

Visual Snow Syndrome

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Financial Disclosures

- No relevant disclosures

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Case presentation

- 14 yo WF
- Cc: “pixelated” vision
 - TV static in her entire visual field
 - 24/7
 - Afterimages
 - Photophobia
- PMH
 - Migraine headaches

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Case

- Va 20/20 OU
- Pupils normal, no APD
- Ext'l/SLE normal
- Motility normal, ortho
- HVF normal
- IOP normal
- Fundi normal

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Case

- Cc: "Pixelated" vision, TV static
- Dx?
- Work-up?
- Treatment?



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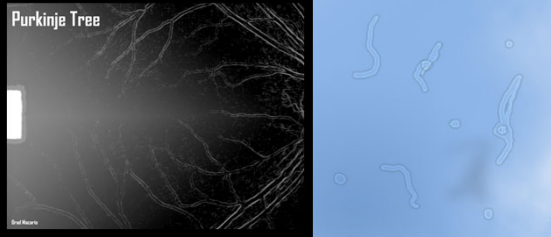
DDX: Spots & Dots

- Diagnosis – benign vs not-so-benign
- Benign
 - Vivid imagination
 - Normal entoptic phenomenon
 - Vitreous floaters
 - Purkinje tree
 - Blue field phenomenon

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Entoptic phenomenon

- Visualize an internal structure in the eye



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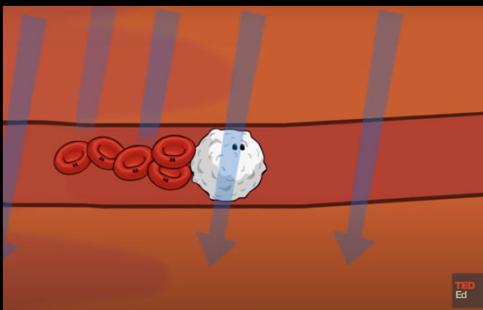


Blue Field Entoptic Phenomenon
Scheerer's phenomenon
Blue sky sprites

By Unimemodjefin - Own work, CC BY-SA 3.0,
https://commons.wikimedia.org/wiki/File:Sky_sprites_27200667

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Blue Field Entoptic Phenomenon



https://youtu.be/Y5a_m0by4Q

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Entoptic Phenomenon: Phosphenes

no light stimulus
on of the retina

Backlightingcenter.com

DukeUniversity.edu

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DDX: Spots & Dots

- Diagnosis – not-so-benign
 - Migraine
 - Occipital lobe epilepsy/lesion
 - Paraneoplastic/autoimmune retinopathy
 - White dot syndromes

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Migraine Visual Aura

KobaArt.com

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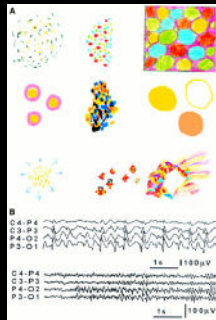
Migraine Visual Aura



[Migrainebuddy.com](http://migrainebuddy.com)

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Occipital lobe epilepsy/lesion



P Panayiotopoulos. J Neurol Neurosurg Psychiatry 1999; 66: 536-540

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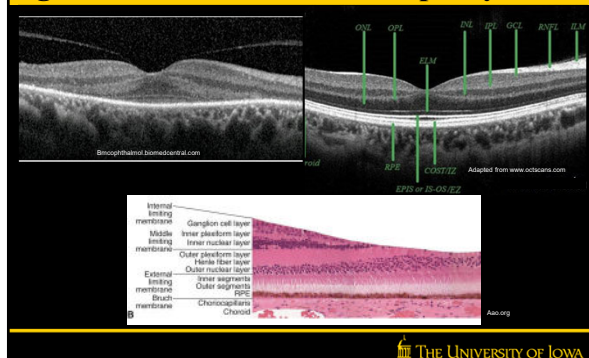
Paraneoplastic/AI retinopathy



<https://www.youtube.com/watch?v=9CCGPFUad10>

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Paraneoplastic retinopathy: e.g., cancer associated retinopathy



White Dot Syndromes e.g., multiple evanescent white dot syndrome

- Photopsias



EYE FOUNDS.ORG

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Visual Snow: Aeropsia

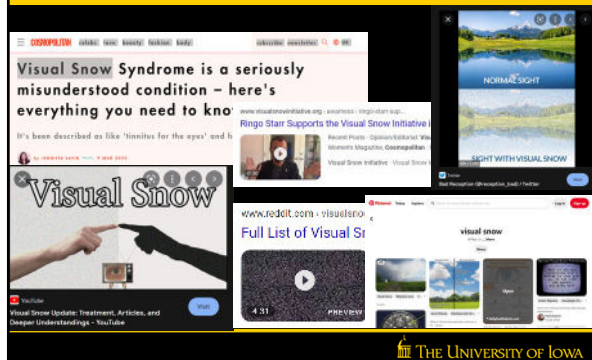
- TV static
- Constant 24/7
- Flickering dots/lights
- Superimposed on environment



Twitter.com

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Visual Snow: Who Knows About It?



Visual Snow



Visual Snow



Visual Snow

- <http://www.visionsimulations.com/>

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Symptoms of Visual Snow

- A million dots
- Black cracks and lines
- Blobs of white and gray
- Blue squares
- Carpet background
- Circles
- Clouds
- Comets
- Dots, b/w & colored
- Grainy vision
- Heat waves
- Lights, flash/flicker
- Lines of ants
- Photopsias
- Rain-like pattern
- Snow
- Squiggles
- TV static

Liu et al. Neurology 45:854-858, 1995

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Visual Snow: many names

- Prolonged migraine aura status (Haas 1982)
- Sustained visual aura
- Persistent positive visual phenomena (1995)
- Persistent migraine aura
- Migraine aura status
- Persistent migrainous visual phenomena
- Persistent migraine aura w/o infarction (IHC 2003)
- Primary persistent visual disturbance (2005)
- Visual snow (2014)

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Persistent Aura w/o Infarction

- Persistent aura without infarction
 - Aura symptoms > 1 week w/o infarction
 - Rare
 - Bilateral
 - Lasts months to years
 - Most have h/o migraine or variant

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Primary Persistent Visual Disturbance

doi:10.1111/j.1468-2982.2004.00858.x

Diffusion- and perfusion-weighted MR imaging in persistent migrainous visual disturbances

HR Jäger¹, NJ Giffin & PJ Goadsby

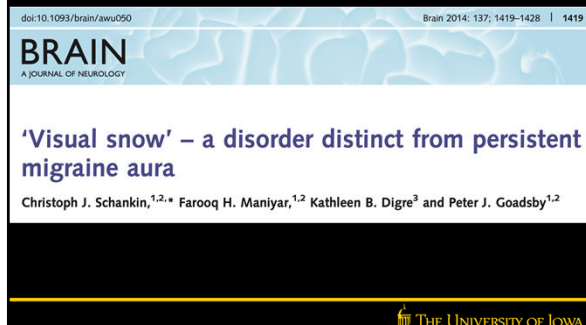
¹Headache Group and ²Division of Neuroendocrinology, Institute of Neurology, The National Hospital for Neurology and Neurosurgery, London, UK

Cephalalgia

Jäger HR, Giffin NJ & Goadsby PJ. Diffusion- and perfusion-weighted MR imaging in persistent migrainous visual disturbances. *Cephalalgia* 2005; 25:323-332. London. ISSN 0333-1024

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Visual Snow



Visual Snow Syndrome: Criteria

- Dynamic, continuous, flickering tiny dots
- Entire visual field
- 2 of the following 4
 - Enhanced entoptic phenomenon
 - Photophobia
 - Nyctalopia
 - Palinopsia

Curr Opin Neurol 2018;31:55-58 THE UNIVERSITY OF IOWA

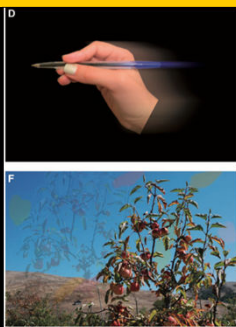
VSS: Criteria

- Entoptic phenomenon
 - Floaters 81%
 - Blue field entoptic 79%
- Photophobia 74%
- Nyctalopia 68%

JNO 38(4):514-521, 2018 THE UNIVERSITY OF IOWA

Palinopsia

- Perseveration of visual image
- Stimulus gone
- Different than normal retinal afterimage

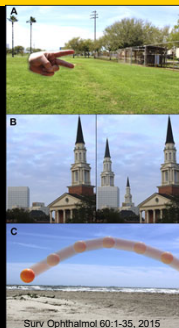


1422 | Brain 2014; 137: 1419-1428

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Palinopsia

- Palinopsia 69%
- Stationary 86
- Trailing moving objects 60



Surv Ophthalmol 60:1-35, 2015

JNO 38(4):514-521, 2018

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VSS: Criteria

- Other frequent & persistent visual phenomenon
- Halos
- Metamorphopsia
- Colored clouds
- Water running down a window

Curr Neurol Neurosci Rep 2018; 18:52
Brain 2014; 137: 1419-1428

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Visual Snow



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VSS: Demographics

- Web-based survey of 1,104 pts w/VS
- Xage 29
- M=F
- Onset early in life
- All of their life 40%

Neurology 2020;94:e564–574. doi:10.1212/



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VSS: Comorbidities

- Comorbidities
 - Migraine 60%
 - Often severe migraine triggers
 - Tinnitus 62%
 - Concentration difficulties 60%
 - Anxiety 44%
 - Depression 19%

JNO 38(4):514-521, 2018

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VSS: Testing normal

- Eye and neuro examination
- Ancillary testing: VF, OCT, ERG, VEP
- MRI
- Mistakenly considered psychogenic

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VSS: Pathophysiology

- Unknown
- Bilateral, symmetric, constant, spacing
- Cortical
- Normal?
 - Visual noise?
 - Similar to tinnitus?
 - Result of abnormal inhibition?

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VSS: Pathophysiology

- Spontaneous: majority (57.6%)
- Inciting events
 - Trauma
 - Change in migraine/aura
 - Illness
 - Hallucinogen persisting perception d/o
 - Associated eye ds, e.g., RP, IIH

Cephalalgia 41:1015-1026, 2021

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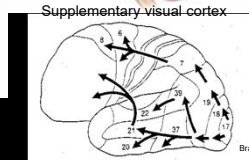
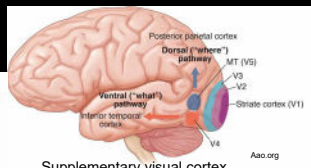
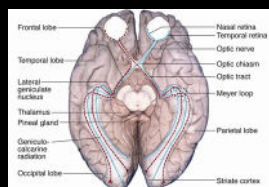
VSS: Pathophysiology

- 3 major theories:
 - Hyperexcitability
 - Hypoconnectivity
 - Thalamocortical dysfunction
 - Combination of all three

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VSS: Pathophysiology

Theories



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VSS: Pathophysiology

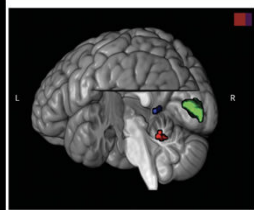
- Cortical hyperexcitability
 - Magnetoencephalography
 - Functional MRI abnormalities
 - Hypermetabolism on PET
 - Hypometabolism on SPECT
 - Elevated spontaneous neural firing
 - Inconsistent studies VEP and EEG



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VSS: Pathophysiology

Figure 3 Summary of cortical volumetric changes in patients with visual snow syndrome, as shown by voxel-based morphometry



Render illustration of the 3 brain regions of increased gray matter volume in patients with visual snow syndrome with respect to healthy controls. Left V1 cluster is illustrated in green, left V2 cluster in blue, left cerebellum cluster in red. For statistical values of each area, see the text. Image was created in MNI-COL and superimposed on standard brain template.

Neurology 95(13):e1702-31708, 2020

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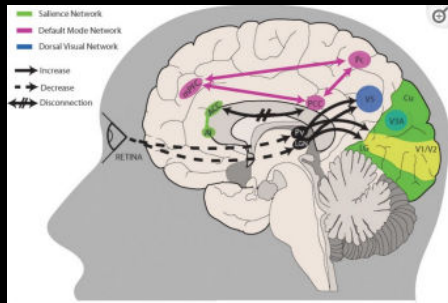
VSS: Pathophysiology

- fMRI connectivity whole brain map (BOLD)
- Hyperactive neural connectivity
 - e.g., V1 to V5, pulvinar to lingual gyrus
- Hypoactive
 - e.g., salience network
 - Resulting in awareness of stimuli that most would consider irrelevant

Hum Brain Mapp. 2021;42:2032-2044

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VSS: Pathophysiology



Front Neurol, 2019; 10: 1325

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VSS: Treatment

•Review of the literature

Medication	Overall trials	Ineffective	Effective (total response)
Lamotrigine	36	28	8 (1)
Topiramate	13	11	2
Valproate	12	11	1 (1)
Propranolol	6	5	1
Sertraline	5	4	1
Verapamil	3	2	1
Naproxen	2	1	1
Baclofen	1	0	1

Prog Brain Res 2020;255:311-316

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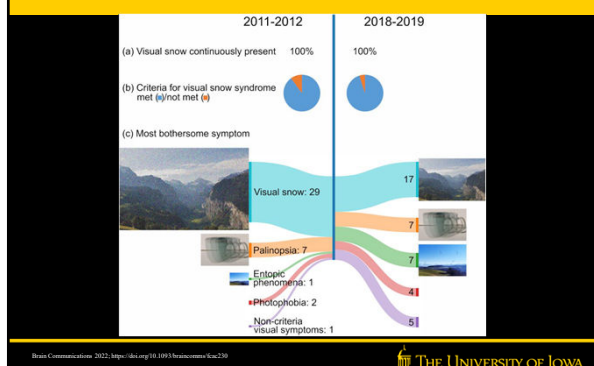
VSS: Treatment

- Reassurance and validation
- Colored filters (blue 80% pts)
- Education
 - Visionsimulations.com
 - Visualsnowinitiative.org
 - Wikipedia.com
 - Eyeonvision.org

Neurology 2019;93:e2000403
J Clin Neurosci 2016; 28:123-127

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Prognosis



VSS: Over the years experience

- 100 patients over 30 years
- Patients wide age range, most 20-30 yrs
- Highly observant, articulate
- Symptoms stereotypical
- Examinations normal
- Treatments unsuccessful
- Tale of a neurologist

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VSS: Conclusions

- Visual snow: it's real
- History is key, exam normal
- Reassurance
- Education



palinopsia.worldpress.com

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VSS: Conclusions

- Criteria
 - Bilateral constant flickering tiny dots
 - 2 of the following 4
 - Palinopsia
 - Enhanced entoptic phenomenon
 - Photophobia
 - Nyctalopia

paderospha.wordpress.com

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VSS: Conclusions

- DDX

Benign	Not-so-benign
Entoptic Phenomenon	Migraine
Phosphenes	Seizure
	Paraneoplastic/AIR
	White dot syndrome
- Treatment
 - Reassurance
 - Education
 - Visualsnowinitiatives.org
 - Visionsimulations.com
 - Eyeonvision.org

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