

An illness that attack your brain and nerve cells -Parkinson's plus syndromes

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Abstract

Parkinson's plus syndromes are illnesses that attack your brain and nerve cells. As the name suggests, they're linked to Parkinson's disease and cause a lot of the same symptoms, but they can bring on other problems as well. Brain makes a chemical called dopamine that helps control your movement. Parkinson's disease is by far the most common of these, but about 15% of people who have a problem making dopamine will have one of the Parkinson's plus syndromes. Parkinson's plus syndromes are more serious and harder to treat than "classic" Parkinson's disease. The four main types are: Progressive Supranuclear Palsy (PSP), Dementia with Lewy Bodies, Multiple System Atrophy and Corticobasal Degeneration

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Introduction

Parkinson's plus syndromes, also called "atypical Parkinson's," are illnesses that attack your brain and nerve cells. As the name suggests, they're linked to Parkinson's disease and cause a lot of the same symptoms, but they can bring on other problems as well. Brain makes a chemical called dopamine that helps control your movement. Parkinson's disease is by far the most common of these, but about 15% of people who have a problem making dopamine will have one of the Parkinson's plus syndromes.

Researchers aren't sure what causes Parkinson's or Parkinson's plus syndrome. There might be some genetic or environmental risk factors that can increase the likelihood of developing Parkinson's plus syndrome. For example, some scientists theorize that exposure to toxins could cause your risk, but more research needs to be done to prove this link.

Types

Parkinson's plus syndromes are more serious and harder to treat than "classic" Parkinson's disease. The four main types are:

Progressive Supranuclear Palsy (PSP)

This is the most common Parkinson's plus syndrome. It causes some of the same issues with

movement and your muscles as Parkinson's disease, like stiffness and problems with walking or balance, but it doesn't usually make your limbs shake. It also can make it harder to move your eyes -- it starts in the part of your brain that controls your eye muscles. Looking down can be especially hard. It can cause mood changes, affect your ability to think of words, and make it hard to swallow as well. PSP causes trouble with balance and stability that can mimic Parkinson's disease. Unlike Parkinson's disease, people with PSP don't experience tremors. They do have difficulty with eye movement and are likely to experience more trouble with speech, swallowing, and mood than people with Parkinson's disease.

Dementia With Lewy Bodies

LBD is a progressive brain condition caused by structures called Lewy bodies that form in your brain. People with LBD might have symptoms that resemble Parkinson's disease, dementia, or a combination of both. This is the second most common form of dementia after Alzheimer's disease. Lewy bodies are clumps of protein that build up in your nerve cells. When that happens, it affects your ability to think clearly, speak, and remember things. It can make you confused and cause hallucinations (when you see things that aren't there). The symptoms get worse over time.

Multiple System Atrophy

MSA is a progressive condition that affects your nervous system. It causes stiffness and loss of balance similarly to Parkinson's disease. Over time, the effects of the disease on your nervous system can lead to difficulty with essential body functions such as digestion, breathing, and your heartbeat. This affects what's known as your autonomic nervous system, which controls things like your blood pressure and digestive system. Symptoms can include things like fainting, losing control of your bladder, and constipation. It also causes more typical Parkinson's symptoms, like shaking, stiffness, and problems with balance or speech. MSA is a progressive condition that affects your nervous system. It causes stiffness and loss of balance similarly to Parkinson's disease. Over time, the effects of the disease on your nervous system can lead to difficulty with essential body functions such as digestion, breathing, and your heartbeat.

Corticobasal Degeneration

It is a condition that causes parts of your brain to become smaller. This causes many symptoms that overlap with Parkinson's disease, such as tremors and balance problems. Over time, it can lead to difficulty with both speaking and writing.

This is the rarest of the four main types. It kills brain cells in the cerebral cortex -- the wrinkly grey matter on the outside of your brain -- and causes the cortex to shrink. It also attacks what's called the basal ganglia, a part of your brain that controls movement.

Its symptoms are like the ones caused by Parkinson's disease, including the loss of muscle control, sometimes starting on only one side of your body. But it also can hurt your ability to think, see, and speak clearly. As the disease gets worse, it gets harder to walk and swallow. CBGD is a condition that causes parts of your brain to become smaller. This causes many symptoms that overlap with

Parkinson's disease, such as tremors and balance problems. Over time, it can lead to difficulty with both speaking and writing.

Unique symptoms of PSP include:

- falling backward
- blurred vision and difficulty reading
- difficulty moving the eyes up and down
- slurred speech
- difficulty swallowing
- depression or other mood issues
- behavioural changes
- laughing or crying at inappropriate times

Unique symptoms of MSA include:

- breathing problems that get worse at night
- syncope, or passing out
- dizziness
- slurred speech
- low blood pressure
- bladder problems
- sleep disturbances

Unique symptoms of CBDG include:

- one-sided movement trouble
- involuntary muscle contractions
- rapid muscle jerks
- trouble with concentration
- trouble with communication
- behavioural changes

- trouble coordinating movements, or apraxia
- loss of control over an arm called “alien limb syndrome”

Unique symptoms of LBD include:

- difficulty processing information
- difficulty following instructions
- decreased awareness of surroundings
- hallucinations
- delusions
- sleep disturbances
- mood changes

Other things that can point to a Parkinson’s plus syndrome rather than the classic form include:

- Early signs of dementia
- Falling often
- Trouble moving your eyes
- Your symptoms get worse then level off for a while

Diagnosis

Parkinson’s plus syndromes can look a lot like other conditions that affect your nervous system, so it can sometimes take a while to find out for sure what’s going on.

If your doctor thinks you might have Parkinson’s or a Parkinson’s plus syndrome, they’ll recommend that you see a neurologist, a doctor who specializes in problems with the nervous system. Your neurologist will examine you and see how you move and follow directions. They then might suggest blood tests and a brain scan to rule out other conditions.

If those don’t show a reason for your symptoms, they may ask you to try a medication called carbidopa-levodopa. Your brain can turn that into dopamine. If your symptoms get better, that may be enough for your doctor to diagnose Parkinson’s disease. If it doesn’t help much or at all, or it helps for a while then stops working that can be a sign of a Parkinson’s plus syndrome.

Treatment

Doctors don’t know exactly what causes any of the Parkinson’s plus syndromes, and there’s no cure for them. Treating them usually is about managing the symptoms. That can include the following:

- Medication can help some people move more easily and feel less stiff. Some drugs also can help with the problems caused by multiple system atrophy, like fainting or constipation.
- A cane or walker can help you get around.
- Speech therapy can help you communicate better.
- Exercise and physical therapy can make your muscles stronger and more flexible.
- Occupational therapy can help make everyday tasks easier.

Parkinson's plus syndrome is the name for a group of neurological conditions that are very similar to Parkinson's disease. Because these conditions cause symptoms that are very similar to Parkinson's, they are often incorrectly diagnosed. However, these conditions can even be treated using many of the same medications and therapies as Parkinson's.

The symptoms of Parkinson plus can vary and depend on the condition you have. Many people will have symptoms that are also found in Parkinson's disease, such as:

- balance problems
- tremors
- stiffness or muscle rigidity
- difficulty walking and standing
- difficulty controlling your movements
- fatigue
- confusion

The conditions that make up Parkinson's plus are not actually Parkinson's disease and do have unique symptoms.

People with Parkinson's plus syndrome are often diagnosed with Parkinson's disease in the early years of their condition. However, their condition won't progress like Parkinson's disease. It might progress faster, and they might start to develop symptoms that aren't present in Parkinson's disease.

There is no definitive test for Parkinson's or Parkinson's plus syndrome. Instead, a doctor might conduct a series of tests that will look at your balance, ability to walk, and coordination. These are generally simple in-office tests involving the doctor watching you walk, sit, stand, and perform other movements. You'll likely also do some memory and cognition tests with the doctor.

The doctor might also order some imaging tests to get a closer look at your brain. These may include:

- **MRI scan.** An MRI uses magnetic waves to create images of your body.
- **PET scan.** A PET scan uses a special dye to look for damage to your brain.
- **CT.** A CT scan can check your brain activity.

What causes it?

Researchers aren't sure what causes Parkinson's or Parkinson's plus syndrome. There might be some genetic or environmental risk factors that can increase the likelihood of developing Parkinson's plus syndrome. For example, some scientists theorize that exposure to toxins could cause your risk, but more research needs to be done to prove this link.

Although the underlying cause isn't known, we do know what changes to your body can cause each Parkinson's plus syndrome:

- **PSP.** When you have PSP, a build-up of protein in your brain cells causes them to deteriorate. Your condition will progress as this continues.
- **MSA.** As with PSP, proteins accumulate in the cells of your brain that control your central nervous system and other vital functions.
- **CBGD.** A protein called tau builds up in your brain cells when you have CBGD. This build up causes the symptoms of CBGD.
- **LBD.** Protein clusters called Lewy bodies grow in your brain when you have LBD. Over time, the Lewy bodies cause changes to your brain that impact your ability to function.

What are the current treatment options?

While there is no specific cure for Parkinson's plus syndrome, there are treatments that can control your symptoms. A doctor can develop a plan for your overall health and to treat your specific symptoms. Medications that treat the symptoms of Parkinson's disease often do not work as well for Parkinson's plus syndrome.

Treatment options might include:

- **Walking and balance assistance.** You might receive physical and occupational therapy to help keep you moving. Therapists can help you build strength and prevent falls. They can also help you learn to use canes, walkers, and other mobility aids, if needed.
- **Swallowing and speech assistance.** A speech therapist can help you adjust to changes that might make it hard to swallow and speak. They can help you communicate and can recommend foods and beverages that are easier to swallow.
- **Medications for cognitive issues.** Your doctor might prescribe a variety of medications that can help with your focus and memory. Many of these medications are also used for conditions such

as Alzheimer's or dementia.

- **Medications for trouble with movement.** You might be prescribed medications that can help you control your muscles and movement. These medications might also address stiffness and balance problems.
- **Medications to help with mood symptoms.** If you're experiencing depression, anxiety, or other mood-related concerns, your doctor might prescribe medications that can help with these symptoms.

What's the outlook for people with Parkinson's plus?

Although there currently isn't a treatment to halt the progression of Parkinson's plus syndrome, there are treatments that can help you manage your symptoms and improve your quality of life.

The exact outlook for Parkinson's plus syndrome depends on the person and the specific condition they have. Someone who is otherwise healthy when they're diagnosed will typically have a longer life expectancy than someone who is already facing other health conditions when they're diagnosed. Your doctor will monitor your condition over time and can let you know how it's progressing.

Treatment

No cures currently exist for atypical Parkinsonism. The goal of treatment is to manage symptoms for as long as possible. The appropriate medication for each disorder depends on your symptoms and how you respond to treatment.

For LBD, some people find relief from symptoms with cholinesterase inhibitors. These drugs increase the activity of neurotransmitters that affect memory and judgment.

For PSP, levodopa and similar drugs that act like dopamine, are helpful for some people.

Participating in physical or occupational therapy can also help with most of these conditions. Keeping physically active may help relieve symptoms. Check with your doctor if any specific exercises might be good for you.

Risk factors

Certain risk factors are known for PD, but little has been established for atypical Parkinsonism. The known risk factors for PD include:

- **Advancing age.** This is the most common risk factor for PD.
- **Biological sex.** Those assigned male at birth tend to develop PD more often than those assigned female at birth.
- **Genetics.** Many studies are exploring the genetic link to PD.

- **Environmental causes.** A variety of toxins have been linked to PD.
- **Head trauma.** Injuries to the brain are thought to contribute to PD onset.

Much research is ongoing to establish risk factors for atypical Parkinsonism disorders, especially in genetics.

Some atypical Parkinsonism disorders have obvious risk factors. For example, drug-induced Parkinsonism is related to certain drugs, and vascular Parkinsonism stems from previous strokes.

But the risk factors for the other Parkinsonisms are the subject of a lot of current research. Scientists are looking into why each of these conditions occur and how to slow or stop their progression.

Possible complications

Perhaps the most serious complication from any of these conditions is dementia.

You may first develop mild cognitive impairment (MCI), which may not interfere too much with your daily activities. If your thinking skills and memory gradually decline, you may need the assistance of family, a home health aide, or an assisted living facility.

Because these conditions affect balance and coordination, fall risk becomes an important concern. Having PD or atypical Parkinsonism means avoiding falls and fractures. Make your home safer by getting rid of throw rugs, lighting hallways at night, and installing grab bars in the bathroom.

Atypical Parkinsonian syndromes are progressive diseases. This means that their symptoms will continue to worsen over time. While no cures exist for these disorders yet, there are treatments that can help to slow their progression.

It's important that you take your medications exactly as prescribed by your doctor. If you're ever unsure about your treatment, call your doctor's office.

PD and atypical Parkinsonism affect each person differently. Those differences include the type and severity of symptoms, as well as life expectancy.

One study found that assuming an average age of about 72 years at diagnosis, people with atypical Parkinsonism lived on average 6 more years.

Life expectancy estimates can vary greatly depending on your overall health. The healthier you are when you're diagnosed, the better your chances of living longer with atypical Parkinsonism.

Conclusion

Patients with Parkinsonism-plus syndromes represent a relatively small portion of Parkinsonism patients seen in general and movement disorders clinics. Given the wide spectrum of disease phenotypes associated with these disorders and the often subtle clinical differences, establishing the correct diagnosis, especially at disease onset, can be difficult despite serial clinical observations and

repeated neurological examination. Autopsy confirmation is often necessary. Advancement in molecular genetics may provide a better understanding of some of these rare syndromes and holds substantial promise for more rational classification and therapy. Moreover, an accurate diagnosis of these disorders is necessary to understand their cause and pathogenesis. This may allow development of biologic therapeutic strategies to stop or slow disease progression, such as inhibition of tau or α -synuclein aggregation.

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