COCHLEAR IMPLANTATION IN THE DIFFICULT EAR
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Grand Rounds Presentation
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BACKGROUND

• Minimum requirement for cochlear implantation is the presence of an implantable cavity in proximity to stimulable neural elements whose projections connect to the auditory cortex (Fishman)

• Who fits this criteria?

• Who doesn't fit this criteria?

• What makes a patient difficult?
ANATOMY AND EMBRYOLOGY OF THE EAR
ANATOMY AND EMBRYOLOGY REVIEW

http://www.youtube.com/watch?v=1JE8WduJKV4&feature=youtu.be

Click the black box once or twice to view the content – or – Go to the URL below
Quick review: Motion of the Basilar Membrane (BM) is sensed by the Inner Hair Cells (IHCs).

IHC motion activates or inhibits chemical transmitter release.

Nearby neurons activate based on these changes and transmit to the central auditory complex.
• Formation of Otic Placode as Ectodermal Thickening in 3rd gestational week - beginning of the combined cochlear and vestibular membranous labyrinthine system

• Placode invaginates from surface to form the otocyst in 4th gestational week

• 3 infolds develop in the 5th week: Primordial endolymphatic sac and duct; the utricle and semicircular canals, and saccule and cochlea

• Cochlear duct at 6th week grows from primordial bud from basal region spiraling apically to reach its full two-and-a-half to two-and-three-quarter turns ending around 8-10th week.

• Organ of Corti completes its formation in 25th week
EM of Mouse at 9 days Gestation
• Semicircular canals form as 3 small folded evaginations on the primordial vestibular appendage

• Develop as out-pouchings whose centers compress and fuse to form semicircular duct structure

• Canals continue to enlarge and complete formation in sequence starting with Superior, Posterior, and finally Lateral Canal
EMBRYOLOGY

• Osseous otic capsule develops from cartilage precursor

• Cochleovestibular nerves and ganglia develop with the membranous labyrinth and cochleovestibular end organs

• Cochleovestibular nerves and Ganglia are of neural crest origin and migrate between epithelial layer and basement membrane of the otic vesicle
The Goals of Cochlear Implantation

• Principle cause of hearing loss is damage to sensory hair cells

• Severs connection to the central auditory system

• Function of CI is to bypass missing or damaged hair cells stimulating directly the surviving neurons

• Peripheral neurons of spiral ganglion cells undergo retrograde degeneration; however cell bodies much more robust
The Goals of Cochlear Implantation

• Nodes of Ranvier in spiral ganglion cells are putative sites of excitation for CI

• Surviving cell counts vary from location to location and from cochlea to cochlea

• Minimal number of auditory neurons necessary for speech recognition with a CI is unknown but thought to be minimal
<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Ganglion Cell Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Labrynthitis Ossificans</td>
<td></td>
</tr>
<tr>
<td>Meningogenic</td>
<td>6,310-28,977</td>
</tr>
<tr>
<td>Tympanogenic</td>
<td>21,700</td>
</tr>
<tr>
<td>Otosclerosis</td>
<td>10,127-22,525</td>
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<tr>
<td>Congenital Causes</td>
<td></td>
</tr>
<tr>
<td>Mondini</td>
<td>7,677-16,110</td>
</tr>
<tr>
<td>Normal</td>
<td>29,802-38,352</td>
</tr>
</tbody>
</table>
Insertion of the Cochlear Implant

Click the black box one or twice to connect to the URL below – or – go to the link below

http://www.youtube.com/watch?v=1JE8WduJKV4&feature=youtu.be
COCHLEOVESTIBULAR MALFORMATIONS

- Embryologic Considerations
- Classification Schemes
- Surgical Considerations
- Outcomes

- Lots of background research in the subject
- Jackler wrote article on classification
A BRIEF BACKGROUND

• Jackler et al in 1987 proposed the widely accepted classification used today

• “In recent years, use of the term ‘Mondini’s dysplasia’ has come to mean virtually any congenital malformation of the osseous labyrinth that is detectable on radiographic examination
Mondini Deformity

- Controversy Remains (Sennaroglu)

- Classic Mondini Deformity
  - 1.5 turns of the cochlea, dilated vestibule, and an enlarged vestibular aqueduct

Fig. 2. Incomplete partition type II (classic Mondini deformity). (A) Axial computed tomography image demonstrating dilated vestibule (V) and enlarged vestibular aqueduct (arrow). (B) Inferior section through the round window showing confluence of middle and apical turns.
WHERE IT GOES WRONG

- Approximately 20% of cases of Congenital SNHL

Fig. 7. Axial (A) and coronal (B) images of a patient with a common cavity malformation who was implanted under fluoroscopic guidance. Note the common cavity (cc) and internal auditory canal (iac). (From Fishman AJ, Roland JT Jr, Alexiades G, et al. Fluoroscopically-assisted cochlear implantation. Otol Neurotol 2003;24(6):882-6; with permission.)
Cochlear Malformations (Sennaroglu)

• Michel Deformity

• Cochlear Aplasia

• Common Cavity Deformity

• Cochlear Hypoplasia

• Incomplete Partition type I (IP-I): cochlea lacking entire modiolus and cribiform area

• Incomplete Partition Type II (IP-II) (Mondini Deformity)
Michel Deformity
- Week 3

Cochlear Aplasia
- Late Week 3

Common Cavity
- Week 4

IP-1
- Week 6

IP-II (Mondini Deformity)
- Week 7
Genetic Associations

- Fibroblast Growth Factor-3 (FGF-3)
- Nkx5 to 1 homeobox gene - semicircular canals
- Pax-2 - cochlea
Comparison of Classification Schema

Fig. 2. Historical classification of cochleovestibular anomalies. (A) Classification of Jackler et al.\(^5\) with weeks of developmental arrest. (B) Simplified classification scheme proposed by Phelps.\(^{15}\) (C) Further subclassification of Mondini-like deformities by Zheng et al.\(^{16}\) (D) New classification scheme suggested by Sennaroglu et al.\(^{17}\)
Schematic Representation of Different Stages

NORMAL COCHLEA

COCHLEAR APLASIA

COMMON CAVITY

INCOMPLETE PARTITION

HYPOPLASIA

Fig. 6. The normal cochlea and cochlear malformations. (A) Normal cochlea, midmodiolar section. Mo, modiolus; CA, cochlear aperture; B, basal turn; M, middle turn; A, apical turn; arrowheads, interscalar septa. (B) Normal cochlea, inferior section passing through the round window niche (RWN). Arrowhead, interscalar septum between middle and apical turns. (C) Cochlear aplasia with normal vestibule. (D) Cochlear aplasia with enlarged vestibule. (E) Common cavity. (F) Incomplete partition type I. (G) Incomplete partition type II. (H) Incomplete partition type III. (I) Cochlear hypoplasia, bud type (type I). (J) Cochlear hypoplasia, cystic cochlea type (type II). (K) Cochlear hypoplasia, with less than 2 turns (type III). (Modified from Sennaroglu L. Cochlear implantation in inner ear malformations—a review article. Cochlear Implants International 2009; doi: 10.1002/cii.416; with permission.)
Evaluation of Patients with Inner Ear Dysplasia

- Readily Identifiable on HRCT
- Remember, 20% of pts with congenital SNHL have osseous abnormalities
- Percentage of patients requiring CI with osseous dysplasia is higher (Papsin 2005)
MRI

- T2 MRI useful in evaluating cochlear patency and septations within a cavity
- Can ascertain absence of a cochlear nerve with sagittal views of the IAC in CISS or FIESTA sequences
• Can we Implant these Patients?
• Are there any contraindications?
• What complications?
Surgical Considerations

• Type of malformation will generally dictate surgical approach

• With exception of Michel’s aplasia, all dysplasias can be implanted

• Most can be approached with a standard transmastoid facial-posterior tympanostomy approach
Electrode Choice

• Malformed Cochlea often does not have neuroepithelium in normal location
• Few instances where normal electrode arrays are not ideal
• Most common anomaly for intrameatal electrode placement is X-linked deafness associated cochlear anomaly
Device Programming

• Frequent adjustment in programming necessary

• Stimulation of facial nerve common requiring deactivation of electrodes in question
Performance

• Consensus that speech perception results can be in same range in implant recipients with normally formed cochleas

• Retrospective matched-pairs analysis of 17 patients with malformed cochleas compared with 17 patients with normal cochleas by Eisenman and colleagues

  • Outcomes similar 24 months after implantation

  • Malformation cases take longer to reach end point
Performance

- Children grouped according to deformity (normal, Common Cavity, Incomplete Partition, Hypoplastic Cochlea, and EVA)
- IP children more likely to have progressive onset of hearing loss compared to other groups
- IP children with consistently higher scores
Performance

- Comparison of groups using different closed and open-set speech scores
- Open-set (GASP-word, PBK-Word, PBK-Phoneme)
- Closed-set (TAC, WIPI)
• Open-set Speech Perception Tests

• Closed-set Speech Perception Testing

Performance

• Hypoplastic cochleae or common cavity children generally with worse performance than IP patients

• Anatomic and Physiologic Basis:
  • Fewer active electrodes implanted
  • Narrower dynamic ranges
  • Impaired neural synchrony
  • Lack of Tonotopic Organization
Common Cavity Implantation

• Cavity lacks the central modiolus (remember the goal to place the CI perimodiolar)

• Rudimentary forms of normal cochlear structures are present

• Equivalent to organ of Corti and stria vascularis

• Most importantly: auditory neural tissue lies peripherally
Surgical Technique

• Utilize continuous facial nerve monitoring

• Standard Mastoidectomy with subperiosteal pockets for receiver-stimulator package

• Exposure of the posterior surface of the cavity in the antrum with the drill

• Cochleostomy drill-out (1.0-1.5 mm in diameter)

• Some authors recommend intraoperative imaging guidance during insertion
Fig. 3. Placement of preformed electrode array into common cavity (right ear).

Surgical Technique

• Controversy on whether to obliterate the external meatus or perform a canal-wall down mastoidectomy in these cases

• Most authors implant a straight array with concentric banded electrodes

• This allows for the outer wall to be contacted
Complications

- CSF Leak
- IAC Implantation
- Facial Nerve Complications
CSF Leak

- Bony cochlear lamina cribrosa often missing or replaced by fibrous band
  - Defects of modiolus and lamina cribrosa at fundus of IAC responsible for mixing of perilymph and CSF
- Degree of Dysplasia does not correlate with risk of leakage
- Common Cavity Malformations with highest risk of CSF/perilymph leakage
CSF Leak Management

• Rate the risk by evaluating pre-operative imaging
• Wider cochleostomy allows for more packing material
• Performing Valsalva maneuvers with head-down to confirm cessation of leak
• Rarely, Lumbar drainage may be necessary
Meningitis

• CI and inner ear malformation increase risk of developing meningitis post-operatively

• These patients NEED preoperative pneumococcal vaccination
IAC Insertion

- Absence of cochlear lamina cribrosa
- Unrecognized insertion can lead to CSF leak, vertigo, facial nerve stimulation, suboptimal hearing outcomes
- Short or compressed electrode arrays can limit but do not eliminate risk
Fig. 8. Transorbital plain radiograph, intraoperative view. Note that the array has passed into the internal auditory canal (iac). The arrow denotes the junction between the common cavity and the iac as seen in this orientation. Inset outlines the lumen of the common cavity (cc) and the iac. (From Fishman AJ, Roland JT Jr, Alexiades G, et al. Fluoroscopically assisted cochlear implantation. Otol Neurotol 2003;24(6):882–6; with permission.)
Facial Nerve Complications

- 15% of pts with anomalies have aberrant facial nerve courses.
- Typically anterior from the first genu down the promontory to the round window.
- Occasionally, the nerve is split with one branch in normal position.
- Coelho & Rolando recommend drilling along tegmen to antrum and incus identified. Incus removed then facial nerve identified along horizontal segment.
- Used in all patients with CHARGE syndrome and BOR syndrome.

Fig. 5. Intraoperative views demonstrate abnormal facial nerve course in the left-side middle ear of a child with hypoplastic cochlea (HC). The facial nerve is split, and the anterior branch (VIIa) runs over the oval window and onto the promontory (P) obstructing the view of the round window niche, making insertion of the electrode array challenging. The posterior branch (VIIp) runs approximately in the normal position.
# TABLE V.

**Reports of Abnormal Facial Nerves Encountered During Cochlear Implantation in Children With Anomalous Cochleovestibular Anatomy.**

<table>
<thead>
<tr>
<th>Reference No.</th>
<th>Authors (y)</th>
<th>No. of Aberrant Facial Nerves</th>
<th>Associated Cochleovestibular Anomalies</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>53</td>
<td>Molter et al. (1993)</td>
<td>1</td>
<td>1 CC</td>
<td>Nerve felt to overlie site of ideal cochleostomy; described new insertion technique through hypoplastic semicircular canal</td>
</tr>
<tr>
<td>12</td>
<td>Tucci et al. (1995)</td>
<td>1/6</td>
<td>1 HC</td>
<td>Identified on preoperative CT scan; importance of nerve monitor</td>
</tr>
<tr>
<td>54</td>
<td>Hoffman et al. (1997)</td>
<td>8/50</td>
<td>Unknown</td>
<td>Results of a mailed questionnaire (n = 50); 1 temporary paresis reported; 1 permanent paralysis reported; noted importance of CT and nerve monitor</td>
</tr>
<tr>
<td>9</td>
<td>Luntz et al. (1998)</td>
<td>2/10</td>
<td>2 CC</td>
<td>Significant seventh-nerve stimulation in 3 of 10; 1 temporary paresis (CC); importance of nerve monitor</td>
</tr>
<tr>
<td>52</td>
<td>Weber et al. (1998)</td>
<td>2/12</td>
<td>1 HC, 1 ?</td>
<td>Paralysis with unrecognized split; facial nerve (HC); importance of nerve monitor</td>
</tr>
<tr>
<td>48</td>
<td>McElveen et al. (1997)</td>
<td>3</td>
<td>3 CC</td>
<td>Used approach through hypoplastic semicircular canals</td>
</tr>
<tr>
<td>49</td>
<td>Ito et al. (1999)</td>
<td>1</td>
<td>1 CC</td>
<td>Canal wall down mastoidectomy to achieve anterior exposure</td>
</tr>
<tr>
<td>Current series</td>
<td></td>
<td>14/103</td>
<td>2 CC, 10 HC, 2 IP</td>
<td>Four children with craniofacial syndrome; all implanted via standard facial recess approach; nerve monitor routinely used</td>
</tr>
</tbody>
</table>

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Facial nerve abnormality was found in children with common cavity deformity (CC), hypoplastic cochlea (HC), and incomplete partition (IP). Several authors noted the importance of monitoring the facial nerve when performing cochlear implantation.
LABYRINTHITIS OSSIFICANS

- Etiology
- Pre-Operative Imaging
- Surgical Considerations
- Outcomes
Bacterial Meningitis

- Most common cause of postnatal profound HL in children
- Occurs in 6-16% of patients
- Enters cochlea from subarachnoid space through cochlear aqueduct that has failed to obliterate
- Cochlear aqueduct runs through petrous bone from posterior fossa and terminates laterally in scala tympani close to round window

**FIG. 1.** Bacterial causes of meningitis and hearing loss.
• Spread to cochlea rapidly and often without hours of diagnosis

• Spreads apically from basal turn damaging intracochlear structures that are usually profound and irreversible

• Endostial inflammation causes new bone formation within the cochlear lumen

• Lots of variability in amount of osteoneogenesis case to case

• Animal studies show ossification occurring within 72 hours of meningitis
Pre-operative Evaluation

• Often difficult

• Once stabilized, audiological evaluation and immediate amplification implemented

• Serial audiometric testing should be performed

• Deterioration to severe or profound SNHL should prompt swift implantation

  • Reflect of progressive fibrosis and ossification
Fig. 1. (A) Noncontrast axial high-resolution computed tomography of normal right cochlea. The arrow is pointing to the internal auditory canal and cochlea to demonstrate similar fluid hypodensity. (B) Coronal view of normal cochlea. The arrow indicates the hypodense fluid-filled area of the pars ascendens at the end of the basal turn. (C) Axial view of severe cochlear ossification. (D) Coronal view of severe cochlear ossification.
Labyrinthitis Ossificans

• Ossification can occur following 3 main pathways
  • Meninogenic
  • Tympanogenic
  • Hematogenic
• Can directly destroy spiral ganglion cells
Pre-operative Imaging

- HRCT necessary to evaluate bony anatomy of temporal bone
- Attenuation of intracochlear fluid signal in T2-weighted MRI is more sensitive method of assessing blockage
- Contrast-enhanced T1-weighted MRI can detect early fibrosis
Figure 1: MR images of the right (R) and left (L) inner ears of a patient (case 2) after pneumococcal meningitis. Depicted are the axial T1 weighted MR images with contrast enhancement (T1, top row) and the T2 weighted MR images (T2, bottom row). The patient, a boy aged 7 months, suffered from asymmetric hearing loss after pneumococcal meningitis. Auditory brain stem response (ABR) audiometry showed a deaf ear on the right side and a sloping hearing loss (60 dB at 3 KHz) on the left side. Red arrows show contrast enhancement in the cochlea on the T1 weighted images of both ears ((a) and (b)). The contrast enhancement involves the whole cochlea and vestibulum on the right side, but it is limited to the basal turn (BT) on the left. Yellow arrows show loss of fluid in the cochlea on the T2 weighted images on both sides ((c) and (d)). Whereas on the right side, the loss of fluid involves the complete cochlea and the basal turn is barely visible, the loss of fluid only partially involves the basal turn of the left cochlea. IAC: internal auditory canal.
Grading

Fig 1. Axial HRCT of the basal turn of the cochlea: A, grade 0 (normal); B, grade 1 (sclerosis without narrowing); C, grade 2 (sclerosis with narrowing); and D, grade 3 (complete or partial obliteration).
HRCT and Implantation

- Young et al retrospectively examined HRCT and correlated with surgical findings
- HRCT of cochlea suggested ossification in basal turn in 45% (50% sensitivity)
- HRCT of LCC suggested ossification in 70% of cases (77% sensitivity)
Accuracy of MRI

• Isaacson et al presented case series of 45 children with post-meningitic hearing loss

• MRI accuracy for predicting intraoperative cochlear obstruction:

<table>
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<tr>
<th>Statistics using all MRI studies</th>
<th>n/N</th>
<th>%</th>
<th>95% Cls</th>
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<tr>
<td>Sensitivity</td>
<td>16/17</td>
<td>94.1</td>
<td>71-99</td>
</tr>
<tr>
<td>Specificity</td>
<td>7/8</td>
<td>87.5</td>
<td>47-99</td>
</tr>
<tr>
<td>Positive predictive value</td>
<td>16/17</td>
<td>94.1</td>
<td>71-99</td>
</tr>
<tr>
<td>Negative predictive value</td>
<td>7/8</td>
<td>87.5</td>
<td>47-99</td>
</tr>
</tbody>
</table>

CI, confidence interval.
Surgical Technique

• As in normal CI insertion, full electrode insertion into Scala Tympani is ultimate goal

• What to do when this is not possible?
Surgical Options

- Limited Ossification: Drilling continued along basal turn until lumen identified
- If uncalcified osteoid or fibrous tissue, remove with picks, rasps or drills
- Patent lumen found before or at ascending turn, full insertion performed
- Judicious use of test electrode or depth gauge
Basal Turn Drill Out

- 1 mm diamond drill in anteromedial direction for no more than 9.5 mm or until the internal carotid artery visualized
- Allows for placement of enough electrodes for multichannel performance
Scala Vestibuli Insertion

• If standard cochleostomy reveals obstruction after drill-out to pars inferior, extended superiorly closer to oval window

• Scala vestibuli is identified, cochleostomy is enlarged, and filled with glycerin to clear blood and bone debris

• Electrode array is inserted and cochleostomy is packed

*Fig. 1. Intraoperative view (left ear) of obstructed scala tympani and patent scala vestibuli. ST indicates scala tympani; SV, scala vestibuli.*

Otology & Neurotology, Vol. 27, No. 5, 2006
Scala Vestibuli Performance

- Lin et al reviewed 8 patients with scala vestibuli insertion
- Improvement in communication among all patients
Implantation in Totally Ossified Cochlea

- Short Insertion Tunnel Technique reported by Cohen and Waltzman
- Total Drill-out Technique reported by Gantz
- CWU without radical cavity/obliteration
Surgical Technique for Implantation of the Totally Ossified Cochlea

Thomas Balkany, MD, FACS; Philip A. Bird, FRACS; Annelle V. Hodges, PhD, CCC-A; Michal Luntz, MD; Fred F. Telischi, MEE, MD, FACS; Craig Buchman, MD

- Technique combines inferior trough along with open trough technique
- Allows for fixation point and more electrodes to be utilized
Fig. 1. Transcanal view of the middle ear. The malleus handle and incus have been removed and the superiorly based tympanomeatal flap elevated. Note the relationship of the basal turn with the round window, carotid canal, cochleariform process, and oval window.
Fig. 2. Drilling the tunnel. Working through the facial recess, an 8-mm-long tunnel is drilled from the cochleostomy. The abnormal bone within the lumen is followed anteriorly with a 1.5-mm diamond drill. Drilling is discontinued if bleeding occurs.
Fig. 3. Beginning the trough. Using a 1-mm diamond drill, the tunnel is unroofed at approximately 4 mm. The abnormal bone of the basal turn is followed around the modiolus using the carotid canal, cochleariform process, and oval window nerve as landmarks.
Fig. 4. Electrode in position. The electrode can be seen passing through the facial recess, the tunnel, and within the trough. The electrode lead cable is seen passing through the area of the slit bridge on the way from the mesotympanum to the epitympanum. Further fixation occurs in the split incus bridge and with bone wedges in the trough.
Double/Split-Array Implantation

- Double array based on positive correlation between number of electrodes and speech recognition
- Basal array placed in inferior basal-turn tunnel
- Fossa Incudis, Incus and Stape suprastructure removed
- Second cochleostomy placed in second turn by drilling adjacent to anterior oval window ligament
Fig. 3. (A) MED-El Split-electrode device. (B) Cochlear Corporation double-electrode device. (From Cosetti M, Roland JT Jr. Cochlear implant electrode insertion. Operat Tech Otolaryngol Head Neck Surg 2010;21(4):223–32; with permission.)

Fig. 4. Intraoperative view through facial recess of the left ear depicting double-array cochlear implant. The basal array (long arrow) is observed entering anteroinferior to the round window. The apical array (short arrow) is observed at the second-turn cochleostomy just anterior the oval window. Asterisk indicates the head of Malleus; LSCC, lateral semicircular canal. (From Roland JT Jr, Coelho DH, Pantelides H, et al. Otol Neurotol 2008;29(8):1068–75; with permission.)
Double/Split-Array Implantation

Fig. 9. Intraoperative photograph (A) and drawing (B) of a transmastoid view through the facial recess of a right ear after removal of the incus and incus bar. Both basal and apical cochleostomies are seen before insertion of a dual-electrode array. AC, apical cochleostomy; BTC, basal-turn cochleostomy; FN, facial nerve in descending (mastoid) portion; HC, horizontal (lateral) semicircular canal; SF, stapes footplate. (From Cosetti M, Roland JT Jr. Cochlear implant electrode insertion. Operat Tech Otolaryngol Head Neck Surg 2010;21(4):223–32; with permission.)
Speech Performance

**TABLE 2. Manchester spoken language development scale levels**

<table>
<thead>
<tr>
<th>Level</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Level 1</td>
<td>Indicates awareness of a range of environmental sounds and voice</td>
</tr>
<tr>
<td>Level 2</td>
<td>Uses voice to attract adult attention, babbling in play</td>
</tr>
<tr>
<td>Level 3</td>
<td>Imitation of adult language models (pattern perception, vowel approximations)</td>
</tr>
<tr>
<td>Level 4</td>
<td>Expressive vocabulary of less than 50 words</td>
</tr>
<tr>
<td>Level 5</td>
<td>Expressive vocabulary of 50–200 words and/or joining 2 words together</td>
</tr>
<tr>
<td>Level 6</td>
<td>Using simple phrases of 3 words</td>
</tr>
<tr>
<td>Level 7</td>
<td>Simple sentence level</td>
</tr>
<tr>
<td>Level 8</td>
<td>Advancing grammar and complex sentence structure</td>
</tr>
<tr>
<td>Level 9</td>
<td>Expressive language skills at a level in line with “hearing age”</td>
</tr>
<tr>
<td>Level 10</td>
<td>Expressive language skills at a level in line with chronological age</td>
</tr>
</tbody>
</table>

**TABLE 1. Categories of auditory performance**

<table>
<thead>
<tr>
<th>Level</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Displays no awareness of environmental sounds</td>
</tr>
<tr>
<td>1</td>
<td>Awareness of environmental sounds</td>
</tr>
<tr>
<td>2</td>
<td>Responds to speech</td>
</tr>
<tr>
<td>3</td>
<td>Recognizes environmental sounds</td>
</tr>
<tr>
<td>4</td>
<td>Discriminates at least 2 speech sounds</td>
</tr>
<tr>
<td>5</td>
<td>Understands common phrases without lipreading</td>
</tr>
<tr>
<td>6</td>
<td>Understands conversation without lipreading with a familiar talker</td>
</tr>
<tr>
<td>7</td>
<td>Can use the telephone with a familiar talker</td>
</tr>
</tbody>
</table>
Speech Performance

**FIG. 3.** MSLDS in nonossified cochlea.

**FIG. 2.** CAPS in nonossified cochlea.

*Otology & Neurotology, Vol. 32, No. 5, 2011*

**FIG. 6.** MSLDS in ossified cochleas. P indicates partial insertion; S, split electrode; SV, scala vestibuli insertion.

**FIG. 5.** CAPS in ossified cochleas. P indicates partial insertion; S, split electrode; SV, scala vestibuli insertion.
Surgical Algorithm for Ossification

![Algorithm Diagram]

Fig. 2. Obstructed cochleae algorithm (note: only the MED-EL device comes with a test electrode). (From Cosetti M, Roland JT Jr. Cochlear implant electrode insertion. Operat Tech Otolaryngol Head Neck Surg 2010;21(4):223–32; with permission.)
OTOSCLEROSIS

• Etiology and Anatomy
• Surgical Considerations
• Outcomes
• Should we change our management?
Anatomy and Etiology

• Process of Bone Resorption of Petrous Bone followed by replacement with thick irregular sclerotic bone

• Most commonly affect location is around the oval window (Fenestral Otosclerosis)

• 10% of patients, otosclerotic foci will affect otic capsule (Retrofenestral Otosclerosis)

• Far Advanced Otosclerosis (FAO): Severe Mixed Hearing Loss described by House and Sheehy

• Today, Advanced Otosclerosis: patients with SNHL and diminished (<100%) SD scores
Surgical Options

• Stapedectomy is relatively simple, inexpensive procedure

• Even in advanced Otosclerosis with mixed hearing loss, surgical correction of conductive component can be effective enough

• In Severe Mixed Hearing Loss, stapedectomy would be unsatisfactory

• Treatment with CI results in excellent hearing

• CI is expensive and requires experienced surgeons
CT Grading of Otosclerosis

Fig. 4. Axial computed tomography scan of the petrous bone in patients with otosclerosis. (A) Grade 1: solely fenestral involvement, otospongiotic lesion on the anterior border of the vestibulum (white arrow). (B) Grade 2A: double ring effect (black arrow). (C) Grade 3: diffuse confluent cochlear involvement with an unrecognizable cochlea.
CT Grading of Otosclerosis

• Different Grading Systems (Rotteveel and Symons/Fanning)

• Rotteveel Classification

• Grade 1: Solely Fenestral

• Grade 2A: Retrofenestral: Double Ring or Halo Affect

• Grade 2B: Narrowed Basal Turn

• Grade 2C: Both Retrofenestral and Narrowed Basal Turn

• Grade 3: Diffuse Confluent Retrofenestral Involvement
Surgical Considerations

• Fenestral and Basal Turn Ossification

• Necessity for Extra Drilling

• Partial Electrode Insertion

• Scala Vestibuli Insertion

• 80-100% of patients with basal turn ossification led to partial electrode insertion or misplacement during surgery

• Obliteration of Scala Tympani required scala vestibuli insertion in 2-25% of cases
<table>
<thead>
<tr>
<th>Author</th>
<th>N</th>
<th>Fenestral Oss (%)</th>
<th>Basal turn Oss (%)</th>
<th>Extra Drilling (%)</th>
<th>Partial Insertion (%)</th>
<th>Scala Vestibuli Insertion (%)</th>
<th>FNS (%)</th>
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</table>

N = number of operated ears; Oss = ossification; NA = not mentioned; FNS = facial nerve stimulation.
Fig. 3. Axial computed tomography (CT) scan of the petrous bone in a patient with retrofenestral otosclerotic lesions: pre-, peri-, and post-operative. (A) Preoperative CT scan: grade 2C retrofenestral otosclerosis (according to Rotteveel classification): double ring (black arrow) and basal turn narrowing (white arrow). (B) Perioperative CT scan of the same patient with the electrode entering a false lumen due to the thickened round window, sclerotic scala tympani, and otospongotic double ring surrounding the cochlea. (C) Postoperative CT image with a complete electrode insertion in the scala vestibuli.
Advanced Otosclerosis Algorithm

Fig. 2. Algorithm guideline to counsel patients with (advanced) otosclerosis. Algorithm is based on the speech discrimination score, computed tomography (CT) classification, and the air-bone gap, and will guide the surgeon to either cochlear implantation, stapedotomy, or a hearing aid and follow-up.

Laryngoscope 121: September 2011
Merkus et al.: Decision Making in Advanced Otosclerosis
## Performance

### TABLE I.
Results of Cochlear Implantation in Patients With Otosclerosis.

<table>
<thead>
<tr>
<th>Author</th>
<th>N</th>
<th>Patients Hearing Improved (%)</th>
<th>Average Speech Discrimination Score Before Surgery, % (SD)</th>
<th>Average speech Discrimination Score After Surgery, % (SD)</th>
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</table>

N = number of operated ears; SD = standard deviation; NA = not available; MSW = monosyllable words test; SEN = sentences test; TSW = two-syllable words test.
Conclusions

- The only absolute contraindication to implantation of a cochlear implant is the absence of the cochlea.
- IP Implant recipients generally do the best of the cochleovestibular malformations.
- CT and MRI are generally both recommended as imaging modalities in patients with cochleovestibular anomalies and with a history of bacterial meningitis.
- T2-weighted MRI is sensitive at detected intracochlear fluid.
- Cochlear Implantation may be an important adjunct in the treatment of hearing loss in a subpopulation of patients with Otosclerosis.