

Endoscopic Approach to the Round Window in Infants and Toddlers: Electrical Stimulation of the Cochlea

John Germiller MD, PhD

Luv Javia MD

Division of Pediatric Otolaryngology
Cochlear Implant Program
Children's Hospital of Philadelphia
Department of Otorhinolaryngology
Perelman School of Medicine, University of Pennsylvania



I have nothing to disclose.

No, within the last 12 months I have not had any type of financial arrangement or affiliation with commercial interests related to the content of this continuing education activity that requires disclosure.

Introduction

- Future therapies for deafness (e.g., gene and stem cell therapy agents) will require a safe method of delivery into the human cochlea.
- Candidates will be infants and toddlers under 2 years.
- The round window (RW) will likely be the most useful portal of entry.
- A safe, minimally-invasive surgical approach to the RW in infants and toddlers is needed.

Introduction

- Endoscopic techniques facilitate access to the posterior mesotympanum, including the round window (RW) niche.
- Because infants and young toddlers rarely undergo endoscopic ear surgery, an endoscopic surgical approach specifically to access the RW area in this age group has not been reported.

Introduction

- At our center, children with cochlear nerve deficiency undergo electrically-evoked ABR (EABR) testing, to determine candidacy for cochlear implantation.
- We deliver noninvasive electrical stimulation to the cochlea via the RW membrane (instead of promontory stimulation).
- We have been using a minimally-invasive endoscopic tympanotomy to access the RW. Patients are 6 mo - 2 years old.

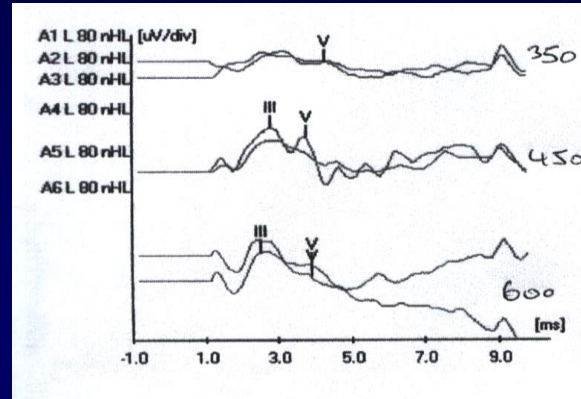
Objective

- Report our experience with accessing the RW in infants and toddlers using an endoscopic approach.
 - Describe our technique
 - Effectiveness (successful RW access)
 - Safety (complications).

Electrically-evoked ABR (EABR)

- Evaluates auditory response to direct electrical stimulation of the cochlea.
- Can be done with an existing CI, or beforehand
 - Determine cochlear nerve status prior to CI or ABI.

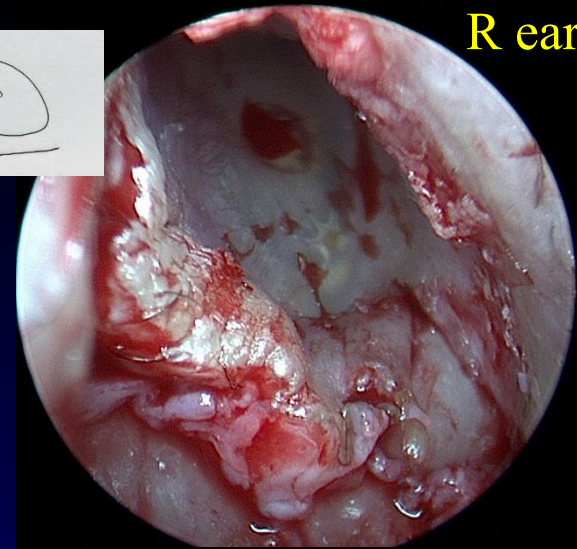
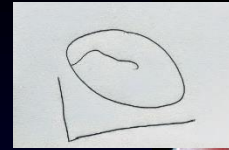
Stimulate cochlea with electric current



Measure response on ABR (wave V)

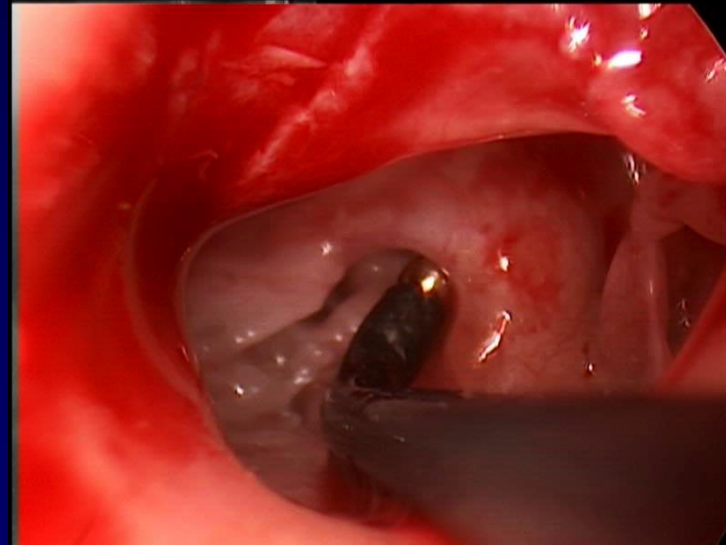
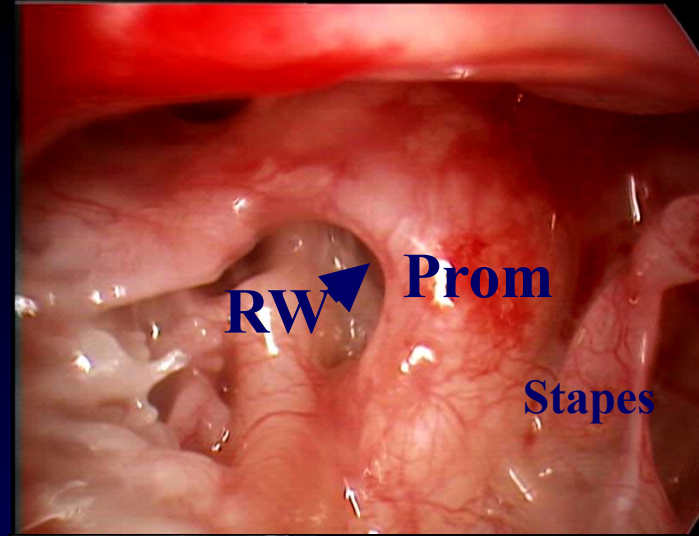
Methods

- Pediatric CI candidates with profound bilateral deafness and apparent cochlear nerve aplasia on high quality MRI.
- Under endoscopic visualization, the posterior TM is elevated
 - Standard 3mm, 14-cm, 0° and 30° endoscopes are used.
 - A short, triangular skin flap better for the very short ear canals in these patients.



Methods

- A 1-mm ball-tip probe is placed directly on the RW membrane and stimulation applied.
- Nerve responses are measured by ABR.
- Stimulation is increased up to the maximum obtainable with current cochlear implants.

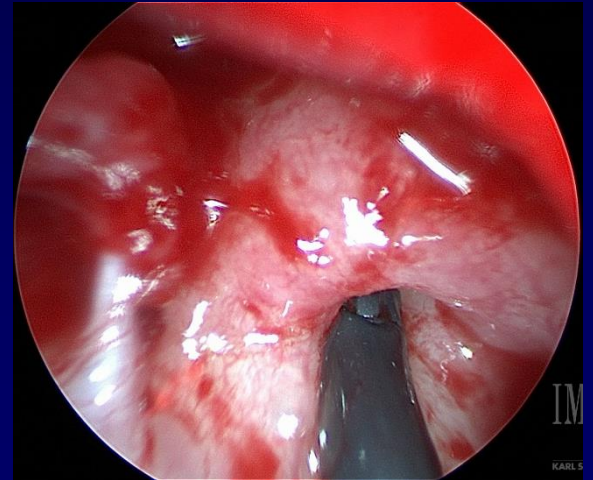
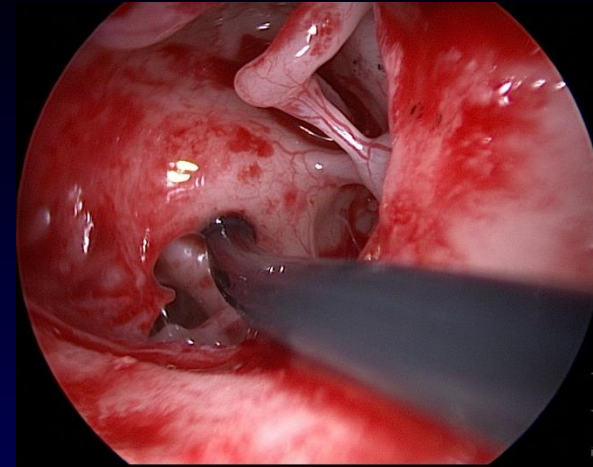


Results

- Procedures were performed unilaterally in 5 children and bilaterally in 9.
- Total 23 ears; median age 23 mo.
- The youngest patient was 9 months of age.

Results

- Endoscopic access, stimulation of the RW, and EABR measurement were successful in all 23 ears.
- Visualization of at least 1/3 of the RWM was typical
- Instruments require mild curvature to reach RW membrane
- None required bone removal (either from the canal wall or RW overhang).



Results

- Chorda spared in every case
- No injuries to the RW membrane occurred.
- All tympanic membranes healed rapidly (follow up 2-3 weeks later)
- One patient experienced transient facial weakness (< 1 hour) due to the electrical stimulation.

EABR Findings

- No responses despite maximal stimulation in 21 ears, supporting cochlear nerve aplasia.
- The remaining 2 ears had responses, and subsequently underwent cochlear implantation.

Conclusions

- Endoscopic tympanotomy is a safe and effective technique to access the inner ear via the round window.
- Access is straightforward and safe, even in young infants.
- This approach may facilitate future therapies requiring access to the inner ear in infants and toddlers.

Acknowledgments

Daniel Lee, M.D.

Paul R. Kileny, Ph.D.

Amy White, Au.D.

Amanda Marchegiani, Au.D.

Joy Peterson, Au.D.

Carmen Hayman, Au.D.

Michael Jackson, Au.D.

Melissa Ferrello, Au.D.



Cochlear Implant Program
Children's Hospital of Philadelphia



Massachusetts
Eye and Ear

Harvard
Medical
School



Thank You



 The Children's Hospital of Philadelphia®

Hope lives here.

