Rhegmatogenous Retinal Detachment

Michael Colucciello, MD

Abstract: Patients often present after trauma with symptoms of vision loss or loss of a field of vision from a rhegmatogenous retinal detachment (RRD). This study aims to equip the health care provider with knowledge on the recognition, evaluation, and management of RRD to improve communication between the patient and consultant. The article highlights the symptoms, signs (including ophthalmoscopic findings), and pathogenesis of RRD. Causes and presentations of RRD are considered. Treatment paradigms are discussed and surgical options for treatment of RRDs are reviewed, including pneumatic retinopexy, pars plana vitrectomy, and scleral buckle surgery.
Keywords: retinal detachment; vision loss; scleral buckle; pars plana vitrectomy; pneumatic retinopexy; rhegmatogenous retinal detachment

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Introduction

Patients often present after trauma with symptoms of vision loss or loss of a field of vision from a rhegmatogenous retinal detachment (RRD). This study aims to equip the health care provider with knowledge on the recognition, evaluation, and management of RRD to improve communication between the patient and consultant.

A retinal detachment (RD) occurs when the neurosensory retina separates from the retinal pigment epithelium.[1] The 4 major types of RD are: rhegmatogenous, tractional, combined tractional and rhegmatogenous, and exudative. This article describes the pathological and clinical aspects of RRDs, which are those associated with retinal tears.

Pathogenesis

Rhegmatogenous (from "rhegma," rent, or fissure) retinal detachments occur when ocular fluid dissects under a full-thickness retinal tear.[1] This full-thickness retinal tear is usually situated in the peripheral retina vitreous base region. This situation differs from the relatively common condition that affects the peripheral retina, known as retinoschisis (prevalence reported as 3.9%[2]), wherein "splitting" of the retinal profile is associated with coalescence of intraretinal cysts.

A normal event that occurs in almost all individuals over time is liquefaction of the vitreous gel (vitreous synchysis). This liquefaction results from a progressive reorganization of the hyaluronic acid and collagen molecular networks, causing vitreous "floater" aggregates. This process leads to contracture of the vitreous (vitreous syneresis) and separation of the posterior vitreous cortex from the surface of the retina. This posterior vitreous detachment occurs when the weak posterior vitreoretinal attachment, comprising only parallel collagen extracellular matrix, is disrupted by the process of vitreous syneresis. Posterior vitreous detachment usually occurs between the ages of 40 to 70 years, (earlier in large [myopic] eyes); posterior vitreous detachment is almost universal by the age of 90 years. Normally, a posterior vitreous detachment does not become associated with a peripheral retinal tear. In certain cases, however, a retinal tear may occur with transmission of forces to the anterior vitreous base after a posterior vitreous detachment.

In contrast to the weak posterior vitreous attachment, the anterior vitreoretinal attachment (vitreous base) is firm. The anterior vitreous collagen fibers are oriented perpendicular to the retinal surface and actually connect with the peripheral retina tissue. Consequently, the anterior vitreous cannot easily separate from the anterior retina. The result of transmission of vector forces to the anterior vitreous base after a posterior vitreous detachment may therefore be a full-thickness retinal tear. This tear may occur during a sudden acceleration-deceleration force during trauma.

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Liquefied vitreous may pass into the subretinal space through a peripheral, anterior retinal tear on traction from the anterior vitreous base. Continuing anterior traction on the horseshoe-shaped retinal tear maintains patency of the tear, allowing the continuous flow of liquid vitreous and fluid under the retina in this location. This incoming fluid overpowers the retinal pigment epithelial pump which normally dehydrates subretinal space, causing an RRD (Figure 1).

An avulsion (tearing away) of the vitreous base (with or without a posterior vitreous detachment) can occur with acceleration-deceleration trauma. This vitreous base avulsion usually occurs in the inferior-temporal (coup) or superior-nasal (contra-coup) regions because the inferior-temporal region is least protected by the orbital rim. A vitreous base avulsion may be associated with a retinal dialysis, which is a peripheral retinal tear at the ora serrata (most anterior aspect of the retina).

Clinical Features

Epidemiology

The RRD annual incidence is 12 cases per 100 000. Most cases occur in persons aged 40 to 70 years, which is the usual age range when the posterior vitreous separates from the retinal surface. It occurs more commonly in men than in women.[3]

Figure 1. Diagram of a peripheral retinal tear under traction from the anterior vitreous base following a posterior vitreous detachment (left) and associated rhegmatogenous retinal detachment (right). This tear is associated with retinal lattice degeneration.
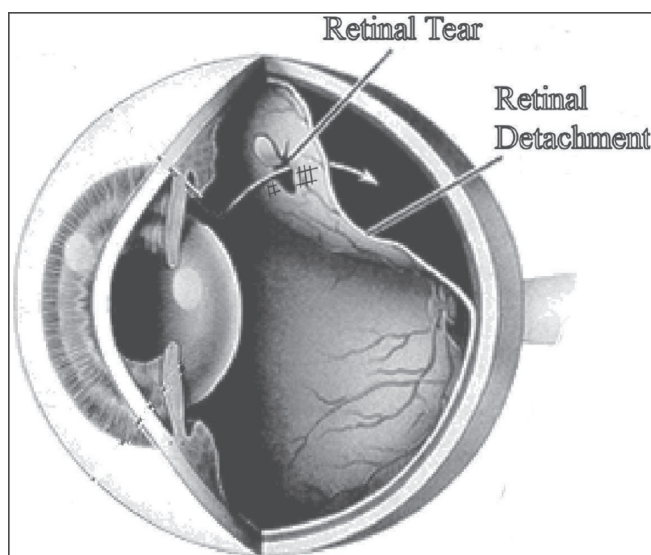
Risk Factors

The most common risk factor is axial myopia with peripheral retinal lattice degeneration (LD).[4] Myopia (nearsightedness) is a condition in which the eye is larger than normal, measured by the amount of minus (D): the greater the minus, the larger the eye.

Lattice degeneration is characterized by strong vitreous adhesions to edges of a focal peripheral (usually circumferential) area of retinal thinning with overlying liquefied vitreous. This retinal thinning is associated with a "lattice-like" configuration of pigmentation and is often seen in patients with myopia.

Lattice degeneration of the retina is the most common of all the hereditary vitreoretinal degenerations; the estimated prevalence is 7% to 11% of the general US population. The percentage of RD cases with LD have been reported to be as high as 32%. The risk of RRD in association with any amount of LD increases with the degree of myopia, especially with a refractive error > -5.00 D. The lifetime risk of RRD in individuals with LD and axial myopia > -5.00 D is 35.9%; those with lesser myopic refractive errors (between -1.00 D and -3.00 D) incur a 5.3% lifetime RD risk.[5]

Rhegmatogenous retinal detachment in LD patients usually occurs after posterior vitreous detachment and is associated with a retinal tear on the edge of a lattice lesion. Rhegmatogenous retinal detachment in LD occurs much less commonly in the setting of atrophic holes in the lattice lesions without a posterior vitreous separation. If the latter does occur, it is usually seen in young patients with axial myopia. Participation in contact sports is not contraindicated by the presence of LD.

A recent study showed a 5-year risk of RD of 23% in fellow eyes in patients with unilateral RRD.[6] Other risk factors are prior intraocular surgery, family history, or retinitis. Familial conditions include Marfan syndrome, Stickler's syndrome (hereditary arthro-ophthalmopathy with Marfanoid habitus), Ehlers-Danlos syndrome, and homocystinuria.

Rhegmatogenous retinal detachment may complicate cataract surgery. One study showed that the risk of RD occurring after cataract surgery is 1.79%.[7] There was no difference in the rate of RD after the (older) extracapsular surgery versus the (newer) phacoemulsification subjects. The majority of RRD cases (88%) occurred 2 or more years after cataract surgery, which emphasizes the importance of long-term follow-up for patients after cataract surgery.

Trauma inducing an acceleration-deceleration force to the head, orbit, or eye may induce vitreoretinal traction with an associated retinal tear which can lead to RD. Activities that can cause direct trauma to the eye, such as boxing, contact martial arts, squash racquets, and paintball, may cause RD or dialysis.[8–11] A retinal dialysis can be detected and treated before it develops into RD. Therefore, governing bodies in some of these sports require regular ophthalmic examination. Other activities that involve sudden acceleration or deceleration also increase risk for retinal tears and detachment. These include bungee jumping, drag racing, and rollercoaster rides.

Symptoms

"Floaters" and photopsias (the perception of light flashes) are not usually associated with a retinal tear or RD. These symptoms are common in individuals because of vitreous synchysis and syneresis, which occur over time. However, evaluation of the retina is frequently performed when these symptoms are noted because they may occur in the setting of a retinal tear. Retinal evaluation is mandatory if a visual field change is seen when these symptoms occur.

Floaters are appreciated as aggregates of vitreous collagen matrix, casting a shadow on the surface of the retina. They are best appreciated when looking at a light background. A ring-shaped floater (termed a "Weiss ring") is the retinal shadow of a separated opaque vitreous condensation that was formerly attached to the edges of the optic disk. "Cobwebs," or large diffuse floaters, can also be appreciated with vitreous synchysis. Small round spots may indicate red blood cells in the vitreous (after rupture of capillaries with an acute posterior vitreous detachment or retinal tear), or deposition of pigment cells on vitreous veils, which can occur after a retinal tear ("Schaffer's sign"). Photopsias occur with mechanical stimulation of the retina by vitreoretinal traction.

A peripheral visual field defect accompanies an RRD. Once the macula (central retinal) becomes involved in the RD (or if a large superior bullous RD obstructs the macula), the central visual field and acuity is affected. The progressive visual field defect coincides with progression of the RD from the peripheral retina to the macular region.

Clinical Examination and Signs

A relative afferent pupillary defect is usually seen with an RRD. This defect is demonstrated by a swinging flashlight test. A light is "swung" from one eye to the other while observing pupillary responses: a relative afferent pupillary defect exists if the pupil responses (constriction to the light exposure) are asymmetrical.

Visual acuity may be normal (Snellen 20/20) in a patient with an RRD if the macula has not become involved in the RD. Visual field deficits are seen in the area of the RD. Intraocular pressure often measures lower than normal in an eye with an RRD. Pigmented cells may be seen in the anterior or posterior chamber.

Ophthalmoscopy of detached retina shows an opaque, elevated, corrugated retina with a convex configuration that undulates freely with eye movements (Figures 2, 3). More than one retinal break is present in 50% of the cases (Figure 4). The RD may be seen by direct ophthalmoscopy; indirect ophthalmoscopy (aided by scleral depression) enables identification of associated peripheral retinal tears. Most retinal tears associated with RD are in the temporal periphery: 60% of all retinal tears are located in the superior temporal quadrant, with 15% in each of the inferior temporal and superior nasal quadrants, and 10% in the inferior nasal quadrant. The peripheral retinal tears occurring as a result of a posterior vitreous separation appear to have a "horseshoe" shape due to the configuration of the vitreoretinal traction (Figure 5).

Although most RRDs are not caused by trauma, the primary care physician may encounter an RD as a result of trauma in the field, including a sports competition. These RRDs may occur with blunt trauma or sudden acceleration-deceleration injuries and may be associated with other injuries, such as

Figure 2. Rhegmatogenous retinal detachment. A peripheral "horseshoe" retinal tear (at 9 o'clock) associated with retinal detachment involving the macular region. Note the retinal folds in the area of the retinal detachment.
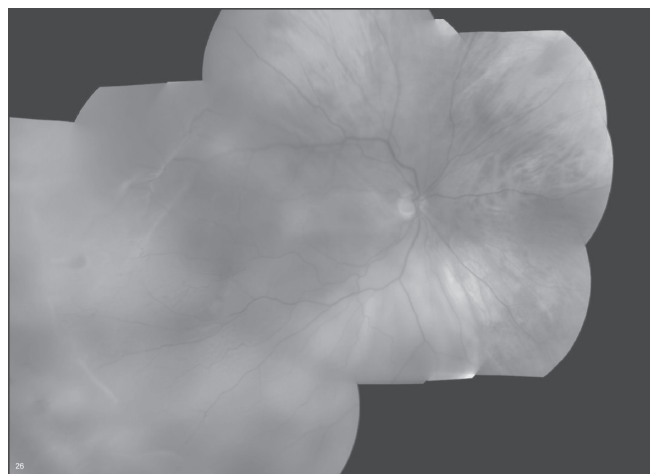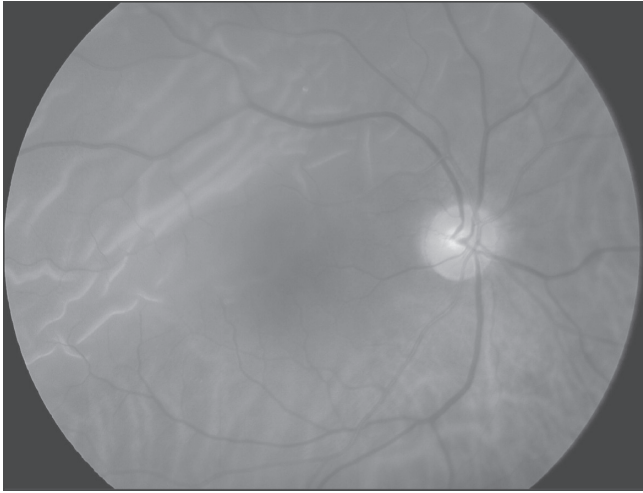
Figure 3. Rhegmatogenous retinal detachment with corrugations in the macular region.

Figure 5. A peripheral retinal tear associated with a rhegmatogenous retinal detachment.

orbital fractures, eyelid ecchymosis and edema, hyphema, or ruptured globe. Clinicians should be extremely suspicious of the presence of an RD in these settings (especially if an afferent pupillary defect exists). Prompt referral to an ophthalmologist or a retina specialist should be made if RRD is suspected. If significant media opacity such as hyphema or vitreous hemorrhage precludes a direct view of the retina, B-scan ultrasonography will be performed by the specialist to rule out RRD (provided there is no concern about a ruptured globe).

Chronic RRD may be associated with an avascular scar response, termed *proliferative vitreoretinopathy* (PVR). The

Figure 4. Two peripheral "horseshoe" retinal tears associated with a rhegmatogenous retinal detachment.
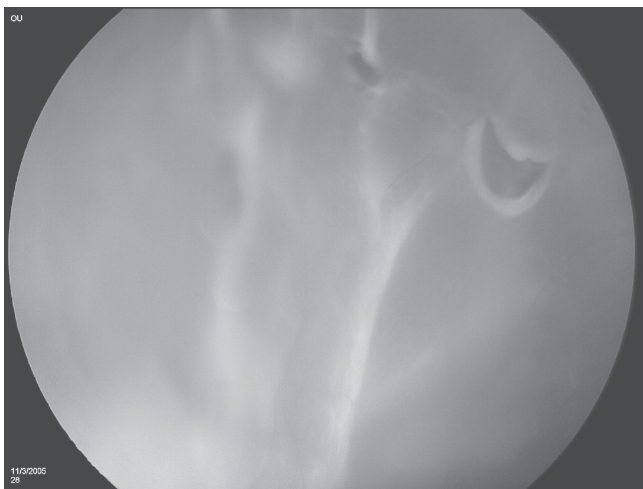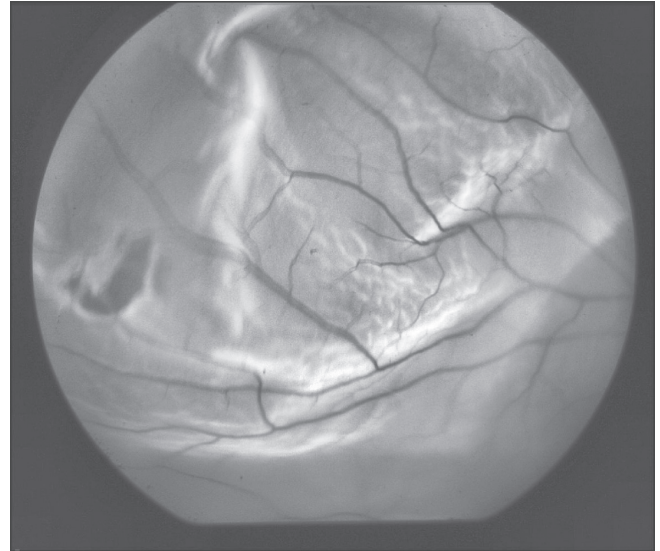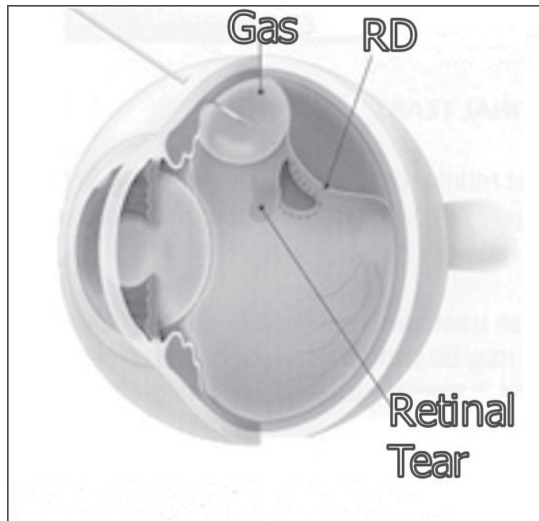

PVR process begins when retinal pigment epithelial cells migrate through a retinal tear and dedifferentiate into a sheet of myofibroblasts. Contraction of this sheet of myofibroblasts adds a tractional component to the RRD, which complicates treatment and worsens prognosis.

Treatment

A retinal tear without an RRD may be delimited by laserpexy or cryotherapy to induce a chorioretinal adhesion. A subclinical RD, defined as subretinal fluid extending at least 1 optic disc diameter away from the nearest retinal break, but not more than 2 disc diameters posterior to the equator, may be treated in a similar fashion. If a clinical RD (a detachment more extensive than found in subclinical RRD) is present, other surgical options may be considered.

The objectives of surgical repair of RRD are to: 1) identify all retinal tears (indirect ophthalmoscopy), 2) treat (induce a chorioretinal adhesion) all retinal tears ("pexy," with either cryotherapy or laser photocoagulation), and 3) close/seal all retinal tears with gas, silicone explant, (heavy) perfluorocarbon liquid, or silicone oil. These goals may be accomplished by different procedures, which may be combined in certain cases to achieve the desired result.

Figure 6. Pneumatic retinopexy retinal detachment repair. Note the internal tamponade of the retinal tear by the gas bubble.

Abbreviation: RD, retinal detachment.

Figure 7. Scleral buckle repair of retinal detachment. Note the retinal tear closed and supported by the circumferential buckling effect.
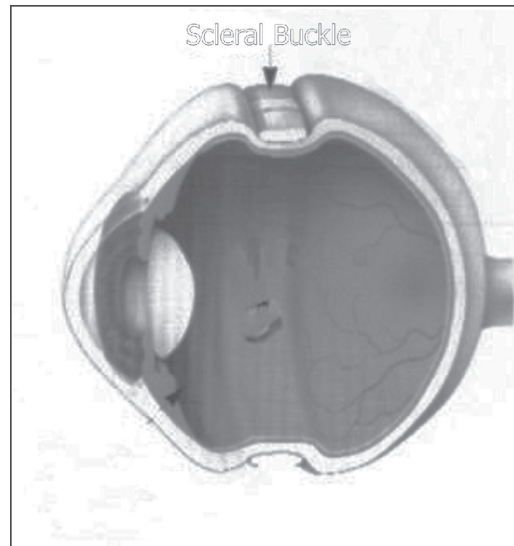
Pneumatic Retinopexy

Unlike the other surgical options to treat RRD, pneumatic retinopexy[12] is an office procedure (Figure 6). Less than 1 mL of a long-acting (relatively insoluble) expansible gas (SF6/ C3F8) is injected in the vitreous cavity via a 30-gauge needle through the pars plana. With appropriate positioning by the patient after the procedure, the gas serves to close retinal tears internally. Cryotherapy is used around the tears to set up a situation that would allow for a chorioretinal adhesion (laser would not "take" in the setting of elevated, detached retina). Alternatively, laser photocoagulation retinopexy may be performed instead of cryotherapy after retinal reattachment occurs (usually 2–3 days after gas injection).

Proper positioning to maximize gas-tear apposition is essential. The positioning requirements mean that this procedure is best used in patients with solitary tears superior to the horizontal meridian.

Scleral Buckle Procedure

This surgical procedure employs a scleral buckle, an explant made of solid silicone (Figure 7). The scleral buckle is sutured to the sclera in either a radial or circumferential fashion (under the rectus muscles) to promote approximation of the retina with the underlying retinal pigment epithelium. Cryotherapy is used about the tears. Subretinal fluid may be drained externally to flatten the retina to allow for closure of the retinal

tears. This procedure is performed after a small radial scleral incision to expose the underlying choroid. A small puncture through the choroid (with a 25-gauge endocautery probe) into the subretinal space allows for spontaneous subretinal fluid drainage as intraocular pressure is maintained with ocular tamponade. As in pneumatic retinopexy, gas may be injected into the vitreous cavity to aid in the closure of tears internally.

Pars Plana Vitrectomy

Standard 20-gauge or small incision (23- or 25-gauge) pars plana vitrectomy may be performed to remove the vitreous gel and cortex, thus removing the vitreoretinal tractional component involved in the genesis of the RRD (Figure 8). The instruments are introduced through sclerotomy sites 3.5 to 4 mm from the corneo scleral limbus (the location of the pars plana) to avoid larger blood vessels and the retina. The presence of vitreous (99% water) is not necessary for optical clarity; the vitreous space is replaced with aqueous humor produced after vitrectomy. During pars plana vitrectomy repair of RD, the flaps of retinal tears are removed or truncated to assure all vitreoretinal traction is relieved. Epiretinal membranes and PVR membranes may be delaminated with micropics or microforceps. Subretinal fluid may be aspirated internally through a posterior drainage retinotomy with fluid-gas exchange or be allowed to drain through existing retinal tears
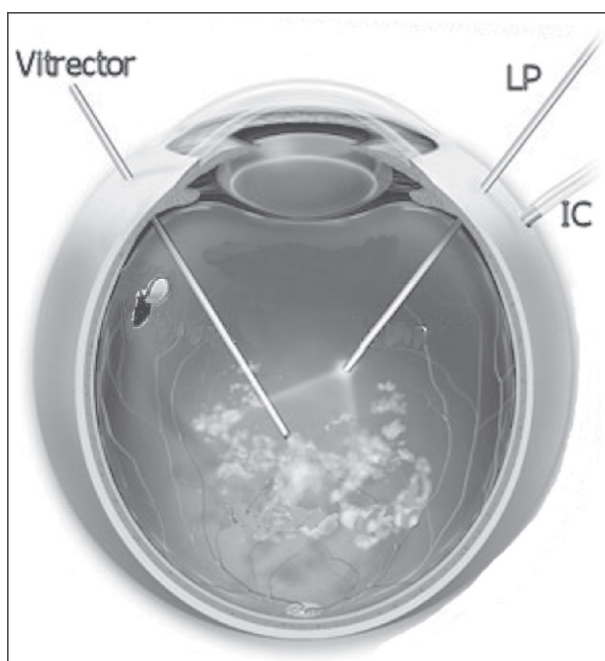
Abbreviation: LP, endoilluminator light pipe, IC: infusion cannula.

through the introduction of heavy perfluorocarbon carbon liquid over the posterior pole. Laser photocoagulation retinopexy is then performed about retinal tears and in the vitreous base. An agent to promote tamponade of the retinal tear(s)—gas (most commonly; this absorbs in the near term) or silicone oil (less commonly; this usually needs to be removed after healing with a separate surgical procedure)—is left in the eye at the end of surgery.

Results and Complications

Single operation anatomical success rates that were reported recently are 77% for the pneumatic retinopexy, 89% for scleral buckle, and 92% for pars plana vitrectomy procedures.[7–9] Each technique has its advocates; ultimately, with one or more procedures, 96% to 98% of all RRDs can be successfully treated. Currently, outcomes appear similar for phakic and pseudophakic eyes.[13] Pseudophakic eyes can do very well with primary vitrectomy repair without scleral buckling.[14]

Postoperative visual acuity is dependent upon whether the macula becomes involved in the RD. Potential visual acuity is worse the longer the duration of the macular detachment. Visual acuity may also be affected adversely postoperatively by

the presence of macular edema or the formation of a macular epiretinal membrane (also called macular pucker or cellophane maculopathy).

Macular edema, which may occur in up to 36% of cases, especially in those with preoperative macular detachment or cataract surgery history, is due to local prostaglandin release postoperatively; this can be treated with topical nonsteroidal anti-inflammatory agents or topical (or intravitreal) steroids.[15] Macular pucker, which has been reported to occur in up to 8.5% of scleral buckle cases[16] and 12.8% of vitrectomy cases[17] is a localized PVR process in the macula. This macular epiretinal membrane tissue can be delaminated surgically via pars plana vitrectomy to re-establish macular architecture. Macular edema and traction can be characterized by optical coherence tomography. This procedure uses noncontact "optical ultrasound," imaging reflections from an 820-nm laser to provide cross-sectional macular images with a resolution of 10 to 20 m. Macular edema, macular pucker, and slowly absorbing macular subretinal fluid postoperatively can all cause metamorphopsia ("waviness" of vision in which straight lines appear wavy).

Proliferative vitreoretinopathy, which may induce tractional retinal re-detachment after initial successful repair is the most common cause of failure of RD surgery.[18] Proliferative vitreoretinopathy complicates 6% to 8% of cases postoperatively. Proliferative vitreoretinopathy incidence generally peaks within 2 to 3 months of the initial surgery. Risk factors for the occurrence of PVR include cases of RRD with larger tears (PVR has an incidence of up to 40%–50% in association with retinal tears larger than 3 clock hours, deemed "giant retinal tears"). Proliferative vitreoretinopathy occurs more commonly in cases associated with numerous tears and in cases with vitreous hemorrhage. Pars plana vitrectomy can be used to treat PVR. Proliferative vitreoretinopathy membrane delamination can be accomplished with intraocular microforceps; relaxing retinotomies can be made. Anatomic success rates of proliferative vitreoretinopathy are 75% to 90%; however, functional results are not as good—visual acuity is generally compromised by macular involvement in these patients.[19]

Endophthalmitis is rarely encountered after retinal surgery. The risk is reported as 0.02% to 0.2% with vitrectomy, < 1% with pneumatic retinopexy, and 0.3% with scleral buckling procedures.[20–22] Complications complications of retinal vascular occlusion and anterior segment ischemia with scleral buckling rarely occur. Complications with an incidence of < 5% include

glaucoma, lens implant dislocation, ptosis, cataract at the time of surgery, choroidal hemorrhage, and retinal tear formation.

There is a 64% incidence of indicated cataract surgery within 1 year of vitrectomy according to a recent study;[23] another study showed a much reduced incidence in cataract progression (only 7%, mean follow-up 27 months) in those undergoing surgery before 50 years of age.[24]

Strabismus and diplopia can occur after scleral buckle surgery associated with the required manipulation of rectus muscles ($<$ 5% incidence); these conditions can usually be treated.[25] An increase in myopia because of a change in shape of the eye after scleral buckling is anticipated; this myopia can be treated by refractive correction.

Conclusion

Rhegmatogenous retinal detachment is an important cause of vision loss. This condition can occur without antecedent trauma or can complicate acceleration-deceleration injuries to the head. A high index of suspicion for the presence of RRD as a cause of visual field loss, floaters, and photopsias can result in directed examination and treatments that can save the patient's vision for the long term.

Conflict of Interest Statement

Michael Colucciello, MD discloses no conflicts of interest.

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