



# Visual snow: A neuro-ophthalmic syndrome

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## ABSTRACT

**Background:** The visual snow syndrome (VSS) is a rare disease characterized by persistent positive visual phenomena that are enigmatic and may be underestimated. It manifests as the perception of tiny, simultaneous, diffuse, and mobile dots frequently throughout the visual field. Although it affects brain networks, ocular examination is also indicated. This paper was aimed to review recent studies on VSS to identify the best clinical approach.

**Methods:** In this narrative review, PubMed/Medline, Directory of Open Access Journals, and Embase were searched using the following terms: “Visual snow”, “Persistent aura”, “Persistent positive visual phenomena”, “Visual aura”, “Migraine”, “TV static”, “Migraine aura status”, and “Visual disturbance”. We searched relevant publications from January 2014 to January 2021 to find the best clinical approach.

**Results:** VSS may be reconstituted as a distinct entity, although the symptoms may be consistent with typical migraine visual aura. For approximately two-thirds of patients, symptoms worsen and reduce their ability for activities of daily living. It often causes psychogenic detriment of the patient and their capacity to function socially and vocationally. Comprehensive history-taking and ocular examination are required to rule out other causes. However, all ocular and radiological findings may be normal.

**Conclusions:** VSS is a neuro-ophthalmic disorder characterized by continuous chronic and recurrent visual disturbances that involve the entire visual field and may respond to empiric antimigraine or anti-seizure treatments. It may reduce the ability for activities of daily living; therefore, proper diagnosis by eye care professionals is necessary to determine its diagnosis and management.

## KEY WORDS

visual snow syndrome, persistent positive visual phenomena, migraine, TV static, visual disturbance, entopic phenomena, migraine with aura, eye care professional, neurologist

## INTRODUCTION


The visual snow syndrome (VSS) is characterized by the perception of tiny, simultaneous, diffuse constant flickering dots, resembling the “TV static”. The mean age of onset of symptoms is 21 years, but it can occur at any age in patients of any sex or race. Liu et al. first described VSS in 1995, and until 2014, only a few cases had been reported in literature [1-3]. The prevalence of VSS has increased and is concomitant with tinnitus and headaches [4]. Manifestations of VSS are subjectively described as “TV static”, “carpet background”, “bubbles”, “rain-like patterns”, “squiggles”, “looking at a pixelated image”, “clouds”, and “TV snow” [2, 3].

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VSS is not a progressive disorder but is chronic with remission and recurrence [5]. Bilateral cases of VSS involving the visual field have been reported [6]. Patients with VSS tend to first visit ophthalmologists. After migraine-related and ocular causes are excluded, a neurologist is consulted [5, 7]. VSS is not notorious in its pathogenesis, although visual disturbances recur. The appearance and persistence of visual phenomena may psychologically disturb the patient [2, 5, 8]. This frightening condition prompts urgent referral to eye care professionals. The diagnosis may be difficult because the anatomical and functional integrity of the eye is preserved [3].

We aimed at reviewing studies on VSS to identify the best diagnostic approach for eye care professionals.

## METHODS

In this narrative review, PubMed/Medline, Directory of Open Access Journals, and Embase were searched using the following terms: “Visual snow”, “Persistent aura”, “Persistent positive visual phenomena”, “Visual aura”, “Migraine”, “TV static”, “Migraine aura status”, and “Visual disturbance”. We searched relevant articles from January 2014 to January 2021 to find the best clinical approach. The reference lists and related articles from each included article were reviewed to identify other potential studies.

## RESULTS

Our search yielded 55 articles, most of which were reviews. Seven papers [3, 9-14] proposed clinical and electrophysiological tests in diagnostic criteria, including a web-based survey study, case-control study, and retrospective case series. Table 1 summarizes the baseline clinical characteristics of patients with VSS reported in these clinical studies. The entoptic phenomena, migraine with and without aura, and common ophthalmic tests, such as perimetry, electroretinography (ERG), and visual evoked potentials (VEPs), were also included.

VSS is not an isolated entity but frequently related to other entities. In most publications [3, 9-14], it is reconstituted as an entity different from migraine, although often associated with it. For approximately two-thirds of the patients, symptoms progress to a series of episodes. For most patients, VSS affects the activities of daily living or has social consequences. Persistence of visual disturbances can disconcert the patient to such an extent that it interferes with activities of daily living. VSS often occurs in people working in social-network communities, which are constantly increasing.

**Table 1. Summary of findings of selected studies on the visual snow syndrome**

Author (Year of Publication)	n	Entoptic Phenomena *(%)	Migraine (%)	Migraine with aura (%)	NL Ophthalmic Assessments (%)
Puledda et al. (2020) [9]	1,104	61.3	51	37	NM, but participants with abnormal ophthalmic conditions were excluded.
Schankin et al. (2020) [10]	20	100	80	30	Ocular exam (100)
					Perimetry (95)
					VEPs (100)
Yoo et al. (2020) [11]	20	65	50	11	Ocular exam (100)
					Perimetry (100)
					ERG (100)
Metzler et al. (2018) [3]	4	100	50	NM	NM
Beyer et al. (2015) [12]	2	100	50	50	NL VEPs in the first case
					NL ocular exam in the second case
Schankin et al. (2014) [13]	120	83	58	31	Ocular exam (100)
Bessero et al. (2014) [14]	20	100	25	100	Ocular exam (100)
					Perimetry (100)
					ERGs; in those for whom the test was performed (100)
					VEPs; in those for whom the test was performed (100)

Abbreviations: n, number of participants; NM, not mentioned; NL, normal; ERG, electroretinography; VEP, visual evoked potentials. \*Symptoms of the entoptic phenomena were floaters, self-light of the eye, flashes, blue field entoptic phenomenon, or photopsia.

## DISCUSSION

VSS is a neuro-ophthalmic disorder characterized by continuous visual disturbances involving the entire visual field [11]. It often causes psychogenic detriment of the patient and their capacity to function socially and vocationally [15]. VSS results in sensory misperception. The etiology is hyperactive visual cortices or weakened processing of simultaneous afferent information projecting to the cortex. Consequently, other syndromes that result in sensory misperception, such as migraine, persistent perceptual postural dizziness, and tinnitus, overlap with VSS [15].

Ophthalmologists, as the first line of referral, frequently play a decisive role in the diagnosis of VSS [5,7]. The most common positive visual phenomena are associated with migraine. Schankin et al. defined these symptoms as “persistent positive visual phenomena”, distinguished from migraine aura [16].

The first step in the clinical approach is to distinguish between primary and secondary VSS. Ophthalmic evaluation is recommended to rule out secondary etiologies, such as drugs and hallucinogenic substances, physical fatigue after sports, posttraumatic stress disorder as a psychiatric comorbidity, visual hallucinations, ischemic infarcts of the retrochiasm visual pathways, hemianopsia, and Charles Bonnet syndrome [17]. VSS might be grouped with the persistent visual phenomena in migraine; however, based on clinical evidence, VSS may be considered as a distinct entity.

Aura is typically a temporary visual phenomenon that lasts for minutes. The International Classification of Headache Disorders, Version 3, beta version, classifies aura as “persistent aura without infarction” or “migraine aura status” [15, 16]. Therefore, VSS was considered as “persistent positive visual phenomena” [3]. Schankin et al. concluded that VSS is a clinically distinct entity that should be classified as a distinct syndrome. Comorbid migraine may aggravate the clinical presentation with additional visual symptoms and tinnitus [13].

Puledda et al. [9] investigated approximately 1,104 patients with VSS, and no clinical endophenotype was defined. Several studies demonstrated a moderately high prevalence of migraine associated with the VSS [10, 13] (Table 1). For instance, a prospective study involving 120 cases identified a history of migraine without aura in 69% of the subjects and migraine with typical aura in 31% of the subjects [13]. Tinnitus is common in this population as well, occurring in up to 63% of the subjects, although the frequency or durability of tinnitus has not been explained [3]. “Enhanced entoptic phenomena” refer to visual perceptions created by the anatomy of the visual system and may include floaters, blue-field entoptic phenomena, spontaneous photopsia, and self-light of the eye [16].

The symptoms of VSS do not fulfill the diagnostic criteria for migraine aura and may be considered as a separate entity. Once the diagnostic criteria for VSS were published [2], cohorts were conducted to determine VSS. Lauschke et al. [18] reported 32 cases of VSS and found that 29 (91%) cases fulfilled the proposed diagnostic criteria. Bessero et al. [14] reported 20 cases and found that all cases were consistent with the proposed VSS criteria. Schankin et al. [13] proposed that patients with VSS may have clouds in their vision with eyes closed. However, they may describe kaleidoscopes of colors with their eyes open [3, 16]. Therefore, the term “enhanced entopic phenomena” has been proposed to be modified to “other frequent or persistent positive visual phenomena”. This would include blue field entoptic changes, “straight lines moving across the visual field”, “water running down a window”, and “geometric and colored images”, as described in Schankin’s retrospective cases [3, 16].

Functional brain imaging in patients with VSS has revealed hypermetabolism within the lingual gyrus, suggesting that the symptoms are caused by cerebral network or cortical dysfunction [10, 15]. The etiology is unknown; however, VSS has a clear biological basis. The direct relationship between migraine and VSS is based on the overlap between the area of hypermetabolism and that involved in photophobia in migraine [16]. Maniyar et al. [19] demonstrated hyperperfusion in the lingual gyrus in patients with photophobia in the premonitory phase of migraine before the onset of headache. This shows that VSS and typical migraine aura are both associated with dysfunction in the Brodmann area 19. The primary and secondary visual areas of the brain were assumed to be hyperexcited. The presence of extra-visual symptoms in VSS has been investigated using positron emission tomography, which revealed hypermetabolism and increased cortical volume in the extrastriate visual cortex at the junction of the right lingual and fusiform gyri [10].

Magnetic resonance imaging of the brain revealed left occipital bending [20]. In a VEP study, the cortical hyperexcitability improved significantly. Lamotrigine resolved symptoms of VSS, and the patients were able to sleep with a decreased frequency of migraine to two attacks/month [20]. Lauschke et al. proposed the use of color filters in the yellow–blue color spectrum, which subjectively may cause partial improvement. The filter may change the koniocellular pathway and regularize the electroencephalographic rhythms [18].

**Table 2. Recommended workup for visual snow syndrome in the eye care setting**

Consider secondary etiologies, such as toxic causes, for hallucinations or similar symptoms [3].
Identify which persistent positive visual phenomena are presented [3] and compare with a standard dataset.
Evaluate the duration and recurrence of the persistent positive visual phenomena [14].
Consider if persistent positive visual phenomena are associated with migraine [9].
Test the visual acuity with refractive correction or record the best-corrected visual acuity [9, 11, 14].
Perform comprehensive ocular examination, including extraocular muscle movement, pupillary reflex, and detailed anterior and posterior segment examinations, to exclude other causes [3, 9, 10, 11, 13].
Perform further evaluations, such as automated perimetry, ERG, and VEPs [10-12, 14].
Perform head computed tomography or magnetic resonance imaging, if indicated [10, 13, 14].
If all ocular examinations are unremarkable, consult a neurologist [11] and jointly plan other potential causes.

**Abbreviations:** ERG, electroretinography; VEPs, visual evoked potentials.

Pharmacological and non-pharmacological treatment approaches for VSS are undergoing randomized controlled clinical trials. Currently, investigational therapies are posted in clinical trial registries [21-23]. Physical examination of VSS is unremarkable. Visual impairment is the first patient complaint, and patients are referred to the ocular emergency unit. Generally, a comprehensive ophthalmic assessment comprises the evaluation of the extraocular muscles, anterior segment, pupillary reflex, crystalline state, vitreous, retina, and optic nerve. Usually, no ocular structure abnormality is detected, and there are no signs associated with VSS [3].

First, the visual acuity should be tested with prepared glasses [24]. Visual acuity of a logarithm of the minimum angle of resolution (logMAR) of 0.45 (decimal equivalent of approximately 0.4) with glasses reveals the potential for admission [24]. Among 20 patients with VSS, Yoo et al. [11] reported a mean best-corrected visual acuity below 0.1 logMAR with reduced contrast sensitivity in only two patients.

Most patients with VSS show no objective abnormalities on ocular examination. In a study of seven patients with VSS, all examinations, including funduscopy, color testing, visual acuity, perimetry, ERG, VEPs, and optical coherence tomography, were normal, with no ocular pathology [25]. In a review of patients with VSS, Schankin et al. [2] reported that all patients had normal funduscopy and perimetry findings. Bessero et al. [14] reviewed 20 patients with VSS and found that all had normal ocular examinations and automated perimetry. Furthermore, ERG and VEP findings were normal in those for whom the test was performed (Table 1). Based on the literature review [3, 9-14], we would recommend a proper approach for the rapid diagnosis of VSS (Table 2).

VSS made it possible to interconnect patients with these symptoms over the internet. VSS self-assisted groups were intuitive, and the first was the Eye on Vision Foundation founded in 2010 [13, 26]. This was an internet-based survey on specific items, such as age groups, sex, and time of onset and duration of symptomatology, while other issues could be consulted through the Visual Snow Initiative [27]. On social networking platforms, the number of digital communities for VSS has increased significantly on Facebook and Twitter. Participants may elaborate on their own experiences with VSS.

EyeWiki, a free online encyclopedia supported by the American Academy of Ophthalmology, has a comprehensive collection of VSS resources [28]. Lamotrigine, naproxen, carbamazepine, nortriptyline, and sertraline have been reported to reduce symptoms, and several resources are available to discuss different aspects of VSS [28, 29]. In 2018, the VSS conference, promoted by the Visual Snow Initiative, was held at the University of California San Francisco to invite the academic community to the VSS. Before this event, a few original studies were presented at national and international ophthalmic congresses [30].

To our knowledge, this is the first review to summarize diagnostic approaches for VSS management for eye care professionals. This study is limited by the small number of primary studies owing to the nature of VSS with unknown etiology and no proven prevention or treatment modality. Further randomized controlled trials are warranted to shed light on this topic, and further systematic reviews and meta-analyses are required to provide a comprehensive overview.

## CONCLUSIONS

Recent commentaries have indicated that VSS is a neuro-ophthalmic disorder; however, VSS may cause visual disturbance and affect activities of daily living. As it is an enigmatic disorder, it is usually underestimated. Visual phenomena are most frequently migraine-related, although this is not always the case. An appropriate and prompt diagnosis might be worthwhile for the best management of VSS, strongly supported by collaboration between eye care professionals and neurologists.

## ETHICAL DECLARATIONS

**Ethical approval:** This study was a review, and no ethical approval was required.

**Conflict of interests:** None.

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