Chapter 8
Vagus Nerve Stimulation Therapy: An Intellectual Disabilities Perspective

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Introduction

Numerous studies have now been published confirming the high prevalence of epilepsy in people with intellectual disabilities (ID).\(^1\)\(^-\)\(^3\) McDermott and colleagues\(^3\) compared the rates of epilepsy between ID and non-ID groups living in the community. The non-LD group had an epilepsy prevalence rate of 1%. The prevalence of epilepsy within the LD group was 13% for cerebral palsy, 13.6% for Down syndrome (DS), 25.4% for autism, 25.5% for mental retardation, and 40% for adults with both cerebral palsy and ID. Further, the researchers found that during the decades of adulthood, the prevalence of epilepsy declined for those with cerebral palsy and ID. The prevalence of epilepsy increased with advancing years for adults with DS and autism. For each decade, the prevalence of epilepsy was higher in the ID group compared to the non-ID group.

A significant proportion of persons with ID will have intractable epilepsy. Antiepileptic drugs (AEDs) remain the principal form of management for intractable epilepsy with up to 40% of individuals on polytherapy.\(^4\)\(^-\)\(^5\) However, poor seizure control may still be evident, leading to the consideration of alternative treatments. The principal alternative form of treatment is psychosurgery, but this is not readily accessible to persons with ID. Vagus nerve stimulation (VNS) therapy has the potential to be the main practical and efficacious treatment alternative to AEDs for individuals with ID who suffer from intractable epilepsy. To date there is very limited information specifically focused on the role of VNS Therapy to treat epilepsy in the ID population. This review chapter examines the role of VNS Therapy in persons with ID.

Background

In 1985 Dr. Jacob Zabara proposed that electrical stimulation of the vagus nerve had potential benefits in seizure prevention.\(^6\) This hypothesis was based on observations from animal studies investigating the effects of VNS on electroencephalogram
(EEG) recordings. Vagus nerve stimulation was found to induce EEG synchronization, EEG desynchronization, and REM and slow wave sleep in animals, depending on the different stimulus parameters.\(^7\)\(^-\)\(^8\) As epileptic seizures are characterized by paroxysmal, abnormal synchronicity of the EEG, it was postulated that VNS could prevent seizure activity by desynchronizing the electrical activity of the brain.\(^6\) Subsequently, animal experiments confirmed the anticonvulsant efficacy of VNS\(^9\),\(^10\) and led to the development of an implantable VNS device by Cyberonics, Inc. (Houston, TX) for use in the treatment of patients with pharmaco-resistant epilepsy.

The first actual vagus nerve stimulator was implanted in a patient with epilepsy in 1988.\(^11\) Following open and randomized controlled trials (see Binnie\(^12\) for review) VNS Therapy was approved in Europe in 1994 and in United States in 1997 for use as an adjunctive therapy in reducing the frequency of seizures in patients whose epileptic disorder is dominated by partial seizures (with or without secondary generalization) or generalized seizures which are refractory to antiepileptic medications.

Vagus nerve stimulation therapy has dramatically changed the management of refractory epilepsy. It is now the most widely used nonpharmacological treatment and has been reported to be beneficial in reducing seizures, preventing seizures soon after VNS Therapy has been initiated; in reducing long-term seizure frequency, and improving quality of life. To date, over 20,000 patients have now had VNS implants for refractory seizures.

**The VNS Therapy System™**

The main component of the VNS Therapy System™ consists of a titanium-encased pulse generator (Figure 8.1) that delivers mild automatic intermittent electrical stimuli to the cervical portion of the left vagus nerve via a bipolar lead (the left vagus nerve has less cardiac regulation and therefore is used rather than the right vagus nerve). The pulse generator contains a battery and programmable computer chips. The stimulus is transmitted via the vagus nerve to the brain. The appropriate settings for the stimulation are determined by a telemetric wand (Figure 8.1), which is held over the pulse generator and controlled by computer software. Patients and/or caregivers can control the degree of stimulation (and side effects) by a hand-held magnet (Figure 8.1).

There have been improvements in VNS design and in the duration battery life of the pulse generator since the first commercially available model. The current device, model 102 (6.9 mm, weight 25g), is less bulky than the original NCP 100 model, resulting in a better cosmetic result following implantation and longer battery life. Depending on stimulus parameters and use, the pulse generator can now run for 6 to 11 years. However, once the battery is depleted, the entire pulse generator must be removed and replaced. The implantation procedure takes approximately one hour and is usually carried out under general anesthesia so as to limit the risk