Successful Placement of a BAHA Implant in a Patient with Epidermolysis Bullosa: A Case Report and Review of the Literature

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Background

• Osseointegrated implants and their implantation techniques have evolved with time
  – No difference in complication rates\textsuperscript{1,2}
  – No difference in post-op pain or soft tissue reaction\textsuperscript{3,4}
• Are they useful/feasible in patients with chronic skin disorders?
Case

- 26 year old caucasian female
- History of recessive dystrophic epidermolysis bullosa
- Bilateral mixed hearing loss with EAC stenosis
- Concern that hearing aids may lead to further stenosis
• Complete EAC stenosis on the right and pinpoint introitus on the left

• Audiogram with headphones

• Decision made for BAHA Connect
Case

- LMA for anesthesia as general surgery to perform esophageal dilation.
- 5mm punch 55mm posterior to right EAC
- 4mm implant with 6mm abutment placed.
- Operative time: 14 minutes
- Bacitracin and healing cap.
- 1 year follow up: 1 mild local cellulitis treated with oral Bactrim.
Epidermolysis Bullosa

• Blistering following even minor trauma to skin
• 4 primary categories based on cleavage planes:\(^5\):
  – EB Simplex (EBS): epidermis
  – Junctional EB (JEB): dermal-epidermal junction
  – Dystrophic EB (DEB; most severe): dermis
  – Kindler Syndrome: multiple levels
Epidermolysis Bullosa

- Recessive Dystrophic EB (RDEB) most severe form with greatest number of mobidities:
  - Mitten deformities
  - Dental caries
  - Sepsis
  - Squamous Cell Carcinoma
  - Growth Restriction
  - Esophageal Scarring/Webbing/Obstruction
Epidermolysis Bullosa

- Sparse in the ENT literature
- Thawley et al 1977 – excised stenotic tissue and grafted
- Kastanioudakis et al 2000 – EAC stenosis after repeated AOE
- Lin and Fayad 2000 – BAHA for CHL in patient with EB. No details in case
- Hore et al 2009 – largest case series of ENT manifestations with 4 ear-related complaints. ¼ had successful cochlear implant
Punch Technique

• 2016 prospective cohort study comparing punch incision with epidermal flap and soft tissue reduction

• 40 patients
  – 20 punch
  – 20 flap

• Shorter surgical time, no difference skin reaction rates, no severe adverse events in punch group.

Conclusion

• Can use BAHA safely in patients with EB

• May also be good for others with blistering skin disorders
  – Pemphigus vulgaris
  – Bullous pemphigoid

• Prospective research is needed
Thank you!