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Pick disease

Pick disease is a rare form of dementia that is similar to Alzheimer disease, except that it tends to affect only certain areas of the brain.

Causes

People with Pick disease have abnormal substances (called Pick bodies and Pick cells) inside nerve cells in the damaged areas of the brain.

Pick bodies and Pick cells contain an abnormal form of a protein called tau. This protein is found in all nerve cells. But some people with Pick disease have an abnormal amount or type of this protein.

The exact cause of the abnormal form of the protein is unknown. Many different abnormal genes have been found that can cause Pick disease. Some cases of Pick disease are passed down in families.

Pick disease is rare. It can occur in people as young as 20. But it usually begins between ages 40 and 60. The average age at which it begins is 54.

Symptoms

The disease gets worse slowly. Tissues in parts of the brain shrink over time. Symptoms such as behavior changes, speech difficulty, and problems thinking occur slowly and get worse.

Early personality changes can help doctors tell Pick disease apart from Alzheimer disease. (Memory loss is often the main, and earliest, symptom of Alzheimer disease.)

People with Pick disease tend to behave the wrong way in different social settings. The changes in behavior continue to get worse and are often one of the most disturbing symptoms of the disease. Some persons have more difficulty with decision making, complex tasks, or language (trouble finding or understanding words or writing).

General symptoms are listed below.

Behavioral changes:

- Not able to keep a job
- Compulsive behaviors
- Inappropriate behavior
- Inability to function or interact in social or personal situations
- Problems with personal hygiene
- Repetitive behavior
- Withdrawal from social interaction

Emotional changes:

- Abrupt mood changes
- Decreased interest in daily living activities
- Failure to recognize changes in behavior
- Failure to show emotional warmth, concern, empathy, sympathy
- Inappropriate mood
- Not caring about events or environment

Language changes:

- Cannot speak (mutism)
- Decreased ability to read or write
- Difficulty finding a word
- Difficulty speaking or understanding speech (aphasia)
- Repeating anything spoken to them (echolalia)
- Shrinking vocabulary
- Weak, uncoordinated speech sounds

Nervous system problems:

- Increased muscle tone (rigidity)
- Memory loss that gets worse
- Movement/coordination difficulties (apraxia)
- Weakness

Other problems:

- Urinary incontinence

Exams and Tests

The doctor will ask about your medical history and symptoms.

Your health care provider might order tests to help rule out other causes of dementia, including dementia due to metabolic causes. Pick disease is diagnosed based on symptoms and results of tests, including:

- Assessment of the mind and behavior (neuropsychological assessment)
- Brain MRI
- Electroencephalogram (EEG)
- Examination of the brain and nervous system (neurological exam)
- Examination of the fluid around the central nervous system (cerebrospinal fluid) after a lumbar puncture
- Head CT scan
- Tests of sensation, thinking and reasoning (cognitive function), and motor function

A brain biopsy is the only test that can confirm the diagnosis.

Treatment

There is no specific treatment for Pick disease. Medicines may help manage mood swings.

Sometimes, patients with Pick disease take the same medicines used to treat other types of dementia.

In some cases, stopping or changing medicines that worsen confusion or that are not needed can improve thinking and other mental functions. Medicines include:

- Analgesics
- Anticholinergics
- Central nervous system depressants
- Cimetidine
- Lidocaine

It is important to treat any disorders that can cause confusion. These include:

- Anemia
- Decreased oxygen (hypoxia) levels
- Heart failure
- High carbon dioxide levels
- Infections
- Kidney failure
- Liver failure
- Nutritional disorders
- Thyroid disorders
- Psychiatric conditions such as depression

Medicines may be needed to control aggressive, dangerous, or agitated behaviors.

Some patients may benefit from hearing aids, glasses, cataract surgery, or other treatments.

Behavior modification can help some people control unacceptable or dangerous behaviors. This consists of rewarding appropriate or positive behaviors and ignoring inappropriate behaviors (when it is safe to do so).

Psychotherapy (talk therapy) does not always work. This is because it can cause further confusion or disorientation.

Reality orientation, which reinforces environmental and other cues, may help reduce disorientation.

Depending on the symptoms and severity of the disease, the patient may need monitoring and help with personal hygiene and self-care. Eventually, there may be a need for 24-hour care and monitoring at home or in a special facility. Family counseling can help the person cope with the changes needed for home care.

Care may include:

- Adult protective services
- Community resources
- Homemakers
- Visiting nurses or aides
- Volunteer services

Persons with Pick disease and their family may need to seek legal advice early in the course of the disorder.

Advance directive, power of attorney, and other legal actions can make it easier to make decisions regarding the care of the person with Pick disease.

Support Groups

Some communities may have support groups, such as for Alzheimer disease and elder care.

Outlook (Prognosis)

The disorder quickly and steadily becomes worse. Patients become totally disabled early in the course of the disease.

Pick disease commonly causes death within 8 to 10 years, usually from infection, but sometimes from failure of the body systems.

When to Contact a Medical Professional

Call the health care provider or go to the emergency room if mental function gets worse.

Prevention

There is no known prevention.

Alternative Names

Semantic dementia; Dementia - semantic; Frontotemporal dementia; Arnold Pick disease

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