

WAIVER GUIDE

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Supersedes Waiver Guide of Nov 2007

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CONDITION: OPTIC DISC (NERVE HEAD) DRUSEN

I. Overview

Optic disc drusen (ODD) are congenital and developmental anomalies of the optic nerve head commonly seen as an incidental finding during routine eye exams and are depositions of acellular, often calcified, hyaline material within the optic nerve thought to be the result of mitochondrial calcification within axons.¹ These hyaline bodies tend to lie beneath the surface of the optic nerve head but may become visible later in life as yellow-white, refractile bodies always superficial to the lamina cribrosa.¹⁻² Drusen may imitate the appearance papilledema with elevation and blurring of disc margins, but can be differentiated clinically by eliciting symptoms of increased intracranial pressure such as headache, especially increased upon awakening or after recumbency, and/or a pulsatile swishing sound heard by the patient with each pulse (pulsatile tinnitus). Symptoms of increased intracranial pressure are not expected with disc drusen and should initiate urgent neuro-imaging and possible lumbar puncture (after mass lesion has been excluded). The pathogenesis of ODD is unproven but likely stems from small optic disc size and mechanical obstruction to axonal transport.¹⁻² It's theorized that impaired ganglion cell transport mechanisms lead to abnormal axonal metabolism and mitochondrial damage, ultimately causing axonal deterioration and extrusion of calcified bodies.²⁻³ ODD may be associated with retinitis pigmentosa and pseudoxanthoma elasticum.¹ Other conditions that share a similar name, e.g. macular drusen associated with age-related macular degeneration, are not pathologically related to optic disc drusen.

ODD follows an autosomal dominant inheritance pattern and has a prevalence of 0.4 to 2 per 100 in the general population with a ten-fold increase in prevalence in family members of patients with drusen.² Men and women are affected equally with an average age at diagnosis of 22 years.² Bilaterality occurs in 67-85% of cases.² Most patients with ODD remain asymptomatic with normal visual acuity, although transient visual obscurations secondary to disc ischemia have been reported in 8.6% of study patients.^{1-2,4} Visual field defects with surface optic disc drusen are a common finding, occurring in 71 to 87% of cases but may be less common with buried drusen.^{1-2,4} Visual field defects are slowly progressive and often manifest as enlarged blind spots (60%) and arcuate defects (59%), typically sparing central vision.^{1,2,6} Potential complications related to ODD are ischemic optic neuropathy, central retinal artery occlusion, and retinal vein occlusion.² Unfortunately, there is no effective treatment established for optic disc drusen and visual field defects attributed to nerve fiber loss are permanent.

Diagnostic tests used to help identify optic disc drusen include.¹⁻²

1. Direct ophthalmoscopy
 - blurred or scalloped disc margins, translucent drusen when visible at surface.
 - the major retinal vessels are often anomalous (increased in number, branching and tortuosity)
2. Ophthalmic B-scan ultrasound
 - Preferred diagnostic test, non-invasive without radiation
3. Computed tomography of the orbit (recommend 3 mm cuts or thinner)
 - shows calcification of drusen at optic nerve head
4. Fluorescein angiography
 - can show late-hyperfluorescent staining of drusen bodies and leakage of juxtapapillary choroidal neovascular membranes
 - Buried and surface drusen display autofluorescence when photographed with the fluorescein filter in place pre-injection.
5. Scanning laser ophthalmoscopy and optical coherence tomography
 - may help detect buried drusen not visualized at the surface

Diagnostic tests used to help monitor effects of optic disc drusen include.

1. Refraction to best Snellen visual acuity
2. Color vision
3. Amsler grid
4. Afferent pupillary responses
5. Direct and/or stereo biomicroscopic ophthalmoscopy for disc and nerve fiber layer evaluation
6. Automated perimetry visual field testing (Humphery 30-2)
7. Contrast sensitivity
8. Color vision testing with anomaloscopy
9. Visual evoked potentials
10. Optical coherence tomography or scanning laser polarimetry



typical of an optic nerve with ODD and scalloped border.⁷ (reproduced with permission from NOVEL)

II. Aeromedical Concerns.

Clinically and aeromedically, the main concern with optic disc drusen is their propensity to induce slowly progressive loss of visual field. As high as 87% of individuals with optic nerve head drusen can expect to have visual field abnormalities. Furthermore, transient disturbances in central acuity and visual field may occur in association with optic nerve head drusen. Color vision defects have also been described in 41% of USAF aviators with ODD in preliminary data collected at the Aeromedical Consultation Service. Less commonly, the association of drusen with spontaneous hemorrhages in and around the optic disc may also result in acute changes in visual function.

Once the diagnosis of drusen is established, careful evaluation of optic nerve function is imperative. This should include visual acuity, visual field testing, Amsler grid, and color vision testing. Visual field loss has the most potential for aeromedical grounding and as such visual field testing should be performed on a regular basis to ensure visual fields remain adequate and consistent with mission effectiveness and flying safety. Optic disc photodocumentation should be obtained for comparison during future monitoring. It is also important for patients to self-monitor their vision periodically with Amsler Grid testing. Periodic surveillance to assess visual function in aircrew with optic nerve head drusen is appropriate, since drusen-related optic nerve problems are often asymptomatic. Routine cases should be monitored every six to twelve months.

III. Waiver Considerations.

Optic nerve head drusen is a disqualifying condition for flying classes I/IA, II, IIU, or III. It is not listed as a disqualifying diagnosis for ATC/GBC or SMOD personnel, but for

ATC/GBC personnel, it would be disqualifying if it results in a visual field defect. Aeromedical Consultation Service (ACS) evaluation is required for initial waiver of optic nerve head drusen for cases eligible for waiver. FC I/IA with optic nerve head drusen is not eligible for waiver. Optic nerve head drusen in untrained FC II and FC III are typically not eligible for waiver. ACS review is required for waiver renewal; depending on the results of local work-up, an ACS evaluation may be required. Waiver criteria for trained aircrew with optic nerve head drusen include acceptable visual performance on ophthalmologic examination including visual acuity, color vision and stereopsis, absence of transient visual loss, no aeromedically significant visual field deficit within the central 30 degrees of either eye, and a full binocular visual field.

Table I – Waiver criteria for aviators with optic nerve drusen

Flying Class (FC)	Waiver Potential Waiver Authority	ACS Review/Evaluation
I/IA	No AETC	No†
II IIU	Yes* MAJCOM**	Yes
III	Yes* MAJCOM	Yes
ATC/GBC	Waiver not required#	N/A
SMOD	Waiver not required#	N/A

* Waiver for untrained flying class II and III is unlikely.

** Waiver authority for FC IIU cases is AFMSA.

† ACS evaluation only required if diagnosis is in question.

Waiver will be required if the condition includes visual field or color vision defects.

A review of AIMWTS through Jan 2011 showed 98 cases of optic nerve head drusen. There were 17 FC I/IA cases (16 disqualifications), 44 FC II cases (0 disqualifications), 35 FC III cases (10 disqualifications – all initial FC III cases), and 2 ATC/GBC cases (0 disqualifications). All 26 of the DQ cases were disqualified for the diagnosis of optic nerve drusen.

IV. Information Required for Waiver Submission.

For optic nerve head drusen, the aeromedical summary for initial waiver or waiver renewal should include the following items:

- A. Complete aeromedical history to include pertinent positives and negatives (e.g. headaches, pulsatile tinnitus, hypertension, diabetes, family history of drusen, etc.)
- B. Presence or absence of visual symptoms and their operational impact (e.g. transient visual obscurations, perceived scotomas or metamorphopsia)
- C. Results of complete optometric or ophthalmologic eye examinations, to include, refraction to best Snellen visual acuity, color vision, Amsler grid, Humphrey threshold visual field testing (preferably central 30-2) – recent and previous, and stereoscopic optic disc evaluation.
- D. Diagnostic tests supporting diagnosis (e.g. ophthalmic B-scan ultrasound, computed tomography of the orbit, etc.)

ICD-9-CM Codes for Optic Nerve Head Drusen	
377.21	Drusen of optic disc

V. References.

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4. Sadun AA, Currie JN, and Lessell S. Transient Visual Obscurations with Elevated optic Discs. *Ann Neurology*, 1984; 16:489-494.
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