Noninvasive Vagus Nerve Stimulation in a Pediatric Patient with Visual Snow Syndrome

Trevor Gerson, MD; Jennifer Bickel, MD

Children's Mercy Kansas City

Background

- Visual Snow Syndrome (VSS) consists of symptoms such as constant visual static, photophobia, poor night vision (nyctalopia), and after images (palinopsia), and entoptic phenomena such as visual floaters, experienced for > 3 months¹ Proposed mechanisms include cortical hyperexcitability, specifically of the lingual
- gyrus²
- There are currently only a few therapies reported in the literature which may be helpful in the treatment of VSS, including lamotrigine and valproic acid, which may have significant side effects³
- Noninvasive Vagus Nerve Stimulation (nVNS) is FDA-approved for the treatment and prevention of migraine and cluster headache that, among other mechanisms, decreases cortical excitability⁴ (Figure 1)
- Given that the proposed mechanisms of VSS overlap with nVNS, it was hypothesized that nVNS may be efficacious in this disorder, and that the benefits may significantly outweigh any risks



Figure 1. Systems, mechanisms of action, and clinical effects of nVNS in headache. Abbreviations: ANS, autonomic nervous system; CSD, cortical spreading depression; NT, neurotransmitter; nVNS non-invasive varuus never simulation: "CIM trigeminal.⁵

LOVE WILL.

Patient Case

- A 14 year old patient presented to the Headache Clinic with a history of Cyclic Vomiting Syndrome (CVS) and was found on further questioning to fit diagnostic criteria for VSS
- Symptoms included constant snow in her vision, palinopsia, nyctalopia, and photophobia, with difficulty reading music and using graph paper in school
- After a trial in the clinic of a single set of 2 stimulations bilaterally, she had significant improvement in VSS symptoms for 24 hours
- nVNS was trialed daily, with two stimulations of two minutes each bilaterally. This was continued for 5 months
- The patient developed a 10 point severity scale for VSS, with an average intensity of 7.6 prior to using Gammacore and a minimum of 4.2 after an individual use (sustained at a 4-5 daily).
- She was able to read music and use graph paper easier during this period. Nyctalopia improved, as well.
- No changes in photophobia or palinopsia
- Average intensity of stimulation was 23-26, with a maximum of 35.
- No adverse events were experienced
- Notably, there was no effect on Cyclic Vomiting Symptoms, in terms of preventing or aborting an episode
- The patient did not wish to continue use of the device after the trial as she had difficulty adjusting to not having her visual snow symptoms, as well as lack of improvement in quality of life related to CVS

Discussion

- This patient had significant benefit in her visual snow symptoms with the use of a noninvasive device, and without any significant adverse effects, though no effect on CVS
- The benefit seen in this patient lends credence to theories of VSS
 pathophysiology
- Larger prospective trials would be needed to confirm efficacy in VSS
- This case raises the question of tolerance of treated visual snow syndrome
 and the need for transition to normalized vision
- Symptom-severity scales would be necessary for further research

Conclusion

- Noninvasive Vagal Nerve Stimulation may be an effective
 - treatment for Visual Snow Syndrome

Acknowledgements

• The authors would like to thank electroCore, Inc for providing the devices used in this study.

References

- 1. The International Classification of Headache Disorders, 3rd edition (beta version). Cephalalgia.
- 2013;33(9):629-808
- Puledda F, Schankin C, Digre K, Goadsby PJ. Visual snow syndrome: what we know so far. Curr Opin Neurol. 2018;31(1):52-58.
- Traber GL, Piccirelli M, Michels L. Visual snow syndrome: a review on diagnosis, pathophysiology, and treatment. Curr Opin Neurol. 2020;33(1):74-78.
 Chen SD, Aul de Mercie AL, et al. More a series dissublished appreciate patient of the series of the series
- Chen SP, Ayl, de Morais AL, et al. Vagus nerve stimulation inhibits cortical spreading depressic 2016;157(4):797-805. doi: 10.1097/j.pain.000000000000437.
- Silberstein S, Yuan H, Najib U, et al. Non-invasive vagus nerve stimulation for primary headache: a clinical update. Cephalalgia. Under review, with permission



