomic nervous system is functioning normally.

There is often no specific cure for the underlying disease, though some forms of dysautonomia go away on their own or are self-limited, meaning they don’t get worse. Usually, doctors will prescribe drugs that target specific symptoms—such as blood-pressure drop upon standing and bladder dysfunction—and will give advice about lifestyle modifications that may improve the symptom severity.

For more information and to join a support group, contact the National Dysautonomia Research Foundation at ndrf.org or by phone at 651-267-0525..

Paola Sandroni, M.D., Ph.D., is associate professor of neurology at the Mayo Clinic in Rochester, MN.