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HEADACHE (R HALKER, SECTION EDITOR)

Visual Snow Syndrome: Proposed Criteria, Clinical Implications, and Pathophysiology

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Abstract

Purpose of Review In this article, we review illustrative case descriptions of both primary and secondary visual snow from our clinic. We discuss recently proposed criteria for visual snow syndrome and offer a slight modification of these criteria. We also discuss the theories on the pathophysiological mechanisms of visual snow, as well as the current approach to treatment.

Recent Findings Visual snow is a condition where patients see constant, innumerable flickering dots throughout the visual field, similar to "TV static." Though visual snow was originally described in 1995, there were still fewer than 10 cases in the literature prior to 2014. In the last 4 years, this has grown to approximately 200 cases and there has been a concentrated effort to better understand and characterize this condition. It has become apparent that patients who see visual snow frequently have additional visual and non-visual symptoms, and the consistency of these symptoms has led to proposed criteria for visual snow syndrome.

Summary When seeing a patient with visual snow, it is important to rule out a possible underlying secondary etiology. Patients with visual snow syndrome frequently have comorbid migraine, but visual snow appears to be a separate entity from persistent migraine aura. The pathophysiology of this syndrome is not clear, but recent neurophysiologic and neuroimaging studies have helped advance our understanding.

Keywords Visual snow · Persistent aura · Persistent positive visual phenomena · Visual aura · Pathophysiology · Migraine

Introduction

"Visual snow" is a rare condition with persistent positive visual phenomena that appear similar to grainy or pixelated television static, as one might see on an analog television channel out of tune [1••, 2, 3•, 4, 5]. Descriptions from patients are varied but include a theme of persistent, innumerable flickering dots throughout the visual field that are typically black and white, though they may sometimes be colorful. The intensity may vary in ambient lighting, be more noticeable on plain backgrounds, and may be more attenuated on textured backgrounds [1••, 3•, 5–7].

Visual snow was originally described in three patients within a case series of ten migraineurs presenting with a spectrum of positive visual symptoms, which Liu et al. referred to as "persistent positive visual phenomena" [3•]. In addition to the "TV static" description, patients in his series described "bubbles," "carpet background," "rain-like patterns," "clouds," "squiggles," and other visual symptoms that Liu et al. classified as definitely, probably, or (in the case of visual snow) possibly related to the underlying history of migraine [3•]. There have now been approximately 200 cases of visual snow described [6, 8••, 9–12]. As more cases have been reported, some authors have separated visual snow from other persistent and repetitive visual symptoms associated with migraine [1••, 13]. Schankin et al., specifically, have concluded that visual snow is a clinically distinct entity that should be classified as its own syndrome, and have proposed diagnostic criteria for a diagnosis of visual snow syndrome (VSS) [1••].

In this paper, we review illustrative case descriptions of both primary and secondary visual snow from our clinic. We

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review the recently proposed criteria for VSS and propose a modification of one subsection of these criteria. We also discuss possible pathophysiological mechanisms of visual snow and the current approach to treatment.

Primary Visual Snow

Case No. 1 from Our Clinic

A 42-year-old woman with a history of migraines presented with colorful "television snow" in her vision since her early childhood (as long as she could remember). This was present throughout her visual field constantly without fluctuation in intensity, though she did notice more colorful visual static when she was in the dark or when there was a white background such as a blanket of white snow. This was not affected by her migraines or any preventative medications she had taken for her migraine headaches. She also had a long history of mild tinnitus.

Case No. 2 from our Clinic

A 42-year-old woman with a history of migraine headaches occurring about 4 days per week presented with a 2-year history of "snow vision" described as a "hazy or granular TV image" throughout her vision, though it was worse in the periphery. Superimposed on this, she sometimes saw "particles" in her vision, "glittery vision," or a "continuous kaleidoscope of colors rolling throughout" her vision. She also described persistent photophobia, and when she closed her eyes it was like "fireworks are going off." In the 2 years after her initial visit, she was able to get good control over her migraine headaches with topiramate, verapamil, and onabotulinumtoxinA injections, but the visual symptoms persisted.

These first two cases from our clinic are fairly typical presentations of primary visual snow (no other known underlying cause). Similar to our patients, studies have shown a relatively high prevalence of migraine in patients with visual snow, with one prospective study of 120 patients demonstrating a history of migraine with or without aura in 58% of patients, and migraine with typical aura in 31% of patients [1••]. Tinnitus seems to be common in this population as well, occurring in up to 63% of patients, though the frequency and persistence of tinnitus has not been described [1••, 4•].

Also similar to our first two cases, it is common for patients with visual snow to present with other concurrent visual symptoms. After recruiting 120 patients with visual snow through social media and self-help groups and interviewing each by phone, Schankin et al. attempted to characterize the subset of patients with no obvious etiology for their visual snow [1••]. They included 78 patients with normal neurologic and ophthalmologic evaluations and found the following associated visual symptoms occurred relatively frequently within this population (see Fig. 1 for illustrations of some of these):

- Palinopsia with "after images" (86%) or "visual trails" after moving objects (60%)
- Excessive floaters (81%)
- Blue field entoptic phenomenon (79%)

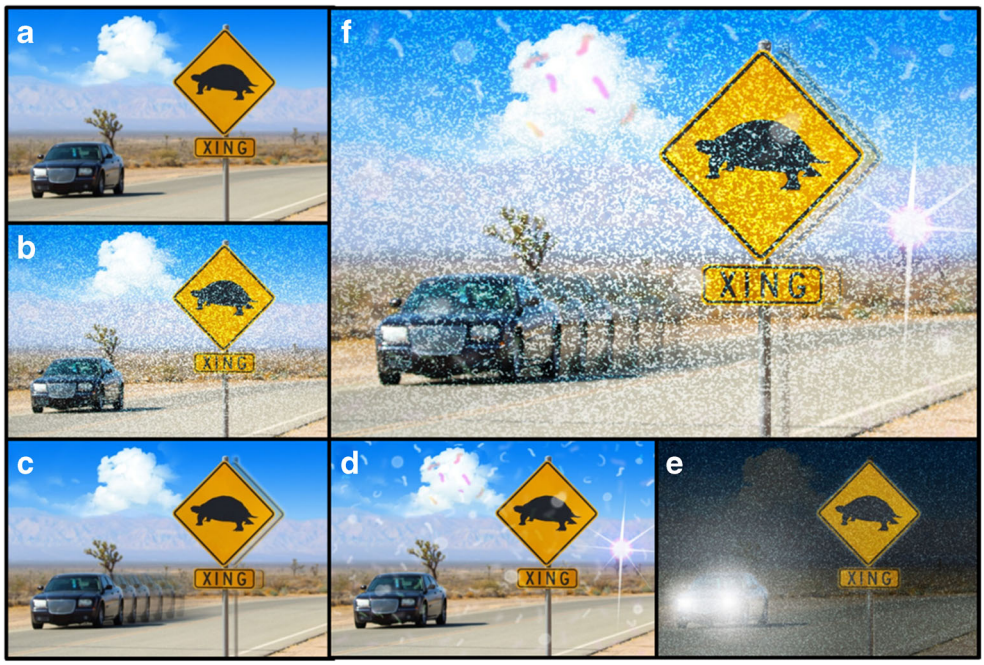

Fig. 1 Illustrative examples of the revised criteria for visual snow syndrome (VSS). **a** Normal vision. **b** Visual snow. **c** Palinopsia (both visual trailing behind the car and a ghost-like afterimage next to the sign). **d** Other frequent or persistent positive phenomena (including increased floaters, blue field entopic phenomena, photopsia, and colored blobs). **e** Nyctalopia (impaired night vision). **f** Composite illustration showing the possible level of visual disability these symptoms may cause when combined. *Used with permission from Mayo Foundation for Medical Education and Research. Original photo modified and used under institutional Shutterstock license

– This is described by patients as "little bright dots travelling on a wiggly path" when looking at a blue background and is thought to be leukocytes flowing within the macular microvasculature [14]

- Photophobia (74%)
- Spontaneous photopsia (flashes of light) 63%
- Self-light of the eye (53%)

– This is described by Schankin et al. as "clouds, swirls, or waves with eyes closed" [1••]

- Nyctalopia (impaired night vision) (68%)

Based on the characteristic presentation of these patients, their clinical features distinctive from migraine aura, and their lack of response to migraine therapy, Schankin et al. proposed diagnostic criteria for VSS. This syndrome would require the persistent symptom of visual snow lasting longer than 3 months, with at least two additional visual symptoms: palinopsia, enhanced entoptic phenomena, photophobia, or nyctalopia [1••]. "Enhanced entoptic phenomena," defined by Schankin et al. refers to visual perceptions created by the anatomy of the visual system and can include any of the following: floaters, blue field entoptic phenomena, spontaneous photopsia, or self-light of the eye. Finally, the symptoms cannot meet definition for migraine aura and cannot be better explained by another disorder. After the syndrome criteria were published, two groups published additional cohorts of patients to test these criteria. Lauschke et al. described 32 additional cases of visual snow and found 29 (91%) fulfilled the proposed diagnostic criteria [4•], while Bessero et al. described an additional 20 patients and found all 20 to be consistent with the proposed VSS criteria [5].

In the last few years, Dr. Schankin and his colleagues have contributed significantly to the characterization, classification, and understanding of this unique subset of patients [1••, 8••, 13, 15••]. Their clinical experience matches ours, and the availability of VSS criteria has added validity to the syndrome and the patients experiencing it. However, in our clinic, we have had difficulty applying the section labeled "enhanced entoptic phenomena." Classically, the term "entoptic" has been understood to refer to phenomena arising from within the structure of the eye. The proposed VSS criteria use a somewhat revised definition of the term, instead, referring to Tyler's 1978 paper that extends this to include "phenomena arising from [any] structure of the visual system as a result of specific stimulation" [16]. Several issues arise with this extended definition of the term. First, by this description, "enhanced entoptic phenomena" would then also include palinopsia and photophobia, as well as the visual snow itself, making their individual listing unnecessary. Second, because this extended definition is not universally used, clinicians using the VSS criteria to assess their patients may be tempted to assume that any otherwise undefined positive visual symptoms, such as the flashes of light (called "phosphenes" by some authors and "spontaneous photopsias" by Schankin, et al.), must originate from within the eye. It is well established that similar flashes of light can occur by stimulation of the visual cortex [17, 18] or within the hemianopic field after injury to the occipital lobe [19]. Similarly, the term "self-light of the eye" references a description by Marshall in 1935 of two types of light seen by the eyes when they are closed; one is the uniform grayness in a completely dark-adapted eye (i.e., after a long sleep), and the other is "luminous clouds, generally of a violet color, moving in waves centripetally or centrifugally" [20]. Marshall proposed this self-light was related to energy from the pigment particles of the retina and retro-retinal circulation [20], though the true etiology for this phenomenon is not clear [21]. As Schankin et al. suggest, patients with visual snow may have similar swirls or clouds in their vision with their eyes closed. However, similar to our clinic patient, they may also describe kaleidoscopes of colors even with their eyes open [1••]. For these reasons, we would respectfully propose that the phrase "enhanced entopic phenomena," be changed to "other frequent or persistent positive visual phenomena" (see Table 1). This would still include increased floaters and blue field entoptic changes, but it also provides a more accurate section for many of the otherwise unclassifiable phenomena described by patients with visual snow, including those found in Schankin's own retrospective case descriptions (i.e., "straight lines moving across the visual field," "water running down a window," "geometric and colored images") [1••].

Table 1 Proposed criteria for visual snow syndrome (modified from Schankin et al. 2014) [1••]

1. Visual Snow: dynamic, continuous, tiny dots in the entire visual field lasting longer than 3 months
2. Presence of at least two additional visual symptoms from the following categories:
   a. Palinopsia: afterimages or trailing of moving objects
   b. Photophobia
   c. Nyctalopia (impaired night vision)
   d. Other persistent positive visual phenomena* including (but not limited to): enhanced entoptic phenomena (excessive floaters or blue field entoptic phenomenon), kaleidoscope-type colors with eyes open or closed, spontaneous photopsias
3. Symptoms are not consistent with typical migraine visual aura
4. Symptoms are not better explained by another disorder

*Modified from Schankin et al. [1••], replacing "enhanced entoptic phenomena" with "other persistent positive visual phenomena"

Secondary Visual Snow

Case No. 3 from Our Clinic

A 21-year-old mechanical engineering student with a prior history of daily marijuana and episodic alcohol abuse presented with a 16-month history of persistent visual symptoms that he first noticed when he woke up in the hospital after a night of cocaine use. Since that hospitalization, his vision has been as if he was "looking at a pixelated image—like looking at a poorly tuned television" with abnormal colors and movement. His depth perception seems abnormal to him, and he has noticed intermittent floaters and halos around objects.

Case No. 4 from Our Clinic

A 17-year-old boy presented with persistent visual symptoms that started abruptly after a sports-related concussion the year before. In addition to a host of post-concussive symptoms including fatigue, "brain fog," and persistent dizziness, he described seeing static "like on TV" throughout his visual field all the time, with associated palinopsia. He estimates getting headaches about 4 days per week, but the visual symptoms seem separate from the headaches, and have not responded to numerous pharmaceutical trials.

Like the first two cases, cases 3 and 4 represent patients with visual snow, including symptoms similar to those described in the VSS, but they are presumed to be secondary to specific etiologies. Case 3 represents a well-described phenomenon called hallucination persisting perception disorder (HPPD). Patients with HPPD describe a variety of positive visual symptoms including persistent geometric hallucinations, visual snow, halos, illusions of movement, flashes of light, and palinopsia in association with hallucinogenic drugs such as lysergic acid diethylamide (LSD), cannabis, synthetic cannabis, mescaline, and MDMA/Ecstasy [22–26]. These hallucinations are persistent even months or years after stopping the drug, and are often resistant to treatment [26].

Prescription drugs have also been associated with persistent visual symptoms such as palinopsia, bilateral shimmering/flickering vision, photophobia, and flashing lights, though specific descriptions of visual snow have not yet been described [27–33]. In the cases where the visual symptoms started in association with trazodone [27], nefazodone [27, 29], mirtazapine [28], and topiramate [31], the symptoms improved after discontinuation of the medication. In the cases described associated with the use of clomiphene, the symptoms were persistent even years after the drug was stopped [32, 33].

Similar to case 4, one of the three patients with visual snow originally described by Liu et al. experienced symptoms immediately after head trauma [3•]. A history of head trauma at onset was not specifically addressed in most of the available case series on visual snow. However, reviewing the same types of social media sites from which Schankin et al.'s case series was recruited, we were able to find another four additional descriptions of visual snow starting just after a head injury (https://www.neurotalk.org/traumatic-brain-injury-and-post-concussion-syndrome/222690-vision-static-snow.html, https://www.tapatalk.com/groups/thosewithvisualsnow/visual-snow-following-brain-injury-anyone-else-t4243.html). In their series of 20 patients with visual snow, Bessero et al. reported two with a history of minor head injury; however, they occurred one and several weeks before the onset of visual snow, so it was unclear if the injuries were related [5]. Lauschke et al. did not specifically discuss a history of trauma but listed post-traumatic stress disorder as one of the comorbid psychiatric symptoms present in a subset of their patients [4•]. The exact percentage of patients with head trauma at the onset of their symptoms is unclear, but we are in the process of examining our institutional experience to answer this question.

The possibility of visual hallucinations after injury to the visual pathways is not a new concept, with one series reporting positive spontaneous visual phenomena in 41% of patients with ischemic infarcts of the retrochiasmal visual pathways, within the area of hemianopsia [34]. This included achromatic or chromatic phosphenes, simple hallucinations such as moving fans, stars, or geometric forms, as well as more complex visual hallucinations. Palinopsia has been reported in numerous lesions in the post-geniculate visual pathway including infarcts, space-occupying lesions, demyelinating lesions, and infections [35].

Charles Bonnet syndrome (CBS) is a syndrome of visual hallucinations occurring in patients who have lost their sight. CBS is often due to bilateral eye disease, but it can occur theoretically with any lesion of the visual pathway that reduces sensory input to the visual cortex. The term is most often used in reference to patients with complex visual hallucinations, seeing things like people or scenes. However, a significant number of patients describe simple or elementary hallucinations such as flashes of light, geometric shapes, palinopsia, or tessellopsia (repeated grid-like geometric shapes) [36]. In one study of patients with CBS related to senile macular degeneration or other ocular pathology, 23% described multiple particles through the visual field like "rain drops appear(ing) over everything" with a striking similarity to visual snow descriptions [36]. In their review on visual snow, Puledda and colleagues point out that many of the associated symptoms, including photopsias, night blindness, and shimmering around objects can be associated with retinal or vitreous pathology, including neoplastic and paraneoplastic retinopathy [8••]. For this reason, they recommend a thorough ophthalmologic evaluation to rule out underlying eye pathology in patients presenting with these symptoms [8••].

An electroencephalogram (EEG) is also typically performed when evaluating patients with visual snow to rule

out an underlying epileptic etiology [1••, 3•]. Though flickering/flashing lights and even the description of "TV static" have been described with epileptic visual auras, the symptoms tend to be within one visual hemi-field and are transient, typically seconds to minutes in duration [37, 38]. Epileptic palinopsia seen both ictally and post-ictally also tends to be transient and within one hemi-field [35].

Ophthalmologic Testing with Visual Snow

While ophthalmologic evaluation is recommended to rule out a secondary etiology for their symptoms, most patients with visual snow have no objective abnormalities on ophthalmologic testing. One study of seven visual snow patients performed extensive testing (including funduscopic exam, color testing, visual acuity, perimetry, electroretinography (ERG), visual evoked potentials (VEPs), and optical coherence tomography (OCT)) and did not find any ocular pathology [11]. In their review of 78 patients with VSS, Schankin et al. reported all 78 had normal funduscopic exams and perimetry, 14 had normal ERGs, and 22 had normal VEPs [1••]. Bessero et al. reviewed medical records of 20 patients with visual snow and found all 20 had normal automated perimetry, ten had normal ERGs and seven had normal VEPs [5].

Imaging with Visual Snow

Imaging studies looking at cerebral blood flow (CBF) in patients with migraine with aura have demonstrated an initial wave of hypoperfusion lasting a few hours (often associated with the prodrome and/or aura), followed by a delayed and more prolonged hyperperfusion that can outlast the headache [39–43]. Jager et al. looked at MR perfusion and MR diffusion in four patients (two with persistent visual aura and two with visual snow phenomenon) and found no changes in perfusion, leading them to hypothesize that these persistent visual symptoms were pathophysiologically different from typical migraine aura [44]. However, there must be some variability based on timing, symptoms, or other factors, as there have been other cases of prolonged or persistent visual aura involving one or both visual hemi-fields where both SPECT and MR perfusion images have shown hypoperfusion either in the contralateral or bilateral posterior cerebral cortex [3•, 45–47]. In fact, two of the original three cases of visual snow described by Liu also had hypoperfusion in the bilateral parietal (patient 7) and parietooccipital (patient 8) lobes on SPECT [3•].

More recently, [^{18}F] FDG PET imaging studies comparing 17 patients with visual snow to 17 healthy controls, showed significantly increased metabolic activity in the right lingual gyrus ($p < 0.001$) and the anterior lobe of the left cerebellum ($p = 0.001$) in patients with visual snow [15••]. This is noteworthy as H_2O^{15} PET imaging studies have shown light stimulation activates the bilateral cuneus, lingual and posterior cingulate cortex in migraineurs (but not controls) [48]. Boulloche et al. hypothesized that the light activation of the cortex in migraine patients demonstrated the lack of habituation and/or cortical excitability in these patients [48]. The same authors then performed a study looking at H_2O^{15} PET during a migraine, after treatment with sumatriptan, and interictally. They found that low levels of light increased cortical blood flow during the migraine attack (before or after treatment) but not interictally [49••]. Of relevance when comparing these studies, Schankin et al. point out that the [^{18}F] FDG PET imaging studies were performed in the dark with the patients' eyes closed, while the patients experienced visual snow and possibly self-light of the eye, but not photophobia or palinopsia [50].

Theories on the Pathophysiology of Visual Snow

Not only is the pathophysiology underlying visual snow unclear, the localization is a matter of some debate. As there is no retinotopic organization of the field of flickering dots (the central visual dots are not necessarily bigger or more spread out than the peripheral dots), many feel it is unlikely that the primary visual cortex is the source of the hallucination [6, 8••]. Though some authors have interpreted this to mean that the pathology must be anterior to the optic radiations (perhaps in the eyes themselves as yet another entoptic phenomenon) [6], others feel the even spacing of dots should localize to somewhere in the visual pathway "beyond the lateral geniculate nucleus" [4•, 51]. As pointed out by Puledda et al., it seems unlikely that a whole-field visual phenomenon can be explained by any simple lesion or disorder of the visual pathway [8••].

Role of the Thalamus

One pathophysiologic theory regarding the origin of visual hallucinations has emphasized the role of the thalamus. Ffytche used photic stimulation to induce Purkinje hallucinations (colors, geometrical shapes, motion) in volunteers while monitoring them with fMRI and EEG. He found a wide network of areas with increased brain activity including the bilateral ventral occipitotemporal regions centered on the fusiform gyrus, extending medially to the lingual gyrus and laterally to the inferior temporal gyrus [52••]. Interestingly, while the occipitotemporal activity was significantly increased during visual hallucinations compared to control conditions, the primary visual cortex (V1) was not significantly more active, and the lateral geniculate nucleus (LGN) was actually significantly decreased compared to controls [52••]. Ffytche hypothesized that during the visual hallucinations, the LGN had switched from its tonic mode

(where retinal input is sent to the cortex in a proportional fashion) to its burst mode (where input and output become partly disassociated), in effect creating a "thalamic blindness." He theorized that this burst mode might create a transient LGN de-afferentation allowing hallucinations to form in a fashion similar to CBS hallucinations [52••]. Some authors have suggested that the coherent low-frequency thalamic oscillation and the subsequent reduced lateral cortical inhibition create a "thalamocortical dysrhythmia" that might underly multiple neurologic processes including tinnitus and phantom limb pain, in addition to visual snow [4•, 53].

Cortical Hyperexcitability/Reduced Habituation

Another common theme within discussions on the pathophysiology of visual snow is the concept that either cortical hyperexcitability or reduced habituation may allow patients to visualize normally subthreshold stimuli [51]. Several studies have demonstrated cortical hyperexcitability, or at least hyperresponsivity, using electrophysiologic testing [12, 54, 55]. In normal patients, repetitive stimulation with visual stimuli elicits physiological habituation [56]. In contrast, Unal-Cevik and Yildiz described a migraineur with visual snow who demonstrated potentiation to repetitive pattern-reversal VEPs. Interestingly, the patient showed partial improvement both electrophysiologically and clinically after treatment with lamotrigine [54]. As this reduced habituation has been seen in migraine patients interictally, it was unclear whether the effect was specific to the patient's visual snow [57]. To provide further clarification on this point, Luna repeated a similar experiment on a 22-year-old with 2 years of visual snow and no clear comorbid migraine and again found potentiation with repetitive VEPs [12]. Moving away from traditional VEPs, Chen et al. used visually evoked magnetic field recordings to study six patients with persistent visual aura, including two patients with visual snow. They found these patients had patterns consistent with persistent visual cortex hyperexcitability, without the interictal-ictal variation typically seen in migraine [55].

In migraine patients who show similar lack of habituation with repetitive VEPs, it has been noted that the VEP habituation is inversely related to the first-block amplitude. This was not seen in controls and may suggest that the habituation deficit is a consequence of initial visual cortex hypoactivation [58]. Some have postulated that in migraine patients between attacks, decreased thalamocortical activity may contribute to low cortical reactivity (possibly related to reduced lateral inhibition), leading to a lack of habituation and subsequent "building up of a globally exaggerated response" with hyperresponsivity [59].

Finally, some authors have theorized that visual snow and many of the associated visual symptoms are a type of default that is not seen in healthy subjects due to an active suppression system, possibly within the supplementary visual cortex [50]. Given the 18F-FDG PET findings of lingual hypermetabolism described in the imaging section, Schankin et al. have questioned whether the pathophysiology of VSS might specifically involve dysfunctional visual processing in the lingual gyrus [8••].

Stochastic Resonance

When pondering the pathophysiology behind visual snow in the context of the newly defined VSS, it is of interest to consider the frequently associated symptoms including tinnitus, dizziness, photophobia, and other visual symptoms such as the tendency to see physiologic phenomena (blue field entoptic phenomena, excessive floaters) [4•, 15••]. A conceptual model that might apply to the association of these additional symptoms is the concept of stochastic resonance. Stochastic resonance (SR) is a nonlinear phenomenon where the addition of noise can improve the signal-to-noise ratio, improving the ability to detect a weak stimulus, at least up to an optimal level of noise. Beyond this level, additional noise intensity degrades detection or information content [60, 61]. This has been shown to occur in all sensory modalities, including the visual system. Unimodal SR, occurring when the signal and noise both involve the same sense, has been described in vision, where visual noise enhances visual perception (background visual static improves contrast detection sensitivity, motion sensitivity, and the perception of ambiguous figures) [61, 62]. Interestingly, there also appears to be a cross-model of SR in the sensory systems, with several studies showing that the introduction of mild to moderate noise in one system increases the sensitivity of another sensory system. For instance, when random auditory "white noise" is introduced, this seems to enhance sensitivity to visual flicker [63] or subthreshold visual flash of light stimuli [64]. In fact, an optimal amount of auditory noise can decrease luminance and contrast visual thresholds, as well as enhance tactile sensation [61]. Some authors have theorized that this ubiquitous cross-model of SR might be used by the sensory perceptual systems to enhance sensory perception [65, 66•]. However, this might also represent a plausible model for the observed cross-modal central sensitization seen in migraine, such as optokinetic stimulation increasing allodynia and photophobia in migraineurs [67]. In the case of patients with visual snow, this might be a model for the high comorbidity of tinnitus, dizziness, migraine, and other symptoms. Perhaps the presence of tinnitus enhances the visual snow or enhanced entopic phenomena, or vice versa, with noise in one sensory system "priming" the other systems. Interestingly, just as too much noise can actually degrade information content in the model for SR, it has been shown that the same level of noise that was ideal for enhancing subthreshold visual stimuli actually reduced the detection of suprathreshold visual stimuli [64,

68]. Ultimately, noise above a certain threshold may degrade sensory content, possibly contributing to the disability of select patients.

Treatment

In many cases of visual snow, patients are treated similarly to those with migraine aura, often with limited or no success [8•, 51]. Similar to our cases 1 and 2, patients may achieve good control over the migraine without any improvement in their visual snow symptoms [51]. Some migraine medicines may even worsen visual snow symptoms as there has been one case of a patient's visual snow intensity consistently worsening approximately 2 h to 1 day after taking triptans (case presented in poster form at the American Headache Society meeting in 2016) [69].

There have been some cases of patients with partial attenuation of visual snow symptoms with lamotrigine [54, 70], but this has not been consistently effective [71]. Evans and Aurora reported a case of visual snow with partial improvement on topiramate [9] and Liu et al. reported some attenuation of visual snow symptoms with sertraline [3•]. A combination of nortriptyline and carbamazepine helped palinopsia but not the other visual symptoms in one patient with visual snow [3•]. Though not specifically reported to be helpful for visual snow, there are some cases of persistent visual aura responsive to acetazolamide and calcium channel blockers [51]. Based on the limited information available, Bou Ghannam et al. have recommended considering lamotrigine and possibly acetazolamide as first-line options to try with visual snow, followed by verapamil [51].

In addition to pharmacological therapies, there may be a potential role for individualized colored prescription glasses in treatment of visual snow. One study offered 12 participants with visual snow various lenses to see if a particular color filter improved their symptoms. Ten of these patients felt their symptoms were partially relieved with filters in the yellow-blue spectrum [4•]. As the koniocellular neurons in the thalamic LGN are activated by blue, the authors hypothesized that the blue-yellow filters may alleviate symptoms by altering an underlying imbalance between the koniocellular and magnocellular/parvocellular pathways, possibly helping (directly or indirectly) with the presumed thalamocortical dysrhythmia underlying visual snow [4•]. Interestingly, there is suggestion that similar tinted lenses may be useful in other disorders of cortical hyperexcitability, such as migraine and photosensitive epilepsy [72, 73].

Conclusion

When seeing a patient with visual snow, it is important to rule out a potential secondary etiology. If ophthalmologic and neurologic testing is normal, and the patient has not ingested psychotropic drugs, the diagnosis of visual snow syndrome should be considered. The patient should be assessed for other visual symptoms such as photophobia, palinopsia, nyctalopia, or other persistent positive visual symptoms. The pathophysiology of this syndrome is not clear, but recent neurophysiologic and neuroimaging studies have helped advance our understanding. Unfortunately, therapeutic options for this syndrome are limited, but it may be reasonable to try a trial of lamotrigine and/or individualized tinted lenses.

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Compliance with Ethical Standards

Conflict of Interest Abby I. Metzler declares no conflict of interest.
Carrie E. Robertson has served on advisory boards for Amgen and Alder.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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This is a very well-written review of what we understand about the pathophysiology and characterization of visual snow.

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