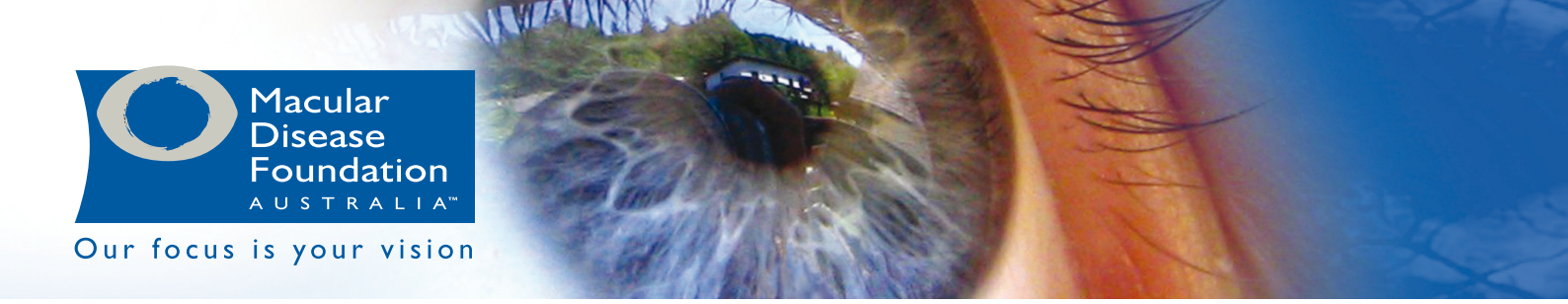

Macular Disease Foundation AUSTRALIA™

Our focus is your vision

Macular Telangiectasia ("MacTel")

Macular Telangiectasia, commonly known as MacTel, is a curious and poorly understood disorder of the blood vessels which supply the central part of the retina that lines the back of the eye (the macula). The fovea is the centre of the macula, and has no blood vessels at all because they would interfere with central vision. With MacTel, the blood vesselas around the fovea become dilated (swollen) and incompetent, like varicose veins but on a much smaller scale. The retina deteriorates and the structure becomes scarred due to the development of liquid-filled cysts, which impairs nutrition of the light-sensitive photoreceptor cells affecting central vision.

While MacTel does not usually cause total blindness, it can lead to 'legal blindness' where there is major loss of the central vision, which is required for reading and driving. MacTel generally develops slowly, over a period of 10 to 20 years.

What is the macula?

The **macula** is the name given to the area at the very centre of the retina. This region is responsible for detailed central vision and most colour vision. It is responsible for the ability to read, recognise faces, drive a car, see colours clearly, and any other activity that requires fine vision. The rest of the retina is called the peripheral retina. It is used to see general shapes and provides 'get-about' vision, which is also called side vision or peripheral vision.

There are two main types of MacTel

Type 1 MacTel involves localised dilations, or aneurysms, of blood vessels in the macula, though the aneurysms may also occur in blood vessels outside the centre of the retina. Aneurysms in the blood vessels cause damage and swelling to the cells of the macula and blood supply is further disrupted. Bleeding from the dilated capillaries sometimes occurs, as well as lipid (fatty) deposits from the leaking blood. New blood vessel growth ("neovascularisation") in response to damage is not usually observed in type 1 MacTel.

Type 2 MacTel is also called idiopathic juxtafoveal telangiectasia or idiopathic perifoveal telangiectasia. Idiopathic means 'of unknown cause'. Juxtafoveal or perifoveal refer to the abnormalities in the blood vessels near, or around, the fovea of the macula. Type 2 MacTel involves widespread dilation and leakage of these vessels.

As damage occurs, new blood vessels form within the macula and subsequently break or leak. The end result is the formation of scar tissue over the macula and the fovea. This series of events drastically impacts the area of central vision.

Features of MacTel

In the early stages of MacTel, the signs can be very subtle, meaning that the diagnosis may be missed by optometrists and ophthalmologists.

Signs and symptoms of MacTel may include slow loss of vision, distorted vision, trouble reading, and **scotoma** (a spot in the visual field in which vision is absent or deficient).

The condition may remain stable for extended periods, sometimes interspersed with sudden decreases in vision.

Patients with MacTel tend to have a significantly higher prevalence of systemic conditions associated with vascular disease, including history of hypertension, history of diabetes, and history of coronary disease.

Treatment of MacTel

Treatment options are limited. To date, no treatment has been shown to prevent progression. The variable course of progression of the disease makes it difficult to assess the efficacy of treatments.

Retinal laser photocoagulation is generally not helpful as the leaking vessels are normally located very close to the fovea and laser would damage the central vision even further.

The use of vascular endothelial growth factor (VEGF) inhibitors which have proven so successful in treating age-related macular degeneration, has been investigated in pilot studies for treatment of subretinal neovascularisation in MacTel. Although anti-VEGF drugs given before the development of new blood vessels under the retina can dramatically reduce the vascular leak, the loss of photoreceptor cells (and hence vision) may still proceed in treated eyes.

The MacTel Project

The MacTel Project is a collaboration involving more than 30 centres from around the world. The project encompasses a natural history observation study that identifies MacTel patients and follows the progression of their disease; a genetics study of MacTel patients and their family members designed to identify genes and genetic variants that may be associated with susceptibility for MacTel; and an eye donor program to study the histology and pathology of MacTel eyes. Patients interested in registering in the MacTel Project should contact the Foundation for details.

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