



# SYMPATHETIC OPHTHALMIA



**Eye Learn**  
All about the Eye

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1. A 30-year-old female got up at night with severe pain and watering in left eye. She gave history of a nail injury to her eye 1 year back. How would you approach and manage such a case? 10 J2016
2. Sympathetic Ophthalmitis (2000)
3. Immunological Considerations in Sympathetic Ophthalmitis. (2007)

### Introduction

- Sympathetic ophthalmia (SO) is a rare, bilateral, diffuse, granulomatous, non-necrotizing panuveitis that may develop after either surgical or accidental trauma to 1 eye (called the exciting eye), followed by a latent period and the appearance of uveitis in the uninjured fellow eye (the sympathizing eye).

### Epidemiology

- Earlier estimates of the incidence of SO ranged from up to 0.5% in eyes with nonsurgical trauma and 10/100,000 cases after intraocular surgery.
- Until recently, accidental penetrating ocular trauma was the most common precipitating event for SO.
- Ocular surgery—particularly vitreoretinal surgery—has now emerged as the main risk for the development of SO.
- In the early 1980s, the prevalence of SO in patients who had undergone pars plana vitrectomy was reported to be 0.01%, increasing to 0.06% when the procedure was performed in the context of other penetrating ocular injuries.
- Improved access to emergency surgical care following penetrating ocular trauma and improved microsurgical technique have undoubtedly influenced this etiologic shift from penetrating injury to surgical trauma.
- Shows no sex predominance and a lower risk in children (resulting in part from a reduced incidence of pediatric ocular injuries) as well as an increased risk in older patients (likely stemming from an increased frequency of ocular surgery and retinal detachment in this population).
- SO has traditionally been reported to develop in 80% of patients within 3 months of injury and in 90% within 1 year

### Etiopathogenesis

- The precise etiology of SO is unknown.
- In majority of patients, there is a history of surgery or penetrating ocular injury complicated by incarceration of uveal tissue.
- The disorder may result from altered T-lymphocyte responses to previously sequestered ocular self-antigens and antigens derived from the RPE or choroid.
- The penetrating wound itself may facilitate exposure of uveoretinal antigens to conjunctival lymphatic channels and thereby initiate this immunopathologic response.
- There may be a genetic predisposition to the development of the disease, as patients with SO are more likely to express HLA-DR4, -DRw53, and -DQw3 haplotypes.
- It should be noted that the immunogenetics of SO and VKH syndrome are virtually identical, as the same associations have been found in both diseases.

### Clinical features

1. Patients with SO typically present with asymmetric bilateral panuveitis, in which the exciting eye exhibits more severe inflammation than the sympathizing eye, at least initially.
2. Signs and symptoms in the sympathizing eye vary in their severity and onset; they range from minimal problems in near vision, mild photophobia, and slight redness to severe granulomatous anterior uveitis.
3. Both eyes may show mutton-fat KPs, thickening of the iris from lymphocytic infiltration, posterior synechiae formation
4. Elevated IOP due to trabeculitis or hypotony as a result of ciliary body shutdown.
5. Posterior segment findings include moderate to severe vitritis with characteristic yellowish white, mid-equatorial choroidal lesions (so-called Dalen Fuchs nodules) that may become confluent.
6. Peripapillary choroidal lesions and exudative retinal detachment may also develop (Fig 6-59) . Structural
7. Traumatic etiology, the presence of active intraocular inflammation, and exudative retinal detachment correlated with poorer vision in the sympathizing eye.
8. Extraocular findings similar to those observed with VKH syndrome, including cerebral spinal fluid pleocytosis, sensory neural hearing disturbance, alopecia, poliosis, and vitiligo, may be noted, although they are uncommon.



## Complications

Cataract	Chronic CME	Optic atrophy	Peripapillary and macular CNV
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## Investigations

- The diagnosis of SO is clinical, and the disorder should be suspected in the presence of bilateral uveitis following any ocular trauma or surgery.

### A. FA

1. During the acute stage of the disease, FA reveals multiple hyperfluorescent sites of leakage at the level of the RPE during the venous phase, which persists into the late stage of the study.
2. Pooling of dye is observed beneath areas of exudative neurosensory retinal detachment.
3. Less-common fluorescein angiographic patterns are determined by the status of the overlying RPE.
4. Multiple chorioretinal lesions may be present and may appear hypofluorescent early in the study, simulating the pattern seen in APMPE, or hyperfluorescent with late staining.
5. These lesions are commonly referred to clinically as “Dalen-Fuchs nodules,” even though that term refers to the histopathologic finding of clusters of epithelioid cells located between the RPE and the Bruch membrane.

### B. ICGA

- ICG angiography reveals numerous hypofluorescent foci, which are best visualized during the intermediate phase of the angiogram; some of these foci may become isofluorescent in the late stage of the study

### C. OCT

- OCT imaging may demonstrate a shallow, serous retinal detachment and/or intraretinal edema, and monitoring these findings can help determine the efficacy of treatment.

### D. USG-B Scan

- B-scan ultrasonography frequently reveals choroidal thickening.

### E. Histology

1. The histologic features of SO are similar for both the exciting and sympathizing eyes.
2. Findings include diffuse, granulomatous, non-necrotizing infiltration of the choroid that classically spares the choriocapillaris in the early stage.
3. Dalen-Fuchs nodules are present in about one third of patients.
4. The nodules are not specific to SO and can also be found in VKH syndrome and sarcoidosis

## Differential diagnosis

1. Other causes of panuveitis, including TB, sarcoidosis, syphilis, and fungal infections, as well as traumatic or postoperative endophthalmitis.
2. Lens-associated uveitis has been reported with SO in up to 25% of cases and may present with a similar clinical picture.
3. The clinical presentations of SO and VKH syndrome may be strikingly similar; however, a history of prior ocular injury is by definition absent in patients with VKH syndrome.

## Treatment

- The course of SO is chronic, with frequent exacerbations; left untreated, SO leads to loss of vision and phthisis bulbi.
- Every attempt should be made to salvage eyes with a reasonable prognosis for useful vision through meticulous and prompt closure of penetrating injuries
- **Enucleation**
  1. Enucleation within 2 weeks of injury to prevent the development of SO should be considered in patients with grossly disorganized globes with no discernible visual function.
  2. Although controversial, enucleation may still be preferred to evisceration as the operation of choice for the removal of ocular contents in severely injured eyes because it eliminates the possibility of residual uveal tissue, which may predispose to the development of sympathetic disease
  3. Regardless of visual potential, once SO has become established, enucleation of the exciting eye has not been shown to be beneficial in altering the disease course of the sympathizing eye.
  4. In fact, the exciting eye may eventually become the better-seeing eye.



- **Medical management**

1. The initial treatment of SO involves systemic corticosteroids, with the frequent addition of corticosteroid-sparing drugs such as azathioprine, methotrexate, mycophenolate mofetil, cyclosporine, chlorambucil, and cyclophosphamide, as extended therapy is anticipated in most patients.
2. Topical corticosteroids, together with cycloplegic and mydriatic agents, are essential in the treatment of the acute anterior uveitis associated with SO.
3. Periocular corticosteroids are used to manage inflammatory recurrences and CME.
4. Intravitreal corticosteroids, including the intravitreal fluocinolone acetonide implant, represent an option for patients intolerant of systemic corticosteroid therapy.

**Prognosis**

- With prompt and aggressive systemic therapy, the visual prognosis of SO is good; 60% of patients achieve a final visual acuity of 20/40, although up to 25% may decline to 20/200 or worse in the sympathizing eye.

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