

Lyme Disease and Cognitive Impairments

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Introduction:

The patient is a college graduate with Lyme encephalopathy (LE). While stopped at a traffic light, she described her thought processes as having a "fog-like" sluggishness. When the light changes, she knows the change from red to green has significance, but at that moment cannot recall that green means go and red means stop.

This is one of many examples of cognitive impairments associated with Lyme disease. Although some cognitive symptoms are indirectly a result of other neurological or emotional impairments, others are a direct result of dysfunction of the cerebral cortex where cognitive processing occurs.

Laboratory tests such as SPECT scans, MRI's, PET scans, and psychological testing have demonstrated physiological and anatomical findings associated with dysfunction of the cerebral cortex in patients with Lyme and tick-borne diseases. The examination of human and animal brains have further supported these findings.

The cognitive impairments from Lyme disease are very different than we see in Alzheimer's disease. Lyme disease is predominately a disease of the white matter, while Alzheimer's is predominately a disease of the gray matter. Memory association occurs in the white matter, while memory is stored in the gray matter. White matter dysfunction is a difficulty with slowness of recall, and incorrect associations.

In contrast, gray matter dysfunction is a loss of the information which has previously been stored. For example, and Alzheimer's patient may not recall the word "pen", while an LE patient may have a slowness of recall or retrieval of a closely related word.

Some of the symptoms I will describe are also found in encephalopathies associated with other illnesses, such as chronic fatigue syndrome, lupus stroke, AIDS, or other diseases which affect the brain. Although no single sign or symptom may be diagnostic of Lyme disease in a mental status exam, we instead look for a cluster and a pattern of signs and symptoms that are commonly associated with Lyme disease.

Everyone with LE has their own unique profile of symptoms. The assessment of these signs and symptoms is one facet of the total clinical assessment of Lyme disease.

There are many ways of categorizing cognitive functioning. Let's begin with a simple model of perception, encoding these perceptions into memory, processing what we perceive, imagery, and finally organizing and planning a response. Simple mental functions such as flexing the index finger of the right hand, correlates with a relatively simple brain circuitry.. More complex functions such as flying an airplane requires the action of a more integrated neural circuitry. The

difference between these two actions is like the difference between playing middle C on a piano vs. a symphony playing an entire concert.

Attention Span:

Many Lyme disease patients have acquired attention impairments which were not present before the onset of the disease. There may be difficulty sustaining attention, increased distractibility when frustrated, and a greater difficulty prioritizing which perceptions are deserving of a higher allocation of attention. If we compare attention span to the lens of a camera, we need the flexibility to constantly shift the allocation of attention dependency upon the current life situation. For example, we shift back and forth between a wide angle and a zoom lens focus to increase or decrease acuity of attention depending on the needs of the current situation.

A loss of this flexibility results in some combination of a loss of acuity (hypoacusis), and/or excessive acuity to the wrong environmental perceptions (hyperacusis). Hyperacuity can be auditory (hearing), visual, tactile (touch), and olfactory (smell).

Auditory hyperacusis is the most common. Sounds seem louder and more annoying. Sometimes there is selective auditory hyperacusis to specific types of sounds. Visual hyperacusis may be in response to bright lights or certain types of artificial lighting.

Tactile hyperacusis may be in response to tight fitting or scratchy clothing, vibrations, temperature and merely being touched may be painful. Some patients prefer to wear loose fitting sweat suits and are frustrated that being touched can be painful. Olfactory hyperacusis may result in an excessive reactivity to certain smells, such as perfumes, soaps, petroleum products, etc.

Memory

Memory is the storage and retrieval of information for later use. There are several different memory deficits associated with LE. Memory is broken down into several functions - working memory, memory encoding, memory storage and memory retrieval.

Working memory is a component of executive functioning. An example of working memory is the ability to spell the word "world" backwards. Sometimes there are impairments of working memory as it pertains to a working spatial memory, i.e. forgetting where doors are located or where a car is parked.

Encoding is the placement of a memory into storage. We cannot retrieve a memory that was not encoded correctly into memory in the first place. One patient described being upset that someone had eaten yogurt in her kitchen during the night. Her activity during the night was not encoded into memory.

Short term (recent) memory is the ability to remember information for relatively brief periods of time. In contrast, long term memory is information from years in the past (or remote).

In LE, there is first a loss of short term memory followed by a loss of long term memory very late in the illness. Patients may have slowness of recall with different types of explicit (or factual) information, such as words, numbers, names, faces or geographical/spatial cues. Not as common, there may also be slowness of recall if implicit information, such as tying shoes, or doing other procedural memory tasks.

Errors in memory retrieval include errors with letter and/or number sequences. This can include letter reversals, reversing the sequence of letters in words, spelling errors, number reversals, or word substitution errors (inserting the opposite, closely related or wrong words in a sentence).

Processing

Processing is the creation of associations which allow us to interpret complex information and to respond in an adaptive manner. Some LE patients say they feel like they acquired dyslexia or other learning disabilities, which were not present previously. Examples of processing functions that may be impaired in the presence of LE include the following:

Reading comprehension: The ability to understand what is being read.

* Auditory comprehension: The ability to understand spoken language.

* Sound localization: The ability to localize the source of a sound.

* Visual spatial perception: Impairments result in spatial perceptual distortions.

One example is microscopia, in which things seem smaller than they really are.

One patient lost depth perception, and had several accidents when the car in front of her stopped. A problem associated with visual spatial processing is optic ataxia, in which there is difficulty targeting movements through space. For example, there may be a tendency to bump into doorways, difficulty driving and parking a car in tight spaces, and targeting errors when placing and reaching for objects.

One patient with optic ataxia, was stopped by a policeman while driving two miles to my office because he kept swerving across the center line. Before Lyme disease he could consistently shoot 13 to 14 out of 15 free throws from the basketball foul line. Now he averages 3 of 15, and misses some shots by several feet.

* Transposition of laterality: The ability to rotate something 180 degrees in your mind. For example, the ability to copy, rather than mirror, the movements of an aerobics instructor facing you.

* Left-right orientation: The ability to immediately perceive the difference between left and right. Although this is a part of congenital Gertsmann's syndrome or angular gyrus syndrome, acquired left-right confusion is the result of an encephalopathic process.

* Calculation ability: The ability to perform mathematical calculations without using fingers or calculators. Many LE patients describe an increased error rate with their checkbook.

- * Fluency of speech: The ability of speech to flow smoothly. This function is dependent upon adequate speed of word retrieval.
- * Stuttering: The tendency to stutter when speech is begun with certain sounds.
- * Slurred speech: A slurring of words, which can give the appearance of intoxication.
- * Fluency of written language: The ability to express thoughts into writing.
- * Handwriting: The ability to write words and sentences clearly.

Imagery

Imagery is a uniquely human trait. It is the ability to create what never was within our minds. When functioning properly, it is a component of human creativity, but when impaired, it can result in psychosis. Imagery functions that can be affected by LE include:

- * Capacity for visual imagery: The ability to picture something, such as a map, in our head.
- * Intrusive images: Images that suddenly appear which may be aggressive, horrific, sexual or otherwise.
- * Hypnagogic hallucinations: The continuation of a dream, even after being fully awake.
- * Vivid nightmares: A tendency towards nightmares of a vivid Technicolor nature.
- * Illusions: Auditory, visual, tactile and/or olfactory perceptions which are distorted or misperceived.
- * Hallucinations: Hearing, seeing, feeling and/or smelling something that is not present. In LE, sometimes this takes the form of hearing music or a radio station in the background. Unlike schizophrenic hallucinations, these are accompanied by a clear sensorium, and the patient is aware hallucinations are present.
- * Depersonalization: A loss of a sense of physical existence.
- * Derealization: A loss of a sense that the environment is real.

Organizing and Planning

Organizing and planning a response is the most complex mental function, and is dependent upon all the functions already described. These functions, along with attention span and working memory, are referred to as executive functioning. Organizing and planning functions that can be affected by LE include:

- * Concentration: The ability to focus thought and maintain mental tracking while performing problem solving tasks.
- * "Brain fog": Described by many LE patients. Although difficult to describe in objective, scientific terms: it is best described as a slowness, weakness, and inaccuracy of thought processes. Prioritizing, organizing, and implementing multiple tasks with effective time management.
- * Simultasking: The ability to concentrate and be effective while performing multiple simultaneous tasks.

- * Initiative: The ability to initiate spontaneous thoughts, ideas and actions rather than being apathetic or merely responding to environmental cues.
- * Abstract reasoning: The capacity for complex problem solving.
- * Obsessive thoughts: May interfere with productive thought.
- * Racing thoughts: May interfere with productive thought.

An assessment of each of these areas of functioning is a critical component in the clinical assessment of LE. The cognitive assessment is only a part of the assessment of LE. Other components include the psychiatric assessment, the neurological assessment, a review of somatic symptoms, epidemiological considerations and laboratory testing when indicated.

I have gradually developed a structured cognitive assessment which focuses upon the areas mentioned after examining many patients with late stage neuropsychiatric Lyme disease. I have also incorporated concepts from others that have made major contributions in this area, such as Drs. Rissenberg, Nields, Fallon, Freundlich and Bleiwiss.

It is difficult to explain exactly how Lyme disease causes cognitive impairments. The variability of these symptoms suggests an episodic release of an endotoxin or cytokine which may contribute to the cognitive dysfunction. This is an area where considerable research is needed, and is beyond the scope of this article.

The symptoms described are often very difficult for patients to describe, and are difficult for many physicians to understand. As a result, patients with these impairments are sometimes erroneously viewed as being hypochondriachal, psychosomatic, depression, or malingering.

These symptoms are real and must be explained: that cannot be discounted as being imaginary.

There are many treatment strategies. Antibiotics and a number of different psychotropics are helpful to many. I have found Aricept to be helpful in the treatment of "brain fog" and problems with slowness of retrieval.

To those of you who have LE, be realistic about your limitations and the validity of these limitations. Use strong areas to compensate for areas of weakness. Avoid excessive stress which compounds the problem. Be aware that certain tasks challenge many higher level attributes. Maintain hope and retain an effective working relationship with your family, support system and treatment team.