

Common Retinal & Ophthalmic Disorders

Cataract
Central Serous Retinopathy
Cystoid Macular Edema (Retinal Swelling)
Diabetic Retinopathy
Floaters
Glaucoma
Macular degeneration
Macular Hole
Macular Pucker - Epiretinal Membrane
Neovascular Glaucoma
Nevi and Pigmented Lesions of the Choroid
Posterior Vitreous Detachment
Proliferative Vitreoretinopathy (PVR)
Retinal Tear and Detachment
Retinal Artery and Vein Occlusion
Retinitis
Uveitis (Ocular Inflammation)
White Dot Syndromes

Anatomy and Function of the Eye

(Short course in physiology of vision)

Cataract

Overview

Any lack of clarity in the natural lens of the eye is called a cataract. In time, all of us develop cataracts. One experiences blurred vision in one or both eyes – and this cloudiness cannot be corrected with glasses or contact lens. Cataracts are frequent in seniors and can variably disturb reading and driving.

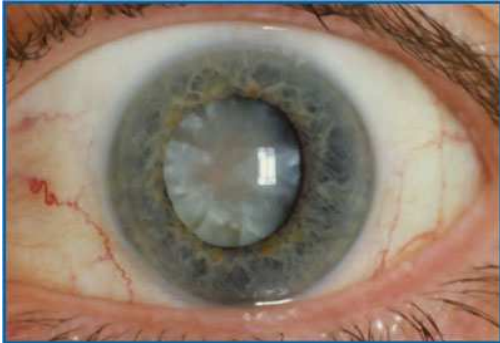


Figure 1: Mature cataract: complete opacification of the lens.

Cause

Most cataracts are age-related. Diabetes is the most common predisposing condition. Excessive sun exposure also contributes to lens opacity. Less frequent causes include trauma, drugs (eg, systemic steroids), birth defects, neonatal infection and genetic/metabolic abnormalities.

Natural History

Age-related cataracts generally progress slowly. There is no known eye-drop, vitamin or drug to retard or reverse the condition.

Treatment

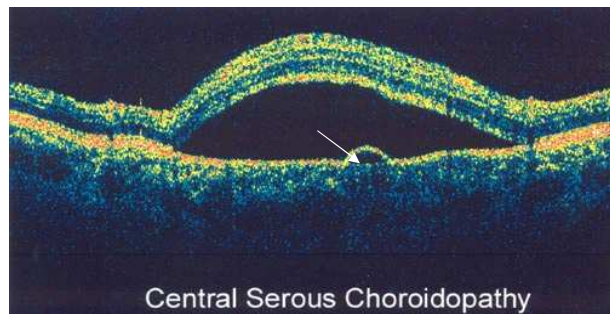
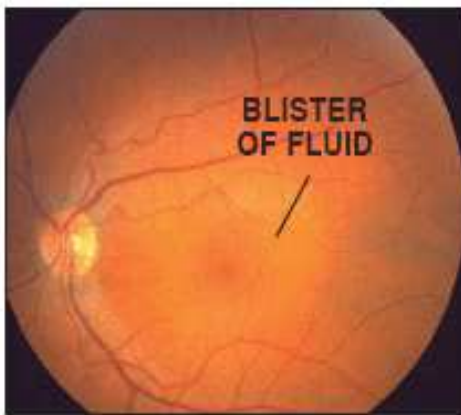
Surgery is the only option. Eye surgeons will perform cataract extraction when there is a functional deficit – some impairment of lifestyle of concern to the patient.

Central Serous Retinopathy (CSR)

Overview

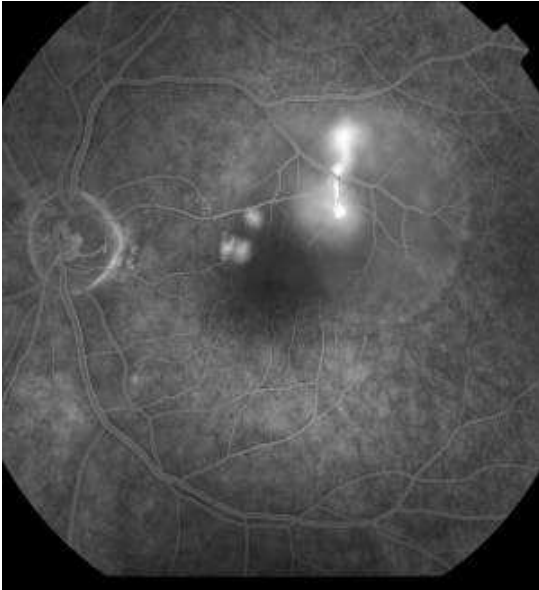
Central serous retinopathy is a condition in which a blister of clear fluid collects beneath the macula to cause acute visual blurring and distortion (Figure 2).

Central serous retinochoroidopathy



Left: Accumulation of clear fluid beneath the retina.
Right: Optical coherence tomography demonstrates separation of retina over a small detachment of the pigment epithelium (arrow).

Central serous retinopathy-angiography



Fluorescein dye study - highlights the site of focal leakage of dye underneath the retinal blister. Leaking dye fills the localized retinal detachment.

Cause

No precise cause has been identified. Males in the 20 - 40 year-old age group are most notably affected, especially if they are high strung type “A” personalities. Steroid use has been implicated as a cause in some individuals.

Natural history

The blisters tend to dry up and disappear in the majority of cases over several months. Central serous recurs in about 25% of cases.

Treatment

With persistent or recurrent disease, **fluorescein angiography** may help identify a well defined leak at the level of the RPE. If outside the **fovea**, a gentle application of **laser** helps to stimulate a healing response to seal the leak. Recent articles suggest that the **intra-vitreous** injection of Avastin may be beneficial.

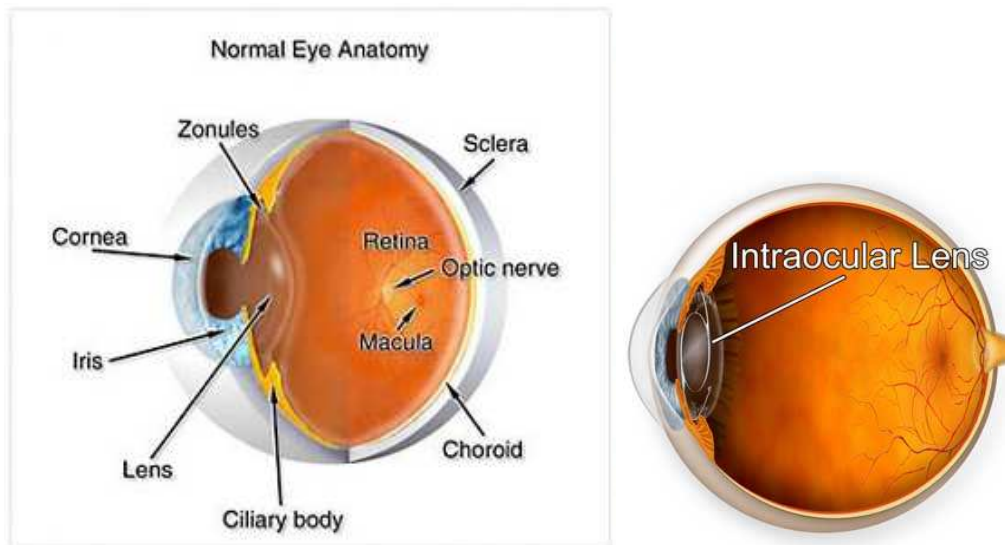
Cystoid Macular Edema (CME)

Overview

Macular edema (swelling) is a consistent finding in many disorder of the retina and is especially common in **diabetic retinopathy**, **venous occlusive disease of the retina** (**branch vein occlusion** and **central retinal vein occlusion**) and **retinitis pigmentosa**. **Macular edema** can also develop in a small percentage of patients undergoing **cataract surgery**.

Uncomplicated **cataract surgery** will rarely lead to CME, but when surgery does not go well, CME may limit the recovery of normal acuity. Most cases of CME that follow cataract extraction have disturbed anatomical relationships of the **iris**, **intraocular lens** and **vitreous**. The ideal position for a **lens implant** is within the natural **lens capsule**. Insertion of the lens in the **sulcus** (between the **iris** and the posterior capsule) or in the **pupil** or **anterior chamber** is more likely to produce irritation and macular edema.

Diagram: IOL anatomy

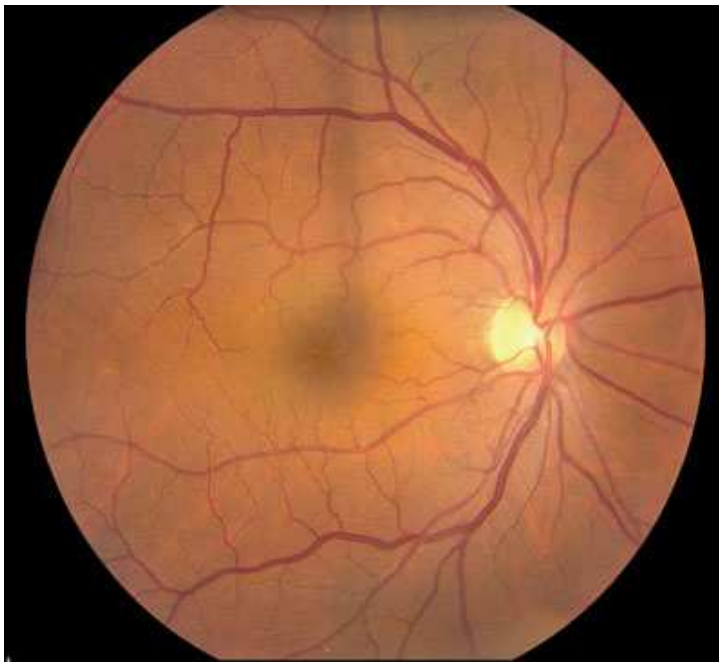


Cause

Common causes: **Cataract surgery/vitreous** loss; **Diabetes**; **Retinal venous occlusion**

Disruption of the **anterior vitreous surface** (“vitreous loss”) during cataract surgery, which produces adhesions to the iris, lens and cornea, is a consistent cause of CME (Irvine-Gass Syndrome). Combined with inflammation related to **retained lens material**, vitreous loss promotes the release of intracellular messenger proteins, which enhance **retinal vascular permeability** and promote leakage and swelling in the macular region (figure 3).

Figure 3. Cystoid macular edema (CME)



Petaloid distribution
of retinal cysts in
the macula.

CME: Fluorescein angiography (left) and OCT (right) delineate cystic retinal swelling



Diabetes damages the [parafoveal](#) capillaries causing either closure or increased permeability with diffuse or focal disease.

Both [branch vein occlusion](#) and [central retinal vein occlusion](#) may cause persistent leakage in the [macula](#) because of vascular congestion.

Natural History

[Macular edema](#) that follows cataract surgery often improves within one year of the procedure and occasionally within the second year. After two years, there is little likelihood of resolution.

[Diabetic macular edema](#) is progressive and aggravated by instability in [glucose control](#) and hypertension (high blood pressure).

[Macular edema](#) secondary to [branch vein occlusion](#) and [central retinal vein occlusion](#) may improve with the natural development of [collaterals](#) and remodeling of the occluded segment.

Chronic (persistent) [macular edema](#) from any cause may lead to permanent damage of the retinal architecture.

Treatment

Irvine-Gass Syndrome (macular edema and ocular inflammation associated with vitreous loss at the time of cataract surgery) Treatment of macular edema may include topical, [periocular injection](#), and [intra ocular injection](#) of [steroids](#) and concomitant use of [non-steroidal anti-inflammatory agents \(NSAIDs\)](#) and [cycloplegia](#) (pharmacological relaxation of the pupil and ciliary body).

[Diabetic macular edema](#) is treated with [focal and grid laser](#) applied to the leaking capillary bed. [Intra vitreal steroids](#) and [Avastin](#) injections are ancillary therapies.

[Macular edema](#) associated with [branch retinal vein occlusion](#) is treated with [laser](#) therapy to the involved retinal sector. [Intra vitreal triamcinolone](#) has been shown beneficial for [central retinal vein occlusion](#). [Avastin](#) (delivered with [intra vitreal injection](#)) may be considered for macular edema secondary to [venous occlusive disease](#).

Diabetic Retinopathy

Overview

[Diabetic retinopathy](#) is caused by alterations in blood flow in the narrowest retinal blood vessels in response to longstanding elevations of blood sugar. The most common group of manifestations, called [background or non-proliferative retinopathy](#), is caused by leakage or closure of the retinal capillaries.

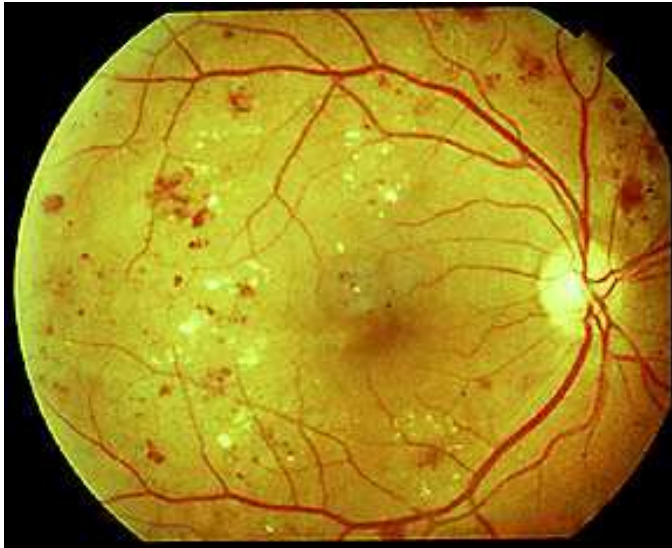
In [proliferative diabetic retinopathy](#), extensive destruction of the retinal capillary network reduces the delivery of oxygen, and stimulates the growth of new blood vessels (via [vascular endothelial growth factor – VEGF](#)) on the surface of the retina. This proliferation of blood vessels (called [neovascularization](#)) may be accompanied by [scar tissue](#) and can lead to bleeding within the central cavity of the eye ([vitreous hemorrhage](#)) as well as [tractional retinal detachment](#).

Well-controlled blood sugar levels retard or prevent the development of [diabetic retinopathy](#).

Cause

[Diabetes](#) leads to damage of both small and large diameter blood vessels throughout the body (figure 4). The severity and prevalence of [diabetic retinopathy](#) is associated with the duration of the diabetic condition and the lack of [glucose control](#) and is accelerated by high blood pressure. Smoking and [heart disease](#) add additional risk for damage to the retinal circulation in diabetics.

Figure 4. Background diabetic retinopathy



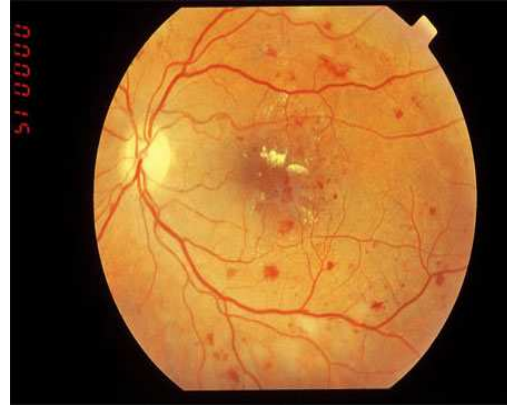
Posterior retina shows scattered intraretinal hemorrhages and exudate.

Natural history

Background (non-proliferative) retinopathy: 95% of diabetics develop the slowly progressive retinal manifestations of background disease.

Early background changes begin in the **macular** region. After 10 years of diabetes, about two-thirds of all adults will show abnormal vessels and small retinal spots of blood. Leakage of blood and/or its fluid components from incompetent vessels into the center of the **macula** causes loss of visual acuity (figure 5). This swelling is called **macular edema**.

Figure 5. Impaired vision in diabetic retinopathy



Areas of blurred vision correspond to swelling and bleeding sites within the retina.

The [capillaries](#) that serve to sustain the macular region may also clot and close – reducing the supply of oxygen and nutrients to the retinal cells. This [ischemia](#) (reduced supply of oxygen) can produce cell death and loss of function in the retina, and become manifest as blurred vision.

Severe changes of [background diabetic retinopathy](#) (figure 6) and [proliferative diabetic](#) disease typically develop after 10-16 years of diabetes.

Proliferative retinopathy: 5% of diabetics develop the potentially catastrophic vascular alterations with proliferative disease. Diabetics requiring [insulin](#) to control their blood sugars are at greatest risk. In proliferative disease, new fragile blood vessels grow onto the surface of the retina ([neovascularization](#)) in response to insufficient blood flow and oxygenation of the tissue. These new blood vessels may spontaneously burst and release blood through the center of the globe ([vitreous hemorrhage](#)), with a sudden and dramatic loss of vision (figure 7).

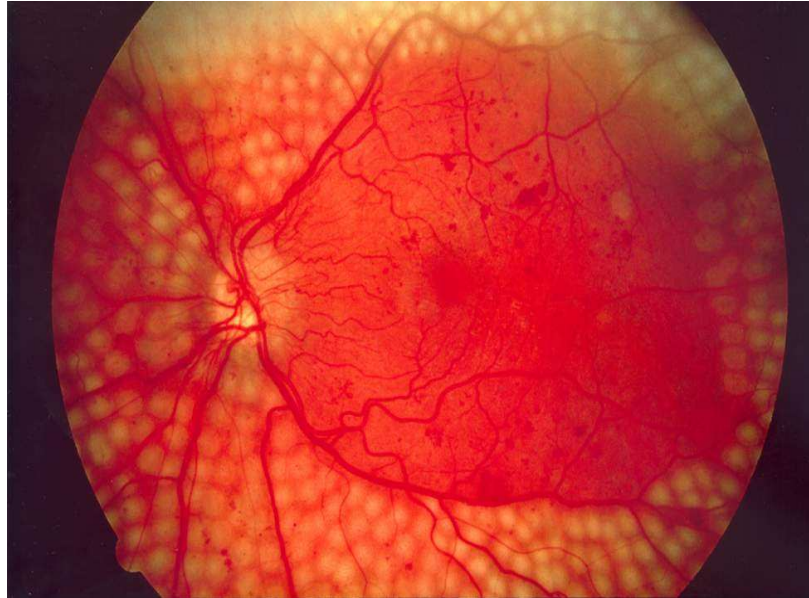
Neovascularization is accompanied by a scaffold of scar tissue that contracts in latter stages of the disease. This scarring process exerts tension on the retina and may cause it to be stretched – and sometimes detached from its normal position (tractional retinal detachment).

Treatment

Macular edema (background diabetic retinopathy). The mainstay of treatment for background disease is to directly cauterize the abnormal capillaries with precisely directly spots of laser (focal laser). In most cases, the surgeon applies a grid of laser spots to the entire region of swollen retina, with special care not to damage the critical central region (foveola). Recent studies support combination treatment with intravitreal injections of Avastin followed by laser.

Proliferative diabetic retinopathy. Neovascularization is treated by the application of 1600 or more individual laser burns to the peripheral retina outside the macular region. (Figure 8.) With the high speed Pascal laser, this may be accomplished in a few minutes with minimal or no discomfort. With conventional lasers, treatment may be staggered over two or more sessions, with or without the injection of local anesthetics around the globe. Freezing treatment (cryopexy) is occasionally used to supplement the laser, especially if the view to the retina is obscured by blood (vitreous hemorrhage), cataract or corneal opacities.

Figure 8. Panretinal laser photocoagulation for proliferative diabetic retinopathy



Hundreds of individual laser burns are applied outside the macula.

Vitreous hemorrhage. Significant vitreous hemorrhage may require evacuation of the vitreous cavity (**pars plana vitrectomy**). Occasionally agents that reduce vascular permeability (steroids or anti-angiogenic compounds) are used alone or in combination with laser, peripheral cryopexy and vitrectomy.

Tractional retinal detachment. Retinal detachments caused by intraocular scarring are repaired with **vitrectomy**. Tiny chisels, scissors, and forceps are employed to separate and remove the scarred elements from the vitreous and retinal surface.

In many cases, **anti-VEGF** agents such as **Avastin**, are used to pretreat or as adjunct therapy for proliferative diabetic retinopathy and its complications.

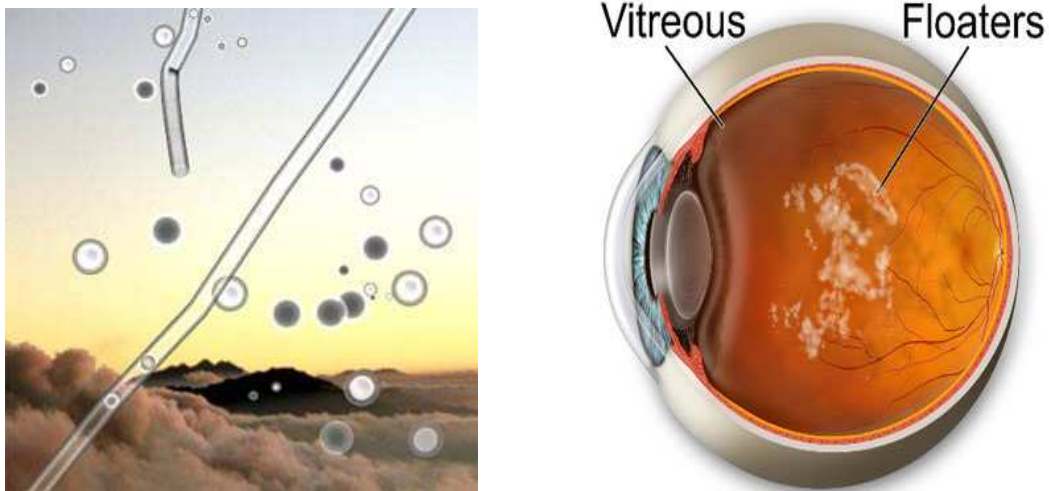
Floaters

Overview

Floaters are clumps of protein in the vitreous cavity of the eye that cast a shadow on the retina, as they float through the space. Everyone has floaters. Most are spots or squiggles that drift through the field of vision, and are best seen when looking at a monochromatic background like a white sheet of paper or a blue sky (figure 9).

The sudden onset of floaters may herald the onset of a retinal tear or detachment, especially if accompanied by flashes of light and or a dark shadow or curtain over the field of vision. A floater may be the first sign of bleeding inside the eye.

Figure 9. Floaters - visual imagery



Strands and clumps protein cast a shadow on the retina as they float through the vitreous (left). Schematic of vitreous floaters (right).

Cause

Floater develop as the vitreous becomes liquified (more watery) with age, allowing fragments of its natural protein filaments to drift. Aging and myopia make the vitreous more watery – and prone to floaters. Posterior vitreous detachment and vitreous hemorrhage may underlie the rapid onset of multiple floaters.

Natural History

Floater appear more regularly as one ages past 40, and are especially frequent in nearsighted individuals, and those who have undergone major eye surgery.

Posterior vitreous detachment, common in seniors, high myopes (highly nearsighted individuals), and those having had cataract surgery, produces a sudden burst of floaters. Sporadically, a posterior vitreous detachment tears or detaches the retina, with the potential to blind the involved eye.

Bleeding into the eye, albeit from diabetes, retinal vascular disease or a new retinal tear, may produce numerous floaters.

Over time, gravity will cause persistent floaters to sink in the vitreous cavity and become less noticeable.

Treatment

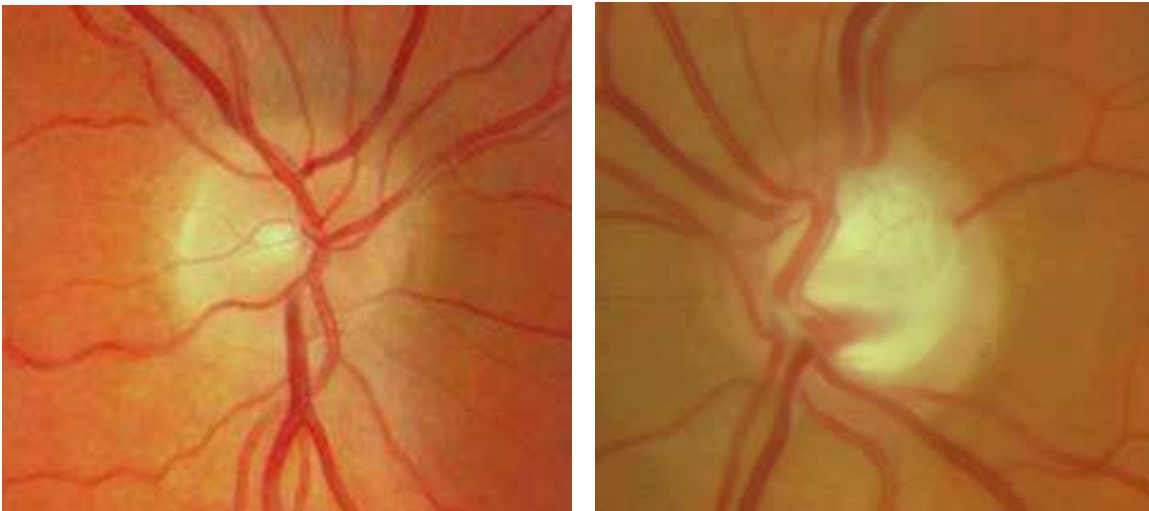
The sudden onset of floaters should be evaluated with a dilated eye examination (to look for retinal tears and detachment).

Glaucoma

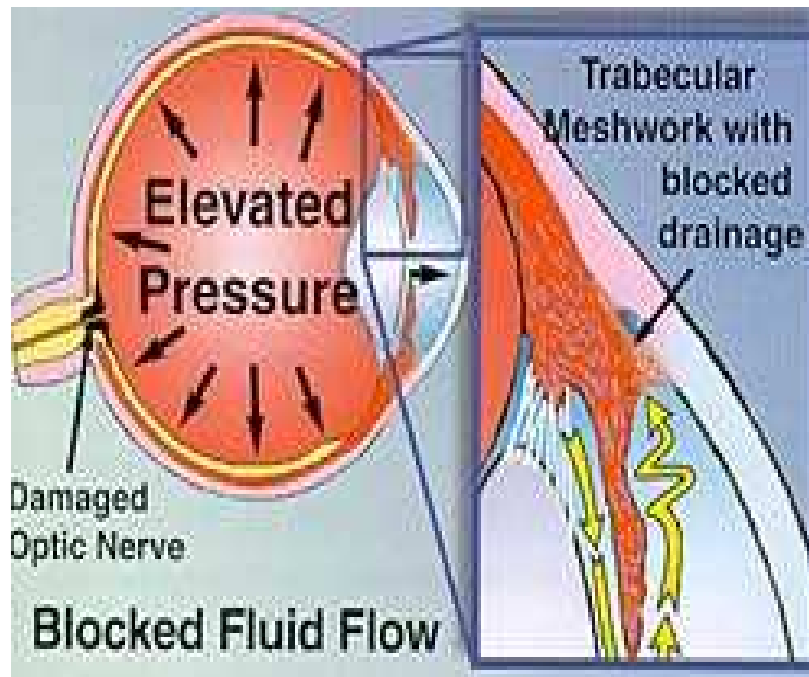
Overview

Glaucoma is a group of conditions with pressure associated damage to the optic nerve. The normal eye maintains a pressure between 4 and 24 millimeters of mercury. If the pressure is elevated outside this range, the optic nerve may become damaged. Most cases develop as one approaches middle-age and progress slowly and insidiously (figure 10).

Figure 10: Chronic glaucoma – Right, normal optic nerve; Left, excavated damaged optic nerve



Outflow obstruction at the angle structures (trabecular meshwork) give rise to glaucoma



Cause

Primary open angle glaucoma – the most common variety. It tends to run in families. Normal drainage of fluid through angle structures (trabecular meshwork and Schlemm's canal) is impeded.

Narrow angle/angle closure glaucoma – narrowing or closure of the angle and trabecular mesh work may elevate the pressure on an acute or chronic basis.

Pseudo-exfoliative glaucoma – associated with the peculiar deposition of protein on anterior eye structures (iris, lens and zonules).

Congenital glaucoma – noted soon after birth with enlargement and clouding of the cornea and tearing.

Low tension glaucoma – shows optic nerve alterations and damage characteristic of glaucoma despite notable elevation in the eye pressure.

Neovascular glaucoma

Natural history

Progressive damage to the optic nerve from glaucoma constricts the field of vision – with loss of perception in the visual periphery. Patients with advanced glaucoma show “tunnel vision.” They can see well straight ahead but nowhere else. Fortunately, in most cases of chronic primary open angle glaucoma, damage and impairment develops gradually. In patients with high pressures in the 30s and 40s (millimeters of mercury), loss of function can occur in weeks to months.

Angle closure glaucoma is uncommon, affecting about one in ten thousand individuals, but develops acutely with a sudden onset of pain in the eye, nausea and a sensation of halos appearing around lights. The iris becomes pushed up against the cornea, blocking the outflow channels and causing an abrupt elevation of the intraocular pressure to dangerous levels. Optic nerve damage develops rapidly unless the angle is deepened.

Treatment

Primary (chronic) open angle glaucoma – One or more topically applied eye drops applied once, twice or even three times daily control the eye pressures in most patients. Patient's with uncontrolled pressures or progressive damage to the optic nerve may undergo a [filtering procedure](#) which creates a new outflow channel for the over-pressurized globe.

A prophylactic hole in the peripheral iris called a laser iridotomy can prevent angle closure in susceptible individuals with narrow angle anatomy. This is a painless office based procedure in which a highly focused pinpoint laser beam breaks open a tiny hole in the peripheral iris.

Macular degeneration

Overview

Age-related macular degeneration (AMD) is a progressive disorder that causes central visual loss in seniors in their late 60s and beyond. It is the leading cause of irreversible blindness in the developed world. The visual impairment ranges through a wide spectrum, with minor difficulty in reading in its earliest forms, and profound loss of central vision precluding reading, driving and identifying faces with advanced disease.

Retinal specialists categorize AMD in two forms—wet and dry. Dry AMD is more common—more than 8 out of 10 AMD patients have dry AMD. About 1 in 10 develop the severe wet form. Over 160,000 individuals in the United States are diagnosed with wet AMD every year.

Cause

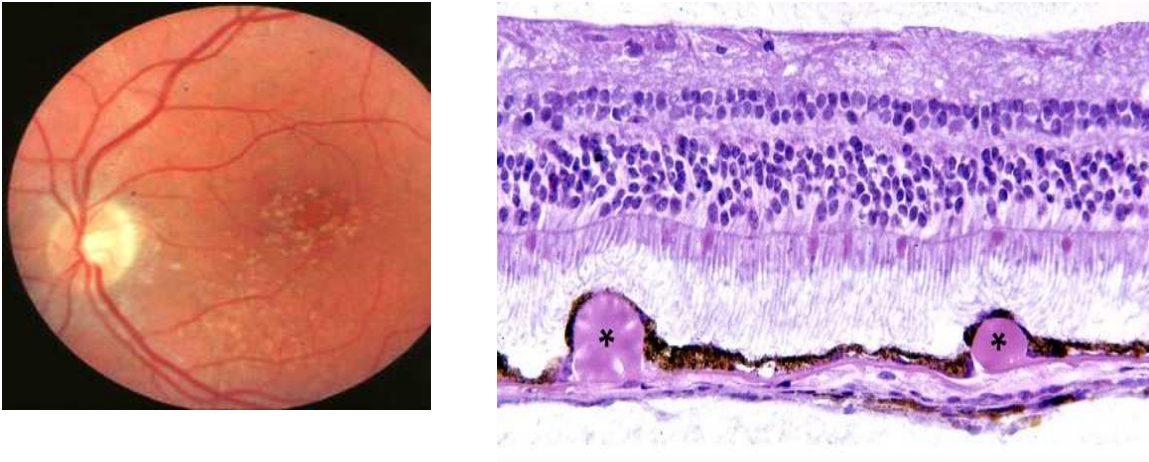
Past analyses have highlighted factors of pigmentation and light toxicity. (AMD is more likely to develop in individuals with light eyes and skin pigmentation and those with notable sun exposure.) New studies have stressed the importance of sufficient zinc and anti-oxidants in the diet to retard the progression of damage. Smoking increases the risk for AMD. Obesity and high blood pressure have also been identified as risk factors. Regular fish consumption (once/week) is associated with lesser rates of disease.

Genetic studies have identified genes and markers for relative increased risk of developing AMD. The [complement](#) factor H (CFH), involving genes that regulate inflammation (via the alternate [complement](#) pathway), plays a key role. At the cellular level, heat shock proteins, specifically alpha-B-crystallin levels, reflect local [stress factors](#) at the level of the RPE, and may be a biomarker for the development and progression of the disease.

Natural history

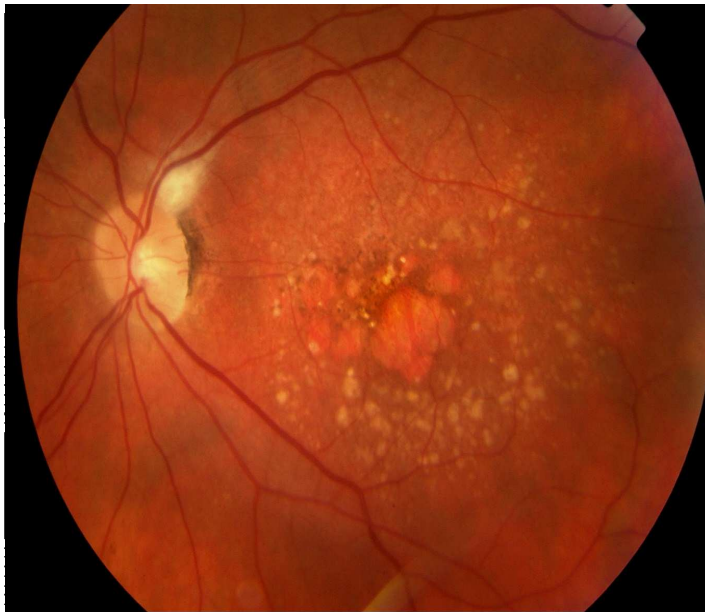
DRY AMD: The most common form of AMD is called the dry type – the loss of function is associated with visible damage to the retina, but without bleeding or leakage of fluid from the layers beneath the retina (figure.) Dry macular degeneration includes a spectrum of disorders of the outer retina and retinal pigment epithelium including drusen (figures) pigment alteration and atrophy, which cause variable loss of central retinal sensitivity. Dry AMD is slowly progressive, but may rapidly deteriorate into the more severe wet form (figures 11-13).

Figure. 11 Drusen



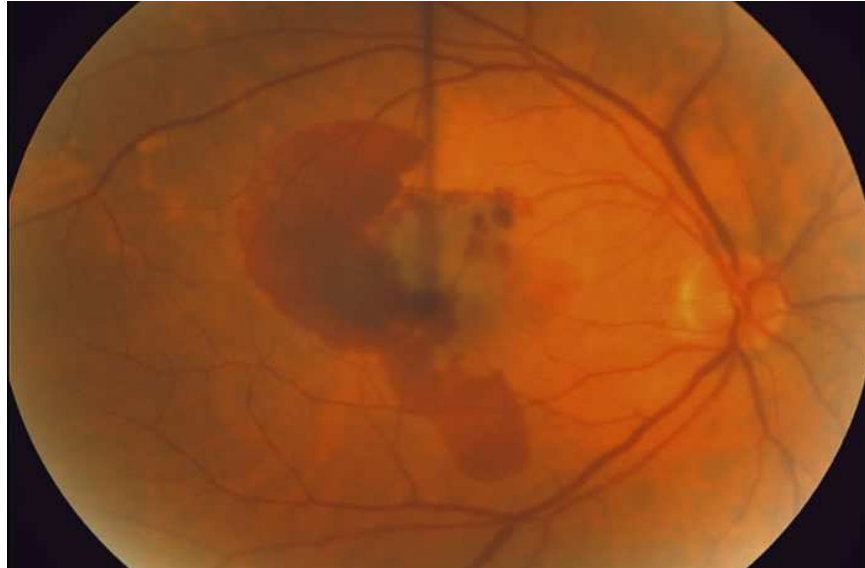
Drusen, nodules beneath the retina, are an early stage of dry AMD (left). Under the microscope, drusen (stars) lie with Bruch's membrane, beneath attenuated and atrophic pigment cells (right).

Figure 12. Dry age-related macular degeneration (AMD): Geographic atrophy of the retinal pigment with surrounding drusen. Central macular involvement is associated with profound loss of visual acuity.



WET AMD: In the more severe and acute wet type, blood, fluid and/or scar tissue accumulate beneath the retina with substantial injury to the central retina. Wet macular degeneration is caused by a failure of the pigment epithelium and the [Bruch's membrane](#) complex to contain blood, fluid and blood vessels away from the subretinal and submacular space. The in-growth of fine blood vessels from the choroid into the subretinal space through breaks in Bruch's membrane (subretinal neovascularization) is a consistent feature of wet AMD (figure 13).

Figure 13. Wet age-related macular degeneration

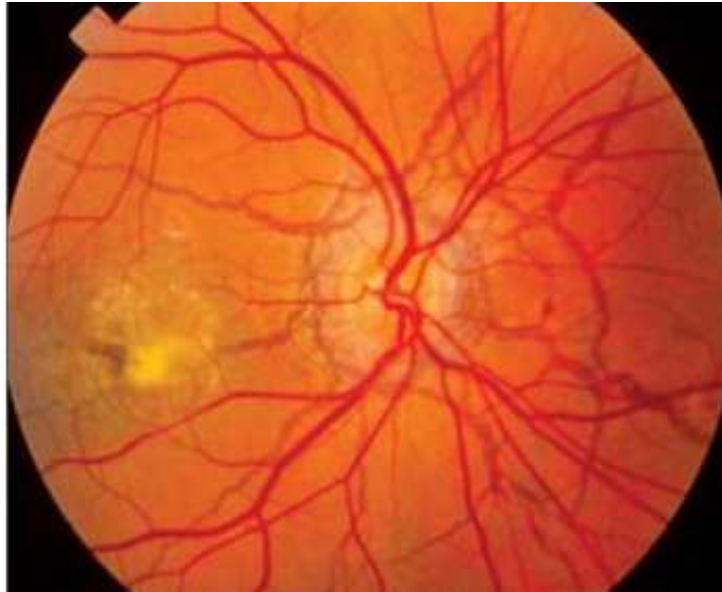


Subretinal and subretinal pigment epithelial bleeding are associated with acute visual loss.

Involvement of the second eye: The risk of the second eye developing wet AMD is about 10%/year or 50% over five years. Large and confluent [drusen](#) (figure 12) increase the risk.

VARIANTS: Macular degeneration may arise from other causes. Almost any injury to the macular region has the potential to lead to sub retinal scarring. [High myopia](#), [presumed ocular histoplasmosis \(POHS\)](#) and [angioid streaks](#) (figure 14) all contribute to choroidal neovascularization. Trauma, laser scars and focal inflammation can also be culprits.

Figure 14. Angioid streaks



Curvilinear breaks in the RPE promotes the development of subretinal neovascularization and bleeding. Angioid streaks are found in sickle cell, pseudoxanthoma elasticum and Paget's disease.

[Sickle cell retinopathy](#), Paget's disease, and pseudoxanthoma elasticum all promote curvilinear breaks in Bruch's membrane (called [angioid streaks](#)) which can permit growth of new vessels (choroidal neovascularization) with secondary bleeding and scarring.

Treatment

Dry AMD. Nutritional therapy with [anti-oxidants](#) and minerals may forestall the progression. Currently there is no treatment to reverse the effects of dry AMD. Studies by the National Institutes of Health called the Age-Related Eye Disease Study (AREDS) indicate that Zinc and vitamins A, C and E supplements and [lutein](#) are particularly valuable. ICAPs (Alcon Pharmaceuticals) and Preservision (Bausch & Lomb) and many over the counter generic multi-vitamins supply the recommended agents.

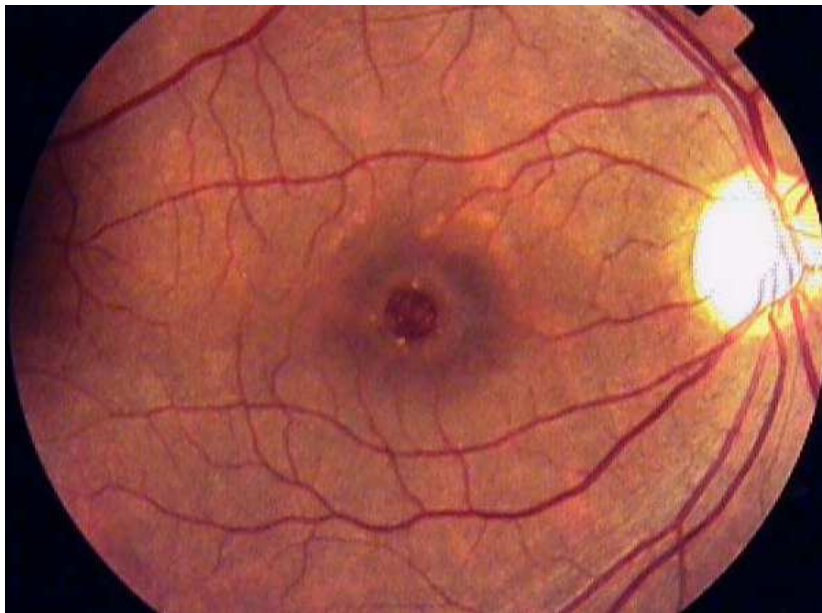
Acute **wet macular degeneration** is primarily treated with repetitive (4-8 weeks apart) intraocular injections of anti-angiogenic agents, chiefly [Lucentis \(ranibizumab\)](#) and [Avastin \(bevacizumab\)](#). Wet AMD that lies outside the most central region of the macula may be treated with laser photocoagulation. Other forms of therapy, including [photodynamic treatment](#) with vertiporphyrin, intraocular steroids, interferon and radiation have shown little or no benefit.

Macular Hole

Overview

Spontaneous **macular holes** develop as a result of tension and traction at the vitreous-retinal interface. They may also be caused by blunt trauma, chronic **macular edema**, and sun-gazing. Vitrectomy techniques combined with an intraocular gas bubble or **silicone oil** can seal the elevated cut edges of macular holes (figure 15) and improve the visual deficit in many cases.

Figure 15. Macular hole



Central full thickness defect in the retina with a cuff of subretinal fluid.

Cause

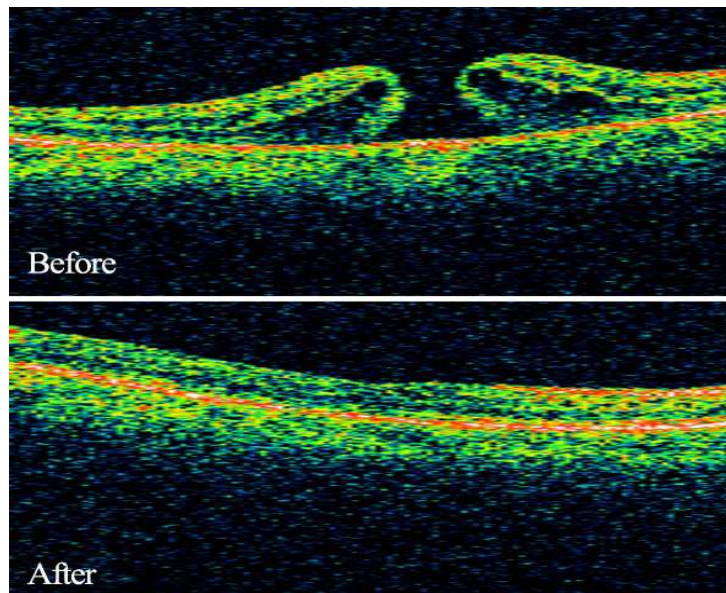
Contraction and separation of the posterior vitreous can tear retina in susceptible individuals. Fine vitreo-retinal adhesions at edge of the **fovea** stretch, elevate and tear the inner retina.

Natural History

It begins with distortion of the central **macula** and can progress from an inner

partial thickness break (closest to the vitreous cavity) to a full thickness hole in the retina. Surgeons evaluate the process with an imaging technique, optical coherence tomography (OCT) (figure 16). The edges of full thickness holes may separate from the underlying pigment layer to form a small, localized retinal detachment.

Figure 16. Macular hole – OCT study



Top: Full thickness macular hole.

Bottom: Same retinal location after surgical repair with vitrectomy.

Treatment

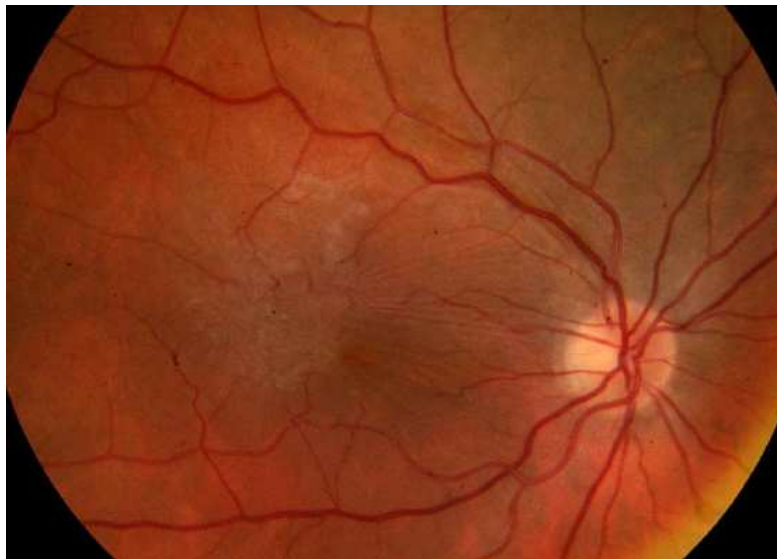
Repair of macular holes with [vitrectomy](#) techniques can improve acuity and minimize the central retinal defect. Surgeons remove the vitreous, pre-retinal membranes and the [inner limiting membrane](#). A gas bubble is introduced into the vitreous cavity to seal the tear, prevent retinal detachment and act as a scaffold for the cells that repair the defect. Patients lie face down for 1-2 weeks while the eye slowly absorbs the bubble.

Macular Pucker – Epiretinal Membrane

Overview

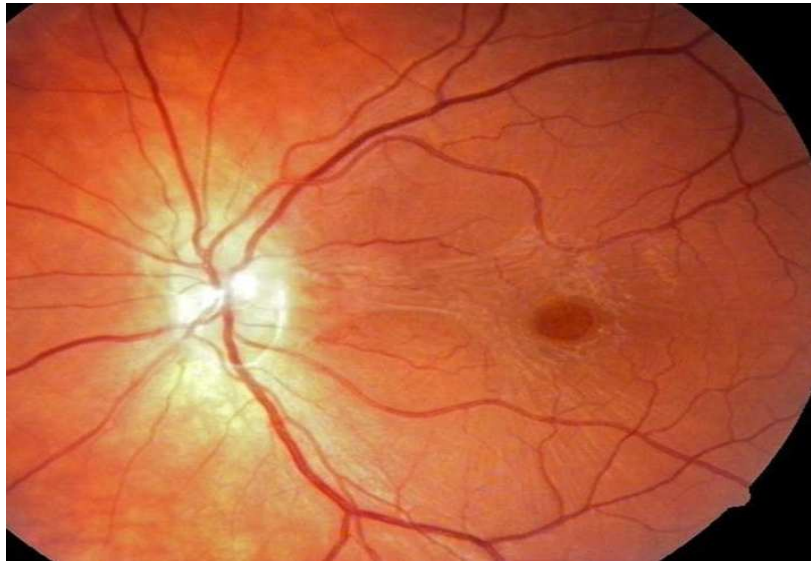
Infection, inflammation, trauma, surgery and disease may all generate pre-retinal membranes, with the potential to distort, wrinkle or “pucker” the retinal surface (figure 17). When associated with minimal or no retinal dysfunction, the term “cellophane retinopathy” is applied to describe the irregular light reflex arising from the retina similar to that reflects off crumpled cellophane. Membranes are frequent with diabetic retinopathy and after retinal tears. They occasionally distort the retinal surface to create a pseudo-hole (a well-defined depression that mimics a [macular hole](#)) (figure 18). Membranes responsible for visual impairment may be scraped off the retinal surface with [vitrectomy](#) techniques.

Figure 17. Macular pucker (epiretinal membrane)



Contracted surface membrane pulls and distorts the retinal surface.

Figure 18. Pseudohole: epiretinal membrane with contour that resembles a macular hole.



Cause

Many retinal cells have the potential to grow as a single or multilayer on the retinal surface. When the cells contract, they distort the highly organized retinal architecture, and reduce the quality of the retinal image sent to the brain. Injury and inflammation promote the growth of pre-retinal membranes.

Posterior vitreous detachment, retinal tears and retinal detachment are responsible for many macular membranes. Retinal tears and treatment for tears (laser and cryopexy) lead to significant puckers in about one in 30 cases.

Proliferative diabetic retinopathy generates new blood vessels (neovascularization) on the retinal surface accompanied by a fibrous scaffold of tissue and epiretinal membranes.

Natural History

Pre-retinal membranes associated with posterior vitreous detachment, retinal tears and detachment, trauma and surgery grow and stabilize over six months.

Membranes associated with disease (diabetes, retinal vascular disease) may continue to progress with their disease.

Treatment

Surgical peeling of retinal membranes is recommended to relieve symptoms and visual distortion and enhance acuity. [Vitrectomy](#) techniques are employed, with fine picks, scissors and grasping forceps.

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Neovascular Glaucoma

Overview

Growth of new blood vessels on the surface of the iris is invariably caused by an insufficient supply of oxygen to the retina. Although it is characteristic of central and branch retinal (hemi-retinal) vein occlusion and diabetic retinopathy, it may follow arterial retinal insufficiency or compromise of the great vessels in the chest and neck. New blood vessels growing over the [trabecular meshwork](#) may obstruct [aqueous](#) outflow and elevate the intraocular pressure to dangerous levels (figure 19).

Figure 19. Neovascular glaucoma: overgrowth of blood vessels on the surface of the iris may obstruct aqueous outflow through the angle structures.



Cause

[Neovascular](#) glaucoma is caused by conditions that cause extensive damage to the retinal circulation, principally diabetes and central retinal vein occlusion. Arterial insufficiency is an occasional cause.

Natural History

[Neovascularization](#) of the iris may progress rapidly and generally requires urgent therapy.

Treatment

Laser or **cryopexy** is applied to the peripheral retina. **Intravitreal Avastin** provides rapid involution of active vessels and is often administered as soon as the condition is recognized.

Nevi and Other Pigmented Lesions of the Fundus

Overview

Five to ten percent of adults harbor flat, pigmented lesions of the [choroid](#) representing the accumulation of benign melanocytes (a group of pigment producing cells). Conversion to a cancerous choroidal melanoma (an actively growing malignant lesion) is highly unusual. Pigmented lesions of the retina and choroid include nevi, and proliferation and hypertrophy of the retinal pigment epithelium (figure 20).

Figure 20. Pigmented choroidal lesions



Left: congenital hypertrophy of the retinal pigment

Right: 3mm x 4.5 mm nevus with overlying pigment alteration.

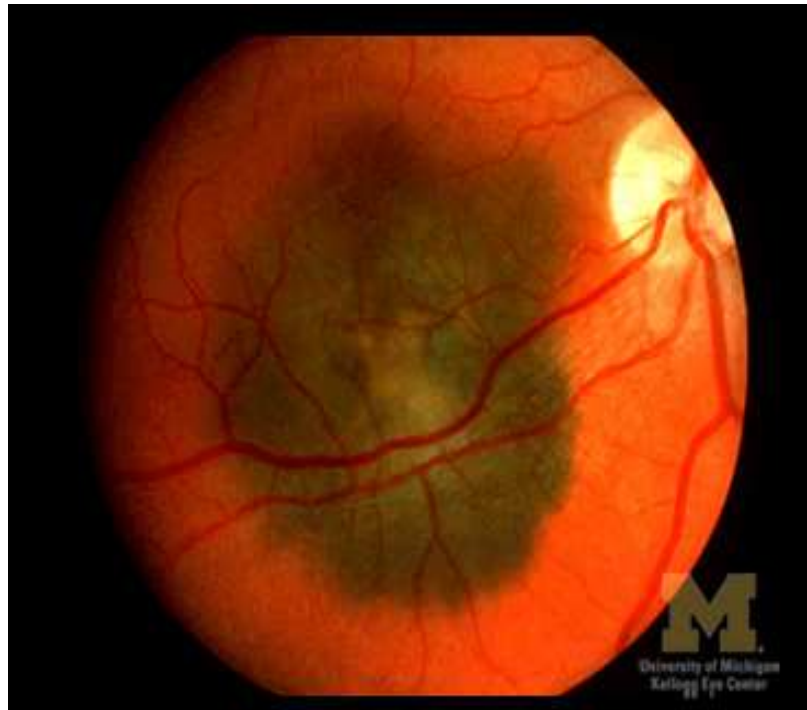
Cause

Nevi and melanomas arise from pigmented cells derived from the [neural crest](#) region.

Natural History

Nevi may grow slowly and show slight elevation. Growth of pigmented nevi and melanomas (figure 21) occurs in a spectrum of behavior consistent with the component cellular subtypes.

Figure 21: Choroidal melanoma: an elevated growing lesion



Treatment

Nevi require no treatment. Suspicious nevi may be observed every few months.

Melanomas may be treated by [radiation](#) (applied locally or by linear accelerator) or enucleation (removal of the eye). Cryopexy, ultrasound and phototherapy are used as ancillary methods. Systemic chemotherapy has not proven effective to date. Oncolytic viruses (a combination vaccine and targeted virus) that destroy melanoma cells are under investigation.

Posterior Vitreous Detachment (PVD)

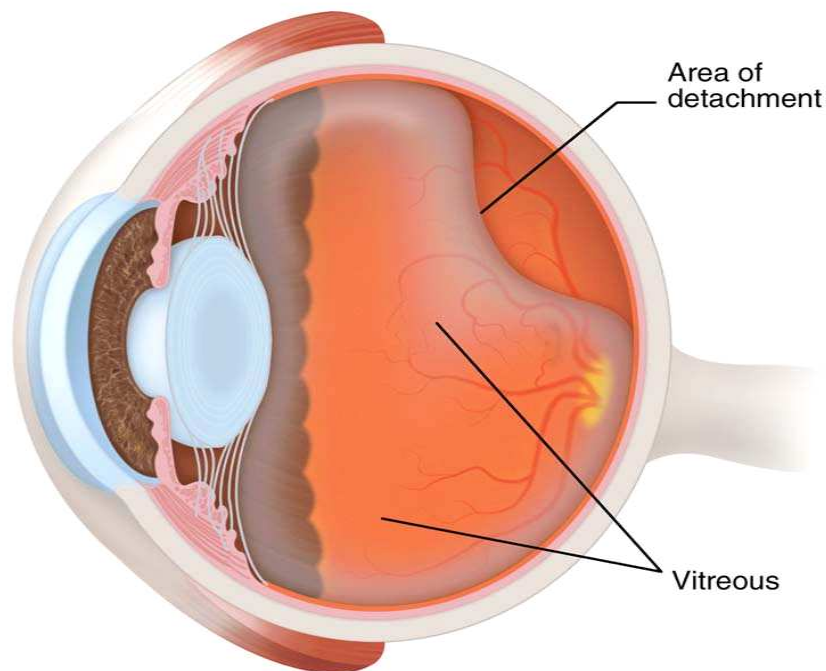
Overview

Flashing lights and new spots or floaters in the fields of vision heralds sudden spontaneous separation of the posterior vitreous. Although usually commonplace in patients in their 60s and 70s, myopia, trauma and intraocular surgery may hasten its onset. These same symptoms may precede a retinal tear or retinal detachment.

Cause

The denser posterior vitreous that lies against the retina is prone to spontaneous separation in high myopes (highly near-sighted individuals) and aging seniors. 25% of all 65-year-olds harbor a detached posterior vitreous. Trauma to the eye, including cataract surgery, may induce the separation (figure 22).

Figure 22. Partial posterior vitreous detachment:
separation of vitreous away from the retinal surface.



Natural History

Most PVDs cause little disturbance except for slowly resolving floaters. But eyes

with new symptomatic PVDs should be carefully evaluated for the presence of a retinal tear, especially if accompanied by intraocular bleeding. In rare instances, microscopic alterations in the surface of the retina caused by PVD lead to a layer of scar tissue (pre-retinal or [epiretinal membrane](#)) with secondary distortion of retinal contour (macular pucker) and disturbed acuity.

Treatment

New PVDs should be carefully evaluated for the presence of a retinal tear, especially if accompanied by intraocular bleeding of uncertain origin.

Proliferative Vitreoretinopathy (PVR)

Overview

About 5% of all tears and retinal detachment produce peri-retinal scarring on the surface or underside of the retina. In the worst cases, the scarring pulls the retina away from the wall of the eye, and reopens tears or creates new retina breaks. The scarred retina becomes too stiff to settle against the choroid and calls for an intricate surgical repair that may include an internal [vitrectomy](#) combined with [scleral buckling](#) and [vitreous substitutes](#) such as gas or silicone oil (figure 23).

Cause

PVR results from an unrestrained healing response after a retinal tear or detachment. The same cells involved in repairing a retinal tear if unchecked, may proliferate and produce an extensive scar. Inflammation, intraocular bleeding and excessive [cryopexy](#) promote PVR.

Natural history

Most active proliferation and scarring develops 4-8 weeks after the injury or operative procedure. PVR produces acute contraction and retinal detachment with precipitous loss of acuity.

Figure 23. Proliferative vitreoretinopathy: Retinal detachment with extensive growth of scar tissue on the surface and beneath the retina.



Treatment

Management of PVR requires a sophisticated combination of maneuvers to remove scar tissue, repair retinal tears, reduce or relieve tractional forces on the retina and maintain the retina in position. Surgeons may rely on one or more techniques: [vitrectomy](#), [sclera buckling](#), [membrane peeling](#), [laser coagulation](#), [retinotomy](#), and [use of vitreous substitutes](#). Each procedure is specifically tailored on a case-by-case basis.

Retinal Artery and Vein Occlusion

(Branch Retinal Artery, Central Retinal Artery, Branch Retinal Vein, Central Retinal Vein)

Overview

Retinal vascular disease leads to retinal damage and visual impairment either through disturbances in flow or leakage from the vascular tree. Blood delivers oxygen and nutrients by passing into the eye through the large caliber arteries. Metabolic exchange occurs at the level of the capillaries, the narrowest of vessels; waste products are removed via the veins. Disease at any point along the vascular tree has the potential to interfere with the vital supply of sufficient oxygen to the retinal cells. Insufficient oxygenation ([ischemia](#)) may stimulate new blood growth (neovascularization) on the surface of the retina or at other sites including the iris and over the [trabecular meshwork](#). Arterial disease, from emboli or clotting, characteristically causes decreased perfusion, with ischemia and/or infarction (cell death). Venous obstruction (closure of veins), whether partial or complete, causes congestion of the vascular tree, increased permeability and leakage, including [macular edema](#), and may also cause ischemia and infarction.

Cause

Although vascular obstruction may develop in young adults, it is a common cause of retinal disease in aging adults, especially those with diabetes and hypertension. In younger individuals, systemic disturbances in clotting and [emboli](#) from the heart may affect the retinal circulation, especially in the presence of a [patent foramen ovale](#). Smoking, birth control pills, pregnancy, migraine and anti-phospholipid antibodies may promote clotting in the arterial circulation. Blood disorders that thicken the consistency of the blood stream (e.g. hyperviscosity), inflammation and infection, and hemoglobinopathies (e.g., sickle cell disease) hasten venous congestion and clotting.

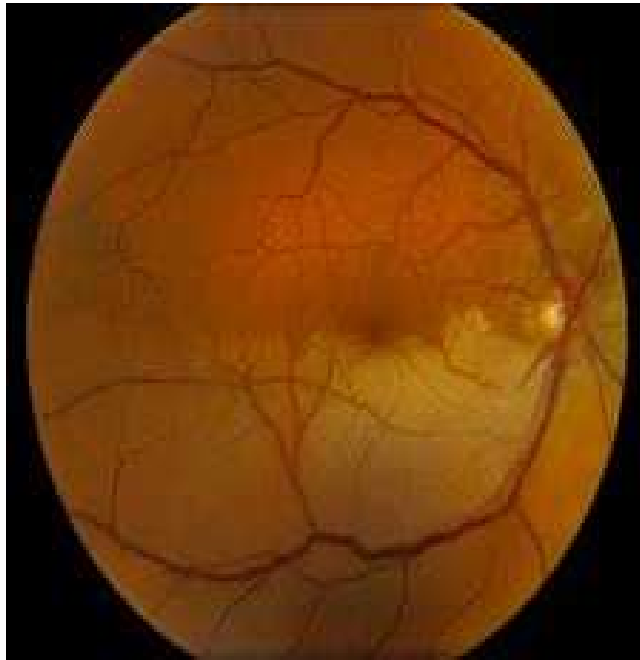
In the elderly, major vessels of the retina may become damaged by high blood pressure, arteriosclerotic disease and diabetes. Retinal arteries and veins share a common wall at sites where their paths cross. High blood pressure induces thickening and hardening of the arterial walls, which in turn may impinge and deform blood flow in contiguous veins. Flow disturbances lead to thrombosis (clotting) with venous obstruction. Connective tissue diseases associated with vasculitis (e.g, temporal arteritis), other inflammatory disease (e.g.) sarcoid have been implicated as predisposing conditions.

Branch Retinal Artery Occlusion

Natural history

Branch Retinal Artery Occlusion (BRAO) may develop if emboli (particulate matter) from the heart or great vessels obstruct the downstream circulation to produce a retinal stroke (infarct) involving all or part of the retinal circulation. Clotting may also develop in the central retinal or branch arteries related to coagulation disorders or local disease. The retinal circulation has no capacity to reroute arterial blood flow to bypass occluded segments. (Figure 24).

Figure 24. Inferior branch retinal artery occlusion: extensive whitening and swelling of the inferior retina



In younger individuals, cardiac abnormalities factor, especially in association with conditions that predispose clotting and migraine, predisposes to retinal stroke. Carotid or cardiac ultrasonography may be used to determine whether a cardiac abnormality is responsible for the occlusive disorder.

Treatment

Supportive care. Evaluation for cause (with special emphasis on the origin of possible emboli).

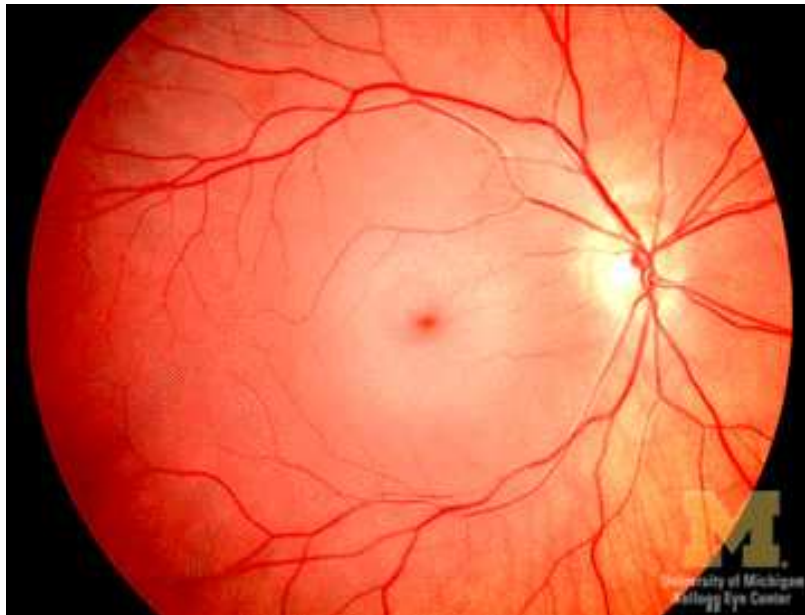
Ocular neovascularization – unusual in branch retinal artery occlusion. Scatter laser photocoagulation and/or trans-conjunctival cryopexy are used to ablate ischemic retina when neovascular complications arise in arterial occlusive disease.

Central Retinal Artery Occlusion

Natural history

Central Retinal Artery Occlusion (CRAO) may develop if emboli (particulate matter) from the heart or great vessels obstruct the downstream circulation to produce a retinal stroke (infarct) involving all or part of the retinal circulation (figure 25). Emboli characteristically arise from arteriosclerotic plaques in the lining of vessels.

Figure 25. Central retinal artery occlusion: extensive whitening (infarction) of the macula with a “cherry red” spot - indicative of a retinal stroke with preservation of the choroidal circulation.



In the elderly, arterial occlusion is most often related to arteriosclerotic disease of the carotids. Carotid or cardiac ultrasonography may be used to investigate whether emboli arising from arteriosclerotic plaques are responsible for the vascular obstruction.

Treatment

Supportive care. Evaluation for cause (with special emphasis on the origin of possible emboli).

Ocular neovascularization – unusual in branch and retinal artery occlusion. Scatter **laser** photocoagulation and/or trans-conjunctival **cryopexy** are used to ablate ischemic retina when neovascular complications arise in arterial occlusive disease.

Branch Retinal Vein Occlusion

Natural history

Branch Retinal Vein Occlusion (BRVO), an obstruction of a segment of the venous circulation, is associated with hypertension. The site of occlusion is generally at the disc or at a site away from the optic nerve where the retinal artery crosses over a retinal vein. The venous occlusion causes a sector of the retina to become engorged with blood: congested veins leak blood and fluid into the retina (**macular edema**), and reduce the central acuity when the macula is involved (figure 26). In time, the occluded segment reopens or retinal collateral vessels develop to reroute blood around the obstruction. Ischemia (lack of oxygenation to the tissue) may lead to **neovascularization** of the retina and subsequent vitreous hemorrhage (figure 27).

Figure 26. (Superior) branch vein occlusion: Sectoral distribution of blood in the upper half of the macula.

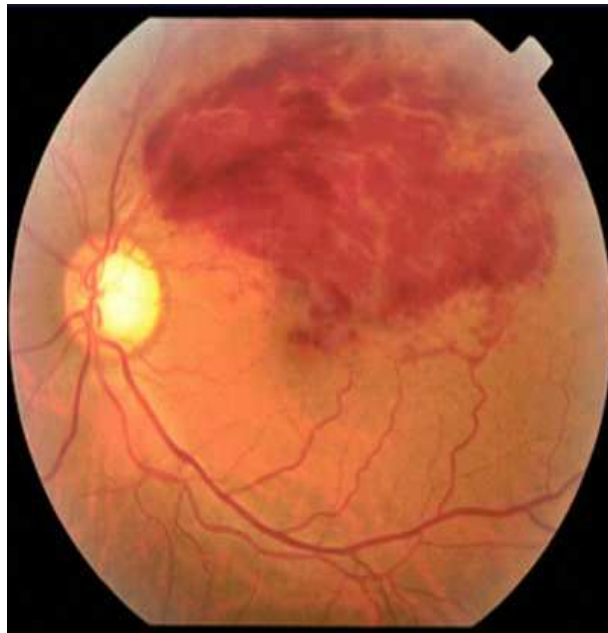


Figure 27. Neovascularization on the retinal surface:
New vessels arising from the optic nerve have grown
over the retinal surface.



Treatment of Branch Retinal (BRVO)) Vein Occlusion

Macular edema in BRVO - Laser photocoagulation has been the gold-standard of therapy since 1984. Direct treatment to the leaking vessels may improve acuity. Ozurdex (dexamethasone), a slow release steroid (Alcon Pharmaceuticals) has been approved for treatment of macular edema in both BRVO and CRVO. Lucentis, administered with intravitreal injection, has been beneficial in controlled experimental trials.

Ocular Neovascularization – Scatter laser photocoagulation and/or trans-conjunctival cryopexy are used to ablate ischemic retina when neovascular complications arise in venous occlusive disease.

(Figures 25 & 27, courtesy University of Michigan, Ann Arbor, Department of Ophthalmology.)

Central Retinal Vein Occlusion

Natural history

Central retinal vein occlusion (CRVO). Diabetes, arteriosclerosis, glaucoma, leukemia, increased blood viscosity and other disturbances in clotting may all contribute to clotting in the central retinal vein. CRVO may be partial or complete. Complete occlusion severely impairs retinal perfusion and causes ischemia, infarction, massive congestion and retinal swelling. [Neovascular glaucoma](#) develops in a substantial number of cases of complete CRVO. Neovascular activity often peaks at three months after the onset of the occlusion.

Partial (incomplete) CRVO may resolve without permanent visual disturbance. Complete CRVO generally causes profound macular and retinal damage (figures 28, 29).

Figure 28. Non-ischemic (partial) central retinal vein occlusion: extensive intraretinal hemorrhage with engorged retina veins.

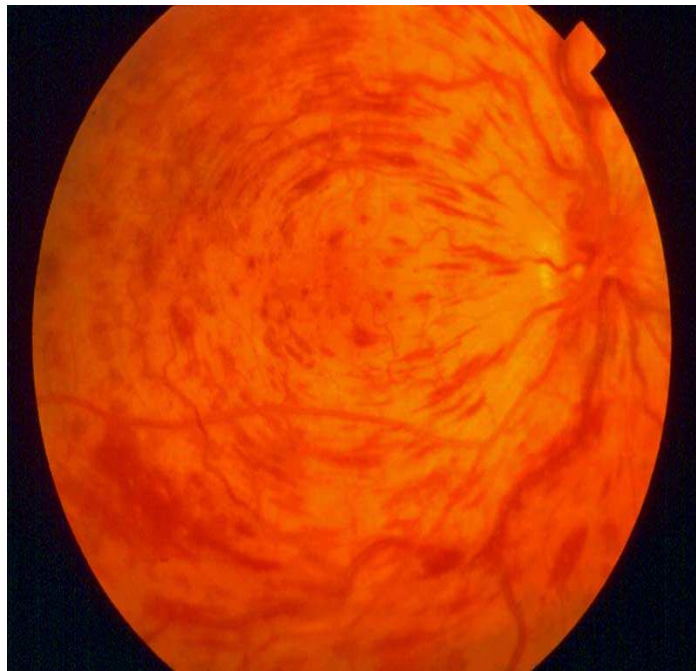


Figure 29. Ischemic (complete) central retinal vein occlusion: retinal hemorrhage, cotton wool spots (localized retinal infarction) and engorged retinal vessels



Retinal **neovascularization** (figure 27) is a regular feature of central retinal vein occlusion and may lead to vitreous hemorrhage and a sudden and catastrophic loss of vision.

Obstruction of the central retinal vein may be partial or complete. The complete form is ischemic, with retinal whitening and often sudden and profound loss of acuity. **Fluorescein angiography** reveals extensive non-perfusion. The ischemia tends to stimulate new blood vessel growth (**neovascularization**) on the iris surface (rubeosis iridis), which can lead to **neovascular glaucoma** (figure 19).

Treatment of Central Retinal (CRVO) Vein Occlusion

Macular edema in CRVO - Ozurdex (dexamethasone), a slow release steroid (Alcon Pharmaceuticals) has been approved for treatment of macular edema in both BRVO and CRVO. Recent studies suggest that intravitreal Lucentis or low dose (1mg) intravitreal **triamcinolone** is useful.

Ocular Neovascularization – Scatter laser photocoagulation and/or trans-conjunctival cryopexy are used to ablate ischemic retina when neovascular complications arise in venous occlusive disease

Retinal Tears and Detachment

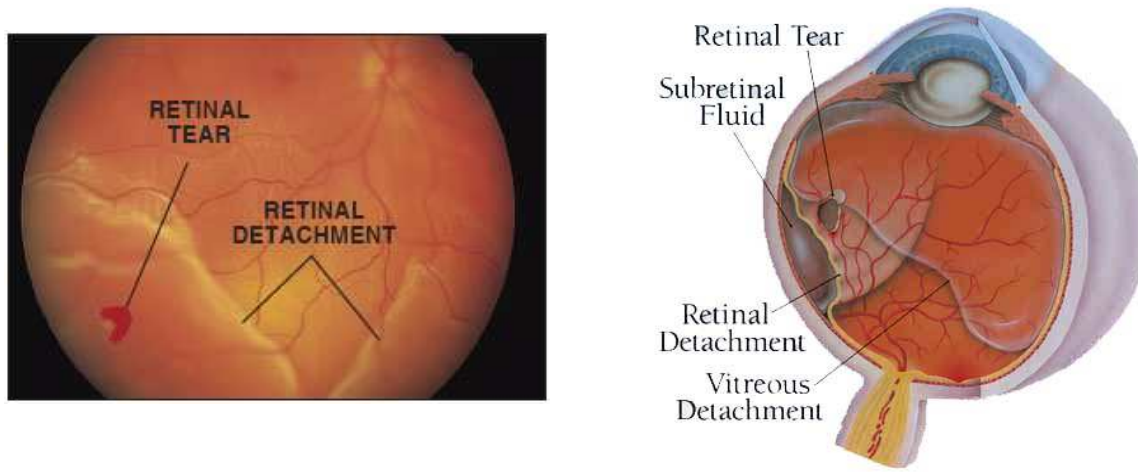
Overview

Most retinal tears develop spontaneously, and give rise to lightening-like flashes (photopsia) and new floaters and variable haziness in central vision. If accompanied by an advancing dark shadow, the adjacent retina may be detaching. The usual causes are posterior vitreous detachment and blunt or surgical trauma (e.g., cataract surgery.)

Cause

A retinal tear by itself does not cause a retinal detachment. Detachments occur because of an additional factor - traction on the retina. Forces generated by a [posterior vitreous detachment](#) may overwhelm the natural adhesion of retinal to the underlying retinal pigment epithelium. Prompt detection and treatment of retinal tears, which invariably develop after a posterior vitreous detachment, may prevent the development of retinal detachment. Retinal tears are most likely to arise in individuals with peripheral lattice degeneration, a condition characterized by a focal pigmentary alteration with strong vitreous to retinal (vitreo-retinal) adhesion and with [high myopia](#). Most retinal tears and detachments develop spontaneously, but they may follow blunt trauma to the head or globe. (Figure 30).

Figure 30. Retinal detachment



Left: retinal detachment with horseshoe tear. Many retinal breaks cause vitreous hemorrhage.

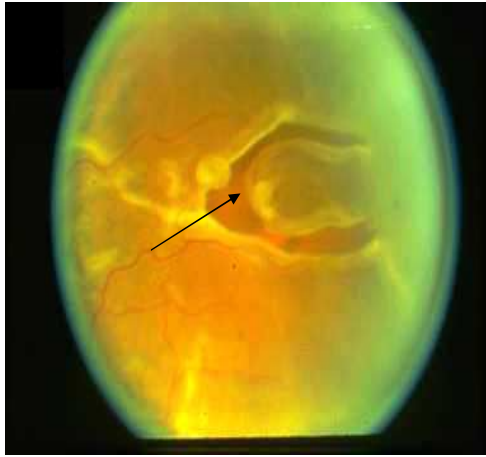
Right: schematic of retinal hole with vitreous and retinal detachment. Much of the retina remains attached.

Natural History

Posterior vitreous detachment causes most retinal tears. Injuries and direct trauma to the globe may also produce a retinal tear. Although some tears fail to produce progressive detachment of the adjacent retina, symptomatic tears – with flashes (photopsia) are more likely to produce a symptomatic retinal detachment with loss of vision.

Retinal tears have characteristic shapes and locations: Horseshoe tears (Figure 31) develop in the equatorial region and show a flap attached to the vitreous body. Operculated holes have an overlying tuft of avulsed retina, with no remaining vitreous adhesions. Giant tears extend for greater than 90 degrees around the circumference of the retina.

Figure 31. Horseshoe retinal tear.



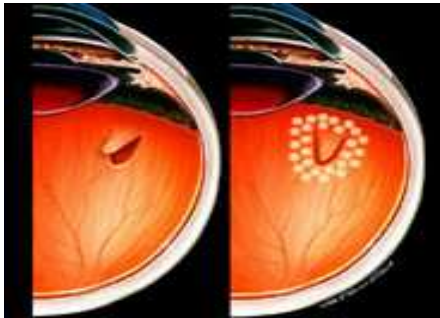
Retinal detachment with horseshoe (flap) tear. Caused by strong vitreous traction at the tip of the flap (arrow).

In unusual cases, retinal tears and detachment remain localized without treatment and produce no effect on visual acuity.

Treatment

Retinal tears are closed by inducing a local scar with [laser](#) (by causing of local burn) or by [cryopexy](#) (freezing) to bond the retinal tear to the underlying [choroid](#). (Figures 32, 33). Retinal detachments are repaired by [scleral buckle](#), [vitrectomy](#) or [pneumatic retinopexy](#) or a combination of [vitrectomy and scleral buckling techniques](#) to reduce traction on the retina and restore the retina to its intended position.

Figure 32. Laser (heat) and cryopexy (freezing) for retinal tears.



Laser (top, before and after) or freezing (below) burns the retina and the underlying choroid to generate a localized scar to seal the hole.

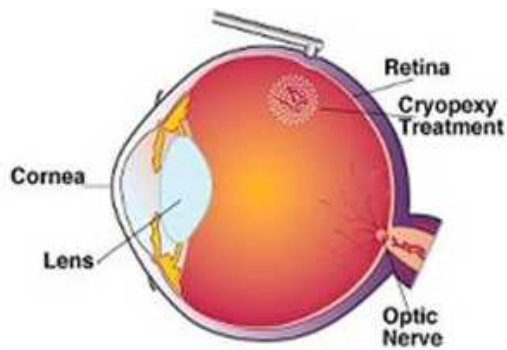


Figure 33. Laser photocoagulation of the retina.



Left: Fresh laser scars around a retinal tear.

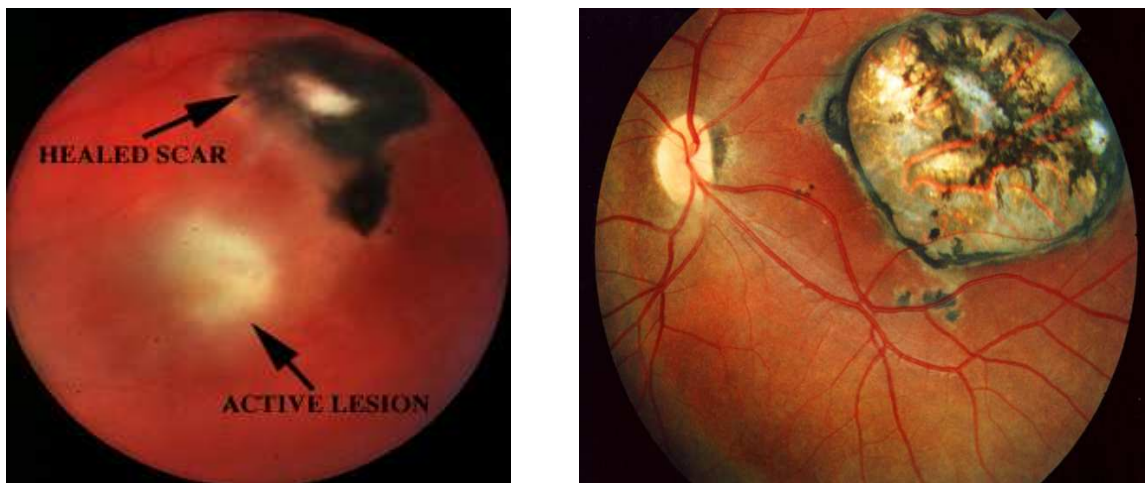
Right: Healed retinal scars six weeks later.

Retinitis

Overview

Retinal infections with parasites, fungi and viral agents may occur in healthy and compromised individuals. Toxoplasmosis (parasitic infection by *Toxoplasma gondi*) is the common retinal infection affecting healthy individuals (figure 34). An impaired immune system places an individual at risk for systemic and retinal infection from herpes group viruses and parasites. Diagnosis is supported by determinations of systemic infection, antibody response and patterns of retinal inflammation. Vitreous and/or retinal biopsy is reserved for unusual and unresponsive conditions.

Figure 34: Retinal toxoplasmosis: (left) active retinitis clouds the vitreous with inflammatory cells; (right) healed congenital lesion



Cause

Retinitis is most often attributed to Herpes group viruses (Cytomegalovirus, Herpes simplex, Herpes zoster and Measles), fungi (Candida and Cryptococcus) Toxoplasmosis and the HIV infection. Rubella and syphilis produce similar infections but primarily involve the choroid.

Natural History

The pattern of infection is related to both cause and natural immunity. Hemorrhage, retinal whitening, [vitritis](#) and [vasculitis](#) may be seen in all forms of retinal infection.

Treatment

Vitreous biopsy, antibody assay, and polymerase chain reaction (PCR) may all be used to support or establish a precise diagnosis. Treatment is tailored to the offending agent.

Uveitis

Overview

The eye may become inflamed after injury, infection, loss of blood supply or spontaneously. Inflammation may involve all layers of the eye, but usually involves the uvea, the middle, pigmented layer of the eye. Uveitis (inflammation of the uvea) involving the retinal region is called choroiditis, that is, inflammation of the choroidal layer, which lies adjacent to the retina. Descriptive terms for ocular inflammation at or adjacent to the retina emphasize the layer(s) showing greatest involvement: retinal pigment epitheliitis (inflammation of the retinal pigment epithelium, retinochoroiditis (inflammation of the retina and choroid), retinitis (retinal inflammation), vitritis (vitreous inflammation) and panuveitis (all layers).

Penetrating injuries, with rupture of the sclera, the white fibrous wall of the eye, may lead to sympathetic ophthalmia, a chronic inflammation of the injured eye, as well as inflammation of the uninjured eye.

Cause

Genetic factors influence and underlie the development of Harada's disease, Behcet's disease and Birdshot chorio-retinopathy. Sarcoid (figure 35) syphilis, tuberculosis and Lyme disease may be also implicated in chronic uveitis.

Figure 35. Sarcoid choroiditis: yellow-white inflammatory nodules in the choroid show through the retina.



Natural History

Chronic uveitis may lead to posterior synechiae (adhesions between the iris and lens), cataract, epiretinal membranes and macular edema. The vitreous may become clouded with proteinaceous debris and clumps of inflammatory cells.

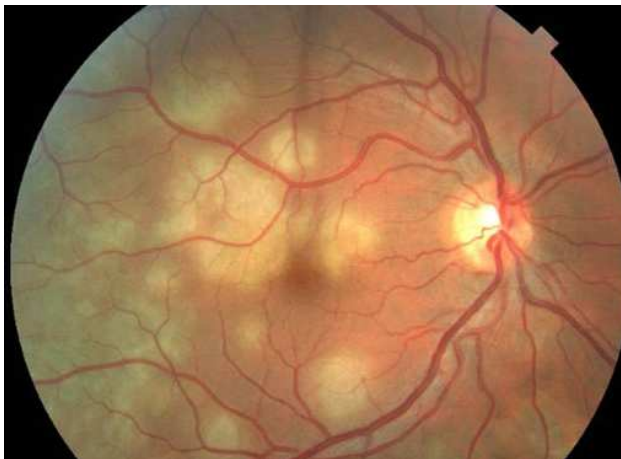
Treatment

Steroid therapy is the primary treatment for uveitis. Methotrexate, Azathioprine, Cyclosporin, Mycophenolate mofetil (Cellcept), Tacrolimus (FK06) and Infliximab are all used for severe or resistant cases.

White Dot Syndromes

White dot syndromes present with white, yellow or gray spots within the retina or choroid. It includes acute syndromes: multiple evanescent white dot syndrome (MEWS), acute idiopathic blind-spot syndrome, acute zonal occult outer retinopathy, acute multifocal posterior placoid pigment epitheliopathy, punctate inner choroiditis and multifocal choroiditis. Chronic white syndromes include birdshot chorioretinopathy, Harada's disease, sympathetic ophthalmia and sarcoid uveitis. (Figure 36).

Figure 36. White dot syndrome: sympathetic ophthalmia
(courtesy Digital Journal of Ophthalmology, Harvard)



Numerous white dots in the macula and posterior pole reflect inflammation in the choroid.

Cause

The white dot syndromes include some of the most peculiar and poorly understood retinal maladies. The root causes of the acute syndromes may include viral and inflammatory disease. In MEWS, the OCT demonstrates disruption of focal areas at the photoreceptor areas. Chronic white dot syndromes reflect autoimmune responses and infectious disease.

Natural History

Acute white dot syndromes frequently follow a flu-like syndrome or viral prodrome – with a spectrum of unilateral or bilateral visual impairment that may last for 6-8 weeks before complete or partial recovery.

Among chronic white dot syndromes, visual impairment may be linked to retinal swelling and [choroidal neovascularization](#), retinal detachment and persistent

inflammation and retinal scarring.

Of greatest concern: ocular lymphoma may present as a white dot syndrome in older adults.

Treatment

Acute white dots syndromes have no accepted therapy. Chronic white dot syndromes are treated with steroids and immunosuppressive agents.

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(Revised 1/22/2011)