

## **PROGRESSIVE CORTICAL VISUAL DYSFUNCTION SYNDROME**

David F. Tang-Wai M.D.,C.M.  
Division of Behavioral Neurology  
Department of Neurology  
Mayo Clinic  
Rochester, MN

### **What is this syndrome?**

Progressive cortical visual dysfunction syndrome is a *rare* progressive degenerative neurologic condition of the brain that primarily affects vision. There have been other terms that have been used in the past that describes this syndrome, including (1) *posterior cortical atrophy*, (2) *visual variant of Alzheimer's disease* and (3) *asymmetrical cortical degeneration–visual-perceptual subtype*. We prefer to use the term *progressive cortical visual dysfunction syndrome* as it best describes the major visual dysfunction as the major problem with other secondary symptoms, and it does not infer an underlying cause/pathology or specify the affected regions of the brain.

### **What are the symptoms of this condition?**

This syndrome starts very slowly and gradually affects daily life. Most persons notice an initial change in their eyesight that they cannot describe to physicians, optometrists or ophthalmologists. They often describe it as *cannot see clearly or sharply* or *blurry*. Most persons first come to their optometrists or ophthalmologists who often do not find any problems with either the eyes or the retina and despite changing the prescription of eyeglasses several times, it does not help the vision.

Some persons will notice difficulty with reading. Specifically, they notice that it is difficult to read and follow a single line in a magazine, book, newspaper etc. Often

persons describe that *words will jump out* or they will need to *re-read the line over*. To help, persons will place a ruler, for example, under the line of text to guide their eyes across the page.

Persons and those that live with them will notice other changes as well. It may be difficult filling out forms – such as not being able to *see* where to write the name, where to write the address, or where to sign. Completing cheques will be difficult for the same reason. All persons will notice that they cannot see objects placed in front of them. For example, groping for the pair of eyeglasses even though they are placed directly in front of the person.

Some persons will have difficulty watching and interpreting too many items or actions simultaneously. The medical term for this is *simultanagnosia*. Many persons have several examples, such as searching for a serving fork in a drawer full of utensils, unable to tell time using either an analog clock or an analog watch, or unable to either watch or follow what is happening on television or a movie because there is too much activity on the screen.

Driving also becomes difficult because of the visual difficulties. Persons report difficulty with depth perception and can drive too close to cars in front of them. Persons report driving too close to the edges of the road or too close to parked cars and often will find that they have scraped the side of the car. Persons report that they are involved in near

accident because they could not *see* the other car, such as turning in an intersection with an on-coming car.

Unfortunately, over time the vision worsens to the point where persons will need help with daily activities that require seeing, such as cooking, driving or writing. Persons eventually become *cortically blind* – where the eyes still function but the brain can no longer interpret what the eyes are seeing.

**Who gets this syndrome?**

This syndrome can affect either women or men. In our experience, more women are affected. Typically symptoms start in a person's late-40's to mid-60's.

To date there is no cause for this syndrome. It can happen by chance.

**In addition to vision problems, will other symptoms develop? What can I expect?**

Unfortunately, as with other neurologic degenerative conditions of the brain, other symptoms may develop over time as the vision worsens. The visual system in the brain is located in the occipital and parts of the parietal lobe. The parietal lobe is also necessary for the ability to do mathematics, the ability to write, the ability to know the difference between the right and the left sides, and the ability to perform simple and complex tasks.

Many daily activities will be very challenging. For example, persons will have difficulty balancing the cheque book or calculate change (the ability to do mathematics), difficulty

handwriting documents/ notes or signing a person's name, and difficulty with dressing or putting on make-up (the ability to perform simple and complex tasks).

Memory and language may also be affected over time. Persons may have difficulty with short-term/day-to-day memory, such as remembering grocery lists, appointments or important dates. Coming up with the correct word in speech or mixing up words may also develop.

**But can I drive?**

**No.** Driving also becomes difficult because of the visual difficulties. Persons report difficulty with depth perception and can drive too close to cars in front of them. Persons report driving too close to the edges of the road or too close to parked cars and often will find that they have scraped the side of the car. Persons report that they are involved in near accidents because they could not *see* the other car, such as turning at an intersection with an on-coming car.

Driving heavily involves the use of the visual system of the brain and its ability to integrate all of the pieces of simultaneous information together, such as looking at the position of the car relative to others, looking at the speedometer, looking at traffic, or looking at potential obstacles or potential hazards. We are concerned for you and your family's safety and welfare and recommend that **you do not drive**.

### **What is causing this?**

At this time, we do not know what causes this syndrome. Published reports of those persons with this condition who underwent a brain autopsy at the time of their death reveal that the majority of them had microscopic changes of Alzheimer's disease (tangles and amyloid plaques) predominantly affecting the visual system.

### **Does this mean I have Alzheimer's disease?**

This is a difficult question to answer. Persons with the typical Alzheimer's *disease* that either is portrayed in the news or that someone hears about, usually develop short-term memory difficulties first with relative preservation of their long-term memory. At brain autopsy, these persons will have the tangles and amyloid plaques (Alzheimer's pathology) predominantly affecting the memory system.

Early in the course of progressive cortical visual dysfunction syndrome, persons initially have visual difficulties with relative sparing of memory. From this standpoint, it is different from Alzheimer's disease. However, at brain autopsy, most persons will have the Alzheimer's *pathology*. Controversy currently exists among physicians to determine if Alzheimer's *pathology* should be regarded as the same as Alzheimer's *disease*.

### **Can it be cured? Are there any treatments?**

There is no cure for this syndrome. The treatments currently available are for those with Alzheimer's disease and can be used for this disorder as well; however, the effectiveness of these treatments is unknown as this is a rare disorder.

There are two methods for treatment: non-pharmacological and pharmacological. Non-pharmacological means involve methods and/or devices to help persons function at home or outside and maintain their independence. Pharmacological means may include either using medications for Alzheimer's disease, such as *Aricept*<sup>TM</sup> (donepezil), *Reminyl*<sup>TM</sup> (galantamine) or *Exelon*<sup>TM</sup> (rivastigmine), or using vitamin E.

Suggested non-pharmacologic means:

- Use of a digital or talking watch
- Learning Braille
- Use of a cane for the blind
- Obtaining large print magazines
- Simplify the organization of the items in a room (for example, the kitchen) to make it easier to find objects

**Will my children get or develop this?**

This is not a contagious syndrome and no one can catch it from casual contact. Little is known about this disorder and there may be a possibility of heredity with a slight risk to offspring. At this time, however, there are no published reports of heredity with this syndrome and it appears to occur sporadically.

### **Who can I tell? What should I say to them?**

Deciding *whether* to tell someone or *who* to tell is a personal choice. Persons usually divulge such information to people that they trust the most and who can understand.

Examples of explanations of this syndrome include (1) my eyes work but there is a problem with the connection to the brain, or (2) the back part of the brain responsible for vision is not working well or is degenerating for unknown reasons.

### **What happens if depression sets in?**

Some persons may eventually feel depressed or sad. Some persons may feel that they are the only affected one in the world as this is so rare. Consequently, persons may not want to go out and socialize, feel that life is no longer interesting or fun, or sleeping too much or too little. If you or your spouse/care-giver notices this pattern, it may be a sign of depression. Please either call your local physician or us. We can help.

### **What can I do now?**

With some modifications, you can continue with your daily activities, except for driving, as you normally would with the knowledge that *over time* it may become more difficult. You may need help with them and if so, ask for assistance. You may also want to attend your local chapter of the Alzheimer's Association. You may also want to consider contacting the Association for the Blind as they may have tips and strategies to help you out. You should try to continue reading – some read with large print books or magazines, read headlines in a newspaper, use a ruler to help with following lines, go out and

socialize with friends, have conversations, take long walks... In essence, you can do anything (within reason).

### **Internet web-sites**

- Unfortunately, there are no web-sites that specifically addressed this syndrome.

### **Internet search words**

- Posterior cortical atrophy
- Alzheimer's disease
- Visual dysfunction
- Simultanagnosia
- Cortical blindness
- Tangles
- Amyloid plaques
- Balint's syndrome

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