Retrofacial Approach to Access the Round Window for Cochlear Implantation of Malformed Ears

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• We have nothing to disclose
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Background

• CI surgery fairly standardized
  – Facial recess approach

• Difficult cases:
  – Subtotal petrosectomy with middle ear obliteration
Case 1: H&P

- Premature
- VATER Syndrome
- Diagnosis of severe to profound SNHL at 5 months
- No benefit from hearing aids
- First CI evaluation at 26 months of age
  - Grade I microtia
  - SRT 80dB (UNAIDED) 40dB (Aided)
Case 1: CT scan

- Abnormal SCC
- Absent stapes Malformed ossicles
- Normal cochlea
- Facial nerve course was difficult to trace

DECISION TO PROCEED WITH LEFT COCHLEAR IMPLANTATION AT THE AGE OF 2.5 YEARS
Case 1: Follow up

- Speech acquisition
- Left implanted ear thresholds: 20dB HL
- Decision to implant right ear at the age of 6 years
  - Same approach
  - Implantation was done through round window
Case 1: Video right CI
Case 2: H&P

- Referred after newborn screening test
  - Severe to profound SNHL on ABR
- Grade II microtia
- Medial EAC stenosis on the right
- Bilateral preauricular pits and tags
- Bilateral optic nerve hypoplasia
- Congenital LEFT facial palsy HB III/VI

GENETIC DIAGNOSIS: HEMIFACIAL MICROSomia; CONNEXIN 26 MUTATION
CASE 2: Imaging

- 6 months old CT scan of the temporal bones
  - Narrow IAC
  - Small cochlear nerve apertures
  - Dilated vestibule

- 9 months old MRI:
  - Left cochlear nerve aplasia
  - Right cochlear nerve hypoplasia

DECISION TO IMPLANT RIGHT EAR AGE 15 MONTHS OLD
Case 2: Imaging

- Narrow IAC
- Small cochlear Aperture
- Dilated Vestibule
Case 2: Imaging

- Arrows indicate bilateral bifid facial nerve
Case 2: Intraoperative

- Very prominent chorda tympani/bifid facial nerve
- Poor exposure through facial recess
- No stapes present
- Facial nerve was coursing inferomedial (6 mm) to horizontal SCC
- Subtotal petrosectomy and closure of EAC
  - RW Still not visualized
- Retrofacial approach decided
Case 2: Intraoperatively

- Long arrow: Middle Ear
- Short arrow: Round window
- Arrowhead: hypotympanic air cell
Discussion

- 2 case reports in the literature
  - Both for aberrant facial nerve
- ANS meeting Spring 2014: Show of hands of 10 neurotologists having used this approach
- Incidence of abnormal facial nerve 0.3%
- Incidence of dehiscent facial nerve: up to 55%
- Tympanic segment anomalies are not isolated: Stapes+++ 
- Bifurcations of tympanic segment are most common: level of second genu+++
Discussion

• Width of facial recess is rarely variable
• Narrow facial recess defined by distance between chorda tympani and facial nerve of less than 1mm
  – Chorda tympani is difficult to evaluate in standard thin-slice temporal bone CT scans

PREOPERATIVE IMAGING CANNOT ALWAYS PREDICT DIFFICULTY OF FACIAL RECESS
Discussion

• Options for Difficult cases:
  – Subtotal petrosectomy with mastoid obliteration: risk of secondary cholesteatoma
  – Removal/Replacement of EAC: Fish and May Difficult
  – Cases of decompression and mobilization of facial nerve (Raines CH et al): risk of facial nerve injury
  – Transmastoid labyrinthotomy technique in common cavity or cystic cochleovestibular deformity: McElveen et al
The case for the retrofacial approach

- Common approach for an otologist/neurotologist
- Used in chronic ear surgery to access sinus tympani but also allows access to the mesotympanum
- No mobilization of the facial nerve needed
- No complex reconstruction
- Lower risk of iatrogenic long term complications
Conclusion

• Careful preoperative planning and imaging review is always necessary
• Ultimate decision might need to be made INTRAOPERATIVELY
• GOOD KNOWLEDGE OF MIDDLE EAR AND MASTOID ANATOMY AND EXPERTISE WITH VARIOUS APPROACHES ARE A MUST.
THANK YOU.
References


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