Visual Conditions Commonly Associated with Deaf-Blindness

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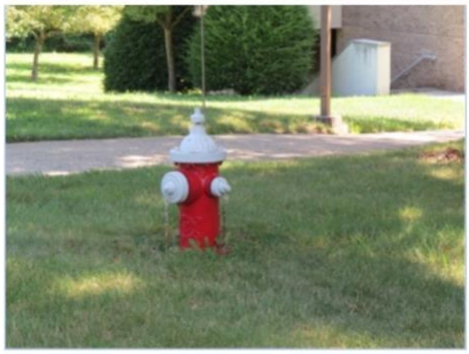
Macular Degeneration



Macular degeneration, often referred to as age related macular degeneration (AMD) is a leading cause of vision loss in people 55 and older in the United States, affecting approximately 30-50 million people globally. Stargardt disease is a form of macular degeneration found in children and young adults but is relatively rare. AMD causes loss of central vision. It is a progressive condition, which does not result in total blindness, but can greatly impact one’s ability to function. Symptoms include distorted vision, difficulty adjusting vision when moving from a dark to bright setting or vice versa, decreased visual acuity, central scotomas (missing areas of vision) and difficulty identifying colors. Therefore, common everyday tasks such as reading, driving and recognizing people’s faces can become difficult.

The two main types of macular degeneration are referred to as dry and wet. Approximately 90% of people with AMD have the dry form.

The "dry" form of macular degeneration is caused by slow break down light sensitivity cells in the macula. This may lead to distortion of vision and in advanced stages a person can experience blind spots in their central vision or total loss of central vision. The cause is unknown, but the progression of vision loss tends to be significantly slower than that in the wet form. At this time, there is no treatment for dry AMD.

The "wet" form of macular degeneration is caused by the growth of abnormal fragile blood vessels underneath the retina which often leak blood and fluid into the retina causing distortion and scarring of the retina. This can result in rapid and severe vision loss. There are some treatment

options to slow down the progression of wet macular degeneration. These include anti-angiogenesis drugs, photodynamic therapy and laser therapy.

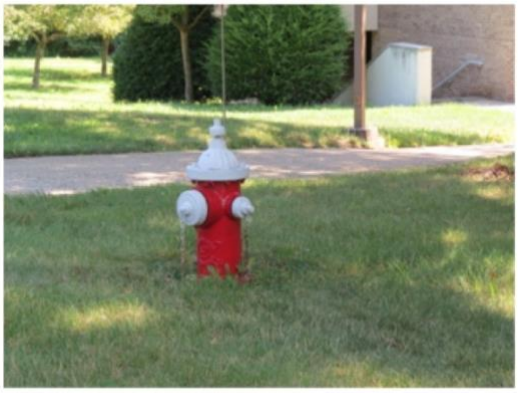
Risk factors for age related macular degeneration include age, race (more common in Caucasians) and family history.

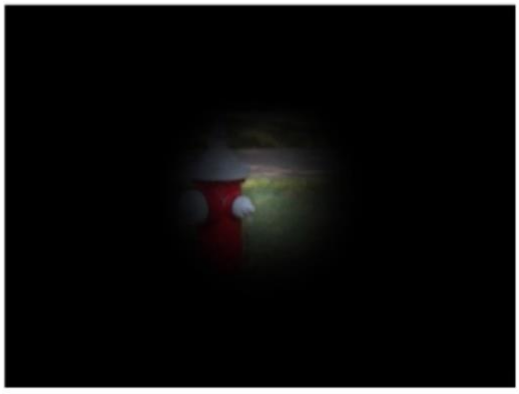
Practical Implications

A person who is dealing primarily with AMD will most likely be able to move around, communicate and complete many daily tasks independently or with some accommodations because they will compensate for their visual condition through use of their residual vision and their hearing. For example, a person with macular degeneration may not see a new person enter the room, but they may hear them or see other people looking toward the door.

A person who has AMD in combination with a hearing loss or deafness faces a greater challenge as they can't rely on their hearing to compensate for their vision loss. This can result in mobility challenges, communication breakdowns and the inability to complete daily tasks. For people who are Deaf who communicate through sign language, AMD can cause serious issues with receptive communication as there may be several spots in their visual field where they can't see the signs. Strategies such as tracking or tactile sign language may help ensure clear communication.

Retinitis Pigmentosa





Retinitis pigmentosa (RP) is a group of rare, genetic disorders that cause a breakdown and loss of cells in the retina. The disorder affects 1 in 4000 people worldwide. Retinitis pigmentosa is caused by genetic mutations and harmful changes to any one of more than 50 genes that are responsible for the process of making proteins that are needed in the cells of the retina. The retina is the light sensitive tissue that lines the back of the eye. Common symptoms include difficulty seeing in dim light and at night and a loss of peripheral vision or narrowing of the field of vision. This has also been described as tunnel vision.

Symptoms typically begin in late childhood with difficulty getting around in the dark and slow adjustment to changes in lighting. Field loss may not be noticeable at first, but certain behavioral signs can indicate the onset of field loss. The progression of loss of vision can vary greatly from person to person.

Eventually retinitis pigmentosa can lead to total blindness.

There is no cure for retinitis pigmentosa, but there are some treatments that professionals believe can slow the progression including Vitamin A supplements. There are also adaptive vision aids that can maximize existing vision.

A retinal implant, known as a “bionic eye,” is being used in some cases of RP. The prosthetic device does not cure the disease but can restore partial sight for individuals with RP.

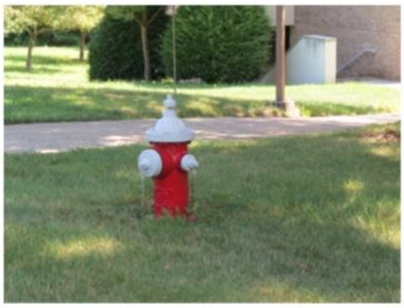
It is a common misperception that RP together with a hearing loss is a result of Usher syndrome. Usher syndrome is a genetic condition (see description under Conditions Related to Deaf-Blindness) that involves hearing loss and RP. However, it is important to note that RP can happen in conjunction with a hearing loss and not be a result of Usher syndrome.

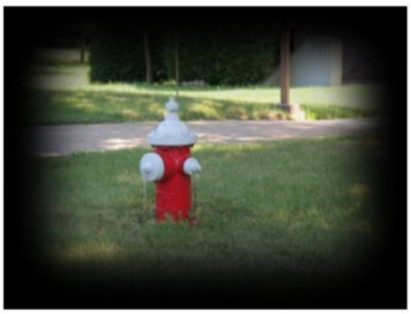
A person who is dealing primarily with RP will likely have many challenges related to mobility and completing daily or work-related tasks. Communication will be impacted because the person may not be able to access facial or body expressions, maintain eye contact or follow the social flow of a conversation with a group. When a person’s fields become severely restricted there are concerns about tripping over objects on the ground or bumping into things on the right, left or above, beyond the visual fields.

Practical Implications

Having RP along with a hearing loss can create intense challenges. A person who is hard of hearing with RP may lose access to the facial expressions or lip movements that they use to assist them in conversations. Someone who is deaf with RP may eventually need to move to tactile sign language because they can no longer see the signs. Strategies such as signing at a distance or signing in a small space may be necessary when RP progresses. At some point, many people who are deaf with RP move to tactile sign language. Sometimes this progression is slow, beginning by only using tactile sign language at night or in low lighting and then moving to full time tactile sign as needed.

Glaucoma





Glaucoma is a group of diseases that causes damage to the optic nerve due to an increase of pressure in the eye. This leads to progressive loss of peripheral vision.

There are several varieties of glaucoma found in adults and children.

Anyone can develop glaucoma, but there is a higher incidence in African Americans over age 40, all adults over aged 60, and those with a family history of glaucoma. However, with early detection and treatment, many people can protect their eyes against serious vision loss.

Glaucoma is typically detected by an eye doctor during a comprehensive eye exam that includes dilation of the eyes. Early detection and treatment can help delay the progression of the disease and vision loss. Glaucoma

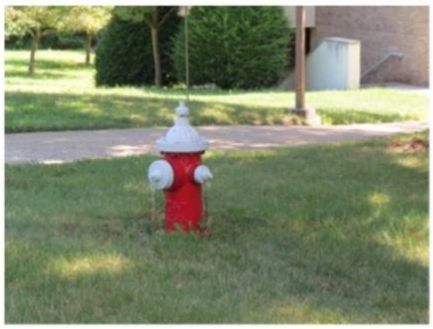
initially affects a person's peripheral vision, eventually leaving them with only central vision. Left untreated or in severe cases when treatment does not work, a person's central vision will likely diminish, and blindness may occur.

There is no cure for glaucoma. Treatment for glaucoma focuses on decreasing the intraocular pressure using medicines such as eye drops, surgery or a combination of both. The goal of any treatment is to prevent loss of vision. It is important to follow treatment guidelines to preserve vision and delay progression of the disease.

Practical Implications

Because glaucoma can progress slowly with little or no initial symptoms, many people don't even realize that they are losing their vision until the disease has significantly progressed. Diagnosis can be a traumatic event for many people as they may not have any idea that they have a vision problem at all. Glaucoma involves loss of visual fields that can progress to total blindness. When communicating with someone who has glaucoma it is important to note that their vision may fluctuate from day to day. As with other vision conditions involving field loss, experiencing glaucoma along with a hearing loss or deafness can cause problems with mobility, communication and other everyday tasks. As with retinitis pigmentosa, a person who is hard of hearing with glaucoma may lose access to the facial expressions or lip movements that they use to assist them in conversations. Someone who is deaf with glaucoma may eventually need to move to tactile sign language because they can no longer see the signs. Strategies such as signing at a distance or signing in a small space may be necessary when RP progresses. At some point, many people who are deaf with glaucoma move to tactile sign language.

Diabetic Retinopathy



Diabetic retinopathy is an eye disease caused by complications from diabetes. It is the most common diabetic eye disease and a leading cause of blindness in American adults. Some people experience changes in the blood vessels of the retina where the vessels swell and leak fluid, while others may experience growth of abnormal blood vessels on the surface of the retina. The retina is the light-sensitive tissue at the back of the eye and is necessary for good vision.

All people with diabetes, both type 1 and type 2, are at risk for diabetic retinopathy. Approximately 40-45% of Americans diagnosed with diabetes have diabetic retinopathy and are at some stage in its progression. Yearly comprehensive dilated eye exams are necessary for those with diabetes to prevent a rapid progression of vision loss.

Symptoms of diabetic retinopathy are varied and there are four identified stages. In the early stages, there often are no symptoms and no pain. Later stages may bring swelling to the macula from leaking fluid or blood vessels growing on the surface of the retina which can bleed into the eye and cause vision loss. Diabetic retinopathy usually affects both eyes. Comprehensive eye exams are necessary to diagnose diabetic retinopathy, identify stages and prevent vision loss.

Early stages do not require treatment, but controlling blood sugar levels, blood pressure and cholesterol are helpful in preventing the progression of diabetic retinopathy. Proliferative retinopathy, the most advanced stage, requires treatment using laser surgery. There are other more invasive surgical treatments, if necessary.

Practical Implications

In advanced stages of diabetic retinopathy blind spots and blurry vision can occur. A person with diabetic retinopathy may have daily changes in vision depending on the amount and extent of blood

leakage and the resulting damage. This may cause extreme anxiety and fear due to the unstable nature of the progression. As a result, planning for accommodations can be challenging as there may be daily changes in vision. It is a good strategy to take time to discuss with the consumer their current visual condition before engaging in any in-depth conversations or intense visual tasks to ensure that the accommodations meet the person's needs at that time.

Retinopathy of Prematurity

Retinopathy of prematurity (ROP) is a leading cause of vision loss and blindness in children. It primarily affects babies weighing 2 ¾ pounds or less that are born before 31 weeks' gestation. The smaller the baby is at birth, the more likely the baby is to develop ROP. Because advances in prenatal and neo-natal medicine have increased the survival of premature babies, an increase in the risk of ROP has been reported and is one of the most common causes of visual loss in childhood and can lead to lifelong vision impairment and blindness. Being diagnosed with ROP also puts infants at risk for developing other eye problems later in their life including retinal detachment, glaucoma, myopia and strabismus.

In American, approximately 14,000 babies are born each year with some degree of ROP. Out of the 14,000 babies, approximately 560 are considered legally blind.

ROP is the result of retinal detachment caused by abnormal blood vessels that grow and spread over the retina. When these blood vessels leak they can cause damage and movement of the retina resulting in a detached retina. Cases with fully detached retinas oftentimes have a poor visual prognosis.

Treatments for ROP includes laser therapy and cryotherapy, both of which can destroy peripheral vision but preserve and maintain central vision, allowing the person to conduct daily activities like driving and reading. Both treatments are invasive and reserved to severe cases of ROP in infants.

Practical Implications

Professionals working with young adults through older adults will see varying degrees of adult ROP. Because there have been advances in the treatment of ROP in infancy over the years, those adults with ROP who are younger will be more likely to have better visual outcomes than those older adults who were not treated early. Because ROP can cause a variety of retinal issues with a range of severity, it is difficult to predict the visual condition of an adult with ROP. It is also important to note that because the root cause of ROP is premature birth, other visual conditions may be present as a result including cortical visual impairment (CVI).

Cortical Visual Impairment (CVI)

Cortical visual impairment (CVI) is a neurological issue that affects the visual part of the brain causing a variety of visual problems. Very often, people with CVI will have a normal eye exam because the physical aspects of the eye are intact and function appropriately. It is possible that someone may have an additional visual problem along with CVI, but that would not explain the abnormal visual behaviors that are associated with CVI.

CVI is caused by damage to the areas of the brain related to the visual process. Traumatic brain injury, stroke, decreased oxygen or blood supply to the brain, and certain neurologic disorders are examples of situations that can cause CVI.

The characteristics of CVI vary from person to person. Some of the most identifiable include:

- Specific color preferences (oftentimes red and yellow)
- Problems attending to visual stimuli
- Delay in responding to visual stimuli
- Light gazing
- Eccentric viewing of objects and people
- Better reaction to visual stimuli that is moving or shiny in color.

Practical Implications

In practical terms, someone with CVI will function as a person who is blind or with a significant vision impairment even though there is likely nothing wrong with their eyes. Because CVI is caused by serious trauma to the brain, it is commonly associated with other physical, cognitive and sensory disabilities.

Because of the complex and individualized nature of CVI, it is advised that professionals working with a person who has CVI consult with a vision specialist with knowledge of the situation to identify the best strategies for working with that individual.

Additional Resources

- National Eye Institute
- Macular Degeneration, Eyesight.org
- Glaucoma, American Academy of Ophthalmology
- Diabetic Retinopathy, American Academy of Ophthalmology