

## Vagal Nerve Stimulator as a Treatment Approach in CHARGE Syndrome: A Case Report

**Authors :** Roya Vakili, Lekaa Elhajjmousa, Barzin Omid-Shal, Kim Blake

**Abstract :** Objective: The purpose of this case report is to highlight the successful treatment of a patient with Coloboma, Heart defect, Atresia choanae, Retarded growth and development, Genital hypoplasia, Ear anomalies/deafness, (CHARGE syndrome) using a vagal nerve stimulator (VNS). Background: This is the first documented case report, to the authors' best knowledge, for a patient with CHARGE syndrome, epilepsy, autism, and postural orthostatic tachycardia syndrome (POTS) that was successfully treated with an implanted VNS therapeutic device. Methodology: The study is a case report. Results: This is the case of a 24-year-old female patient with CHARGE syndrome (non-random association of anomalies Coloboma, Heart defect, Atresia choanae, Retarded growth and development, Genital hypoplasia, Ear anomalies/deafness) and several other comorbidities including refractory epilepsy, Patent Ductus Arteriosus (PDA) and POTS who had significant improvement of her symptoms after VNS implantation. She was a VNS candidate given her longstanding history of drug-resistant epilepsy and current disposition secondary to CHARGE syndrome. Prior to VNS implantation, she experienced three generalized seizures a year and daily POTS-related symptoms. She was having frequent lightheadedness and syncope spells due to a rapid heart rate and low blood pressure. The VNS device was set to detect a rapid heart rate and send appropriate stimulation anytime the heart rate exceeded 20% of the patient's normal baseline. The VNS device demonstrated frequent elevated heart rates and concurrent VNS release every 8 minutes in addition to the programmed events. Following VNS installation, the patient became more active, alert, and communicative and was able to verbally communicate with words she was unable to say prior. Her GI symptoms also improved, as she was able to tolerate food better orally in addition to her G and J tube, likely another result of the vagal nerve stimulation. Additionally, the patient's seizures and POTS-related cardiac events appeared to be well controlled. She had prolonged electroencephalogram (EEG) testing, showing no significant change in epileptiform activity. Improvements in the patient's disposition are believed to be secondary to parasympathetic stimulation, adequate heart rate control, and GI stimulation, in addition to behavioral changes and other benefits via her implanted VNS. Conclusion: VNS showed promising results in improving the patient's quality of life and managing her diverse symptoms, including dysautonomia, POTs, gastrointestinal mobility, cognitive functioning as well seizure control.

**Keywords :** autism, POTs, CHARGE, VNS

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