

# Age-related Macular Degeneration

Age-related macular degeneration (ARMD) causes a gradual loss of central vision. It does not affect peripheral vision. Central vision is needed for detailed work and for everyday activities like reading and driving. Depending on the type and severity of ARMD, Vision loss can vary from months to years. The two types of ARMD are referred to as 'wet' and 'dry' ARMD.

'Wet' ARMD is more severe form but responds to treatment better than "dry" ARMD. Visual loss caused by ARMD cannot normally be reversed by medication. New drugs which have come on the market in recent years for wet ARMD can slow down or even halt the progression of visual loss. Therefore early diagnosis is important. ARMD is the most common cause of irreversible sight loss in the western world.<sup>1</sup>

## The eye

To understand the condition and how treatment works it is best to understand the basics of the eye structures and functions.

The retina is located at the back of the eye. The retina is a light-sensitive layer which consists of an inner layer of cells called rods and cones. These cells collect light signals directed onto them and send them as electrical signals to the optic nerve at the back of the eye. Rod cells are concentrated around the edge of the retina. They help see things that are not directly in front, giving a rough idea of what is around. They help with mobility; in short they stop the person from bumping into things not directly in the line of vision. They also enable us see objects in dim light and see movement. Cone cells are concentrated in the centre of the retina where the light is focused by the cornea and lens. This area is called the macula. The macula is a small but vital area of the retina at the back of the eye. It is about 5 mm in diameter. The macula is the part of the retina that is the most densely packed with rods and cones. In the middle of the macula is an area called the fovea which only contains cones.

Cone cells give the more detailed vision needed for activities like reading, watching TV, sewing, driving and recognising people's faces. They are also responsible for most colour vision. There are many more rods than cones, and rods are smaller cells than cones. The cone cells help to see in the daylight, providing the basis for colour vision. The rod cells help to see in the dark (i.e.) night vision.

The optic nerve is made up of thousands of nerve fibres. These fibres pass electrical signals to the brain where they are processed into the images the person is looking at.

The choroid is a layer of tissue behind the retina which contains many minute blood vessels. These tiny blood vessels take oxygen and nutrients to the retina. Bruch's membrane is a thin membrane which forms a barrier between the choroid and the delicate retina. The sclera is the outer thick white layer of the eye.

By looking at an object, light from the object passes through the cornea, then the lens, and then hits the retina at the back of the eye. The light from the object focuses on the macula. A healthy macula is needed for detailed central vision.

## What happens to eye with ARMD?

ARMD occurs when cells in the macula degenerate. Damage to the macula affects central vision needed for reading, writing, driving, recognising people's faces and doing other fine tasks. The rest of the retina used for peripheral vision is not affected. Therefore, without a fully functioning macula, a person can still see enough to get

about, be aware of objects and people, and be independent. However, the loss of central vision will severely affect normal sight. There are two types - 'dry' and 'wet' ARMD.

### **Who does it affect?**

ARMD is the most common cause of vision loss in those aged over 50, however it does not cause complete sight loss. ARMD is rare under the age of 60. If ARMD develops in one eye, there is a high chance (about 6 in 10) that it will also develop in the other eye.

About 1 in 100 people aged 65-75, and about 1 in 8 people aged over 85 have ARMD severe enough to cause serious visual loss. In the over 75 age group, it is twice as common in women compared to men. There are about 7000 new cases of ARMD in Ireland every year with about 60000 people suffering from ARMD in Ireland at the moment.<sup>2</sup> It is possible for macular degeneration to affect younger people, however this is very rare and is caused by a rare inherited genetic disorder.

### **The two types of age-related macular degeneration**

#### **Dry ARMD**

This is the most common type of ARMD accounting for 9 out of 10 cases. In this type the cells in the macula gradually degenerate becoming thinner, ultimately leading to rods and cones in the macula, to degenerate and die. Typically, dry ARMD is a very gradual process and it can take up to 10 years for vision to become seriously affected. Generally those suffering from dry ARMD do not totally lose their ability to read.

#### **Wet ARMD**

Wet ARMD, also called neovascular or exudative ARMD, generally causes severe visual loss over a short time period, sometimes only months. Untreated wet AMD leads to significant visual loss within 3 years.<sup>3</sup> It accounts for about 1 in 10 cases of ARMD. Wet ARMD accounts for 90% of severe sight loss.<sup>4</sup> In wet ARMD, in addition to the retinal pigment cells degenerating, new tiny blood vessels grow from the tiny blood vessels in the choroid. The new vessels break through Bruch's membrane and into the macular part of the retina. These vessels are not normal so are fragile and tend to leak blood and fluid. This leads to damage to the rods and cones and scarring in the macula, causing more accelerated vision loss than would occur in simple ARMD. Rarely if there is a bleed from a new blood vessel, this visual loss can occur suddenly, within hours or days. The fluid from the leaking vessels may accumulate which can lead to retinal detachment.

Both wet and dry ARMD are classified according to severity. Early, intermediate or advanced types refer to the degree of damage to the macula. Wet ARMD is always considered advanced ARMD.

### **Causes**

With dry ARMD, advancing age causes the transport of nutrients to the rods and cones and the clearance of waste materials and by-products made by the rods and cones to gradually slow. This eventually results in tiny abnormal deposits called drusen developing under the retina. With time the retinal pigment cells and their nearby rods and cones degenerate, stop working and die. With wet ARMD, something also triggers new blood vessels to develop from the choroid to cause the wet form of ARMD.

The trigger is not known. It may be that some waste products which are not cleared may stimulate new blood vessels to grow in an attempt to clear the waste.

## **Risk factors**

The exact reason why cells degenerate in people with ARMD is not known. However certain risk factors may increase the risk of developing ARMD:

- Smoking is one of the biggest causes
- A family history of ARMD. While ARMD is not a straightforward hereditary condition, it does appear to run in families.
- Sunlight. This has yet to be proven, however studies suggest that the retina is damaged by UVA and UVB rays from sunlight.
- High blood pressure may be a factor.<sup>5</sup> This has yet to be conclusively proven.
- High alcohol intake may play a role but this has not been proven yet

ARMD seems to be more common in people from Caucasian (white) and Chinese racial backgrounds than from other racial groups. While diabetes can cause eye problems, it is not a cause of ARMD.

A 2010 study demonstrated that a combination of healthy eating, physical activity and not smoking was associated with 71% lower risk for ARMD compared with those who smoked and did not eat healthy or exercise.<sup>6</sup>

The recent Irish study, directed by Professor Stephen Beatty and Dr. John Nolan at the Waterford Institute of Technology and supported by Irish charity Fighting Blindness investigated macular pigment, which is a dietary pigment obtained from eating fruits and vegetables. Macular pigment is believed to protect against AMD as it filters blue light which causes damage to the macula and neutralises unstable molecules which are believed to contribute to AMD. The findings have confirmed that high levels of body fat reduce this protective eye pigment and that weight loss can significantly increase it, suggesting that weight loss may lead to additional protection against AMD.<sup>7</sup>

## **Symptoms**

The main early symptom of dry ARMD is blurring of central vision despite using prescription glasses. In the early stages of the condition the person may notice that words in a book or newspaper may become blurred, brighter light is needed to read, colours appear less bright and the person has difficulty recognising faces.

Many of the symptoms of wet ARMD include the symptoms of dry ARMD described above; however there are some more serious symptoms specific to wet ARMD. The deterioration of central vision in wet ARMD may be preceded by floaters caused by leaking blood vessels. A specific early symptom for wet ARMD is distortion of straight lines. Typically, straight lines appear wavy or crooked. For example, the lines in a note book, the lines between bricks in a brick wall, frame of a doorway or the border of any other straight object may not look straight. A 'blind spot' can also develop in the middle of the visual field. This tends to become larger over time as more and more rods and cones degenerate in the macula.

Visual hallucinations are common with most types of severe visual loss. Visual hallucinations such as seeing shapes or complex colour patterns can occur with severe ARMD. These visual hallucinations typically improve within 18 months even without treatment but in some people they last for years. There are occasions when these visual hallucinations are put down to a mental illness or early Alzheimer's disease in the older person so diagnosis of ARMD is important. The diagnosis of ARMD can therefore provide relief in these cases. ARMD is a painless condition.

It is important to see a doctor or optometrist promptly if visual loss or visual distortion develops. Peripheral vision is not affected with ARMD and so it does not cause total blindness. If the vision is affected in one eye only, there may not be any symptoms as the other good eye often compensates; 70% of people with wet ARMD experience symptoms in both eyes. Older people should have regular eye checks to check for early ARMD and to check for other eye conditions such as glaucoma and cataracts.

## **Other causes of vision loss**

Other than ARMD, other painless causes of vision loss include<sup>8</sup>:

- Refractive errors
- Cataracts
- Some corneal diseases
- Posterior vitreous detachment or retinal detachment
- Retinal artery occlusion or retinal vein occlusion
- Central serous retinopathy
- Cerebrovascular disease (eg) Stroke
- Pituitary and other neurological tumours
- Some drugs or chemicals (including methanol, chloroquine, hydroxychloroquine, isoniazid, thioridazine, isotretinoin, tetracycline or ethambutol)
- Diabetes (diabetic maculopathy).
- Type 2 membranoproliferative glomerulonephritis

## **Diagnosis**

If a doctor or optician notice symptoms suggesting ARMD, referral to an ophthalmologist is normal. This should be done urgently if there are any signs of wet ARMD. If the ophthalmologist finds signs of visual distortion, then ARMD is a likely cause of the visual problem. The ophthalmologist will examine the back of the eye with a slit lamp microscope looking for the typical changes that occur with dry ARMD and wet ARMD. If wet ARMD is diagnosed or suspected, a further test called fluorescein angiography may be done. For this test a fluorescein dye is injected into a vein in the arm. Under a specialised camera, this test can give the ophthalmologist an indication of the extent and severity of the condition, checking if there are any dye leaks into the macula from leaky blood vessels, usually an indication of wet ARMD.

A test called ocular coherence tomography is becoming more commonly used. This is a noninvasive test that uses special light rays to scan the retina. It can give a detailed '3D' view of the macula and can show if the macula is thickened or abnormal. This test is useful if there is doubt about whether ARMD is the wet or dry form. It is also a useful test to assess and monitor the results of any treatment.

## **Treatment**

There is no specific treatment yet for the more common dry ARMD. There are certain actions that can maximise the sight the person still has and to improve eye health. An optician will help advice on maximising sight through using glasses and lenses. Stopping smoking and protecting the eyes from the sun's rays by wearing sunglasses are important.

## **Can diet and dietary supplements slow down ARMD?**

A healthy balanced diet rich in antioxidants can be beneficial. This involves eating a diet rich in antioxidant vitamins and carotenoids found in green leafy vegetables such as spinach and kale, eggs, and fresh fruit.

There is some evidence that a diet high in vitamin A, C, and E as well as a carotenoid called lutein may slow the progression of dry AMD and reduce risk of getting wet AMD.<sup>9</sup> Foods high in vitamin A, C, and E include oranges, kiwi, green leafy vegetables, tomatoes and carrots. Green leafy vegetables, peas, mangos, and sweetcorn are also a good source of lutein. In 2001, a study by the National Institutes of Health and the National Eye Institute in America called the *Age-Related Eye Disease Study* (AREDS) showed that high levels of antioxidants and zinc significantly reduce the risk and symptoms of the ARMD.<sup>10</sup>

### **Vitamin supplements to prevent ARMD**

Supplements especially designed for macular degeneration contain antioxidants which can slow down the progression of both dry and wet ARMD. They are thought to be most beneficial to those with either advanced ARMD or with vision loss (due to ARMD) in one eye. Various products are available to buy over-the-counter (OTC) in pharmacies.

### **How they work?**

There is an important and naturally occurring pigment at the back of the eye, known as macular pigment. Macular pigment has been shown to prevent the development of macular degeneration. This pigment is made up of 3 dietary compounds, known as lutein, zeaxanthin and meso-zeaxanthin. Lutein and zeaxanthin are dietary carotenoids that filter damaging high-energy blue wavelength light from the visible-light spectrum by as much as 90%. Lutein and zeaxanthin are antioxidants and therefore protect against the damage caused by free radicals. Meso-zeaxanthin is the 3rd component of the macular pigment and is only found at the centre where vision is sharpest; lutein is involved in its formation. It is not found in a typical diet but is found in shrimps and certain fish types like halibut, salmon and sea bass. Meso-zeaxanthin is being recognised as a more powerful neutraliser of free radicals than either lutein or zeaxanthin. In Whelehans pharmacy, we recommend Lutein Omega 3<sup>®</sup> as the best all round supplement to prevent macular degeneration. It is also the best value supplement for macular degeneration in Ireland; the 2 month pack costs less than €10 per month, meaning it costs at least 50% less than other leading brands. Lutein Omega 3<sup>®</sup> contains Lutein, Zeaxanthin, Vitamins C and E, Copper, Zinc, combined with fish oil rich in DHA.

Some studies suggest that consumption of fish and foods rich in omega-3 fatty acids may be associated with a lower risk of AMD.<sup>11</sup> However; there is currently insufficient evidence to support their routine consumption as a preventative measure for ARMD.

### **Are supplements allowed on medical card?**

These eye supplements are not licensed medicines and generally are not available directly on the GMS or DPS scheme. However, they are available for free on the Hardship Scheme for GMS (medical card) patients in many areas once they are initially prescribed by a consultant ophthalmologist and the pharmacist can show proof of this to their local health centre. A copy of the ophthalmologist's prescription for the dietary supplement is usually sufficient for this.

### **Other treatment options**

For the less common wet ARMD, there are now treatments may halt or delay the progression of visual loss in some people. Newer treatments may even be able to reverse some of the visual loss. Treatment options include anti-vascular endothelial growth factor (anti-VEGF) drugs, photodynamic therapy and laser photocoagulation. In general with these treatments, about 80% of patients experience a slowing of the progression of vision loss. The aim is to preserve central vision but most patients will

still have some difficulty in reading small print and other visually demanding tasks, even with treatment.<sup>3</sup> The distortion of straight lines (straight lines appear wavy or crooked) can persist in some people even after treatment.

### **Anti-VEGF drugs**

A class of drugs called anti-VEGF drugs has been developed in recent years for wet ARMD. Vascular endothelial growth factor is a chemical that is involved in the formation of new blood vessels in the macula, a cause of wet ARMD. Blocking the action of this chemical prevents the formation of the abnormal blood vessels. Anti-VEGF drugs include ranibizumab (Lucentis<sup>®</sup>), pegaptanib (Macugen<sup>®</sup>), and bevacizumab (Avastin<sup>®</sup>). Others are being developed but, due to the new technologies involved, these drugs are very expensive.

In the UK, The National Institute for Health and Clinical Excellence (NICE) evaluated these therapies in August 2008 and gave approval to ranibizumab. Because it is such an expensive drug, very specific criteria have been set out by NICE to determine which patients are eligible for treatment. NICE continue to monitor clinical trials and other anti-VEGF may be allowed on the NHS in the future. Anti-VEGF drugs are injected using a fine needle directly into the vitreous humour of the eye. Ranibizumab injections are needed every four weeks.

Ranibizumab improves vision in about 1 in 3 people treated. However, treatment in most people will maintain vision and prevent the condition from getting worse. Anti-VEGF drugs do not work at all in about 1 person in every 10 treated. Because of cuts to health budgets in Ireland in recent years, eye consultants in are finding it difficult to get funding to treat patients with Ranibizumab because of its cost. Dr Noel Horgan, a consultant ophthalmic surgeon at the Royal Victoria Eye and Ear Hospital in Dublin was quoted in an article by Fiona Gartland in the Irish Times in May 2012 that he can no longer obtain Ranibizumab even though it was available up to the end of 2011. He said most hospitals in the country no longer stock it because of its cost.<sup>12</sup> Ranibizumab costs approximately €1000 per injection; however it is covered through some private health insurance plans. Your ophthalmologist and health insurer will provide you with more details.

### **Photodynamic therapy**

This is a technique that was developed in the late 1990s. It involves a drug called verteporfin being injected into a vein in the arm. Within a few minutes the verteporfin binds to proteins in the newly formed abnormal blood vessels in the macula. A light at a special wavelength is then shone into the eye for just over a minute. Verteporfin is a photosensitive drug. This means that when light is shone at the blood vessels coated with verteporfin, the verteporfin activates and causes damage, destroying the abnormally growing blood vessels without damaging the nearby rods and cones or any normal blood vessels.

It is generally less successful in maintaining current vision than anti-VEGF drugs and needs to be repeated approximately every three months to continue to suppress newly growing blood vessels. In 1-4% of cases, it can result in a temporary loss of vision and, in a small number of these patients; the loss can be severe and permanent. It depends on exactly where the new blood vessels are growing and their extent. Unlike anti-VEGF treatment, there is no chance that photodynamic therapy will restore vision already lost. However it can prevent vision loss from getting worse. Photodynamic therapy causes less damage to the normal retina than laser photocoagulation.

### **Laser photocoagulation**

For this procedure, a fine laser is directed at the tiny new 'abnormal' blood vessels that are forming. This destroys the developing new blood vessels, preventing the condition from getting worse. A major disadvantage of laser photocoagulation is that people undergoing this treatment will develop a permanent black or grey patch affecting their vision and no sight is restored. Laser photocoagulation is only suitable for a small number of cases. It depends on exactly where the new blood vessels are growing, as the laser may also damage the rods and cones and there is a risk of vision loss.

### **Future treatments**

Anti-VEGF drugs are the most successful treatment option for wet ARMD developed so far. Treatments such as radiation therapy, other drugs, and surgery to the retina are being investigated. For example, a surgical technique where a part of the peripheral retina is grafted into the diseased macular area is being investigated. Major pharmaceutical firms, including Novartis and Pfizer continue to invest significantly into research on the causes of ARMD with the hope of developing better screening tools and improved treatments so treatment may well improve in the near future.

### **Practical help**

Regular sight tests are even more important in over 60's. A person should visit their optician for a sight test every two years, even if there is no change in vision. Early detection of problems often allows more effective treatment.

Practical devices which can make a person's life easier are magnifying lenses, large print books, and bright lamps which may assist reading. Gadgets such as talking watches and kitchen aids aid independent living.

### **Entitlements if you are registered blind**

If your level of vision means you fulfill the registration criteria as a person who is blind or vision impaired, you may be automatically entitled to a companion free travel pass, a disabled person's parking card and Eircom free directory enquiry service. If you are working and have a guide dog user, you are also eligible to get tax credits. You do not have to be 100% blind to be registered blind, having significant vision loss that effects your quality of life can entitle you to be registered blind and benefit from some entitlements because of your disability.

If you are over 66 and already have an old age free travel pass, a companion free travel pass can be issued to you by returning your pass along with a letter from NCBI confirming that you are registered blind. Apply to the Free Travel section of the Department of Social Protection or LoCall 1890 500 000 Ext 48345. For more information on the disabled person driving card, you can contact the Disabled Drivers Association of Ireland at 094 9364054. You can apply for the free directory enquiry service by calling Eircom at 1800 574 574. To apply for tax credits for a guide dog, you will need a letter from the Irish Guide Dogs for the Blind to accompany your claim to your local tax office.

If you are registered as blind with the National Council for the Blind (NCBI), there are a number of other entitlements you can apply for. You may be entitled to the blind person's pension, to qualify for this payment; you must be registered with NCBI, be over 18 years of age, be resident in the Republic of Ireland and undergo a means test.

Other entitlements you can apply for are free electricity allowance or natural gas allowance or bottled gas refill allowance, telephone allowance and free television license. To apply for any for these entitlements, contact the Department of Social Protection at LoCall 1890 500000 Ext 3371 or check their website, [www.welfare.ie](http://www.welfare.ie).

If you get either the blind person's pension or old age contributory or non-contributory pension you may also be eligible to receive the blind welfare allowance administered through the Health Service Executive. This allowance is paid once a month and is subject to a means test. You can apply for this allowance through your local HSE Health Centre.

A public health nurse can arrange home help and other services which can help you live independently and more easily at home. You can also organise for an Occupational Therapist to visit your home to advise on practical ways, including devices which help make living with sight loss easier.

Depending on your level of vision impairment, you may or may not be allowed to drive. You will be required to fill out an eyesight report which must also be completed by you optician or doctor. This form must be submitted with your local motor tax office. Contact your local Motor Tax office for more details. The Road Safety Authority of Ireland has more information on their website, [www.rsa.ie](http://www.rsa.ie).

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