CHAPTER 16

Retinal Detachment and Predisposing Lesions

This chapter includes a related activity. Go to www.aao.org/bcscactivity_section12 or scan the QR code in the text to access this content.

Highlights

- Lattice degeneration is present in 6%–10% of people and may predispose eyes to retinal detachment (RD).
- Although symptomatic retinal breaks typically are treated in most eyes, the complete clinical scenario is important to consider when clinicians are determining whether asymptomatic lesions, including lattice and retinal breaks, should be observed or treated.
- Retinal detachments are categorized as rhegmatogenous, traction (or tractional), or exudative.
- Retinoschisis can be differentiated from rhegmatogenous RD by examination and imaging.
- Retinoschisis without RD typically does not require treatment.

Examination and Management of Posterior Vitreous Detachment

The vitreous gel is attached most firmly at the *vitreous base,* a circumferential zone straddling the ora serrata that extends approximately 2 mm anterior and 3–4 mm posterior to the ora. Vitreous collagen fibers at this base are so firmly attached to the anterior retina and pars plana epithelium that the vitreous typically cannot be separated from these tissues without tearing them. The vitreous is also firmly attached at the margin of the optic nerve head, at the macula, along major vessels, at the margins of lattice degeneration, and at chorioretinal scars.

Most *retinal tears* result from traction caused by posterior vitreous detachment (PVD). The predisposing event is syneresis (collapse) of the central vitreous. Vitreous traction on the retina can produce a *retinal break,* a full-thickness defect in the neurosensory retina; in this setting, the break usually occurs at the posterior edge of the vitreous base (Fig 16-1).

Many patients do not report acute symptoms when a PVD occurs. Symptoms of PVD at the initial examination include the entoptic phenomena of photopsias (flashing lights), multiple floaters, and the appearance of a curtain or cloud across the visual field. Patients with

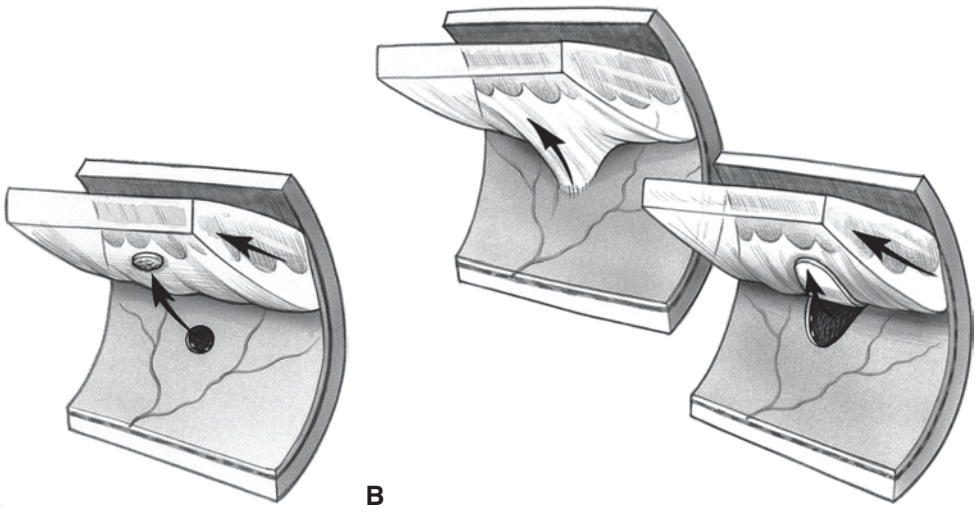

Figure 16-1 Schematic representations of mechanisms of retinal tear formation associated with posterior vitreous separation. **A,** Round or oval hole. **B,** Flap tear: posterior extension of vitreous base with firm vitreoretinal attachment. *(Illustration by Christine Gralapp, based on illustrations by Tim Hengst.)*

these symptoms should be examined promptly, and office staff should be made aware of the urgency of these symptoms. Photopsias are caused by the physical stimulus of vitreoretinal traction on the retina. Floaters are caused by vitreous opacities such as blood, glial cells torn from the optic nerve head, or aggregated collagen fibers, all of which can cast shadows on the retina.

Vitreous hemorrhage may arise from avulsion of superficial retinal or prepapillary vessels or from rupture of retinal vessels that cross retinal tears. Overall, 7%–18% of patients with acute symptomatic PVD have retinal tears. If vitreous hemorrhage is present, 50%–70% of patients have retinal tears, versus 7%–12% without vitreous hemorrhage. Patients with an acute PVD complicated by a retinal tear are 7 times more likely to present with vitreous pigment or granules than are those without a tear.

Indirect ophthalmoscopy with scleral depression or slit-lamp biomicroscopy are used to clinically diagnose PVD and rule out retinal breaks or detachment. Optical coherence tomography (OCT) can also be employed to visualize the state of the vitreous gel on the optic nerve head and throughout the posterior pole. Reexamination of the patient 2–4 weeks after presentation may be appropriate, because as the PVD evolves over time, new retinal breaks may occur. Additional risk factors to consider in follow-up determination include trauma, aphakia, myopia, fellow-eye history, family history of retinal detachment (RD), and signs of Stickler syndrome. All patients should be instructed to return to the ophthalmologist immediately if they notice a change in symptoms, such as increasing numbers of floaters or the development of visual field loss. They should also be told that a PVD may occur in the fellow eye. Four to 8 weeks after initial presentation and examination, if clinical symptoms of flashes and floaters are stable or improving, the risk of new retinal breaks is low.

If a large vitreous hemorrhage precludes complete examination, ultrasonography may be performed to evaluate for flap tears and rule out RD and other fundus lesions.

In addition, bilateral ocular patching and bed rest, with the patient's head elevated 45° or more for a few days, may allow the hemorrhage to settle sufficiently to permit detection of superior breaks. If the cause of the hemorrhage cannot be identified, the patient should be reexamined at frequent intervals, and early vitrectomy may be considered.

Byer NE. Natural history of posterior vitreous detachment with early management as the premier line of defense against retinal detachment. *Ophthalmology.* 1994;101(9):1503–1514.

van Overdam KA, Bettink-Remeijer MW, Klaver CC, Mulder PG, Moll AC, van Meurs JC. Symptoms and findings predictive for the development of new retinal breaks. *Arch Ophthalmol.* 2005;123(4):479–484.

Lesions That Predispose Eyes to Retinal Detachment

Lattice Degeneration

Lattice degeneration, a vitreoretinal interface abnormality, is present in 6%–10% of the general population and is bilateral in one-third to one-half of affected patients. It occurs more commonly in—but is not limited to—myopic eyes; there is a familial predilection. Examination findings characteristic of lattice include well-defined oval or linear regions of retinal thinning classically with white crossing lines. There may be 1 or more lesions, and they may contain areas of hyperpigmentation, hypopigmentation, or atrophic retinal holes.

Lattice may predispose eyes to retinal breaks and detachment. The most important histologic features include varying degrees of atrophy and irregularity of the inner retinal layers, an overlying pocket of liquefied vitreous, condensation, and adherence of vitreous at the margin of the lesion (Figs 16-2, 16-3).

Lattice is found in approximately 20%–30% of all patients who present with rhegmatogenous retinal detachments (RRDs). However, because lattice is not necessarily causative, prophylactic laser treatment is not universally recommended. When lattice is the cause of RD, a tractional tear at the lateral or posterior margin of the lattice or, less commonly, an atrophic hole within the zone of lattice may be found (see Fig 16-3). RDs secondary to atrophic holes typically occur in younger patients with myopia and absence of PVD; they are often asymptomatic until fixation is involved.

Byer NE. Lattice degeneration of the retina. *Surv Ophthalmol.* 1979;23(4):213–248.

Byer NE. Long-term natural history of lattice degeneration of the retina. *Ophthalmology.* 1989;96(9):1396–1402.

Vitreoretinal Tufts

Peripheral retinal tufts are small, focal areas of elevated glial hyperplasia associated with vitreous or zonular attachment and traction, which may be overlooked if careful peripheral examination with scleral depression is not performed. Tractional tufts are classified according to anatomical, pathogenetic, and clinical distinctions into the following groups:

- noncystic retinal tufts (Fig 16-4)
- cystic retinal tufts
- zonular traction retinal tufts (Fig 16-5)

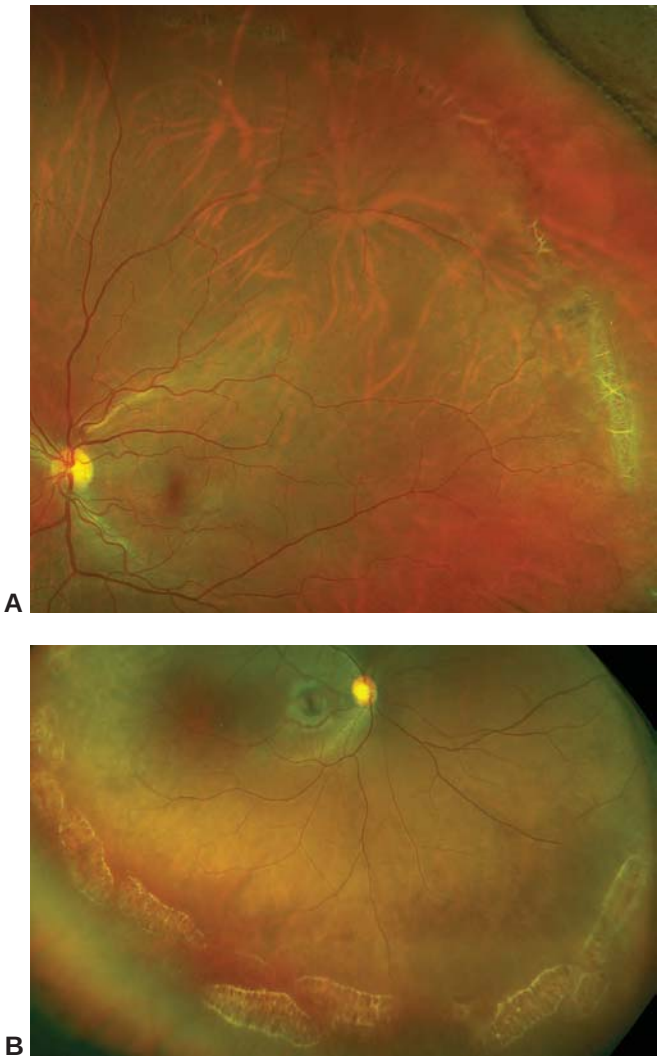

Figure 16-2 Lattice degeneration. **A,** Wide-field color fundus photograph of lattice degeneration. Vascular sheathing is apparent where vessels cross the area of lattice. Characteristic white lattice lines are visible. **B,** Color fundus photograph from another patient shows lattice degeneration inferiorly that appears similar to "snail track" degeneration, with multiple small atrophic retinal holes. *(Courtesy of Hannah J. Yu, BS, and Charles C. Wykoff, MD, PhD.)*

Retinal pigment epithelial hyperplasia may surround the tuft. Cystic and zonular traction retinal tufts, both with firm vitreoretinal adhesions, may predispose eyes to retinal tears and detachment.

Byer NE. Cystic retinal tufts and their relationship to retinal detachment. *Arch Ophthalmol.* 1981;99(10):1788–1790.

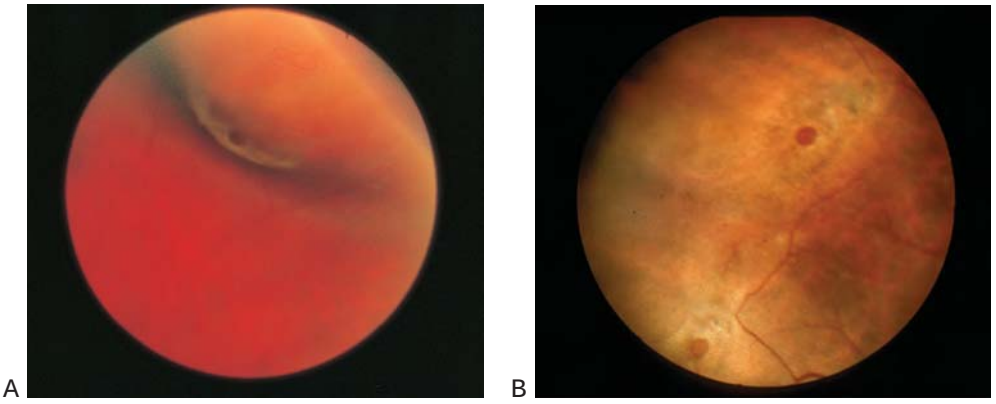

Figure 16-3 Lattice degeneration with atrophic hole. **A,** Fundus photograph of lattice degeneration with a small atrophic hole as viewed with scleral depression. **B,** Fundus photograph of atrophic holes as viewed without scleral depression. *(Part A courtesy of Norman E. Byer, MD.)*

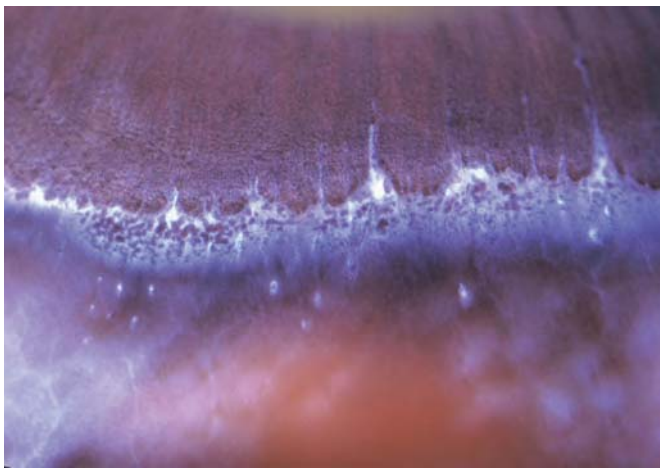

Figure 16-4 Color photograph of a gross eye specimen shows a cluster of white surface nodules with characteristic gross appearance and location of noncystic retinal tufts. *(Reproduced with permission from Foos RY, Silverstein RN, eds.* System of Ocular Pathology. *Vol. 3. iPATH Press; 2004.)*

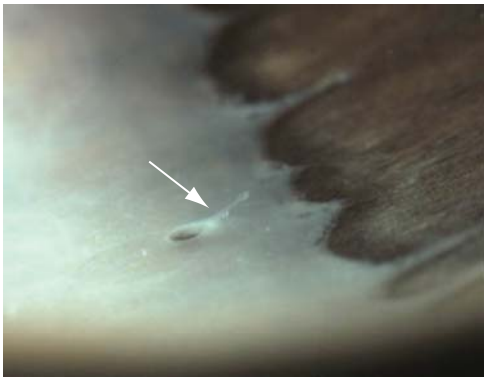

Figure 16-5 Color photograph of a gross eye specimen shows a small zonular traction retinal tuft *(arrow)* with cystic base. Note that the tuft points anteriorly toward the peripheral lens. *(Reproduced with permission from Foos RY, Silverstein RN, eds.* System of Ocular Pathology. *Vol. 3. iPATH Press; 2004.)*

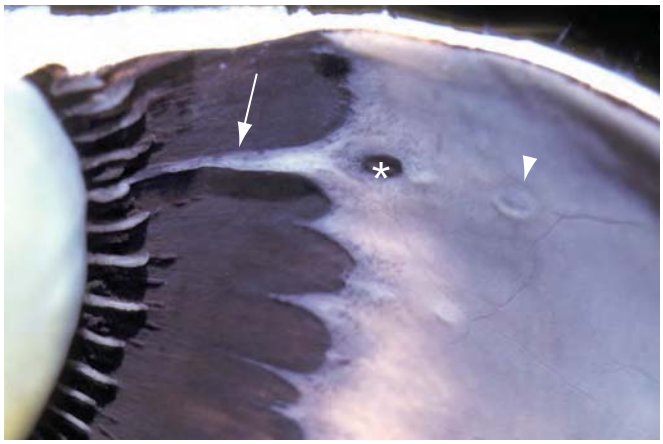

Figure 16-6 Color photograph of a gross eye specimen shows a meridional complex, consisting of an atypical and large dentate process *(white arrow)* that is continuous with a ciliary process of the pars plicata and an area of enclosed pars plana and ora bay *(asterisk)*. Slightly posterior to the complex is a small area in the same meridian that appears to be excavated but is in fact a cyst *(arrowhead)*. *(Modified with permission from Foos RY, Silverstein RN, eds.* System of Ocular Pathology. *Vol. 3. iPATH Press; 2004.)*

Meridional Folds, Enclosed Ora Bays, and Peripheral Retinal Excavations

Meridional folds are folds of redundant retina, usually located superonasally. They are usually associated with dentate processes but may also extend posteriorly from ora bays. Occasionally, tears associated with PVD occur at the most posterior limit of the folds (see Chapter 1, Fig 1-3). Retinal tears can also occur at or near the posterior margins of enclosed ora bays, which are oval islands of pars plana epithelium located immediately posterior to the ora serrata and completely or almost completely surrounded by peripheral retina (Fig 16-6). Occasionally, tears may occur at the site of peripheral retinal excavations, which represent a mild form of lattice degeneration. The excavations may have firm vitreoretinal adhesions and are found adjacent to, or up to 4 disc diameters (DDs) posterior to, the ora serrata. They are often aligned with meridional folds.

Engstrom RE Jr, Glasgow BJ, Foos RY, Straatsma BR. Degenerative diseases of the peripheral retina. In: Tasman W, Jaeger EA, eds. *Duane's Clinical Ophthalmology on DVD-ROM.* Vol 3. Lippincott Williams & Wilkins; 2013:chap 26.

Lesions That Do Not Predispose Eyes to Retinal Detachment

Paving-Stone Degeneration

Cobblestone (or paving-stone) degeneration is characterized by peripheral, discrete areas of retinal atrophy (Fig 16-7); it appears in 22% of individuals older than 20 years. The atrophic areas may occur singly or in groups and are sometimes confluent. On histologic examination, these "paving stones" are characterized by atrophy of the retinal pigment epithelium (RPE) and outer retinal layers, attenuation or absence of the choriocapillaris, and adhesions

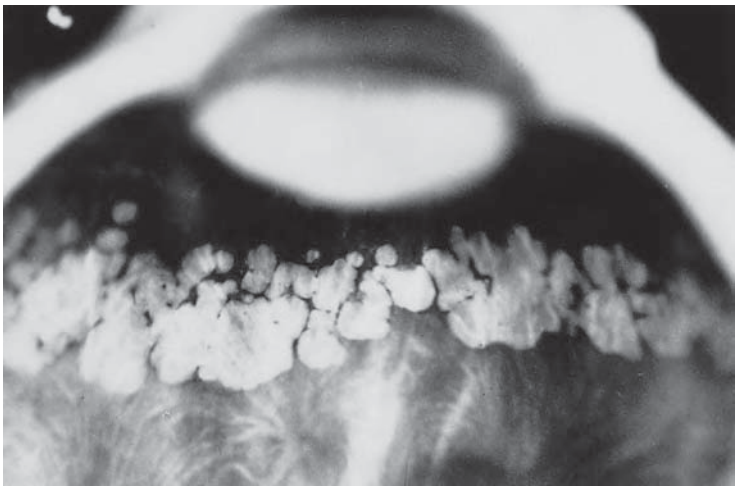

Figure 16-7 Gross appearance of paving-stone degeneration. *(Reproduced with permission from Green WR. Pathology of the retina. In: Frayer WC, ed.* Lancaster Course in Ophthalmic Histopathology, *unit 9. FA Davis; 1988:181.)*

between the remaining neuroepithelial layers and Bruch membrane. These lesions are most common in the inferior quadrants, anterior to the equator. Ophthalmoscopically, they appear yellowish white and are sometimes surrounded by a rim of hypertrophic RPE. Because the RPE is absent or hypoplastic, large choroidal vessels can be visible beneath the lesions.

Retinal Pigment Epithelial Hyperplasia

When stimulated by chronic low-grade traction, RPE cells proliferate. Diffuse retinal pigment epithelial hyperplasia may be observed straddling the ora serrata, in latitudes that correspond roughly to the insertion of the vitreous base. Focal hyperplasia may also occur on the pars plana and in the peripheral retina, especially in areas of focal traction such as vitreoretinal tufts and lattice degeneration. Areas of previous inflammation and trauma may also be sites of retinal pigment epithelial hyperplasia.

Retinal Pigment Epithelial Hypertrophy

Acquired retinal pigment epithelial hypertrophy is a degenerative change associated with aging that commonly occurs in the periphery, often in a reticular pattern. Histologically, it is characterized by large cells and by large, spherical melanin granules. Similar histologic features are present in congenital hypertrophy of the RPE (eg, grouped pigmentation, or "bear tracks"), for which possible associations with systemic findings such as familial adenomatous polyposis (Gardner syndrome) should be considered.

Peripheral Cystoid Degeneration

Typical peripheral cystoid degeneration, characterized by zones of microcysts in the far-peripheral retina, is present in the eyes of almost all adults older than 20 years. Although retinal holes may form in these areas, they rarely cause RD. *Reticular peripheral cystoid*

degeneration, found in approximately 20% of adult eyes, is almost always located posterior to typical peripheral cystoid degeneration. It usually occurs in the inner retina and presents with a linear or reticular pattern that follows the retinal vessels. This form may develop into reticular degenerative retinoschisis. See also the section Retinoschisis.

Retinal Breaks

Retinal breaks are clinically significant in that they may allow liquid from the vitreous cavity to enter the potential space between the neurosensory retina and the RPE, thereby causing an RRD. Some breaks are caused by vitreoretinal traction (tears); others result from atrophy of the retinal layers (holes). Traumatic breaks are discussed in the next section. Retinal breaks may be classified as

- flap, or horseshoe, tears
- giant retinal tears
- operculated holes
- retinal dialyses
- atrophic retinal holes

A *flap tear* occurs when a strip of retina is pulled anteriorly by vitreoretinal traction, often during a PVD or secondary to trauma (Fig 16-8). A tear is considered symptomatic when the patient reports photopsias, floaters, or both. A *giant retinal tear* extends 90° (3 clock-hours) or more circumferentially and usually occurs along the posterior edge of the vitreous base. An *operculated hole* occurs when traction is sufficient to tear a piece of retina completely free from the adjacent retinal surface. A *retinal dialysis* is a circumferential, linear break that occurs at the ora serrata, with vitreous base attached to the retina posterior to the tear's edge; it is commonly a consequence of blunt trauma. An *atrophic retinal hole* is generally not associated with vitreoretinal traction and has not been linked to an increased risk of RD.

American Academy of Ophthalmology Retina/Vitreous Panel. Preferred Practice Pattern Guidelines. *Posterior Vitreous Detachment, Retinal Breaks, and Lattice Degeneration.* American Academy of Ophthalmology; 2019. www.aao.org/ppp

Traumatic Breaks

Blunt or penetrating eye trauma can cause retinal breaks by direct retinal perforation, contusion, or vitreous traction. Fibrocellular proliferation occurring later at the site of an injury may cause vitreoretinal traction and subsequent detachment. Also see Chapter 17.

Blunt trauma can cause retinal breaks by direct contusive injury to the globe through 2 mechanisms: (1) coup, adjacent to the point of trauma, and (2) contrecoup, opposite the point of trauma. Blunt trauma compresses the eye along its anteroposterior axis and expands it in the equatorial plane. Because the vitreous body is viscoelastic, slow compression of the eye has no deleterious effect on the retina. However, rapid compression of the eye results in severe traction on the vitreous base that may tear the retina.

Contusion injury may cause large, ragged equatorial breaks; dialysis; or a macular hole. Traumatic breaks are often multiple, and they are commonly found in the inferotemporal

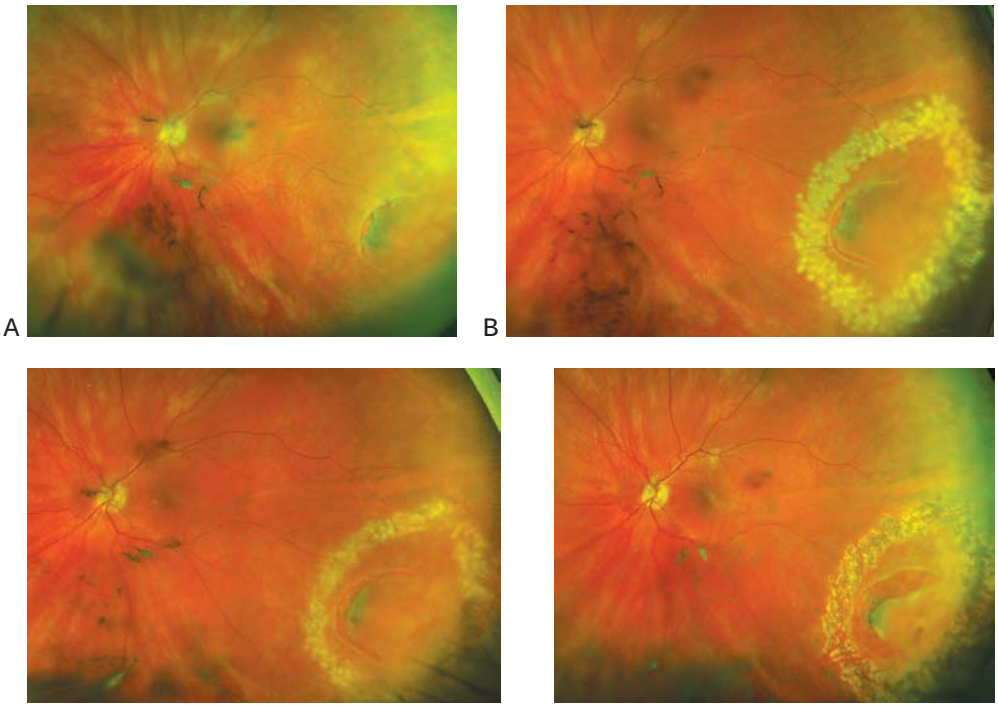

Figure 16-8 Flap retinal tear. Sequence of color fundus photographs of a large, temporally located flap tear with associated small cuff of subretinal fluid and multiple small ruptured bridging retinal vessels with mild vitreous hemorrhage. **A,** At presentation. **B,** Immediately following laser demarcation. **C,** One week after laser demarcation. **D,** One month after laser demarcation. *(Courtesy of Hannah J. Yu, BS, and Charles C. Wykoff, MD, PhD.)*

and superonasal quadrants. The most common contusion injuries are dialyses, which may be as small as 1 ora bay (the distance between 2 retinal dentate processes at the latitude of the ora serrata) or may extend 90° or more. Dialyses are usually located at the posterior border of the vitreous base but can also occur at the anterior border (Fig 16-9, Activity 16-1). Avulsion of the vitreous base may be associated with dialysis and is considered pathognomonic of ocular contusion. The vitreous base can be avulsed from the underlying retina and nonpigmented epithelium of the pars plana without tearing either one; generally, however, one or both may also be torn in the process. Less common types of breaks caused by blunt trauma are horseshoe tears (which may occur at the posterior margin of the vitreous base, at the posterior end of a meridional fold, or at the equator) and operculated holes.

ACTIVITY 16-1 Anatomy Marker Activity: Retinal tears and holes.
Courtesy of Mark M. Miller and Colin A. McCannel, MD.

Trauma in Young Eyes

Although young patients have a higher incidence of eye injury than do other age groups, only in rare instances does the retina detach immediately after blunt trauma because

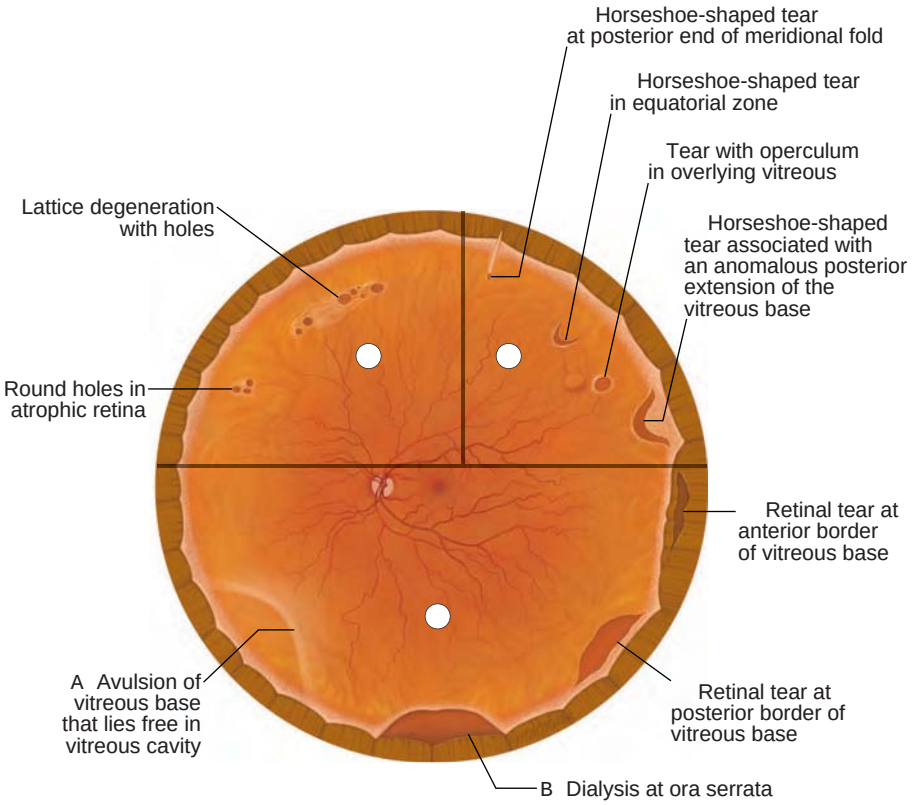

Figure 16-9 Schematic illustration of retinal tears and holes. **Part 1:** Retinal breaks at borders of the vitreous base. **A,** Avulsion of the vitreous base that lies free in the vitreous cavity. **B,** Dialysis of the ora serrata. **C,** Retinal tear at the posterior border of the vitreous base. **D,** Retinal tear at the anterior border of the vitreous base. **Part 2:** Retinal breaks with areas of abnormal vitreoretinal interface (lattice degeneration). **E,** Lattice degeneration with holes. **F,** Round holes in atrophic retina. **Part 3:** Retinal breaks associated with abnormal vitreoretinal attachments. **G,** Flap, or horseshoe, tear at the posterior end of a meridional fold. **H,** Horseshoe tear in the equatorial zone. **I,** Tear with operculum in the overlying vitreous. **J,** Horseshoe tear associated with an anomalous posterior extension of the vitreous base. *(Illustration by Mark M. Miller.)*

young vitreous has not yet undergone synchysis (liquefaction). The vitreous, therefore, does not allow fluid movement through the retinal tears or dialyses. With time, however, the vitreous may liquefy over a tear, allowing fluid to pass through the break to detach the retina. The clinical presentation of the RD is thus usually delayed:

- 12% of detachments identified immediately
- 30% identified within 1 month
- 50% identified within 8 months
- 80% identified within 24 months

Traumatic RDs in young patients may be shallow and often show signs of chronicity, including multiple demarcation lines, subretinal deposits, and areas of intraretinal schisis.

When posterior vitreous separation is present or occurs later after trauma, retinal breaks are often associated with abnormal vitreoretinal attachments and may resemble nontraumatic breaks. RDs may occur acutely in these patients.

Prophylactic Treatment of Retinal Breaks

Any retinal break can cause an RD by allowing liquid from the vitreous cavity to pass through the break and separate the sensory retina from the RPE. However, the majority of retinal holes and breaks do not cause a detachment.

The ophthalmologist may consider prophylactic treatment of breaks in an attempt to reduce the risk of RD (Table 16-1). Treatment does not eliminate the risk of new tears or detachment, an important point for patient counseling.

The goal of prophylactic laser treatment of or cryotherapy for retinal breaks is the creation of a chorioretinal adhesion around each break to prevent fluid from entering the subretinal space. When subretinal fluid is present, treatment is applied so that it fully surrounds the area of subretinal fluid (see Fig 16-8). When insufficient treatment is applied, vitreous traction can lead to anterior extension of horseshoe tears and RD (Fig 16-10). Similarly, when lattice degeneration is treated, typically the entire lesion is surrounded with treatment applications.

When considering prophylaxis, the ophthalmologist weighs numerous factors, including symptoms, family history, residual traction, size and location of the break, phakic status, refractive error, status of the fellow eye, presence of subretinal fluid, and availability of the patient for follow-up evaluation. The following discussion serves only as a broad guideline, because individual patient characteristics and clinical factors must be considered for each patient (see also the discussion of hereditary hyaloideoretinopathies with optically empty vitreous in Chapter 15).

Table 16-1 Prophylactic Treatment of Retinal Breaks

Type of Retinal Lesion	Treatment
Acute symptomatic dialysis	Treat promptly
Acute symptomatic flap, or horseshoe, tear	Treat promptly
Acute symptomatic operculated hole	Consider treatment
Asymptomatic atrophic hole	Often observed without treatment, but consider treatment in specific clinical circumstances
Asymptomatic dialysis	No consensus guidelines, but consider treatment
Asymptomatic horseshoe tear (no subretinal fluid)	Consider treatment
Asymptomatic lattice degeneration, with or without atrophic holes (no subretinal fluid)	Often observed without treatment, but consider treatment in specific clinical circumstances
Asymptomatic operculated hole	Often observed without treatment, but consider treatment in specific clinical circumstances
Eyes with lattice degeneration, atrophic holes, or asymptomatic retinal tear where the fellow eye has had a retinal detachment	No consensus guidelines, but consider treatment

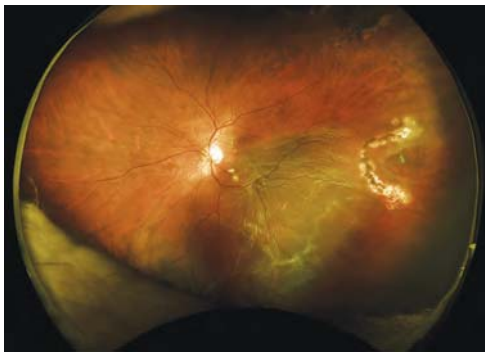

Figure 16-10 Wide-field image shows a horseshoe tear inadequately surrounded by laser applications and subsequent retinal detachment (RD). *(Courtesy of Stephen J. Kim, MD.)*

Symptomatic Retinal Breaks

Overall, 7%–18% of eyes with a symptomatic PVD are found to have 1 or more tractional tears at initial examination. Numerous clinical studies have demonstrated that acute symptomatic breaks are at substantial risk of progressing to RD, especially when there is associated vitreous hemorrhage. Therefore, acute symptomatic flap tears are commonly treated prophylactically.

Acute symptomatic operculated holes may be less likely to cause detachment because there may be no residual traction on the adjacent retina. Typically, prophylactic treatment is performed when evaluation reveals persistent vitreous traction at the margin of an operculated hole, when the hole is large or located superiorly, or when there is vitreous hemorrhage.

Atrophic holes may be incidental findings in a patient who presents with an acute PVD, and treatment may not be necessary for these holes.

Asymptomatic Retinal Breaks

Asymptomatic flap tears progress to RD in approximately 5% of cases. Because of this risk, treatment may be considered but may not be universally recommended in emmetropic, phakic eyes. Asymptomatic flap tears accompanied by additional clinically relevant findings such as lattice degeneration, myopia, or subclinical detachment, or by aphakia, pseudophakia, or history of RD in the fellow eye may confer an increased risk of progression to RD, and treatment may be recommended. Asymptomatic operculated holes and atrophic holes rarely cause RD, so treatment is generally not recommended unless the patient is at high risk for RD.

Lattice Degeneration

Although limited data are available, an 11-year follow-up study of patients with untreated lattice degeneration and no symptomatic tears showed that RD occurred in approximately 1% of cases. Thus, the presence of lattice, with or without atrophic holes, generally does not require prophylaxis in the absence of other risk factors or symptoms. If lattice degeneration is present in a patient with additional risk factors, such as RD in the fellow eye, flap tears, pseudophakia, or aphakia, prophylactic treatment may be considered.

Wilkinson CP. Interventions for asymptomatic retinal breaks and lattice degeneration for preventing retinal detachment. Cochrane Database Syst Rev. 2014;(9):CD003170.

Aphakia and Pseudophakia

Removal of the native crystalline lens can lead to anterior shifting of the vitreous and subsequent retinal traction. Because aphakic and pseudophakic eyes have higher risks of RD (1%–3%) than phakic eyes, such patients should be warned of potential symptoms of RD and carefully examined if they occur. These symptoms overlap with symptoms associated with PVD and include new flashes, floaters, or the appearance of curtains in the visual field. In a population-based 25-year follow-up study comparing eyes that underwent cataract surgery with eyes that did not, the probability of RD following cataract surgery was greatest in the first year (an approximately 11-fold difference, compared with approximately fourfold in years 5 through 20). The cumulative risk of RD steadily increased to 1.79% at 20 years. Risk factors for the development of RD following cataract surgery include male sex, younger age, myopia, increased axial length, posterior capsule tear, and absence of a PVD.

Erie JC, Raecker MA, Baratz KH, Schleck CD, Burke JP, Robertson DM. Risk of retinal detachment after cataract extraction, 1980–2004: a population-based study. *Ophthalmology.* 2006;113(11):2026–2032.

Fellow Eye in Patients With Retinal Detachment

In patients with RD, the fellow-eye risk of detachment is approximately 10% for phakic patients and substantially higher for aphakic and pseudophakic patients. Vitreous status may play a role in determining this clinical risk. Complete retinal evaluation, including peripheral evaluation with 360° scleral depression, may be considered for patients developing concerning symptoms. Prophylactic treatment of retinal breaks and lattice degeneration, even if asymptomatic, may be considered. Additional clinical scenarios, including Stickler syndrome, may indicate the need for fellow-eye prophylactic treatment to decrease the risk of RD development.

Folk JC, Arrindell EL, Klugman MR. The fellow eye of patients with phakic lattice retinal detachment. *Ophthalmology.* 1989;96(1):72–79.

Subclinical Retinal Detachment

The term *subclinical RD* is used in various ways in the literature. It can refer to an asymptomatic RD, or a detachment in which subretinal fluid extends more than 1 DD from the break but not more than 2 DDs posterior to the equator. Because approximately 30% of such detachments progress, treatment may be considered. Treatment may be advised particularly for symptomatic patients and cases involving traction on the break. Presence of a demarcation line suggests a lower risk; however, progression may occur through the demarcation line.

Brod RD, Flynn HW Jr, Lightman DA. Asymptomatic rhegmatogenous retinal detachments. *Arch Ophthalmol.* 1995;113(8):1030–1032.

Retinal Detachment

Retinal detachments are classified as rhegmatogenous, traction, or exudative. The most common are *rhegmatogenous retinal detachments (RRDs).* The term is derived from the

Greek *rhegma,* meaning "break." RRDs are caused by fluid passing from the vitreous cavity through a retinal break into the potential space between the neurosensory retina and the RPE. Less common than RRDs, *traction* (also called *tractional*) *detachments* are caused by proliferative membranes that contract and elevate the retina. Combinations of traction and rhegmatogenous pathophysiologic components may also lead to RD. *Exudative,* or *serous, detachments* are caused by retinal or choroidal diseases in which fluid accumulates beneath the neurosensory retina.

The differential diagnosis of RD includes retinoschisis, choroidal tumors, and retinal elevation secondary to detachment of the choroid. Table 16-2 lists diagnostic features of the 3 types of RD.

Rhegmatogenous Retinal Detachment

The Rochester epidemiology project determined that RRD has an annual incidence of 12.6 per 100,000 persons in a primarily White population. A given individual's risk is affected by the presence or absence of certain factors, including high myopia, family history of RD, fellow-eye retinal tear or detachment, recent vitreous detachment, pseudophakia, trauma, peripheral high-risk lesions, and vitreoretinal degenerations.

In 90%–95% of RRDs, a definite retinal break can be found, often with the help of Lincoff rules (Fig 16-11). In the remainder, an occult break is presumed to be present. If no break can be found, the ophthalmologist must rule out all other causes of retinal elevation. Half of patients with RRD have photopsias or floaters. The intraocular pressure is usually lower in the affected eye than in the fellow eye but may occasionally be higher, for example, when associated with Schwartz-Matsuo syndrome. A Shafer sign, descriptively termed "tobacco dust" because of the small clumps of pigmented cells in the anterior vitreous, is frequently present. The retina typically detaches progressively from the periphery to the optic nerve head; usually it has convex borders and contours and a corrugated appearance, especially in recent RDs, and undulates with eye movements. In contrast, in a long-standing RRD, the retina may appear smoother and thinner. Further, retinal macrocysts, arising from the outer plexiform layer, can be seen in long-standing RRD, particularly of the traumatic type with retinal dialysis (Fig 16-12). Fixed folds resulting from proliferative vitreoretinopathy (PVR) almost always indicate an RRD. Shifting fluid may occur, but it is uncommon and more typical of exudative RDs.

PVR is the most common cause of failure after surgical repair of an RRD. In PVR, retinal pigment epithelial, glial, and other cells grow and migrate on both the inner and outer retinal surfaces and on the vitreous face, forming membranes. Contraction of these membranes causes fixed retinal folds, often appearing as star-shaped folds; equatorial traction; detachment of the nonpigmented epithelium from the pars plana; and generalized retinal shrinkage (Fig 16-13). As a result, the causative retinal breaks may reopen, new breaks may occur, or a traction detachment may develop.

To better compare preoperative anatomy with postoperative outcomes, a classification scheme of PVR was created (Table 16-3). The classification lists 3 grades of PVR (A, B, and C), which correspond to increasing severity of disease. Anterior and posterior involvement (CA, CP) are distinguished and subclassified into focal, diffuse, circumferential, subretinal, and anterior displacement, and the extent of the pathology is described in clock-hours.

Table 16-2 Diagnostic Features of the 3 Types of Retinal Detachments

	Rhegmatogenous (Primary)	Nonrhegmatogenous (Secondary)	
		Traction	Exudative
History	Aphakia, myopia, blunt trauma, photopsia, floaters, visual field defect; progressive, generally healthy	Diabetes, ischemic venous occlusive disease, sickle cell disease, penetrating trauma, prematurity	Systemic factors such as malignant hypertension, eclampsia, renal failure, severe central serous chorioretinopathy
Retinal break	Tears or holes identified in 90%–95% of cases	None or may develop secondarily	None, or coincidental
Extent and characteristics of detachment	Extends ora serrata to optic nerve head early, has convex borders and surfaces and corrugated appearance; gravity dependent	Frequently does not extend to ora serrata, may be central or peripheral, has concave borders and surfaces	Volume and gravity dependent; extension to ora serrata is variable, may be central or peripheral, smooth surface
Retinal mobility	Undulating bullae or folds	Taut retina, peaks to traction points	Smoothly elevated bullae, usually without folds
Evidence of chronicity	Demarcation lines, intraretinal macrocysts, atrophic retina	Demarcation lines	Usually none
Pigment in vitreous	Present in 70% of cases	Present in trauma cases	Not present
Vitreous changes	Frequently syneretic; posterior vitreous detachment, traction on flap of tear	Vitreoretinal traction with preretinal proliferative membranes	Usually clear, except in uveitis
Subretinal fluid	Clear	Clear, no shift	May be turbid and shift rapidly to dependent location with changes in head position
Choroidal mass	None	None	May be present
Intraocular pressure	Frequently low	Usually normal	Varies
Transillumination	Normal	Normal	Normal; however, blocked transillumination if pigmented choroidal lesion present
Examples of conditions causing detachment	Retinal break	Proliferative diabetic retinopathy, retinopathy of prematurity, toxocariasis, sickle cell retinopathy, posttraumatic vitreous traction	Uveitis, metastatic tumor, malignant melanoma, Coats disease, Vogt-Koyanagi-Harada syndrome, retinoblastoma, choroidal hemangioma, senile exudative maculopathy, exudative detachment after cryotherapy or diathermy

Modified from Hilton GF, McLean EB, Brinton DA, eds. *Retinal Detachment: Principles and Practice.* 2nd ed. Ophthalmology Monograph 1. American Academy of Ophthalmology; 1995.

B

n the primary brea lies within cloc -hours of the highest border

Total or superior detachments that cross the 12 o'clock meridian:
In 93%, the primary break is at 12 o'clock or in a triangle, the apex of which is at the ora serrata, and the sides of which extend 1 clock-hours to either side of 12 o'clock.

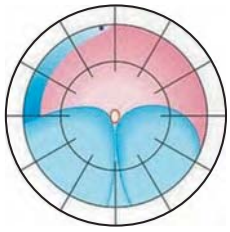

Figure 16-11 Lincoff rules for finding the primary retinal break. Guttering, as illustrated in Rule 4, may be difficult to identify and may require indirect ophthalmoscopy with scleral depression for visualization. *(Reproduced with permission from Kreissig I.* A Practical Guide to Minimal Surgery for Retinal Detachment. *Vol 1. Thieme Medical Publishers, Inc; 2000:13–18. www.thieme.com.)*

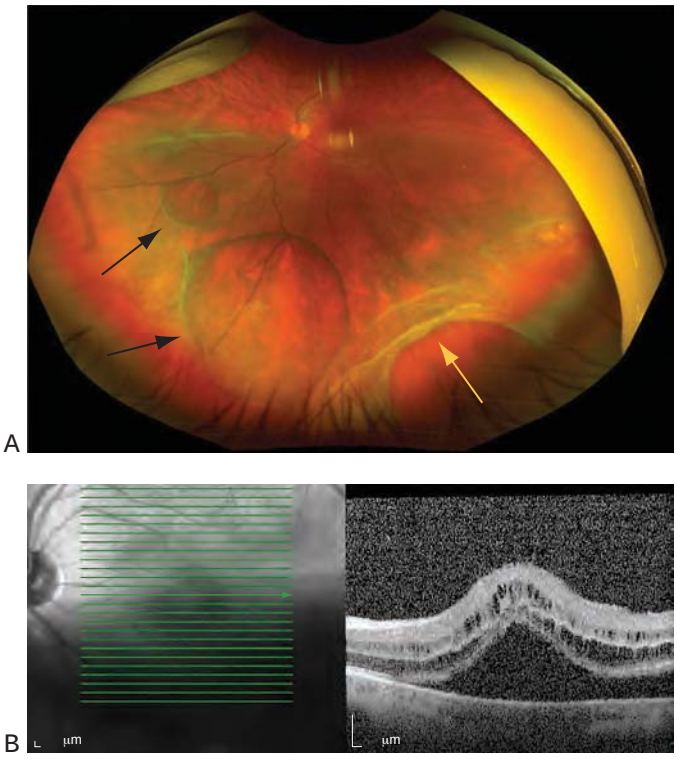

Figure 16-12 Retinal dialysis and macrocysts. **A,** Wide-field color fundus photograph shows a long-standing macula-off RD with an inferotemporal dialysis *(yellow arrow)* and large nasal and inferior macrocysts *(black arrows)* that arise from the outer plexiform layer. **B,** OCT shows that the detachment extends through the macula. *(Courtesy of Avni P. Finn, MD, MBA.)*

Han DP, Lean JS. Proliferative vitreoretinopathy. In: Albert DM, Miller JW, Azar DT, Blodi BA, eds. *Albert & Jakobiec's Principles and Practice of Ophthalmology.* Saunders; 2008:chap 183.

Rowe JA, Erie JC, Baratz KH, et al. Retinal detachment in Olmsted County, Minnesota, 1976 through 1995. *Ophthalmology.* 1999;106(1):154–159.

Management of rhegmatogenous retinal detachment

The principles of surgery for RD are as follows:

- Find all retinal breaks.
- Create a chorioretinal irritation around each break.
- Close the retinal breaks.

The most important element in management of RD is a careful retinal examination. Retinal breaks can be closed by several methods, all of which involve bringing the RPE and choroid into contact with the retina long enough to produce a chorioretinal adhesion that will permanently wall off the subretinal space. This process usually involves 1 of 3 approaches: (1) *scleral buckling,* (2) *vitrectomy,* or (3) *pneumatic retinopexy.* For acute,

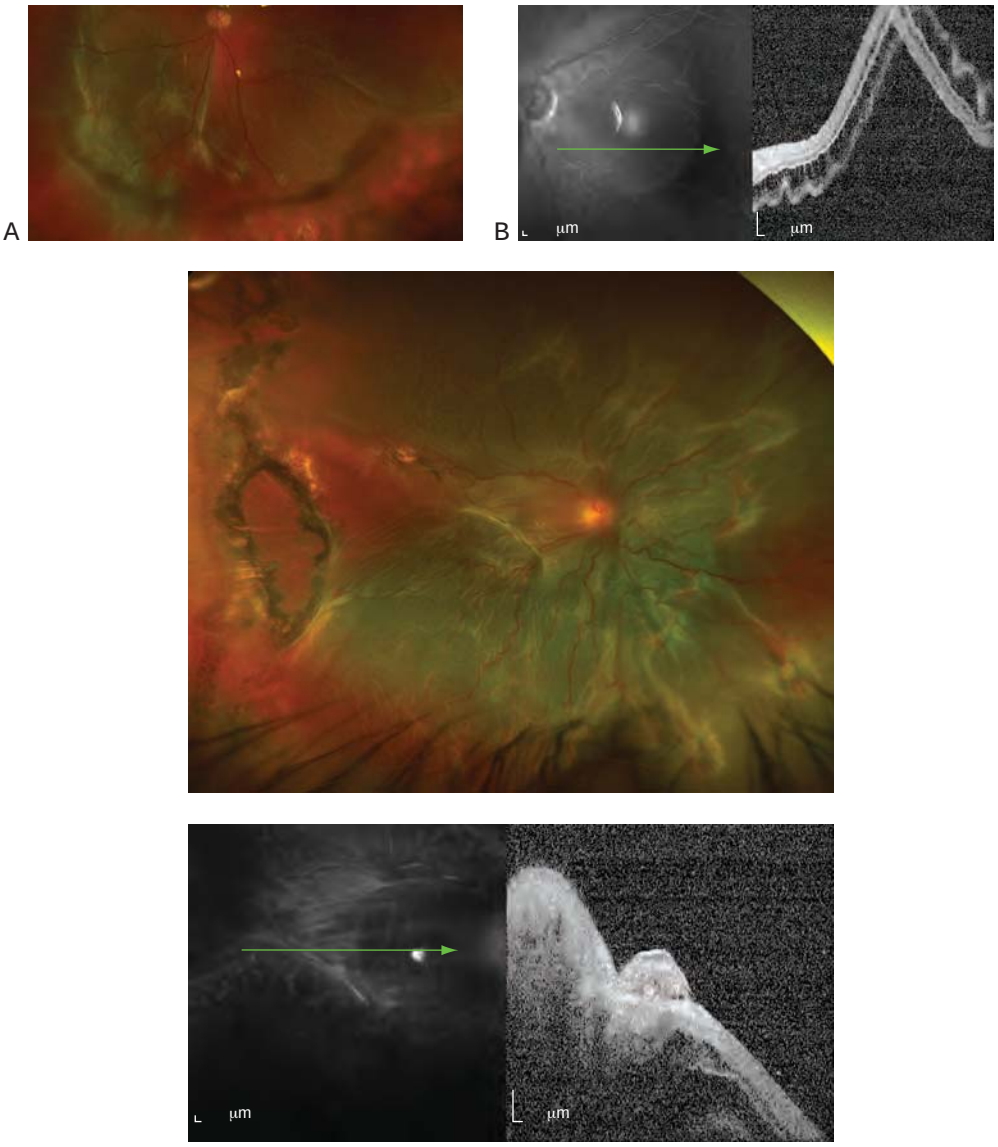

Figure 16-13 Proliferative vitreoretinopathy. **A,** Wide-field color fundus photograph of rhegmatogenous retinal detachment (RRD) with multiple early fixed retinal folds in the inferonasal quadrant. Underlying indentation of the peripheral retinal pigment epithelium is visualized because of a previously placed encircling scleral buckle. **B,** Corresponding near-infrared reflectance image from the same patient *(left). Green arrow* marks the location of the spectral-domain optical coherence tomography (SD-OCT) image *(right),* which shows RD with no appreciable preretinal proliferative tissues posteriorly. **C,** Wide-field color fundus photograph (different patient) of total RRD with extensive and severe preretinal proliferative tissues with retinal contraction, multiple fixed retinal folds, and a large retinal break temporally. **D,** Corresponding near-infrared reflectance image from the same patient *(left). Green arrow* marks the location of the SD-OCT image *(right)*, which shows RD with thick preretinal proliferative tissues posteriorly. *(Courtesy of Tien P. Wong, MD, and Charles C. Wykoff, MD, PhD.)*

Table 16-3 Classification of Proliferative Vitreoretinopathy, 1991

Grade	Features
A	Vitreous haze, vitreous pigment clumps, pigment clusters on inferior retina
B	Wrinkling of inner retinal surface, retinal stiffness, vessel tortuosity, rolled and irregular edge of retinal break, decreased mobility of vitreous
CP 1–12	Posterior to equator: focal, diffuse, or circumferential full-thickness folds,[a] subretinal strands[a]
CA 1–12	Anterior to equator: focal, diffuse, or circumferential full-thickness folds,[a] subretinal strands,[a] anterior displacement,[a] condensed vitreous with strands

[a] Expressed in number of clock-hours involved.

Used with permission from Machemer R, Aaberg TM, Freeman HM, Irvine AR, Lean JS, Michels RM. An updated classification of retinal detachment with proliferative vitreoretinopathy. *Am J Ophthalmol.* 1991;112(2):159–165.

macula-on RDs with symptoms, surgery is typically performed urgently. Preoperatively, minimizing patient eye movement, particularly saccadic movements while reading, and head positioning to orient the RD in a dependent position can minimize the risk of subretinal fluid extension into the fovea. In contrast, in eyes with chronic RDs with pigmented demarcation lines, treatment may be delayed or may not be needed (Fig 16-14). See Chapter 19 for a more detailed discussion of these management approaches.

Traction Retinal Detachment

Vitreous membranes caused by penetrating injuries or by proliferative retinopathies such as diabetic retinopathy can pull the neurosensory retina away from the RPE, causing a traction RD (Fig 16-15). The retina characteristically has a smooth concave surface and contours and is immobile. The detachment may be central or peripheral and, in rare cases, may extend from the optic nerve head to the ora serrata. In most cases, the causative

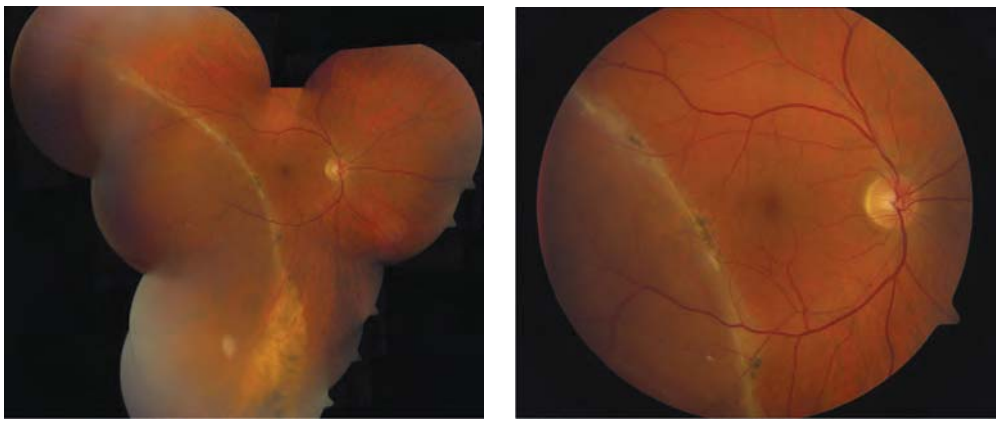

Figure 16-14 Chronic RD. At initial examination *(left),* the patient presented with an asymptomatic large RD with a pigmented and atrophic demarcation line. No progression was observed over several weeks, and surgery was performed electively *(right).* *(Courtesy of Stephen J. Kim, MD.)*

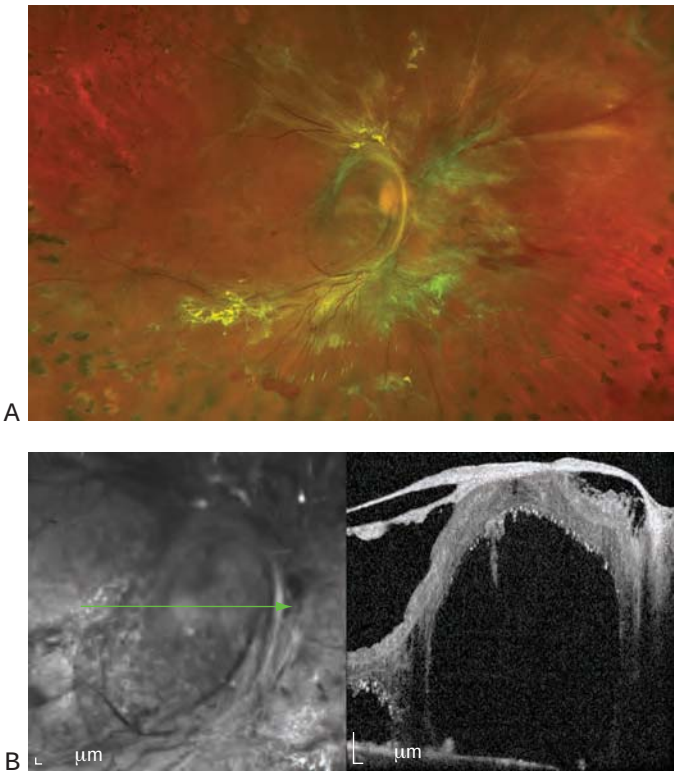

Figure 16-15 Traction RD secondary to proliferative diabetic retinopathy. **A,** Wide-field color fundus photograph of macula-involving traction RD. Extensive fibrotic neovascularization of the optic nerve head extending along the arcade vasculature is visible with associated severe retinal traction. Peripheral panretinal photocoagulation marks are visible. **B,** Corresponding near-infrared reflectance image from the same patient *(left). Green arrow* marks the location of the SD-OCT image *(right),* which shows separation of the neurosensory retina from the underlying retinal pigment epithelium due to thick preretinal proliferative tissues. *(Courtesy of Tien P. Wong, MD, and Charles C. Wykoff, MD, PhD.)*

vitreous membrane can be visualized. If the traction can be released by vitrectomy, the detachment may resolve without evacuation of the subretinal fluid.

In some cases, traction may tear the retina and cause a combined traction and rhegmatogenous detachment. Clinically, the retina loses its concave surface and assumes a convex shape similar to that in an RRD. Retinal mobility is often limited because of the tethering by proliferative tissue. In addition, corrugations characteristic of an RRD are present, and subretinal fluid, which is more extensive than in traction RD, may extend from the optic nerve head to the ora.

Exudative Retinal Detachment

Recognizing that an RD is exudative is crucial because, unlike with other types of RDs, the management of exudative RD is usually nonsurgical. Exudative detachment occurs when either retinal blood vessels leak or the RPE is damaged, allowing fluid to accumulate in the

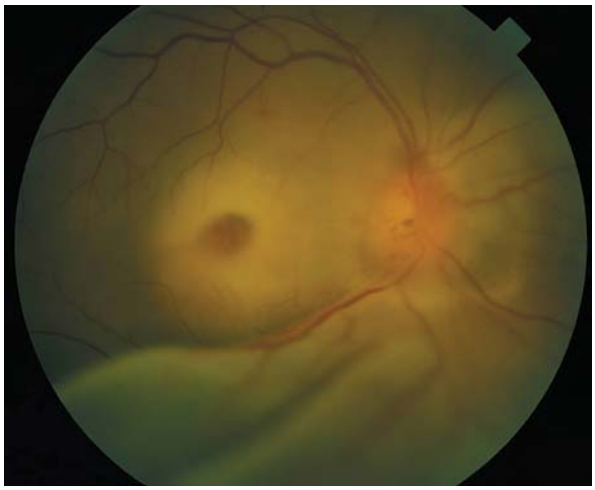

Figure 16-16 Color fundus photograph of an exudative RD that resulted from metastatic breast carcinoma. *(Courtesy of Hermann D. Schubert, MD.)*

subretinal space (Fig 16-16). Neoplasia and inflammatory diseases are the leading causes of large exudative detachments.

The presence of shifting fluid strongly suggests an exudative RD. Because the subretinal fluid responds to gravity, it causes detachment of the area of retina in which it accumulates. For example, when the patient is sitting, the inferior retina is detached. When the patient becomes supine, the fluid moves posteriorly in a matter of seconds or minutes, detaching the macula. Another characteristic of exudative detachments is the smoothness of the detached retinal surface, in contrast to the corrugated appearance in eyes with RRDs. Included in the differential diagnosis is rhegmatogenous inferior bullous detachment associated with a superior retinal tear, in which the subretinal fluid may shift (see Fig 16-11, Rule 4). Fixed retinal folds, which usually indicate PVR, are rarely, if ever, present in exudative detachments. Occasionally, the retina is sufficiently elevated in exudative detachments to be visible directly behind the lens (eg, in Coats disease), a rare occurrence in RRDs.

Differential Diagnosis of Retinal Detachment

Retinoschisis

Typical peripheral cystoid degeneration is present in virtually all adult eyes bilaterally. Contiguous with and extending up to 2–3 mm posterior to the ora serrata, the area of degeneration, which is best visualized with scleral depression, has a "bubbly" appearance. The cystoid cavities in the outer plexiform layer contain a hyaluronidase-sensitive mucopolysaccharide. The only known complications of typical cystoid degeneration are coalescence and extension of the cavities and progression to typical degenerative retinoschisis.

Reticular peripheral cystoid degeneration is almost always located posterior to and continuous with typical peripheral cystoid degeneration, but it is considerably less common. It

has a linear or reticular pattern that corresponds to the retinal vessels and a finely stippled internal surface. The cystoid spaces are in the nerve fiber layer. This condition may progress to reticular degenerative retinoschisis, also known as *bullous retinoschisis* (Fig 16-17).

Although degenerative retinoschisis is sometimes divided into typical and reticular forms, clinical differentiation is difficult. The complications of posterior extension and progression to RD are associated with the reticular form. Retinoschisis is bilateral in 50%–80% of affected patients, often occurs in the inferotemporal quadrant, and is commonly associated with hyperopia.

In *typical degenerative retinoschisis,* the retina splits in the outer plexiform layer. The outer layer is irregular and appears pockmarked on scleral depression. The inner layer is thin and appears clinically as a smooth, oval elevation, usually in the inferotemporal quadrant but sometimes located superotemporally. Occasionally, small, irregular white dots ("snowflakes") are present; these are footplates of Müller cells and neurons that bridge or formerly bridged the cavity. The retinal vessels may appear sclerotic. In all cases, peripheral cystoid degeneration with a typical bubbly appearance can be found anterior to the

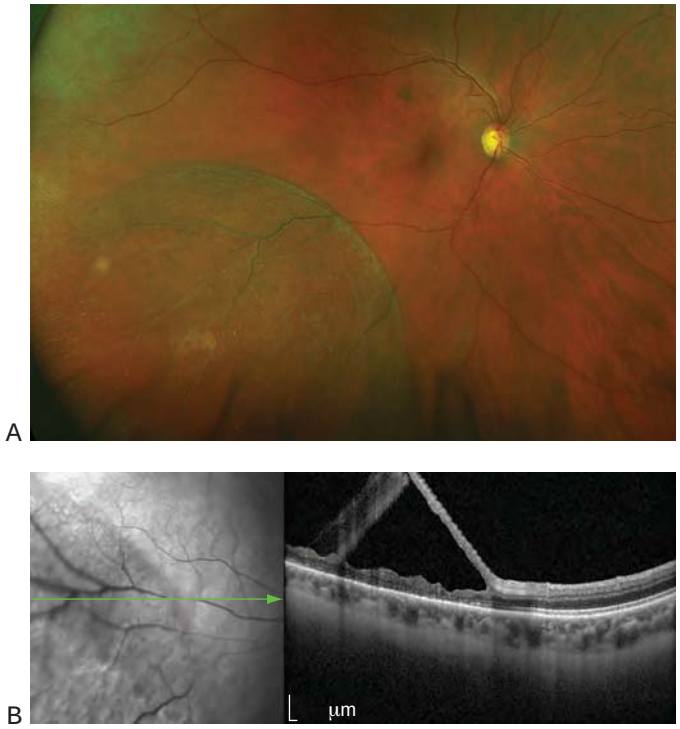

Figure 16-17 Bullous degenerative retinoschisis, also referred to as *reticular degenerative retinoschisis*. **A,** Wide-field color fundus photograph of inferotemporal bullous retinoschisis with visible "snowflakes." **B,** Corresponding near-infrared reflectance image from the same patient *(left).* The area of retinoschisis is visible as the slightly darker area to the left with flat posterior retina to the right. *Green arrow* marks the location of the SD-OCT image *(right),* which shows the transition into the area of retinoschisis with splitting of the retina in the outer plexiform layer and thinning of the inner layer compared with the uninvolved more posterior retina. *(Courtesy of Hannah J. Yu, BS, and Charles C. Wykoff, MD, PhD.)*

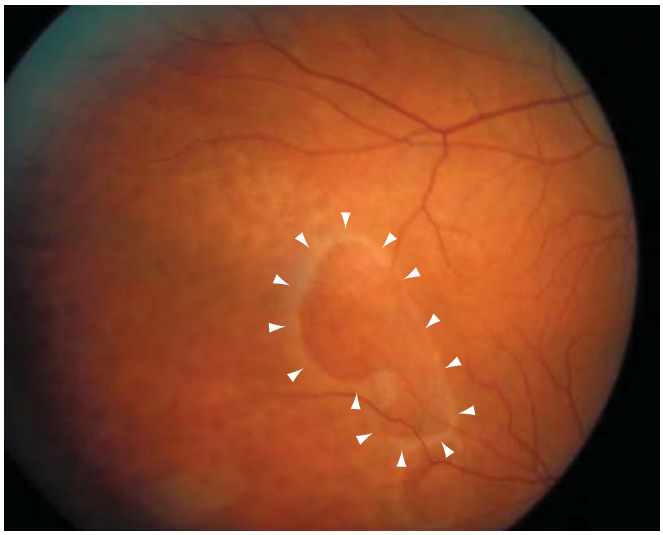

Figure 16-18 Retinoschisis with large, irregular outer-schisis-layer holes *(outlined by arrowheads)* and yellow dots on the inner surface. *(Courtesy of Colin A. McCannel, MD.)*

schisis cavity. The schisis may extend posteriorly to the equator, but complications such as hole formation, RD, or marked posterior extension are rare. The split in the retina almost never extends as far posteriorly as the macula. Laser barricade is generally not effective.

In *reticular degenerative retinoschisis,* the splitting occurs in the nerve fiber layer. The very thin inner layer may be markedly elevated. As in typical retinoschisis, the outer layer appears pockmarked, and the retinal vessels may appear sclerotic. Posterior extension is more common in reticular than in typical retinoschisis. Approximately 23% of cases have holes in the outer wall that may be large and have rolled edges (Fig 16-18).

Reed D, Garg AJ. Degenerative retinoschisis. In: Schachat AP, Wilkinson CP, Hinton DR, Sadda SR, Wiedemann P, eds. *Ryan's Retina.* 6th ed. Elsevier/Saunders; 2018:chap 100.

Differentiation of Retinoschisis From Rhegmatogenous Retinal Detachment

Retinoschisis must be differentiated from RRD (Table 16-4); this can be accomplished by examination and imaging. Retinoschisis causes an absolute scotoma, whereas RRD causes a relative scotoma. Tobacco dust, hemorrhage, or both are present in the vitreous

Table 16-4 Differentiation of Retinoschisis From Rhegmatogenous Retinal Detachment

Clinical Feature	Retinoschisis	Rhegmatogenous Retinal Detachment
Retinal surface	Smooth-domed	Corrugated
Hemorrhage or pigment	Usually absent	Present
Scotoma	Absolute	Relative
Response to photocoagulation	Generally present	Absent
Shifting fluid	Absent	Usually absent
Optical coherence tomography	Subretinal fluid generally absent	Subretinal fluid present

with retinoschisis only in rare instances, whereas they are commonly observed with RRD. In retinoschisis, the retina has a smooth surface and usually appears dome shaped; in contrast, in eyes with RRD, the retina often has a corrugated, irregular surface. In long-standing RRD, however, the retina also may appear smooth and thin, similar to its appearance in retinoschisis. In addition, atrophy of the underlying RPE, demarcation line(s), and degenerative retinal macrocysts may be present in eyes with long-standing RRD, whereas the underlying RPE is normal in eyes with retinoschisis.

Retinoschisis is associated with approximately 3% of RDs. Two types of schisis-related detachments occur. In the first type, holes are present in the outer but not the inner wall of the schisis cavity, so cavity contents can migrate through a hole in the outer wall and slowly detach the retina (see Fig 16-18). Demarcation lines and degeneration of the underlying RPE are common. A demarcation line in an eye with retinoschisis suggests that a full-thickness detachment is present or was formerly present and has spontaneously regressed. This type of retinoschisis detachment usually does not progress, or it may progress slowly and seldom requires treatment.

In the second type of schisis-related detachment, holes are present in both the inner and outer layers of the schisis cavity, which may collapse, resulting in a progressive RRD. Such detachments often progress rapidly and usually require treatment. The causative breaks may be located very posteriorly and thus may be difficult to repair with scleral buckling, often requiring vitrectomy.

Byer NE. Long-term natural history study of senile retinoschisis with implications for management. *Ophthalmology.* 1986;93(9):1127–1137.

Gotzaridis EV, Georgalas I, Petrou P, Assi AC, Sullivan P. Surgical treatment of retinal detachment associated with degenerative retinoschisis. *Semin Ophthalmol.* 2014;29(3): 136–141.

Ip M, Garza-Karren C, Duker JS, et al. Differentiation of degenerative retinoschisis from retinal detachment using optical coherence tomography. *Ophthalmology.* 1999;106(3):600–605.

Xue K, Muqit MMK, Ezra E, et al. Incidence, mechanism and outcomes of schisis retinal detachments revealed through a prospective population-based study. *Br J Ophthalmol.* 2017;101(8):1022–1026.

Macular Lesions Associated With Retinal Detachment

Optic Pit Maculopathy

Optic nerve pits are small, hypopigmented, excavated colobomatous defects of the optic nerve. These oval or round, yellow or whitish defects are usually found within the inferior temporal portion of the optic nerve head margin (Fig 16-19). Most are unilateral, asymptomatic, and congenital, but they can be acquired in the setting of glaucomatous excavation. Optic nerve pits may lead to serous macular detachments with a potentially poor prognosis if left untreated. The macular retinal thickening and detachment typically extend from the optic nerve pit in an oval shape toward the fovea. OCT imaging reveals macular schisis as well as subretinal fluid. Whether the subretinal fluid is liquid vitreous or cerebrospinal fluid is controversial; a proteomic analysis of fluid in 1 adult case

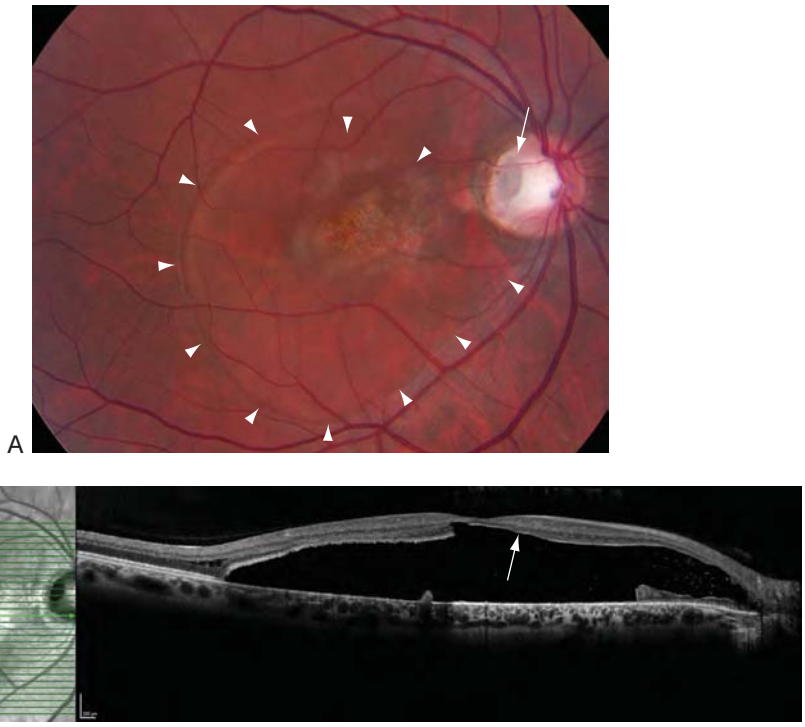

Figure 16-19 Optic nerve pit with macular detachment, retinal thinning, and retinal pigment epithelial atrophy. **A,** Fundus photograph shows an abnormal temporal optic nerve head appearance with an excavation, or pit *(arrow).* The adjacent retina is thickened and elevated, extending into the macula *(outlined by arrowheads).* **B,** OCT scan illustrates subretinal fluid *(pound sign)* associated with the optic nerve pit. Note the degenerated outer segments *(arrow)* of the photoreceptors. *(Courtesy of Colin A. McCannel, MD.)*

confirmed that vitreous was the definite source. Many cases can be observed. When treatment is indicated, many approaches have been reported. The associated schisis as well as subretinal fluid can recur following surgical resolution.

The differential diagnosis of optic nerve pits includes glaucomatous nerve damage, such as optic pit–like changes that may occur at the inferior or superior pole of the optic nerve. Optic nerve pits are included in the differential diagnosis of macular thickening or detachment. Careful examination of the optic nerve head margin is important for recognizing this condition.

Bottoni F, Cereda M, Secondi R, Bochicchio S, Staurenghi G. Vitrectomy for optic disc pit maculopathy: a long-term follow-up study *Graefes Arch Clin Exp Ophthalmol.* 2018; 256(4):675–682.

Jain N, Johnson MW. Pathogenesis and treatment of maculopathy associated with cavitary optic disc anomalies. *Am J Ophthalmol.* 2014;158(3):423–435.

Patel S, Ling J, Kim SJ, Schey KL, Rose K, Kuchtey RW. Proteomic analysis of macular fluid associated with advanced glaucomatous excavation. *JAMA Ophthalmol.* 2016; 134(1):108–110.

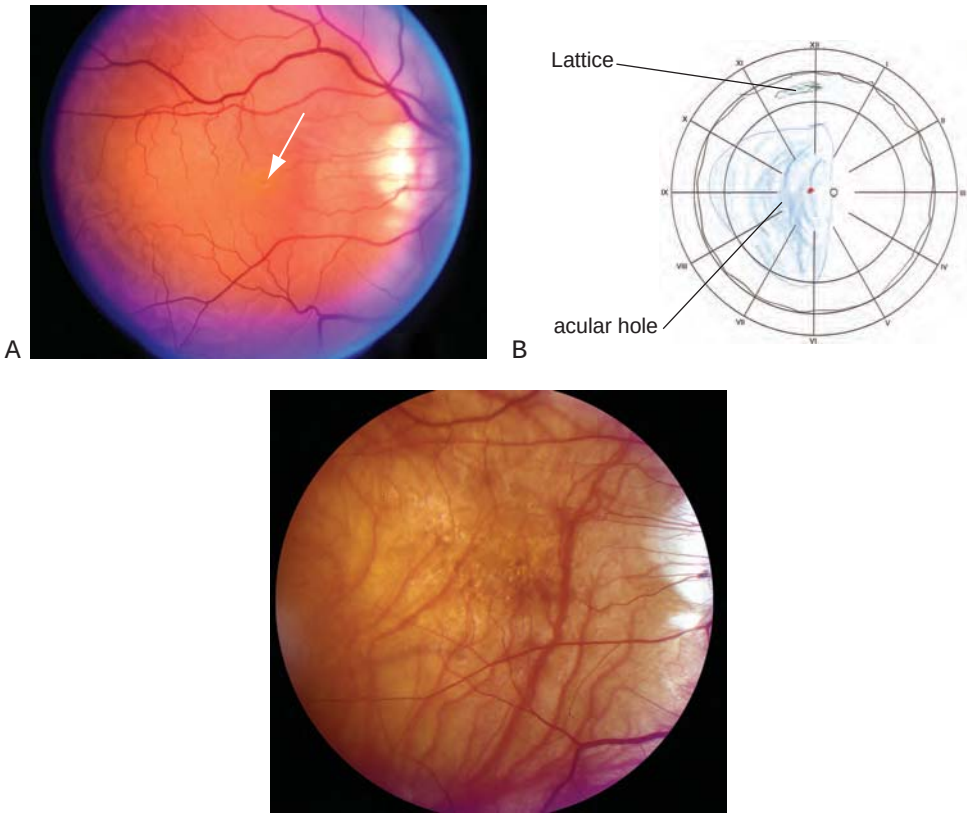

Figure 16-20 A case of RRD caused by a macular hole. **A,** Fundus photograph (50-degree camera) of the detached retina with a hole visible in the center of the macula *(arrow).* **B,** A retinal drawing of the extent of the detachment. The area within the blue outline is detached; the *red dot* symbolizes the macular hole. At presentation, the detachment did not extend to the ora serrata; lack of such an extension is uncharacteristic even for a limited RRD arising from a peripheral break. A lattice lesion with pigmentation is drawn superiorly. **C,** Fundus photograph (30-degree camera) of the macula after successful retinal reattachment using vitrectomy with gas tamponade. Typical myopic fundus features include retinal pigment epithelial changes from myopic degeneration, fair pigmentation, peripapillary scleral crescent, and prominent large choroidal vessels. *(Courtesy of Colin A. McCannel, MD.)*

Macular Holes in High Myopia

A distinct variant of RRD is caused by macular holes, almost always in the setting of a posterior staphyloma in highly myopic eyes (Fig 16-20). Vitreous cavity fluid enters the subretinal space through the macular hole and initiates the detachment. The success rate for surgical repair is lower for this variant than for either macular holes without RRD or typical RRDs.

Ando Y, Hirakata A, Ohara A, et al. Vitrectomy and scleral imbrication in patients with myopic traction maculopathy and macular hole retinal detachment. *Graefes Arch Clin Exp Ophthalmol.* 2017;255(4):673–680.

Ho TC, Ho A, Chen MS. Vitrectomy with a modified temporal inverted limiting membrane flap to reconstruct the foveolar architecture for macular hole retinal detachment in highly myopic eyes. *Acta Ophthalmol.* 2018;96(1):e46–e53.