# Intermediate Uveitis

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## Introduction

Anatomically, the term *intermediate uveitis* (IU) refers to the subset of uveitis where the vitreous is the major site of the inflammation, and the presence of peripheral vascular sheathing and macular edema do not change the classification. The IU may or may not be associated with infection or systemic disease, whereas the term *pars planitis* has been recommended for a particular subset of IU associated with snowbank and snowball formation in the absence of an infectious or systemic disease (Jabs et al. 2005).

## Epidemiology and Etiology

The incidence and prevalence of IU show great variability according to geographic, genetic, and referral patterns of the patients. It is the least common anatomic type of uveitis in most of the series and accounts for 0.9–16% of all uveitis patients (Jones 2015; Nakahara et al. 2017; Silpa-archa et al. 2015; Singh et al. 2004; Yalçındağ et al. 2018). However, pars planitis is an important cause of pediatric uveitis and constitutes 5–26.7% of uveitis in this age group (Ozdal et al. 2015; Yalçındağ et al. 2018).

The IU may be associated with infectious diseases such as tuberculosis, leprosy, Lyme disease, syphilis, toxocariasis, Whipple's disease, Epstein–Barr virus infection, and noninfectious systemic diseases such as multiple sclerosis (MS), sarcoidosis, thyroid disease, and inflammatory bowel disease. The IU cases not associated with infectious or systemic diseases are considered as idiopathic. In a recent study including both pediatric and adult patients, idiopathic IU has been reported in 59% (Ness et al. 2017). Association with systemic diseases is extremely rare, and IU is almost always idiopathic in the pediatric population (Tugal-Tutkun 2011).

Besides systemic disease associations, the presence of HLA-DR15 and HLA-A28 in patients with IU and the presence of HLA-DR2, -DR15, -B51, and DRB1∗0802 in patients with pars planitis suggest an immunogenetic predisposition and autoimmune process in its pathogenesis. Patients with IU who are positive for HLA-DR15 were reported to have systemic findings of other HLA-DR15-related disorders such MS, optic neuritis, and narcolepsy, suggesting a common genetic background (Arellanes-Garcia et al. 2008; Babu and Rathinam 2010; Bonfioli et al. 2005; Ness et al. 2017; Ozdal et al. 2015; Raja et al. 1999).

Although affecting all age groups, IU is mostly seen in young adults within the third and fourth decades (Babu and Rathinam 2010; Bonfioli et al. 2005). Pars planitis, however, predominantly affects children between 6 and 10 years of age and adolescents (Ozdal et al. 2015). The disease has no definite gender predilection. In studies comparing childhood-onset and adulthood-onset cases, boys constituted the majority of childhood cases, while women made up most of the adult cases (Heinz et al. 2014; Paroli et al. 2014).

## Clinical Features

Floaters and blurred vision are the most common symptoms at presentation. Other less common symptoms include pain, photophobia, and red eye. Severe cases may present with significant visual loss due to macular edema or aggregation of floaters in the vitreous. The disease may also be asymptomatic and diagnosed incidentally during routine eye examination, especially in young children (Babu and Rathinam 2010; Donaldson et al. 2007; Paroli et al. 2011). Young children may even present with strabismus secondary to the development of amblyopia or complications which cause leukocoria (Fig. 10.1) (Paroli et al. 2011; Tugal-Tutkun 2011).

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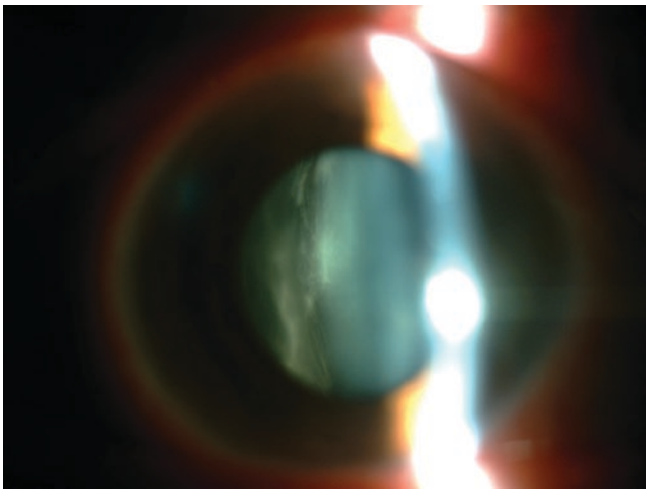

**Fig. 10.1** Slit lamp photograph shows dense vitreous condensation causing leukocoria

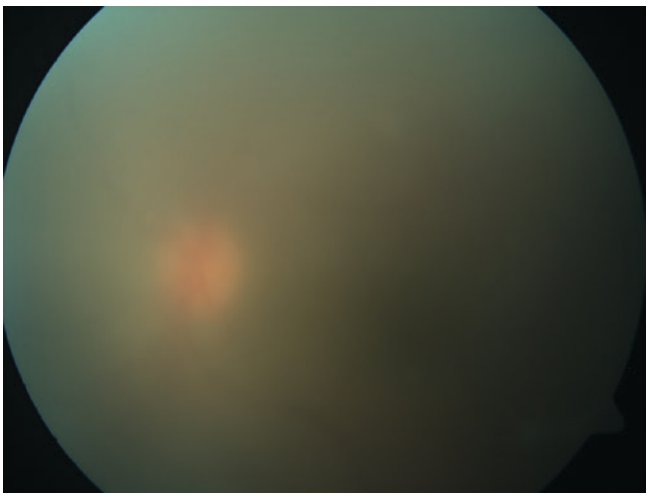

**Fig. 10.3** Fundus photograph shows severe vitreous haze associated with intermediate uveitis

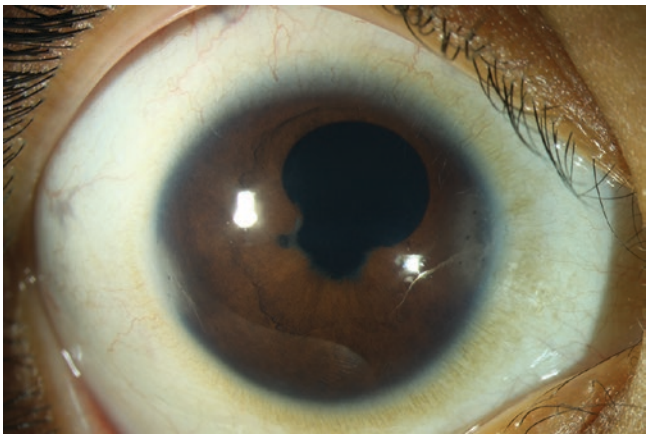

**Fig. 10.2** Slit lamp photograph shows posterior synechiae particularly involving the inferior iris and a band keratopathy in a child with pars planitis

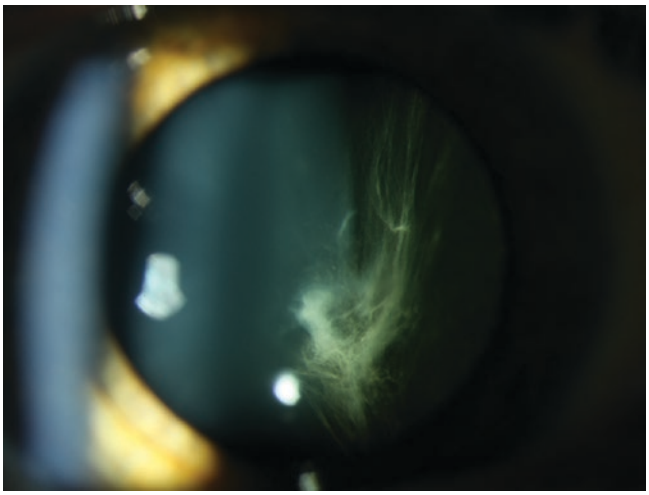

**Fig. 10.4** Slit lamp photograph shows severe vitritis and condensation of the anterior vitreous which is visible in the pupillary area

IU, especially pars planitis, affects both eyes in most of the patients. However, an asymmetric involvement with few vitreous cells in the less affected eye may be seen (Ozdal et al. 2015; Tugal-Tutkun 2011). High rates of bilaterality, ranging between 70% and 90%, have been reported in the Western literature (Babu and Rathinam 2010).

The eye is usually quiet with mild to moderate anterior segment inflammation associated with keratic precipitates (KP) distributed mostly in the inferior part of the cornea. Peripheral corneal endotheliopathy characterized by inferior stromal edema and linearly arranged KPs on the border of edematous and normal cornea has also been reported. Posterior synechiae particularly involving the inferior iris (Fig. 10.2) may be observed especially in childhood pars planitis and usually do not occur in adulthood. In children with long-standing inflammation, a band keratopathy may develop (Bonfioli et al. 2005; Donaldson et al. 2007; Tugal-Tutkun 2011). Compared to adults, anterior segment inflammation, band keratopathy (Fig. 10.2), peripheral corneal endotheliopathy, and posterior synechiae are more frequent in children (Tugal-Tutkun 2011). The characteristic finding of IU is vitritis (Fig. 10.3) which may or may not cause vitreous haze. Vitreous inflammation may be severe and can be visible in the pupillary area at biomicroscopic examination (Fig. 10.4). Yellow-white inflammatory aggregates called as *snowballs* are usually found in the mid-vitreous and inferior peripheral vitreous (Fig. 10.5). A snowball may rarely be located on the surface of the macula. With progression, these aggregates coalesce (Fig. 10.6a, b) forming a plaque of exudates usually located inferiorly and called as *snowbank* (Fig. 10.7a, b). Snowballs and snowbank are diagnostic findings of pars planitis. Vitreous bands and vitreous condensations at the inferior peripheral retina are findings

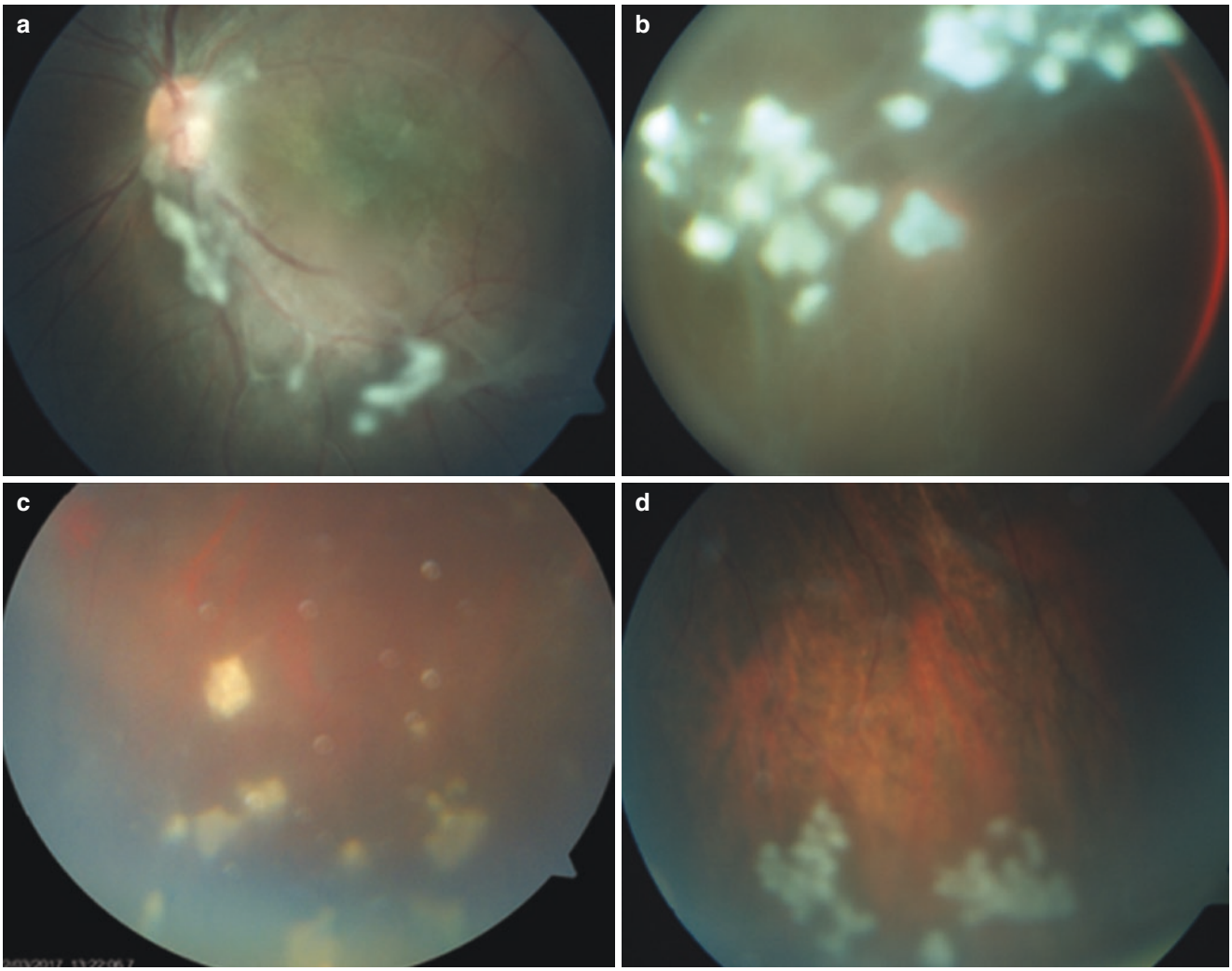


**Fig. 10.5** (**a**–**d**) Fundus photographs show variable location of snowballs in patients with pars planitis

suggesting pars planitis (Fig. 10.8). Sheathing of peripheral retinal venules due to retinal vasculitis is another common clinical finding of IU (Fig. 10.9). Optic disc edema can be observed in around 70% of the cases when fluorescein angiography (FA) is performed (Arellanes-Garcia et al. 2008) (Fig. 10.10a, b).

## Ocular Complications

Cystoid macular edema (CME) is the most common complication of IU and the leading cause of visual morbidity (Fig. 10.11). Chronic edema can lead to further macular complications such as scarring, epiretinal membranes, and macular hole formation (Fig. 10.12) (Bonfioli et al. 2005).

Because of the chronic and asymptomatic course, presentation with ocular complications is a prevalent condition especially among pediatric patients. The most frequent complications of pars planitis include CME, cataract, and severe vitreous opacities. Band keratopathy (Figs. 10.2 and 10.13), seclusion pupillae (Fig. 10.13), glaucoma, epiretinal membrane formation (Fig. 10.14a–d), vitreous condensation, retinal neovascularizations (Fig. 10.15a, b), vitreous hemorrhage (Fig. 10.16), retinal detachment, peripheral retinoschisis, cyclitic membranes, and amblyopia are also well-known consequences of chronic pars planitis in children (Arellanes-Garcia et al. 2008; Ness et al. 2017; Ozdal et al. 2015).

Occasionally, dense vitreous condensation appearing as leukocoria may be misdiagnosed as cataract particularly in young children. Pars planitis is the leading cause of vitreous hemorrhage in children which may be associated with neovascularization of the optic disc or the peripheral retina (Tugal-Tutkun 2011). Optic disc neovascularization is mostly due to severe intraocular inflammation. Retinal neovascularization, elsewhere or in the snowbank, and rarely peripapillary subretinal neovascularization have also been reported in pars planitis (Arellanes-Garcia et al. 2008).

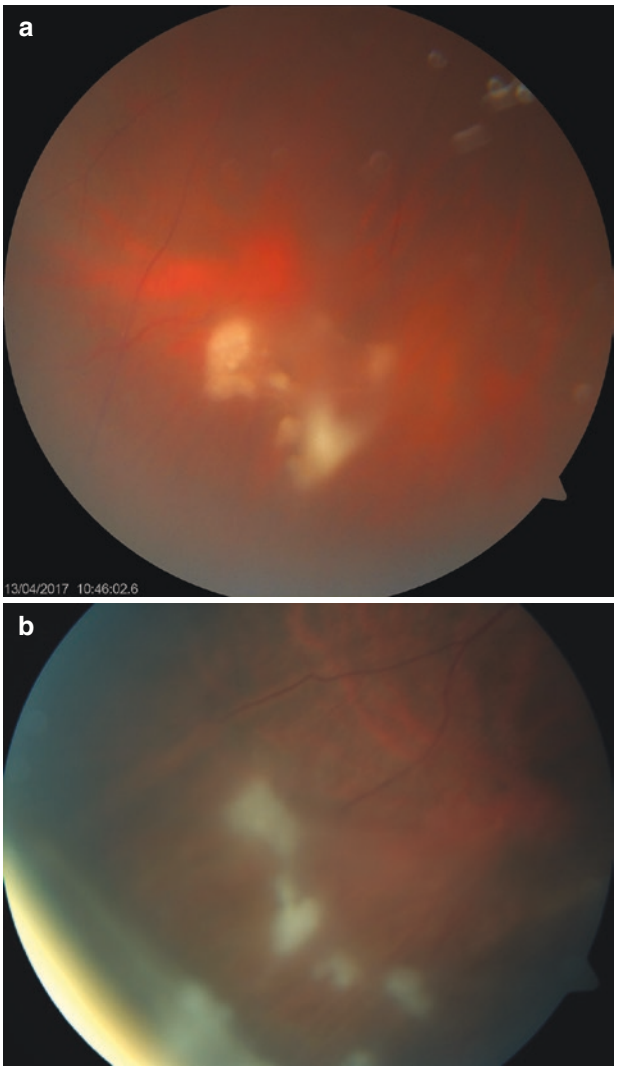


**Fig. 10.6** (**a**, **b**) Fundus photographs show snowballs coalescing and making a plaque located on the inferior retina

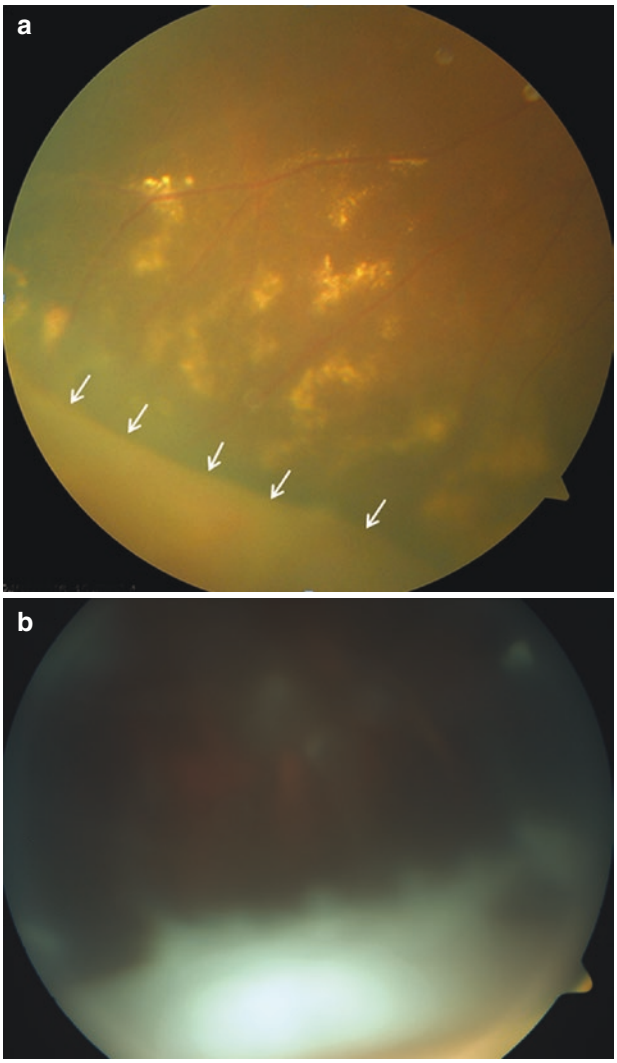


**Fig. 10.7** Fundus photograph demonstrates snowbank exudates with a distinct border located inferiorly (shown with white arrows, **a**) and with irregular border located inferiorly (**b**)

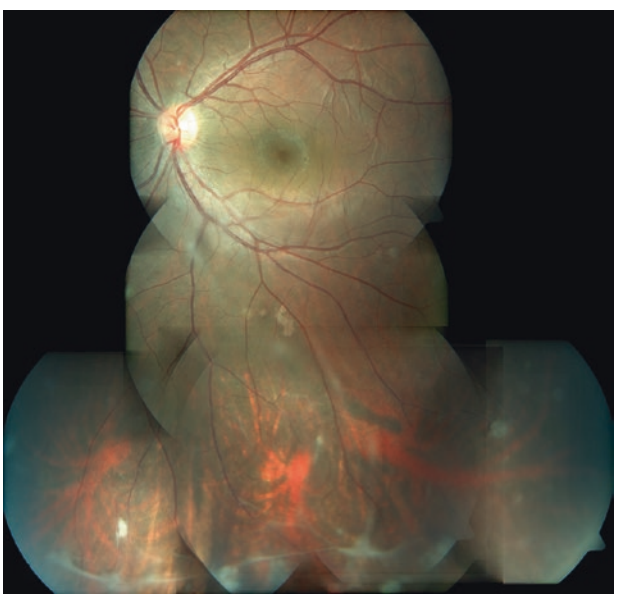

**Fig. 10.8** Fundus photograph shows vitreous bands, condensations, and snowballs at the inferior retina

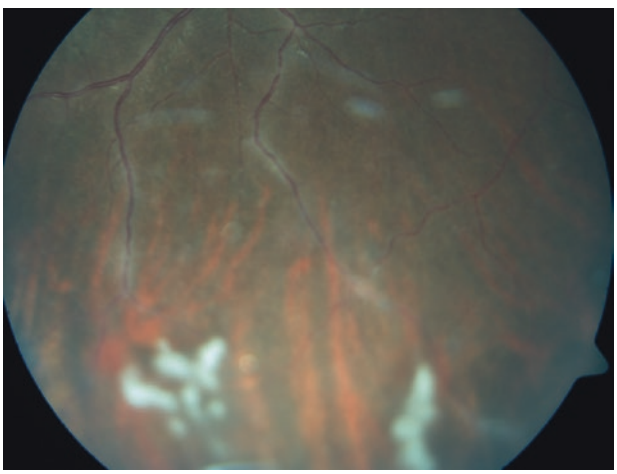

**Fig. 10.9** Fundus photograph shows snowballs and vascular sheathing at the inferior retina

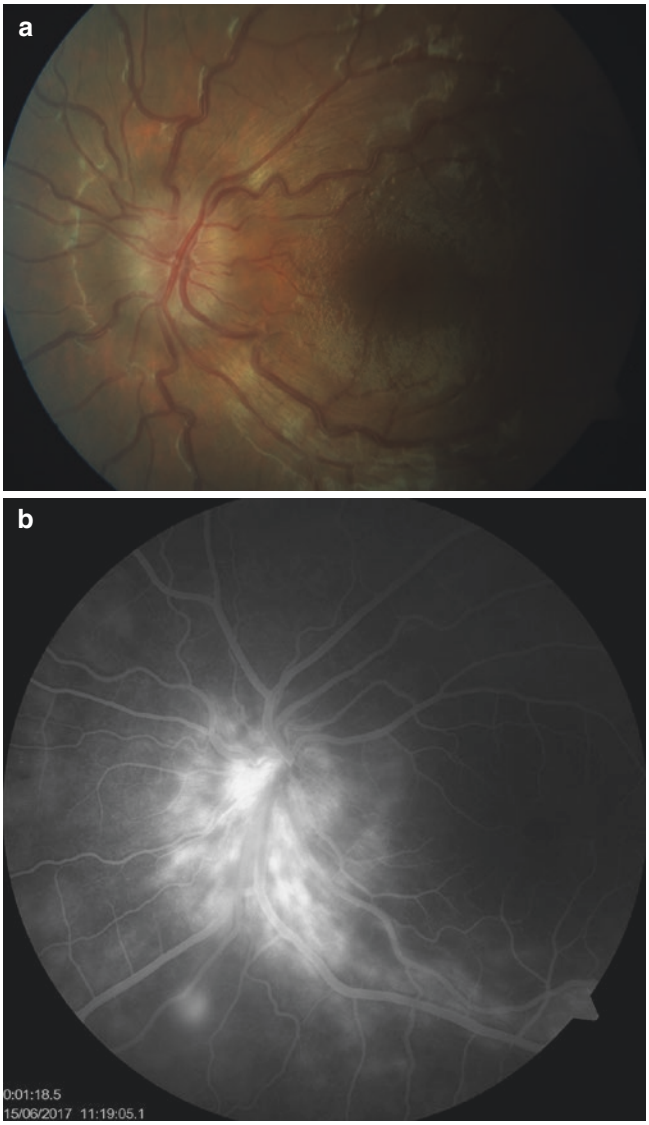


**Fig. 10.10** (**a**, **b**) Fundus photographs showing optic disc edema in patients with pars planitis

Peripheral retinal traction and retinal tear may occur occasionally (Fig. 10.17a, b). Retinal detachment (tractional, rhegmatogenous, or exudative) is also a rare complication of pars planitis (Donaldson et al. 2007; Paroli et al. 2011) (Fig. 10.18). Inferior peripheral retinoschisis, however, is a more frequent complication which occurs almost exclusively in children (Tugal-Tutkun 2011) (Fig. 10.19a–d). Retinoschisis has been reported in 19% of eyes in a recent study from a tertiary referral center. It has been found to be bilateral, inferior, and adjacent to a snowbank (Malalis et al. 2017). Young children with pars planitis are at high risk of amblyopia as a consequence of band keratopathy, vitreous opacities and cataracts obscuring the visual axis, or persistent macular edema. Delayed diagnosis and treatment may result in permanent visual loss.

## Diagnosis

The diagnosis of IU is based on clinical findings. Decreased and/or blurred vision, floaters in the absence of pain, redness, and photophobia are suggestive symptoms. Clinical diagnosis is based on the presence of vitreous cells, snowballs, and pars plana exudation. When ophthalmoscopy with scleral depression is not performed, inferior snowballs or snowbanks can be missed and especially pars planitis may be underdiagnosed. There is no specific diagnostic laboratory test for IU. For a diagnosis of an idiopathic IU or pars planitis, however, systemic associations and mainly infectious causes of IU need to be ruled out with a careful history, systemic evaluation, and laboratory tests.

Systemic investigations include complete blood count, serological tests for syphilis, Lyme and cat-scratch disease, serum angiotensin-converting enzyme (ACE) and lysozyme levels, chest x-ray, purified protein derivative skin test, and brain magnetic resonance imaging (MRI). Because of the significant association with MS, neuroimaging should be performed especially in adult patients, in whom systemic associations are more common. Imaging modalities such as FA, optical coherence tomography (OCT), ultrasound biomicroscopy (UBM), and ultrasonography are also helpful in confirming the diagnosis and/or showing the disease-related complications.

Fluorescein angiography is usually performed to assess the presence of CME, retinal vasculitis, neovascularizations, and retinal ischemia. Retinal vascular leakage, diffuse capillary leakage, and CME are common FA findings (Fig. 10.20a). Peripheral retinal ischemia, however, may be observed as a less frequent FA finding of the disease (Fig. 10.20b). Fluorescein angiography is also valuable in documenting the response to treatment, especially in eyes with CME and peripheral vasculitis. The OCT imaging shows disease-related macular and retinal changes and provides information regarding the reversibility of lesions. It is valuable in detecting macular edema and its sequelae such as cystoid changes, epiretinal membranes, macular hole, and atrophy. As in FA, it is beneficial in monitoring the treatment response. Ultrasonography and UBM are valuable methods providing additional information regarding the ciliary body, pars plana, and retina when visualization of the fundus is obscured due to band keratopathy, cataract, synechiae, vitreous inflammation, or hemorrhage (Babu and Rathinam 2010; Ozdal et al. 2015).

## Sarcoidosis

It has been reported that IU was associated with sarcoidosis in about 10% of patients (Ness et al. 2017). Cystoid macular edema, optic disc edema, and periphlebitis are typical

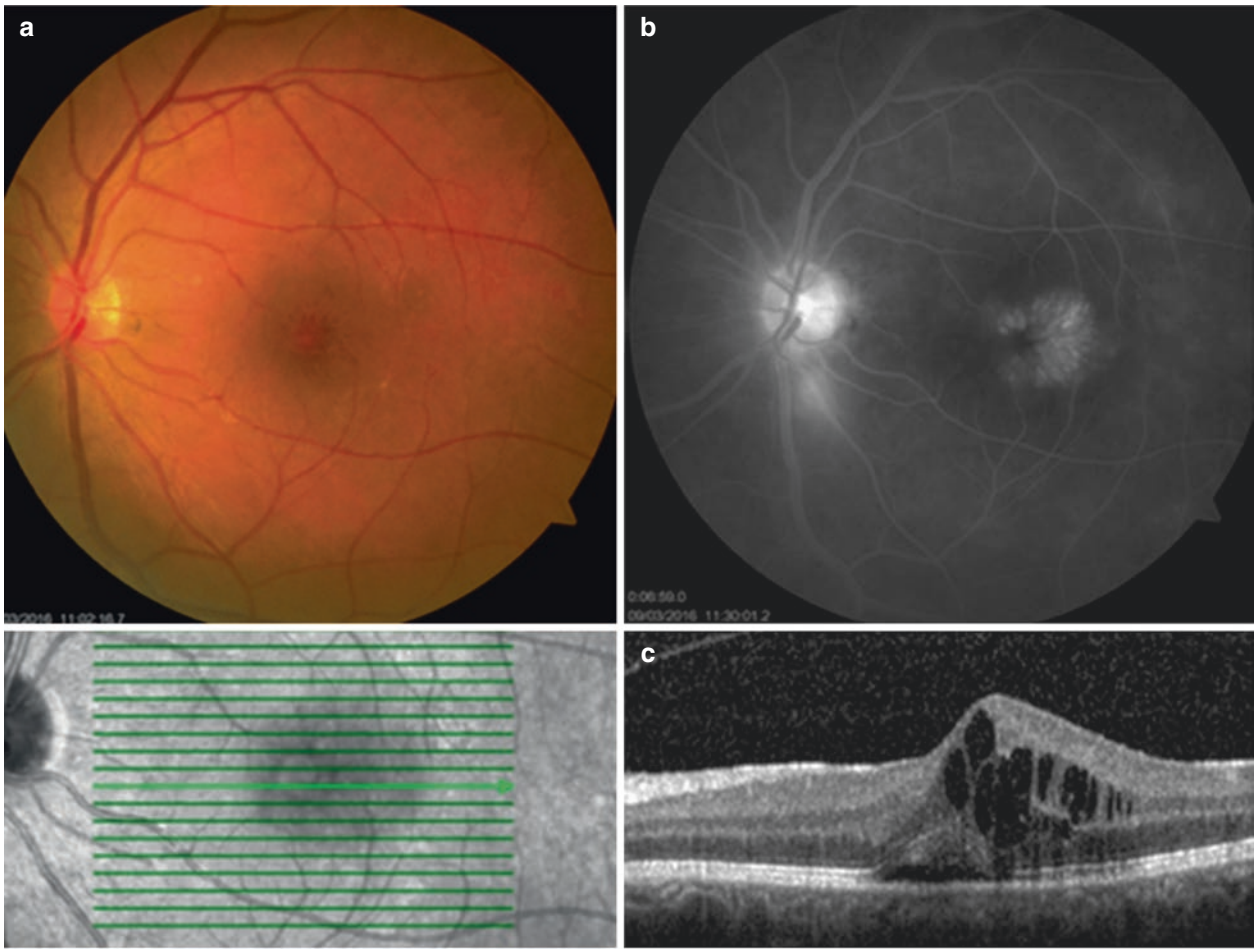


**Fig. 10.11** Fundus photograph (**a**), fluorescein angiography (**b**), and optical coherence tomography (**c**) of a patient with intermediate uveitis complicated with cystoid macular edema

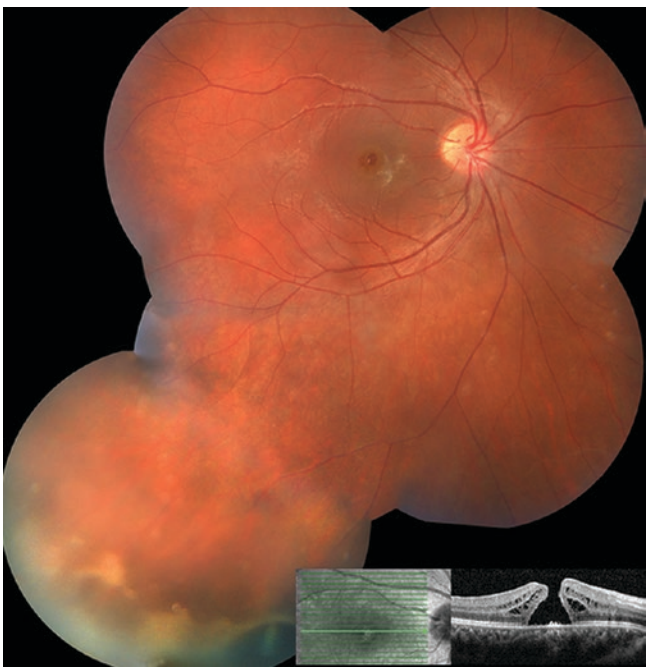

**Fig. 10.12** Fundus photograph and optical coherence tomography of a patient with pars planitis complicated with the development of a macular hole

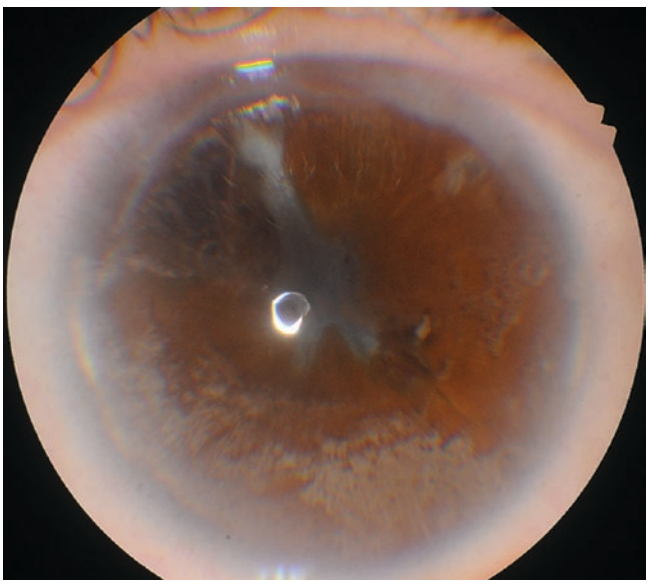

**Fig. 10.13** Slit lamp photograph shows band keratopathy, seclusio pupillae, and secondary cataract associated in the more severely affected eye of a 5-year-old girl with bilateral pars planitis

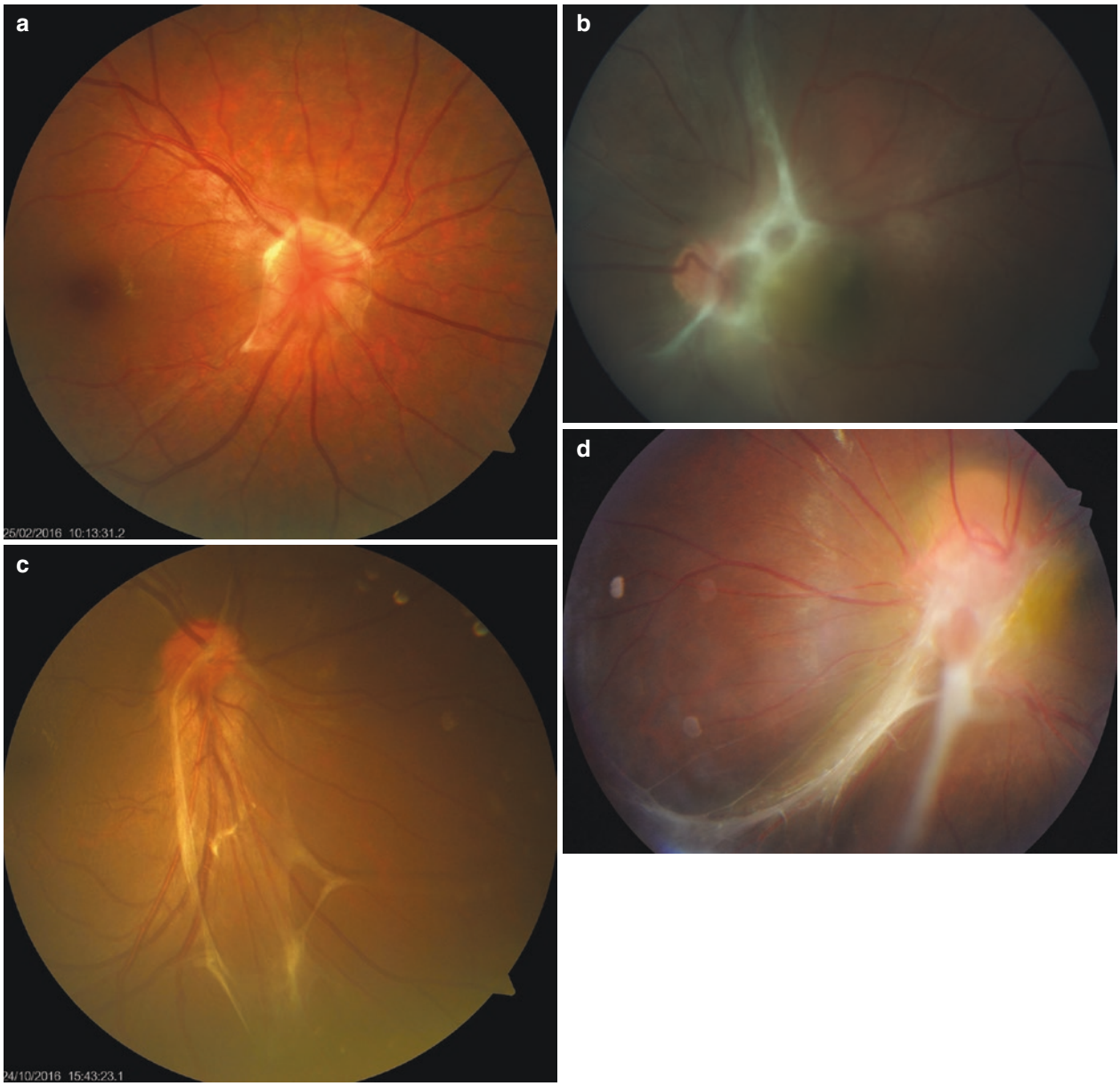


**Fig. 10.14** (**a**–**d**) Fundus photographs showing various epiretinal membrane in intermediate uveitis

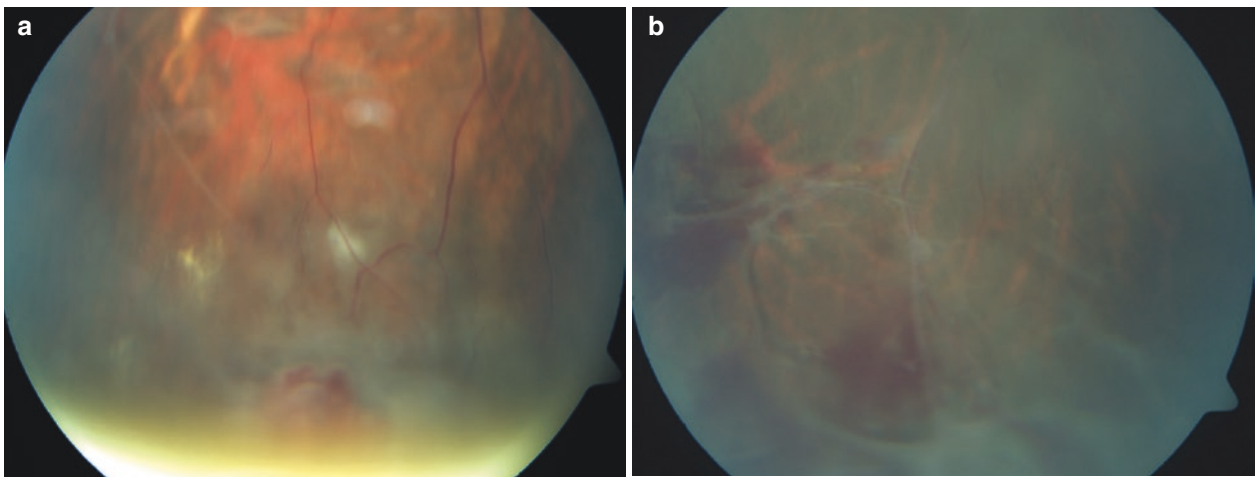


**Fig. 10.15** Fundus photographs showing exudates, periphlebitis, snowballs, and neovascularization at the inferior peripheral retina (**a**) and vitreous bands and retinal hemorrhages from the neovascularizations (**b**) in pars planitis

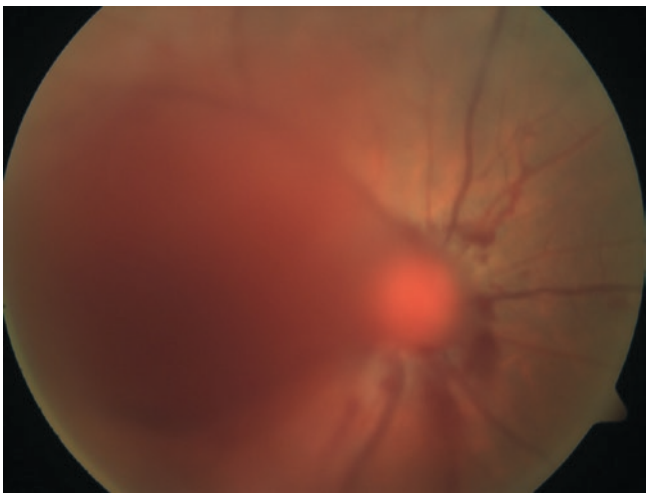

**Fig. 10.16** Fundus photograph shows vitreous hemorrhage from the optic disc neovascularization

findings of sarcoidosis-associated IU (Babu and Rathinam 2010) (Fig. 10.21a, b). Pulmonary manifestations such as hilar lymphadenopathy, serum ACE and lysozyme levels, gallium scan, chest computed tomography, biopsy from conjunctival nodules or skin granuloma, bronchoalveolar lavage, and transbronchial lung biopsy are helpful in diagnosing sarcoidosis (Ozdal et al. 2015).

## Multiple Sclerosis

About 20% of patients with IU have been shown to have MS. A strong association between pars planitis and MS has already been shown. Children with pars planitis may later develop MS during adolescence or in adulthood, and IU may be the first manifestation of MS (Fig. 10.22a, b). Thus, in the presence of clinical signs suggestive of MS such as prominent retinal periphlebitis with or without optic neuritis, a neurological evaluation, MRI of brain, and cerebrospinal fluid analysis should be performed (Ness et al. 2017; Raja et al. 1999). A granulomatous anterior uveitis accompanied by retinal periphlebitis is also highly suggestive of MS.

## Tuberculosis

*Mycobacterium tuberculosis* may induce an intermediate uveitis with nonspecific clinical presentation or with clinical findings of pars planitis. A low-grade chronic inflammation, vitritis, snowball opacities, peripheral vascular sheathing, snow banking, and peripheral retinochoroidal granuloma may be observed. To make the differential diagnosis, accurate history, chest imaging, tuberculin skin test, and interferon gamma release assays are all helpful especially in

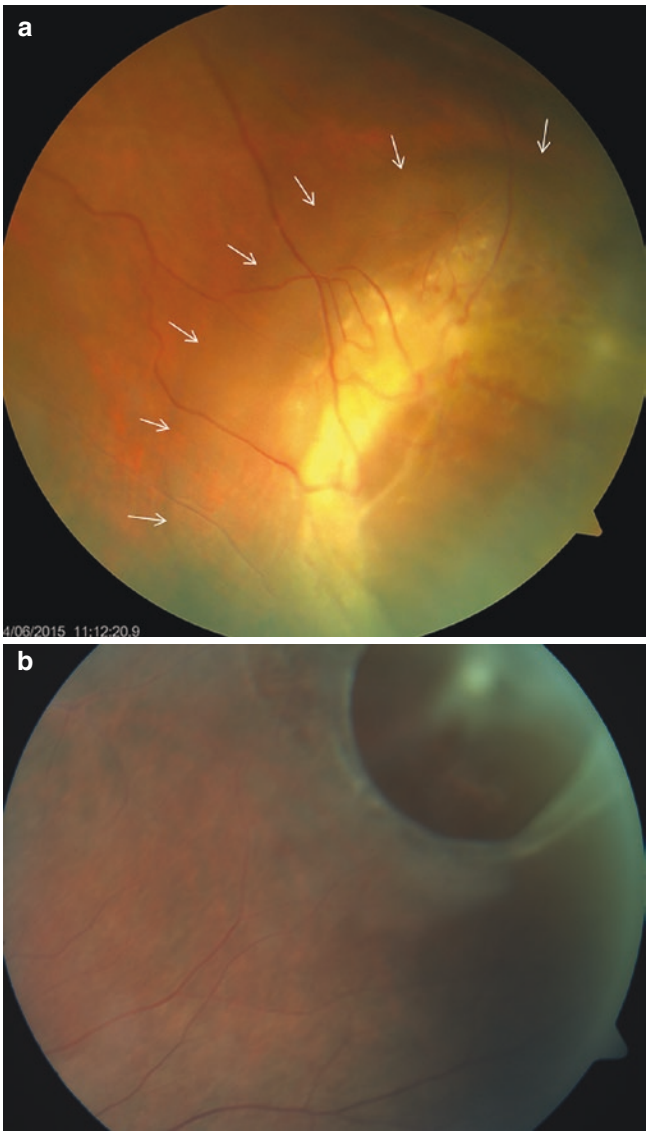


**Fig. 10.17** (**a**, **b**) Fundus photographs showing retinal tear due to peripheral retinal traction and minimal subretinal fluid around the tear

patients living in endemic areas (Babu and Rathinam 2010; Gupta et al. 2015).

## Syphilis

Syphilis may present with all anatomic types of uveitis. Anterior uveitis, both granulomatous and non-granulomatous, is the most common form of syphilitic uveitis. Intermediate uveitis has been observed in 10.3% of the cases (Anshu et al. 2008). Placoid retinitis, neuroretinitis, and retinal vasculitis are other common findings of syphilitic uveitis. The diagnosis of syphilis can be suggested by history and systemic and ocular examination and confirmed by serologic tests. Polymerase chain reaction (PCR) of ocular fluids may be performed when needed.

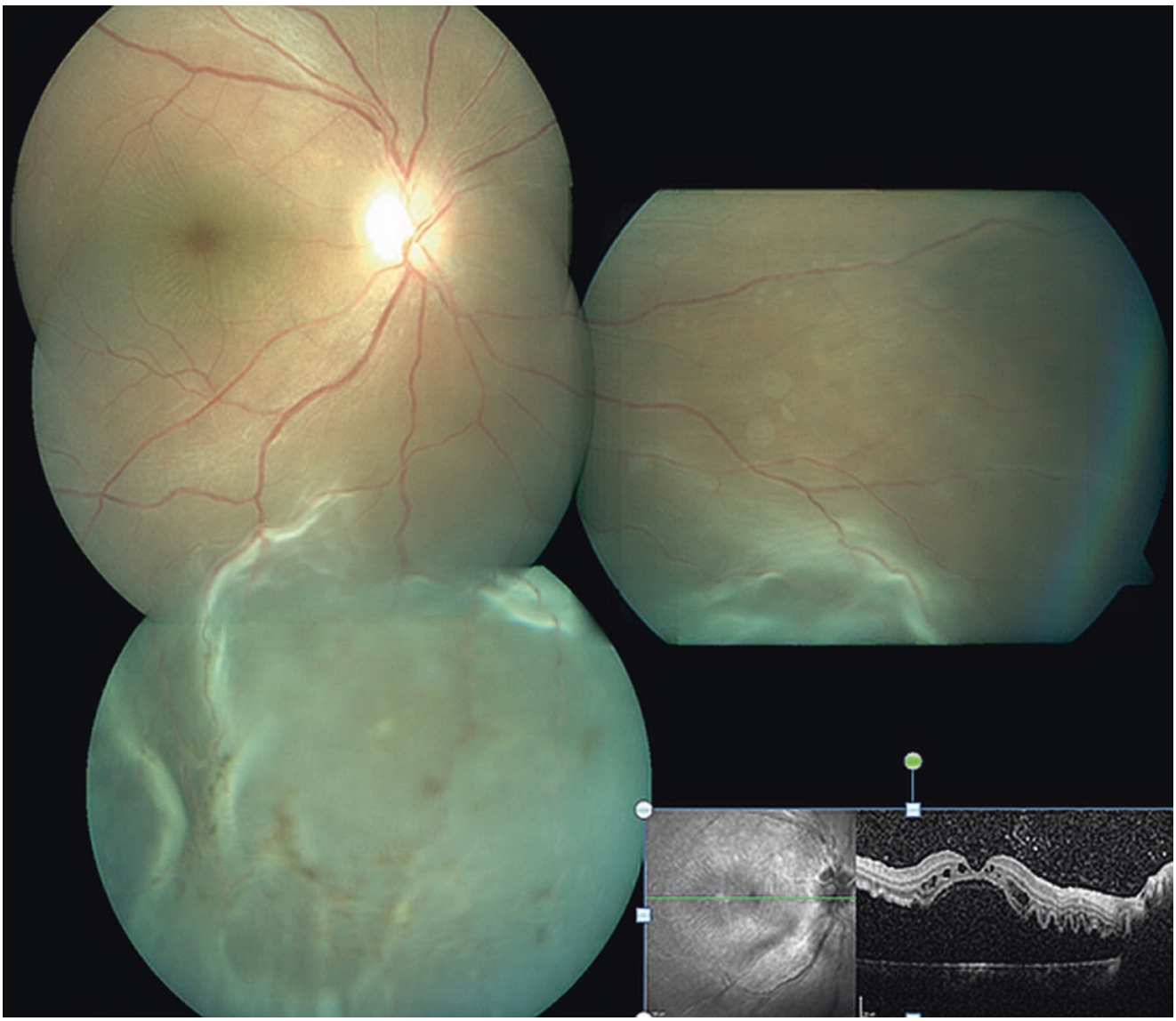

**Fig. 10.18** Fundus photograph shows inferior exudative retinal detachment involving the macula. Optical coherence tomography confirms the macular detachment

### Lyme Disease

Intermediate uveitis associated with Lyme disease caused by *Borrelia burgdorferi* has been reported in both adults and children (Babu and Rathinam 2010) (Fig. 10.23a–d). History of exposure to ticks, presence of rash and chronic arthritis, Lyme indirect immunofluorescence assay, and Lyme enzyme-linked immunosorbent assay are all helpful in the diagnosis (Whitcup 2010).

### Other Rare Causes of Intermediate Uveitis

Serologic tests for cat-scratch disease, a gastroenterologic evaluation for inflammatory bowel disease in patients with a history of chronic or bloody diarrhea, should also be considered (Whitcup 2010). Ocular toxocariasis may present as unilateral IU and peripheral toxocara granuloma may sometimes be difficult to distinguish from snowbank of idiopathic pars planitis. Serology and UBM are valuable for the diagnosis of ocular toxocariasis (Ozdal et al. 2015).

## Differential Diagnosis

### In Children

As chronic anterior uveitis which is idiopathic or associated with juvenile idiopathic arthritis (JIA) has complications similar to pars planitis such as band keratopathy, posterior

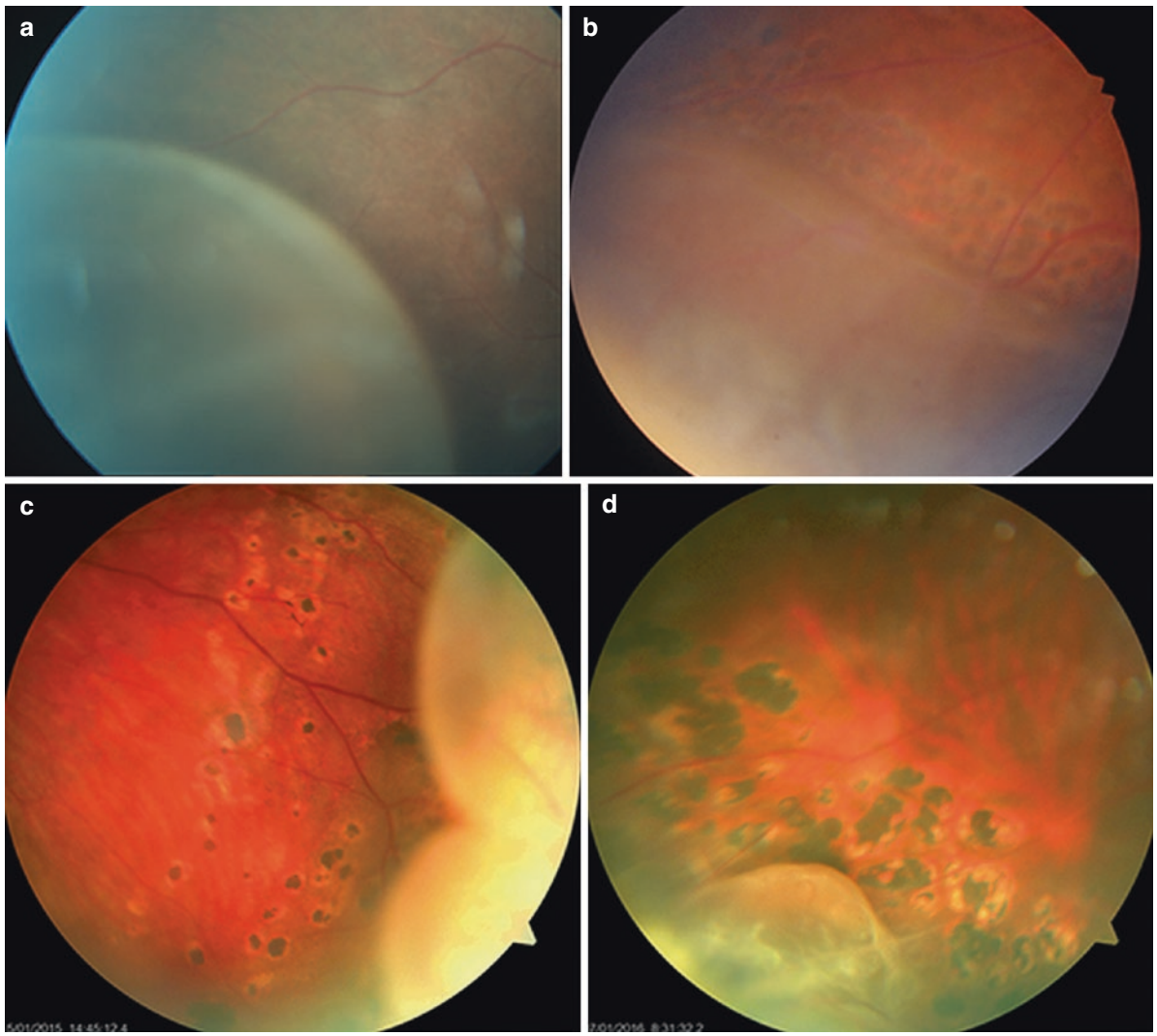


**Fig. 10.19** (**a**–**d**) Fundus photographs show peripheral retinoschisis in different patients with pars planitis

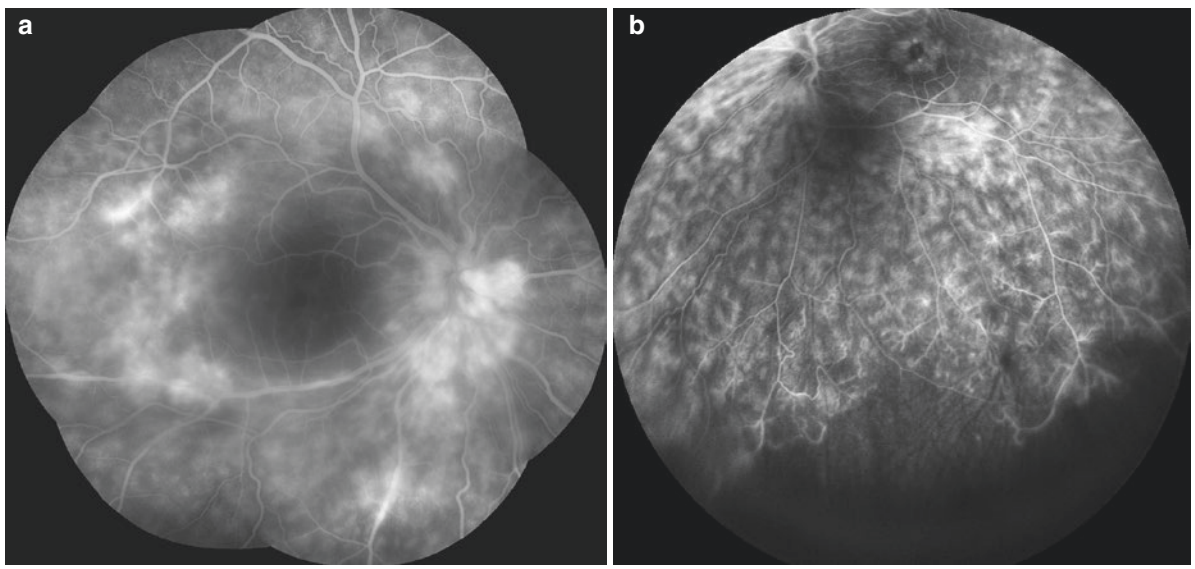


**Fig. 10.20** Fluorescein angiography shows leakage from the veins, capillaries, and optic disc (**a**) and diffuse capillary and peripheral vascular leakage, macular edema, and inferior peripheral retinal ischemia (**b**) in pars planitis patients

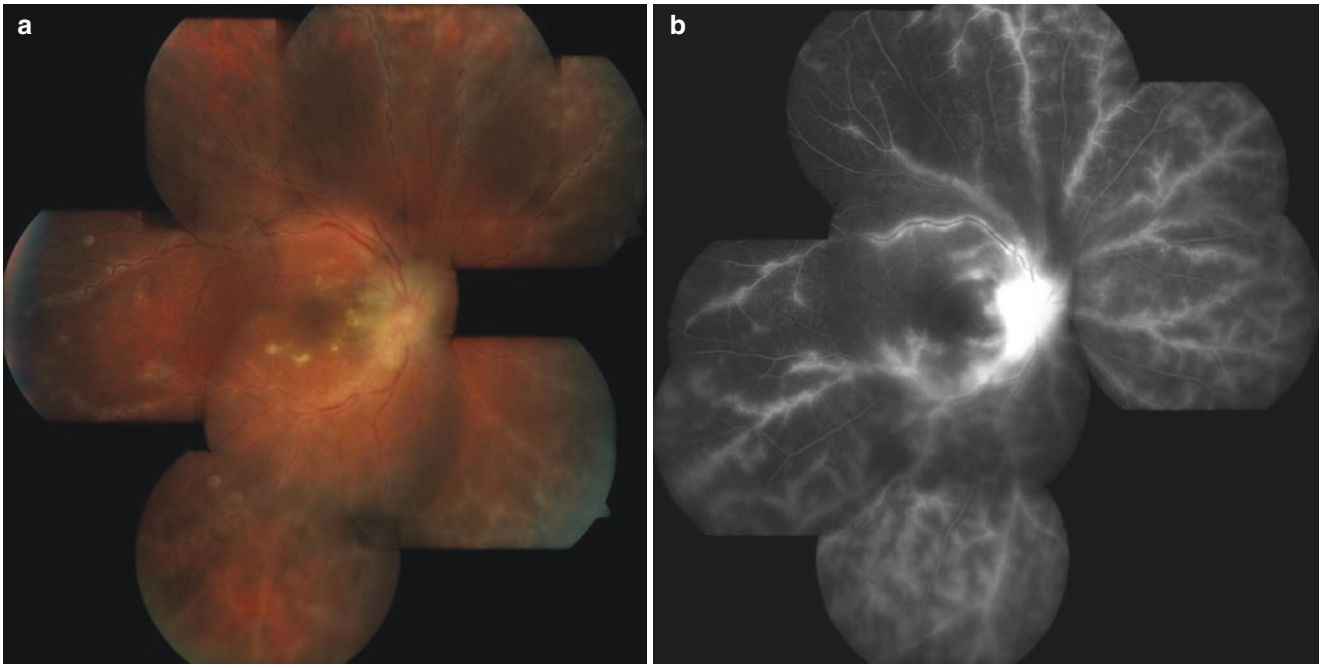


**Fig. 10.21** Fundus photograph shows severe periphlebitis, snowballs, and optic disc edema (**a**) and diffuse vascular and optic disc leakage (**b**) in a patient with sarcoidosis

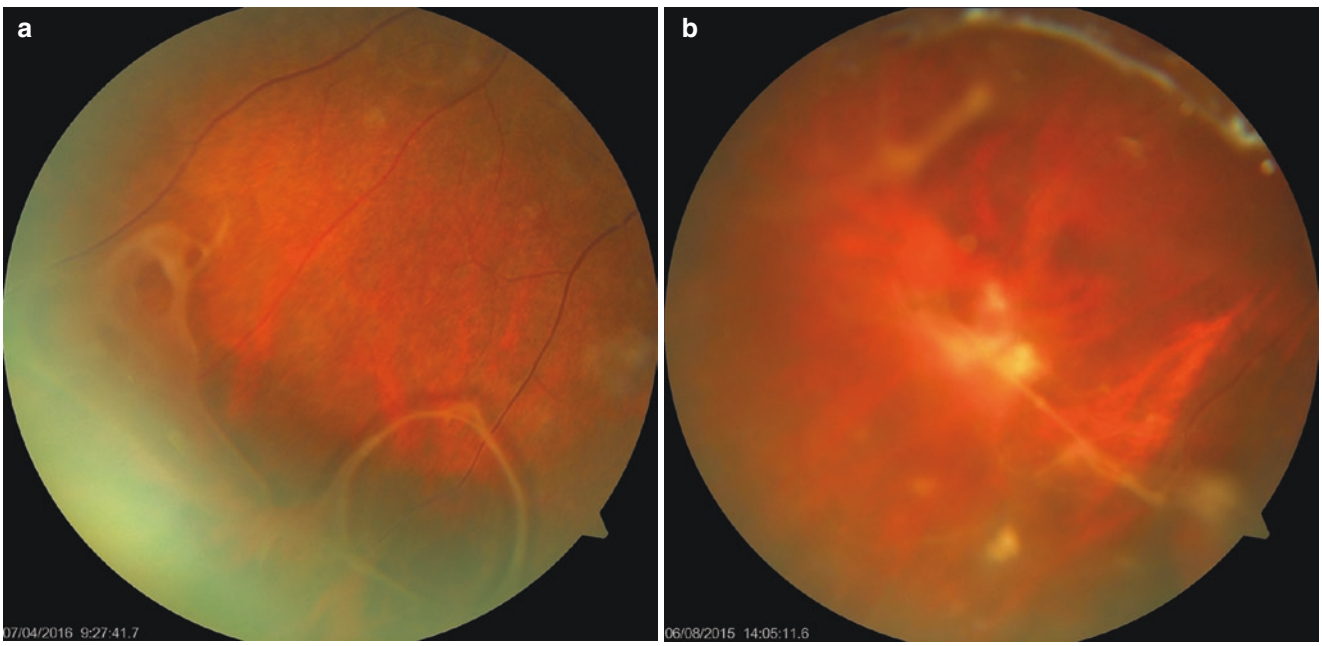


**Fig. 10.22** Fundus photographs showing peripheral vitreous bands (**a**) and snowballs (**b**) in patients with multiple sclerosis-associated intermediate uveitis

synechiae, and cataract, these two entities should be differentiated. An attentive posterior segment evaluation is crucial in making the differentiation. Although rare, masquerades should also be considered in differential diagnosis of children presenting with pars planitis. Retinoblastoma may present as cellular reaction or white deposits in the anterior chamber and vitreous infiltrates. Diagnostic fine-needle aspiration biopsies are needed in such unusual cases (Ozdal et al. 2015).

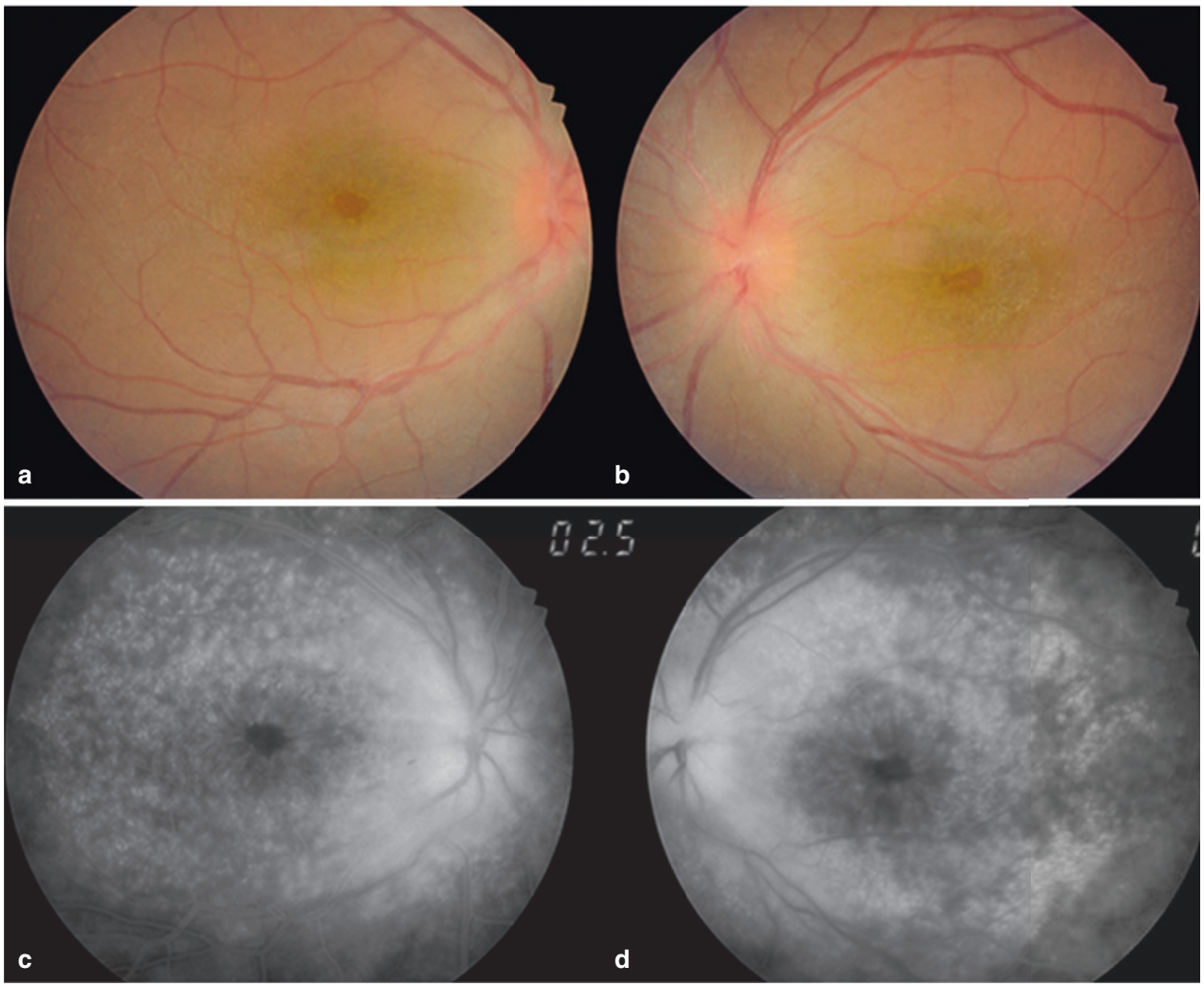


**Fig. 10.23** Fundus photographs (**a**, **b**) show retinal vasculitis, cystoid macular edema, and optic disc edema, and fluorescein angiography (**c**, **d**) shows diffuse vascular leakage, optic disc, and macular edema in a patient with Lyme disease

### In Adults

Inferior pearl-like precipitates occurring with the resolution of acute inflammatory attack in Behçet uveitis may lead to a misdiagnosis of pars planitis. Recurrent ocular inflammatory attacks, acute onset and spontaneous resolution of inflammation, appearance of inferior peripheral precipitates during resolution of vitreous haze, and absence of snowballs, snowbanks, or persistent vitreous condensates are typical characteristics of Behçet uveitis differentiating it from pars planitis. Pearl-like precipitates seen in Behçet uveitis are located on the surface of the retina and are small, uniform, and immobile (Fig. 10.24a, b), whereas snowball opacities are round, white collections located in the vitreous and are mobile (Tugal-Tutkun et al. 2013). Because of prominent vitreous infiltration and condensations, Fuchs' uveitis syndrome (FUS) should also be considered in the differential diagnosis of IU. Clinical findings of FUS including unilaterality, diffusely distributed small, round or stellate KPs, iris atrophy with or without heterochromia, and the absence of macular edema are helpful in making the differential diagnosis (Tugal-Tutkun et al. 2009) (Fig. 10.25a, b). In the elderly, primary intraocular lymphoma may present with diffuse vitreous infiltration, which may mimic intermediate uveitis. Not only severe vitreous inflammation but also poor or partial response to therapy is highly suggestive of primary intraocular lymphoma. As it is usually associated with primary central nervous system lymphoma, a brain MRI, cerebrospinal fluid analysis, and a careful neurologic history may provide useful information. However, cytological evaluation of vitreous samples, identification of cell surface markers by immunohistochemistry, cytokine analysis, retinal biopsy, and gene rearrangement are required for a definitive diagnosis of intraocular lymphoma. An elevated IL-10/IL-6 ratio in aqueous humor or vitreous is highly suggestive of intraocular lymphoma (Davis 2004).

## Management

Exclusion of infectious and noninfectious associations which may present with intermediate uveitis is the most important step before starting a treatment. Infectious causes need a specific anti-infectious treatment. The treatment of pars planitis, how-

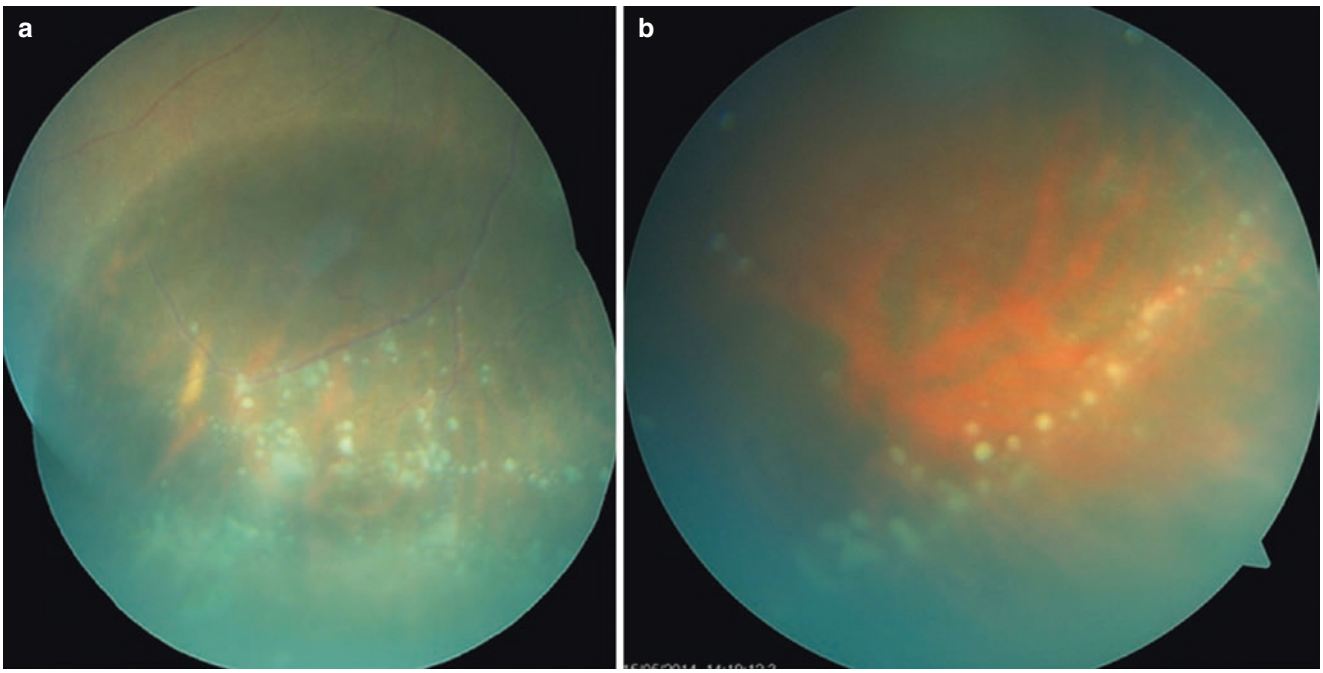


**Fig. 10.24** (**a**, **b**) Fundus photographs show inferior pearl-like precipitates occurring with the resolution of acute inflammatory attack in Behçet uveitis in two different patients

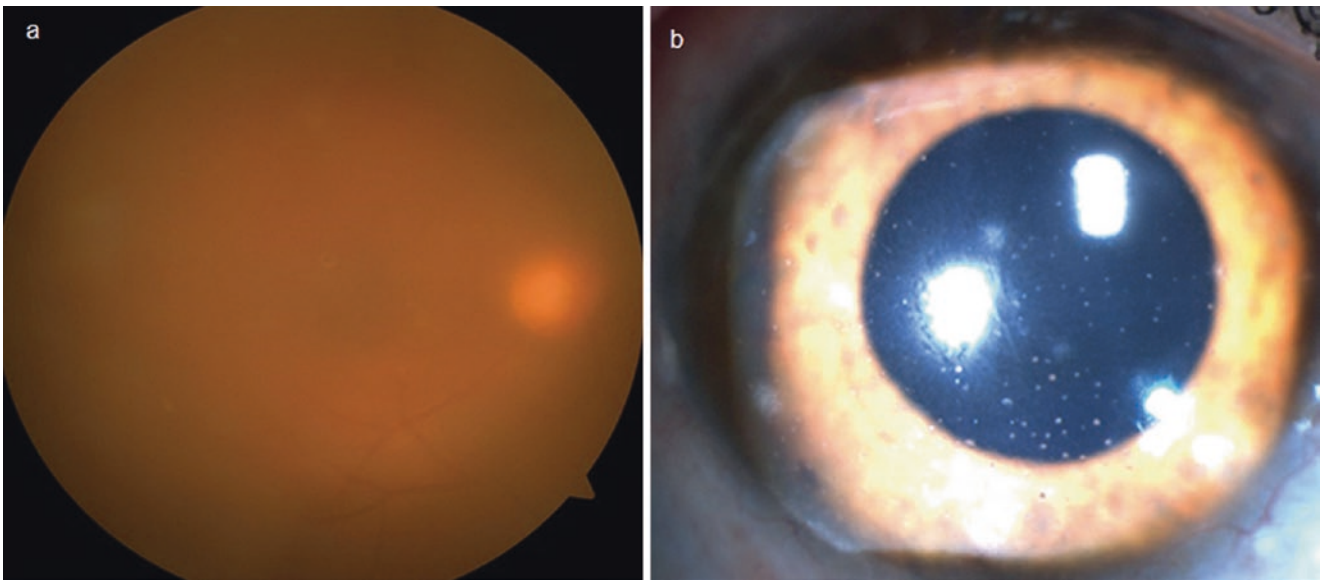


**Fig. 10.25** (**a**, **b**) Fundus photograph shows severe vitreous haze in a patient with Fuchs uveitis, and the slit lamp photograph demonstrates diffusely distributed small, round keratic precipitates as a distinguishing feature

ever, is still a controversial issue. There is no consensus especially for cases with minimal inflammation and relatively good visual acuity. Pars planitis may present as a severe disease leading to several ocular complications and deserve an aggressive treatment. The presence of macular edema, vitreous haze leading to a decrease in visual acuity, complications such as band keratopathy, cataract or retinoschisis in at least one eye, vasculitis, and a severe infiltration of the pars plana are indications for treatment irrespective of the level of visual acuity. A stepladder approach is used while treating patients with pars planitis.

## Medical Therapy

***Corticosteroids:*** The first step of medical therapy includes corticosteroids (CS) which are still the mainstay of treatment. Topical CS are used only if there is anterior segment inflammation. Periocular CS injections are beneficial particularly in patients with unilateral or asymmetrical involvement and in the presence of macular edema. Posterior subtenon injection of 40 mg triamcinolone acetonide is the most frequently used method for employing periocular CS. Intravitreal CS injections

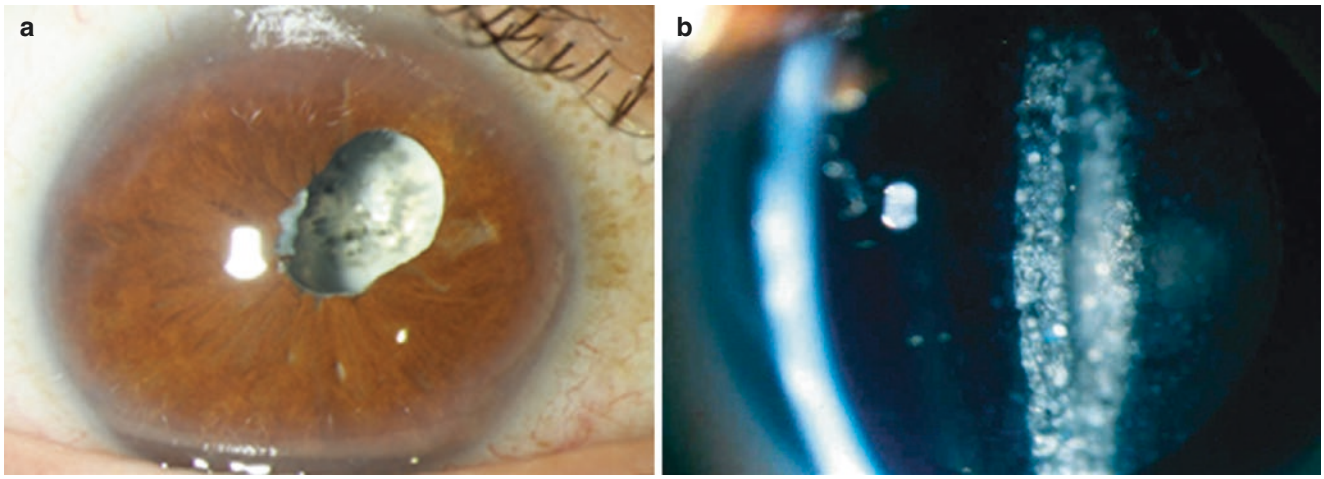


**Fig. 10.26** Slit lamp photographs demonstrate severe posterior capsule opacification (**a**) and dense precipitates on the anterior and posterior surface of the intraocular lens forming a cocoon membrane (**b**) due to inadequate control of inflammation following cataract surgery

have also been found effective in treating intermediate uveitis and associated macular edema. The most common complications of periocular CS are increased intraocular pressure, cataract, and aponeurotic ptosis, and the most common complications of intravitreal CS injections are cataract, increased intraocular pressure, and glaucoma, while rare complications include vitreous hemorrhage, retinal detachment, and endophthalmitis. Because of these high complication rates, intravitreal CS injections should be used as an emergency procedure to save the macula immediately and allow time to organize the long-term management (Ozdal et al. 2015). Intravitreal dexamethasone implant has been shown to be effective and safe particularly in persistent chronic CME and vitritis due to noninfectious intermediate uveitis (Lightman et al. 2013; Palla et al. 2015). In patients with bilateral involvement, severe ocular inflammation or unilateral disease unresponsive to periocular, systemic CS treatment should be considered. A dose of 1–1.5 mg/kg/day of prednisone tapered according to clinical response is preferred by most uveitis specialists. Intravenous pulse methylprednisolone therapy (1 g/day for adults, 30 mg/kg/day for children) may be administered when more rapid and potent action is needed.

***Immunosuppressive agents:*** Steroid-sparing immunosuppressive therapy should be considered as a second step in patients who require long-term treatment. Methotrexate, mycophenolate mofetil, azathioprine, and cyclosporine may be used alone or in combination. Because of its long-term safety profile and well tolerance, methotrexate is the most widely used first-line immunosuppressive agent in children. It is of importance to remember that these agents need 4–8 weeks to become effective and CS should be given concomitantly until the immunosuppressive agent is expected to take action. In patients who present with serious ocular complications, immunosuppressive agents and CS combination may be started as the first step.

***Biologic agents:*** In patients not responding to conventional immunosuppressive agents, anti-tumor necrosis factor-α (Anti-TNF-α) agents should be used as the third step of medical therapy. The use of these agents in refractory ocular inflammation including patients with intermediate uveitis suggests promising efficacy (Ozdal et al. 2015). As pars planitis is associated with an increased risk for MS development and anti-TNF-α agents may potentiate demyelinating disease, extreme caution is needed before starting such therapy in patients with pars planitis (Kump et al. 2013). Interferon-beta has also been shown to be effective in improving macular edema and vitreous haze due to intermediate uveitis (Mackensen et al. 2013).

## Surgical Therapy

Pars plana vitrectomy comprises the fourth step of therapy. It should be considered particularly in patients developing complications such as vitreous condensation, vitreous hemorrhage, retinal detachment, and epiretinal membranes causing retinal traction. Surgical therapy has also been shown to be effective in patients with active inflammation and CME refractory to medical treatment (Stavrou et al. 2001). Pars plana vitrectomy provides the mechanical clearance of inflammatory mediators and debris, anatomical correction of retinal pathology such as vitreoretinal traction, opportunity to obtain vitreous samples and reduction of postoperative anti-inflammatory medication (Babu and Rathinam 2010; Stavrou et al. 2001). Cataract surgery may be safe when an adequate preoperative and postoperative inflammation control and a meticulous surgical technique are employed. If not, severe postoperative complications are inevitably encountered (Fig. 10.26a, b).

## Adjunctive Therapies

Cryotherapy may aggravate blood–ocular barrier disruption and accelerate the rate of retinal detachment in predisposed eyes by inducing vitreous contraction (Kump et al. 2013). Compared to cryotherapy, laser photocoagulation is an easier and safer method with fewer ocular complications. However, it should not be considered as a treatment step alone but may be employed as an adjunctive treatment modality especially in cases associated with peripheral neovascularization, retinal traction, or retinoschisis.

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