

Central Serous Chorioretinopathy

• Author: Kean Theng Oh, MD; Chief Editor: Hampton Roy, Sr, MD [more...](#)

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Background

Central serous chorioretinopathy (CSCR) is a disease in which a serous detachment of the neurosensory retina occurs over an area of leakage from the choriocapillaris through the retinal pigment epithelium (RPE). Other causes for RPE leaks, such as choroidal neovascularization, inflammation, or tumors, should be ruled out to make the diagnosis.

Central serous chorioretinopathy (CSCR) may be divided into 2 distinct clinical presentations. Classically, CSCR is caused by one or more discrete isolated leaks at the level of the RPE as seen on fluorescein angiography (FA). However, it is now recognized that CSCR may present with diffuse retinal pigment epithelial dysfunction (eg, diffuse retinal pigment epitheliopathy, chronic CSCR, decompensated RPE) characterized by neurosensory retinal detachment overlying areas of RPE atrophy and pigment mottling. During FA, broad areas of granular hyperfluorescence that contain one or many subtle leaks are seen.

Pathophysiology

Previous hypotheses for the pathophysiology have included abnormal ion transport across the RPE and focal choroidal vasculopathy. The advent of indocyanine green (ICG) angiography has highlighted the importance of the choroidal circulation to the pathogenesis of CSCR. ICG angiography has demonstrated both multifocal choroidal hyperpermeability and hypofluorescent areas suggestive of focal choroidal vascular compromise. Some investigators believe that initial choroidal vascular compromise subsequently leads to secondary dysfunction of the overlying RPE.^[1, 2]

Studies using multifocal electroretinography have demonstrated bilateral diffuse retinal dysfunction even when CSCR was active only in one eye.^[3] These studies support the belief of diffuse systemic effect on the choroidal vasculature.

Type A personalities, systemic hypertension, and obstructive sleep apnea may be associated with CSCR.^[4] The pathogenesis here is thought to be elevated circulating cortisol and epinephrine, which affect the autoregulation of the choroidal circulation. Furthermore, Tewari et al demonstrated that patients with CSCR showed impaired autonomic response with significantly decreased parasympathetic activity and significantly increased sympathetic activity.^[5]

Corticosteroids have a direct influence on the expression of adrenergic receptor genes and, thus, contribute to the overall effect of catecholamines on the pathogenesis of CSCR. Consequently, multiple studies have conclusively implicated the effect of corticosteroids in the development of CSCR. Carvalho-Recchia et al showed in a series that 52% of patients with CSCR had used exogenous steroids within 1 month of presentation as compared with 18% of control subjects.^[6]

Cotticelli et al showed an association between *Helicobacter pylori* infection and CSCR.^[7] The prevalence of *H pylori* infection was 78% in patients with CSCR compared with a prevalence of 43.5% in the control group. The authors proposed that *H pylori* infection may represent a risk factor in CSCR. While still controversial, other groups have continued to pursue this hypothesis based on case series.^[8, 9]

Epidemiology

Frequency

United States

Kitzmann et al reviewed the incidence of CSCR in Olmsted County, Minnesota. They evaluated the period from 1980-2002. They found the mean annual age-adjusted incidence of CSCR to be 9.9 cases per 100,000 population for men and 1.7 cases per 100,000 population for women.^[10]

International

Liew et al reviewed the epidemiology of CSCR in Australia. They found an incidence rate of 10 cases per 100,000 population in men. The rate of CSCR in this study was 6-fold higher in men than in women.^[9]

Mortality/Morbidity

Serous retinal detachments typically resolve spontaneously in most patients, with most patients (80-90%) returning to 20/25 or better vision. Even with return of good central visual acuity, many of these patients still notice dyschromatopsia, loss of contrast sensitivity, metamorphopsia, or, rarely, nyctalopia.

Patients with classic central serous chorioretinopathy (CSCR) (characterized by focal leaks) have a 40-50% risk of recurrence in the same eye.

Risk of choroidal neovascularization from previous CSCR is considered small (< 5%) but has an increasing frequency in older patients diagnosed with CSCR.^[11, 12]

A subset of patients (5-10%) may fail to recover 20/30 or better visual acuity. These patients often have recurrent or chronic serous retinal detachments, resulting in progressive RPE atrophy and permanent visual loss to 20/200 or worse. The final clinical picture represents diffuse retinal pigment epitheliopathy.

Otsuka et al reviewed a subset of patients who presented with a severe variant of CSCR over a mean follow-up period of 10.6 years.^[13] These patients were characterized by multifocal lesions and bullous retinal detachments with shifting fluid and fibrin deposition. During the follow-up period, 52% of patients experienced recurrences of CSCR ranging from 1-5 episodes. However, 80.4% of eyes (n=46) returned to a visual acuity of better than 20/40 and 52% returned to a visual acuity of 20/20 or better. Eventually, patients reached a state of quiescent disease.

Tsai et al reviewed a population-based cohort of CSCR patients from the Taiwan national health insurance research database from 2000-2007. They identified CSCR as an independent risk factor for ischemic stroke. After adjusting for age, sex, and comorbidities, CSCR had a 1.56-fold increased risk of stroke compared with controls.^[14]

Race

Central serous chorioretinopathy appears uncommon among African Americans but may be particularly severe among Hispanics and Asians.

Sex

Classically, CSCR is most common in male patients aged 20-55 years with type A personality. This condition affects men 6-10 times more often than it affects women.

Age

Patients may present with a later age of onset (>50 y). Spaide et al reviewed 130 consecutive patients with CSCR and found the age range at first diagnosis to be 22.2-82.9 years, with a mean age of 49.8 years.^[15]

Changes in the presentation and demographics of CSCR are observed with increasing age at first diagnosis. Classically, patients tend to be male and present with focal, isolated RPE leaks in one eye.

Patients diagnosed at 50 years or older are found to have bilateral disease, demonstrate a decreased male predominance (2.6:1), and show more diffuse RPE changes. Furthermore, these patients are more likely to have systemic hypertension or a history of corticosteroid use.^[16]

Clinical Presentation

Contributor Information and Disclosures

Author

Kean Theng Oh, MD Consulting Staff, Associated Retinal Consultants, PC

Kean Theng Oh, MD is a member of the following medical societies: [Association for Research in Vision and Ophthalmology](#), [American Society of Retina Specialists](#), [American Academy of Ophthalmology](#)

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Specialty Editor Board

Simon K Law, MD, PharmD Clinical Professor of Health Sciences, Department of Ophthalmology, Jules Stein Eye Institute, University of California, Los Angeles, David Geffen School of Medicine

Simon K Law, MD, PharmD is a member of the following medical societies: [American Academy of Ophthalmology](#), [Association for Research in Vision and Ophthalmology](#), [American Glaucoma Society](#)

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Steve Charles, MD Director of Charles Retina Institute; Clinical Professor, Department of Ophthalmology, University of Tennessee College of Medicine

Steve Charles, MD is a member of the following medical societies: [American Academy of Ophthalmology](#), [American Society of Retina Specialists](#), [Macula Society](#), [Retina Society](#), [Club Jules Gonin](#)

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Chief Editor

Hampton Roy, Sr, MD Associate Clinical Professor, Department of Ophthalmology, University of Arkansas for Medical Sciences

Hampton Roy, Sr, MD is a member of the following medical societies: [American Academy of Ophthalmology](#), [American College of Surgeons](#), [Pan-American Association of Ophthalmology](#)

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Additional Contributors

Brian A Philippotts, MD, MD

Brian A Philippotts, MD, MD is a member of the following medical societies: [American Academy of Ophthalmology](#), [American Diabetes Association](#), [American Medical Association](#), [National Medical Association](#)

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