Pediatric Visual Snow Syndrome (VSS): A Case Series

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ABSTRACT

Visual snow syndrome (VSS) is a relatively rare, unusual, and disturbing abnormal visual condition. The individual perceives “visual snow” (VS) throughout the entire visual field, as well as other abnormal visual phenomena (e.g., photopsia). Only relatively recently has treatment been proposed (e.g., chromatic filters) in adults with VSS, but rarely in the pediatric VSS population (i.e., medications). In this paper, we present three well-documented cases of VSS in children, including their successful neuro-optometric therapeutic interventions (i.e., chromatic filters and saccadic-based vision therapy).

In addition to the presence of VS, and based on currently evolving criteria, the individual must also report two or more of the following to be formally diagnosed with VSS: palinopsia, enhanced entoptic imagery, “nyctalopia”, and photophobia/photosensitivity. These individuals also frequently report having migraine, photopsia, phonophobia, hyperacusis, cutaneous allodynia, tinnitus, balance problems, and tremor.

Due to its presumed extremely low prevalence, there is a relative paucity of reports in the literature on VSS in adults dating back to 1944, and only three case reports in children and adolescents ages 11-14 years over the past few years. In

Figure 1: Cat with overlay of visual snow as depicted by a patient.
these three cases, all conventional medical diagnostic testing was negative (e.g., brain imaging\textsuperscript{15}), and all attempted drug treatments (e.g., sumatriptan\textsuperscript{15}) were unsuccessful. None of the reports in the pediatric literature noted any successful treatments such as chromatic filters or vision therapy. Neither were there any successful medications directed solely at VS, but rather only medications toward migraine symptoms or other coincident symptoms.

Thus, we present a neuro-optometrically-based, pediatric case series (ages 10-15 years) of three patients with well-documented VSS. This report includes both valuable diagnostic and therapeutic aspects for the clinician.

**CASE PRESENTATIONS**

All received a comprehensive optometric vision examination, including refractive, binocular, and ocular health status. The case descriptions will be restricted to the relevant VSS details, as well as the other primary visual diagnoses. The key VSS findings are presented in Table 1 for each patient.\textsuperscript{12}

**Case One**

The patient was a 10-year-old male who first noticed VS soon after an extended bout (~4 weeks) of pneumonia for which he was successfully treated five months prior to his sensorimotor evaluation. The perception of VS was constant and typically monochromatic in nature. His primary VSS-related symptoms included photosensitivity and enhanced entoptic imagery, whereas his secondary VSS-related symptoms included balance problems and cutaneous alldynia. In addition, his other symptoms included intermittent diplopia.

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<td>Age first noticed snow (in years)</td>
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<td>Is visual snow constant (C) or transient (T)?</td>
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<td>Is visual snow chromatic (C) or monochromatic (M)?</td>
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<td>Provocative environments?</td>
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<td>Tinnitus</td>
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<tr>
<td>Balance problems</td>
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<td>Tremor</td>
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was taking nortriptyline (25 mg, 3 times per
day) for depression and leg pain.

The patient was referred by her neuro-
ophthalmologist to the SUNY/University Eye
Center, Vision Rehabilitation Service specifically
for a Cerium colorimetric evaluation. The
basic vision examination revealed esophoria
at distance, low hyperopia, and saccadic
dysfunction, for which she received successful
oculomotor-based vision therapy; that is,
symptoms reduced, and signs improved.
No refractive correction was prescribed at
that time. Then, the Cerium-based (Intuitive
Colorimeter)\textsuperscript{16} colorimetric testing
was performed. She required two different
spectacle tints. One was for lessening the VS,
which reduced it by approximately 75% (pink-
purple, hue = 320, saturation = 25, Purple
A6, Purple Rose A6, Rose C4, 45% overall
transmission). The other was for reducing
the palinopsia duration when reading, which
decreased the visual persistence from 17
seconds to 5 seconds, and thus it improved her
reading comfort (turquoise/green, hue=180,
saturation=25, turquoise C3, green C3, 47%
overall transmission).

**Case Two**

The patient was a 15-year-old female who
first noticed VS at the age of 11 years, after
experiencing her first ocular migraine attack.
The perception of VS was transient and
chromatic. It could be precipitated by bright
lights, fatigue, and computer screen viewing.
Her primary VSS-related visual symptoms included photosensitivity and palinopsia (especially when reading), whereas her secondary VSS-related symptoms included photopsia and balance problems. In addition, her other symptoms included blurred vision, headaches, and loss of place, all when reading, as well as the sensation of
dizziness and not feeling “grounded”. Prior
visual fields and dilated fundus examinations
by two ophthalmologists were normal. She

**Case Three**

The patient was a 15-year-old female who
first noticed VS at the age of 11 years, after
experiencing her first ocular migraine attack.
The perception of VS was transient and
chromatic. It could be precipitated by bright
lights, fatigue, and computer screen viewing.
Her primary VSS-related symptoms included
palinopsia with trailing a few times per day,
enhanced chromatic entoptic imagery once
per day, and constant photosensitivity, whereas
her VSS-related secondary symptom was
intermittent photopsia. The VS and palinopsia
occurred independently. She also reported two
other interesting perceptual phenomena. First,
at times, hallways were perceived to be longer,
and objects appeared to be either closer (i.e.,
pelopsia) or farther (i.e., teleopsia) than their
physical presence, with such size and distance distortions suggesting Alice in Wonderland Syndrome. Second, at times, she noticed a strobe/pulse effect surrounding letters during reading. She had no history of concussion and was not taking any medications.

The patient was self-referred to a private practice, neuro-optometrist for a comprehensive vision evaluation. She was diagnosed with accommodative insufficiency, convergence excess, saccadic oculomotor dysfunction, visual-directional problems, and visual-memory deficits. She was successfully treated with oculomotor-based vision therapy and visual perceptual therapy; that is, symptoms reduced, and signs improved. The saccadic therapy reduced the palinopsia persistence and related trailing, presumably by reestablishing a normal saccadic suppression inhibitory level. In addition, an FL-41 tint was prescribed in spectacle form, which reduced the perception of VS.

**DISCUSSION**

VSS represents a new and exciting challenge to the neuro-optometrist, and others (e.g., the ophthalmologist, neurologist). This syndrome has been a long misunderstood, somewhat perplexing, unique, and even questioned (especially in young children) diagnosis among the medical community, at least since 1944 when visual snow was reported by some adult patients taking digitalis for heart problems.

The neuro-optometrist has expertise in general vision function/dysfunction, with specialty knowledge in optics (e.g., tint spectra, lens design), human visual perception (e.g., motion, saccadic suppression, reading), and vision therapy/neurorehabilitation (e.g., perceptual and motor learning, oculomotor control). Thus, the neuro-optometrist can provide the comprehensive, basic vision examination (i.e., refractive binocular, and ocular health), as well as the vision therapy, and also both the general (e.g., neutral gray) and more advanced (e.g., FL-41, BPI Omega) tint prescribing. In addition, an ever increasing number of optometrists have knowledge and experience in specialized colorimetric testing (i.e., Cerium Intuitive Colorimeter) to prescribe the optimal filter combination for maximum visual comfort and benefit (e.g., reading efficiency) in the patient with VSS. This would especially include the visual symptoms of VS, palinopsia, and photosensitivity/photophobia. Lastly, others may be assistive in the process, such as the occupational therapist and ophthalmologist, to assess vision and related gains with the therapeutic lenses within the context of their own discipline.

Treatment for VSS is important for all individuals, but perhaps even more so in young children/adolescents, for several reasons. First, there is the possible psychological effect of having such unusual and abnormal, undiagnosed, visual percepts. Some adult patients with VSS indicated their initial childhood reservation about telling this to a friend or family member, for fear of disbelief and even possible ridicule. Therefore, a detailed, probing case history may be required to elicit the presence of VS in some patients, especially in those conditions with a relatively high probability of occurrence such as concussion, brain surgery, and taking a new medication. Second, those manifesting palinopsia, which seems to be the majority, frequently find presence of their sustained and superimposed afterimage, especially with trailing, to be both distracting and disruptive when reading, as expected, thus likely leading to reduced reading rate, reduced visual comfort, and poorer comprehension. And lastly, as they become of driving age, the presence of VSS, palinopsia, and/or enhanced entoptic imagery, especially at night, may impact adversely on their driving ability and safety due to impaired visual perception and attentional distraction. If so, they should be advised to obtain counselling from a professional driving trainer with expertise in those having visual impairment.
In an earlier paper on this topic,\textsuperscript{12} we had proposed a “unifying hypothesis”, or mechanism, to understand many/all aspects of VSS, namely neural “disinhibition”. Others had proposed the mechanism to be a “hypersensitivity phenomenon”.\textsuperscript{8} At first blush, this appeared to be descriptive. While not necessarily incorrect, it seems to emphasize the wrong aspect. These patients are not hypersensitive per se, but rather appear to exhibit abnormally-reduced levels of naturally-occurring neural inhibition/have less suppression. This idea is supported by recent laboratory findings.\textsuperscript{21,22} Thus, the patient with VSS sees/experiences what others do not see (e.g., palinopsia) or feel (e.g., cutaneous allodynia).

In the future, a randomized clinical trial (RCT) is warranted to assess efficacy of the two current neuro-optometric, therapeutic interventions, namely tinted spectacles and oculomotor-based vision therapy, separately and combined, in conjunction with a placebo condition.\textsuperscript{23} However, a cross-over, interventional experimental design would also be both acceptable and more efficient.\textsuperscript{21} If proven to provide symptomatic relief (e.g., per the VSS questionnaire)\textsuperscript{12} and/or increased visual/reading efficiency (e.g., per the objective Visagraph reading testing),\textsuperscript{24} additional studies would be helpful to determine retention of the long-term benefits and any visual adaptation effects.

\textbf{REFERENCES}

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Kenneth J. Ciuffreda received his B.S in biology from Seton Hall University in 1969, his O.D. from the Massachusetts College of Optometry in 1973, and his Ph.D. degree in physiological optics from the University of California/School Optometry at Berkeley in 1977. He has been a faculty member at the SUNY/State College of Optometry in New York City since 1979, where he is presently a Distinguished Teaching Professor. He has also had adjunct appointments for many years at Rutgers/ The State University of New Jersey, as well as at the New Jersey Institute of Technology, both in the department of biomedical engineering. He also helped establish a school of optometry in Harbin, China. He has conducted research in many areas: amblyopia, strabismus, reading, myopia, eye movements, accommodation, bioengineering applications to optometry, and more recently with an emphasis in the area of acquired brain injury, both the diagnostic and therapeutic aspects. His goal has been the use of objective recording techniques in the diagnosis and treatment of neurological and ocular conditions. He holds two patents, and has received many awards and honors from the AAO, AOA, NORA, COVD, and various state optometric associations and colleges. He has authored over 400 research papers/ chapters, and 10 books. His hobbies are playing jazz guitar and enjoying the visual aspects of art.