

<p>Hearing History</p> <p><i>Etiology</i></p> <ul style="list-style-type: none"> • Implantation should be pursued quickly when SSD is secondary to bacterial meningitis and in etiologies where better ear progression is a concern. • Cochlear nerve deficiency (CND) is a contraindication to cochlear implantation in SSD. <p><i>Duration of SSD</i></p> <ul style="list-style-type: none"> • At this time families should be counseled that those with longer durations of deafness may experience less benefit than those with shorter durations of deafness. <p><i>Tinnitus</i></p> <ul style="list-style-type: none"> • Adult studies suggest reduction in tinnitus following implantation in cases of SSD but is not well studied in children. <p><i>Experience with alternative hearing technologies</i></p> <ul style="list-style-type: none"> • A trial period with a re-routing device is only recommended when the goal is to mitigate the impact of SSD without providing bilateral auditory input. <p><i>Age at cochlear implantation</i></p> <ul style="list-style-type: none"> • Auditory neural plasticity is known to be greatest during early childhood. Reducing the period of auditory deprivation by implanting early is likely in a child's best interest.
<p>Imaging</p> <ul style="list-style-type: none"> • High resolution 3D MRI is essential to diagnose CND, which has a high prevalence in this population.
<p>Subjective Benefit</p> <ul style="list-style-type: none"> • The following questionnaires are recommended for pre-operative and post-activation evaluation of subjective benefit. Tests should be chosen based on age and targeted behaviors. <ul style="list-style-type: none"> ○ Auditory Skill Development: Parents' Evaluation of Aural/Oral Performance of Children (PEACH) ○ Quality of Life: HEAR-QL ○ Hearing in Everyday Listening Environments: Speech, Spatial, and Qualities of Hearing (SSQ) for parent, child, and/or teacher ○ Academic Performance: Screening Instrument for Targeting Educational Risk (SIFTER) ○ Localization: SSQ for parent, child, and/or teacher or the Auditory Behavior in Everyday Life (ABEL) ○ Tinnitus: Tinnitus Handicap Inventory (THI) or the Tinnitus Functional Index (TFI)
<p>Audiologic Assessment in Order of Priority</p> <p><i>Pre-operatively</i></p> <p>Threshold Assessment</p> <ul style="list-style-type: none"> • Complete behavioral measurement of unaided hearing detection thresholds. • Objective testing for very young children and those who are unable to be tested behaviorally.

Spatial hearing: Speech in Noise (SIN)

- Assess in an unaided condition and/or with alternative hearing technology.
- Test configuration, both with and without device:
 - S_0N_0 : Target and masker presented at 0-degrees azimuth.
 - S_0N_{ci} : Target presented at 0-degrees azimuth and masker presented 90-degrees to the affected ear.
 - S_0N_{contra} : Target presented at 0-degrees azimuth and masker presented 90-degrees to the normal-hearing ear.
- Recommended test materials: See Table 1.

Word Recognition at 60 dBA

- In the sound field unaided
- In the sound field with hearing aid or alternative technology
- In the sound field with the hearing aid alone if applicable or required by insurance, with the contralateral ear masked via insert phone.
- Recommended test materials: See Table 1.

Post-Activation

Threshold Assessment

- Complete behavioral measurement of unaided hearing detection thresholds at regular intervals.
 - This is vital for the normal-hearing ear to assess hearing stability.
 - Recommended for the implanted ear to monitor hearing preservation.
- Assess sound field thresholds with the CI isolated.
 - Plug the contralateral ear and place a circumaural earmuff over the pinna.
 - Thresholds should be screened 10- 15 dB above where CI thresholds were obtained without the speech processor to ensure responses are not from the plugged and muffed ear.

Spatial hearing: SIN

- Test configuration, both with and without device:
 - S_0N_0 : Target and masker presented at 0-degrees azimuth.
 - S_0N_{ci} : Target presented at 0-degrees azimuth and masker presented 90-degrees to the affected ear.
 - S_0N_{contra} : Target presented at 0-degrees azimuth and masker presented 90-degrees to the normal-hearing ear.
- Recommended test materials: See Table 1.

Word Recognition

- Isolate the affected ear from the normal-hearing ear via direct audio input technology.
- Recommended test materials: See Table 1.

Mapping

- The normal-hearing ear should be occluded for behavioral measurement.
- Electrically Evoked Stapedial Reflex Thresholds (ESRTs) may be particularly useful in this population.
- Loudness balancing with the normal-hearing ear may be helpful.

- Clinicians should consider microphone placement when recommending devices.

Counseling

- Realistic family and child-centered goals should be established pre-operatively.
- Alternative technologies should be discussed as part of pre-operative counseling.
- Families should be made aware of developmental risks of untreated SSD.

Aural Habilitation

- Developmentally appropriate therapy is recommended post-activation to strengthen auditory skills with the CI alone and encourage development of binaural skills.