

Birdshot retinochoroidopathy

What is birdshot retinochoroidopathy?^[1] ^[2]

Birdshot retinochoroidopathy is an uncommon, but well-characterised chronic, bilateral posterior uveitis, which is strongly associated with the human leukocyte antigen (HLA) A29 phenotype (up to 96% of cases). Birdshot retinochoroidopathy affects the retina and the choroid layer of the eye.

There are progressive abnormalities which develop in the small choroidal vessels, resulting in leakages and, eventually, atrophic spots. There is an associated low-grade uveitis which can eventually lead to permanent loss of vision.

The antigen trigger is unknown and the exact role of HLA-A29 in the pathogenesis of the disease is not well-understood. It has been suggested that an autoinflammatory response results from presumed molecular mimicry initiated either by prior microbial infection or other damaging factors and that HLA-A29 cross-reactive proliferative responses directed against retinal antigens could lead to loss of immunological tolerance and autoimmunity.

How common is birdshot retinochoroidopathy? (Epidemiology)^[1]

Birdshot retinochoroidopathy is a disease seen almost exclusively in Caucasians with a mean onset age of 53 years and a slight female preponderance. Little data exists on the true national or global prevalence of the disease but it accounts for between 0.6% and 1.5% of cases referred to specialty uveitis centres, or 6-7% of those with posterior uveitis,

Symptoms of birdshot retinochoroidopathy (presentation)^[1] ^[2]

Birdshot retinochoroidopathy presents predominantly in middle-aged Caucasian females who complain of blurred vision, floaters, photopsias, paracentral scotomas and night-blindness.

In the initial stages, visual acuity reduction may be relatively mild (of the order of 6/9–6/12). This gets progressively worse throughout the disease. There is also often some degree of [visual field loss](#).^[3]

Fundoscopy: the vitreous (jelly filling the greater part of the globe, lying behind the lens, through to the retina), reveals floating cells (seen as little pale specks). There are distinctive lesions on the retina, which are well-defined (initially) pale yellow/white spots scattered around the retina, usually much smaller than the disc size. They look somewhat like an explosion of spots scattered over the retina. Over the years, the lesions become ill-defined and confluent. There may be associated [macular oedema](#) and intraretinal haemorrhages.

Differential diagnosis^[2]

Other types of idiopathic multifocal white dot syndromes:

- Acute multifocal posterior placoid pigment epitheliopathy (AMPPPE).
- Serpiginous choroidopathy.
- [Multifocal choroiditis](#) with [panuveitis](#).
- Multiple evanescent white dot syndrome.
- Acute retinal pigment epitheliitis.

Other causes of posterior uveitis including:

- Various causes of [choroiditis](#) (such as toxoplasmosis).
- [Sarcoidosis](#).
- [Syphilis](#).
- Ocular histoplasmosis.

- Postoperative [endophthalmitis](#).
- Problems relating to immunocompromise (eg, cytomegalovirus (CMV) retinitis, [candidal infection](#), herpetic retinitis and so on).
- Retinal vasculitic diseases.

Diagnosing birdshot retinochoroidopathy (investigations) ^[1] ^[2]

The diagnosis is essentially clinical, with eye examination revealing the characteristic fundus picture. The absence of significant anterior inflammatory sequelae (synechiae), the presence of vitritis and/or cystoid macular oedema without pars plana exudation, and HLA-A29 positivity all serve to support the diagnosis.

Investigations such as angiography, ultra-high resolution optical coherence tomography, electroretinogram (measures electrical responses by the various cells of the retina) may be used. If there are real uncertainties over the diagnosis, a biopsy may be performed.

Associated diseases ^[4]

The patient is usually otherwise healthy. There have been studies suggesting links to [glaucoma](#) and rhegmatogenous [retinal detachment](#) but these tend to be weak and findings are probably spurious. Furthermore, vascular disease, sarcoidosis, psoriasis, autoimmune sensorineural hearing loss and [vitiligo](#) have been reported in these patients but, again, a clear association is not currently evident.

Management of birdshot retinochoroidopathy ^[2] ^[4]

If birdshot retinochoroidopathy (or any of the differentials outlined above) is suspected in primary care, a referral to the local ophthalmology department is mandatory. The urgency depends on the symptoms but it is best to discuss it with the ophthalmology team.

- Systemic corticosteroids are commonly used but their efficacy is limited for long-term control of the disease at a low dose, and their side effects prevent long-term use at a high dose. They are usually associated with an immunosuppressive or biological drug, allowing the tapering of oral corticosteroid to a level which is safe for long-term use (below 7.5 mg/day).
- Given the guarded visual prognosis and uncertain natural history, the early introduction of steroid-sparing systemic immunotherapy has been advocated as extended treatment is anticipated in most patients. Studies have demonstrated that both preservation of visual function with a reduction of inflammation, macular oedema, and preservation of global retinal integrity as well as the induction of long-term remission is possible in patients treated in this way.^[1]
- Treatment will be guided by the severity of the disease, the location of the lesions and the presence of any complicating factors.

Complications of birdshot retinochoroidopathy

- [Macular oedema](#) is the most common complication.
- Subretinal choroidal neovascularisation (this increases risks of vessel leaks leading to oedema and bleeds). Treatment is with laser photocoagulation.

Prognosis^[1]

- Birdshot retinochoroidopathy is a chronic progressive disease with the potential for significant visual impairment due to both structural complications and diffuse retinal dysfunction with the duration of disease being a statistically significant risk factor for both.
- In some eyes, central visual acuity may be preserved until late in the disease course with significant irreversible peripheral retinal damage.
- Reported incidence rates for the development of vision loss to 20/50 or worse and to 20/200 or worse are 13% and 4% per eye respectively.
- Cystoid macular oedema, with central visual loss, occurs at 10% per eye.

- The use of long-term immunomodulatory therapy results in an 83% reduction in cystoid macular oedema, may improve and/or stabilise visual function, and in some cases, produce long-term remission.

Prevention of birdshot retinochoroidopathy^[4]

Birdshot retinochoroidopathy cannot be prevented. Once the diagnosis is made, preventative measures revolve around early identification and treatment of vision-threatening complications. All patients complaining of floaters will be followed up, even if central visual acuity is stable.

Further reading

- [Freitas-Neto CA, Boonsoon S, Dhanireddy S, et al](#); Birdshot retinochoroidopathy review. *Arq Bras Oftalmol.* 2015 Jan-Feb;78(1):56-61. doi: 10.5935/0004-2749.20150016.
- [Bergstrom R, Czyz CN](#); Birdshot Retinopathy. *StatPearls*, Jan 2023.

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