# Cochlear™ Nucleus® System
# Single-Sided Deafness/Unilateral Hearing Loss

The FDA approved the Cochlear™ Nucleus® for the treatment of unilateral hearing loss/single-sided deafness in January 2022. Single-sided deafness (SSD) is severe to profound hearing loss in one ear and normal or near normal hearing in the other ear. Unilateral hearing loss (UHL) is hearing loss in one ear and near normal hearing in the opposite ear.

**Cochlear Americas is requesting the HERC revise the guideline note#31, cochlear implantation, to include individuals with a diagnosis of UHL/SSD.**

A comprehensive discussion and table of the clinical evidence for adults and pediatrics was submitted to the HERC staff. This document provides a condensed and edited version of the more extensive dossier.

## Rationale for Revising Guideline Note 31

- CIs have been used successfully to treat bilateral sensorineural hearing loss for more than 30 years. The well-established safety, efficacy and success of cochlear implantation has led to changes in criteria and expansion in indications to include individuals with greater residual hearing, such as those who receive electric & acoustic stimulation devices (2014), and most recently for patients with SSD (2022).
- Traditional thinking presumes only one normal-hearing ear is necessary for developing spoken language. Recent studies have shown that even mild degrees of UHL/SSD have adverse effects on language development (Lieu 2018).
- Risks of education challenges particularly in children, e.g., higher rates of repeating grades, negative cognitive effects, behavioral misunderstandings, demands for higher concentration for listening, and reduced quality of life, have been documented in this population (Kuppler et al., 2013; Wie et al., 2010; Zeitler et al., 2019)
- SSD can impact psychosocial factors such as self-esteem, self-confidence and an individual's sense of security (Borton et al., 2010).

There are three primary benefits patients obtain related to auditory perception seen only in patients with binaural hearing: binaural summation effect, binaural squelch effect and the head shadow effect (Arndt et al., 2011). These effects are defined further:

- Binaural summation – a tone presented to two ears is perceived as louder than the same tone presented to one ear, creating an advantage and/or ease of listening and improved speech perception ability when both ears are provided with equivalent input
- The binaural squelch effect (also called the "cocktail party effect" ) – a signal in the presence of background noise is more easily detected when the signal and noise are presented to both ears versus one ear, thereby enhancing speech understanding ability
- The head shadow effect - a physical effect rather than a central processing mechanism - where the head acts as a barrier, softening the sound coming from one direction, creating an improved signal-to-noise ratio at the ear distant from the noise or vice-versa.

While there are conventional treatment options for UHL/SS, such as bone conduction hearing aids, bone anchored hearing aids, implantable bone conduction hearing aids, and CROS technologies, these options are unable to take advantage of the critical benefits of binaural hearing (Brown et al., 2021; Peters et al., 2021).

Also, those systems route the acoustic signals from the deaf ear to the normal-hearing ear. Therefore, they cannot reliably improve sound localization or hearing in noisy listening environments given that the information is received and processed by the listener through one auditory pathway.

These shortcomings prevent the recipient's ability to restore binaural (two ear) hearing. The listener does not regain the necessary cues to enhance speech understanding in noise (e.g., interaural timing and level difference cues) and/or locate sound. A cochlear implant, on the other hand, has the potential to provide true binaural input, with demonstrative benefits in speech perception, localization, patient satisfaction and quality of life, (Firszt et al., 2018; Arndt et al., 2017; Brown et al., 2021; Buss et al., 2018; Sladen et al., 2017).

## FDA Indications for UHL/SSD Hearing Loss

The expanded device indications include individuals with UHL/SSD who meet the following criteria:

- Individuals ≥5 years who have one ear with a severe to profound sensorineural hearing loss and obtain limited benefit from an appropriately fitted unilateral hearing device and one ear with normal or near normal hearing.
- Limited benefit from an appropriately fit unilateral hearing device is defined as a score of ≤5% on a Consonant-Nucleus-Consonant word test. For individuals between 5 and 18 years of age, insufficient functional access to sound in the ear to be implanted must be determined by aided speech perception test scores of ≤5% on developmentally appropriate monosyllabic word lists when tested in the ear to be implanted alone.
- Failed trial of at least 2 weeks wearing appropriately fit Contralateral Routing of Signal (CROS) hearing aid or another suitable hearing device.

## Contraindications

A Cochlear Nucleus cochlear implant (CI) is unsuitable for individuals with the following conditions:

- Absence of cochlea development
- Absence of a cochlear nerve
- Active middle ear infections
- Tympanic membrane perforation in the presence of active middle ear disease.

For individuals with SSD the following contraindication is also applicable:

- Duration of profound sensorineural hearing loss >10 years

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Cochlear Americas appreciates the opportunity to submit information to the HERC for consideration. Additional information on cochlear implants and SSD may be found in the comprehensive dossier.