DISCUSSION TOPICS

- Embryology
- Etiology
- Presentation
- Associated Anomalies
- Classification
- Management
AURICLE EMBRYOLOGY

- **Week 5** (inner ear week 3)
- 1\textsuperscript{st} and 2\textsuperscript{nd} branchial arches
- Hillocks 1-6
  - 1-3: 1\textsuperscript{st} branchial arch
  - 4-6: 2\textsuperscript{nd} branchial arch
- Multiple theories of embryogenesis of final auricular structure
AURICLE EMBRYOLOGY

- Traditional theory:
  - 1 = tragus,
  - 2,3 = helix,
  - 4,5 = anti helix,
  - 6 = anti tragus and lobule

- Other theories: 4-6 form 85% of the auricle
AURICLE EMBRYOLOGY

THE 6 HILLOCKS OF HIS

EARLY FETUS

LATE FETUS

NEWBORN
AURICLE and EAC EMBRYOLOGY

- **Migration:**
  - Starts anterior
  - Migrates dorsal and cephalic: weeks 8-12
  - Final position: 20 weeks

- **EAC:**
  - 1\textsuperscript{st} branchial cleft
  - Epithelial plug: weeks 4-5
  - Begins recanalization: week 21
  - Open with formed TM: week 28
MICROTIA ETIOLOGY

- Vascular
  - Stapedial artery insult

- Teratogens
  - Retinoic acid inhibitors
  - Thalidomide
  - Mycophenolate mofetil

- Genetic
  - Chromosominal: XO, Trisomy 13, 15, 18, 21, 22
  - Other mutations: Treacher Collins syndrome, neural crest cell migration failure
MICROTIA

- Definition: “The abnormal development of the external ear that results in a malformed auricle”
  - Kelley & Scholes, 2007
- Incidence: 0.83-17.4/10,000 live births
- Strongly associated with hearing loss
  - 80% have conductive hearing loss
  - 20% have sensorineural hearing loss
- Associated with psychological stigma and burden
MICROTIA: PRESENTATION

- More common in males:
  - Male: Female ratio = 2.5:1
- More common in Japanese, Hispanic, Native American
- Prevalence increased at high altitudes
- Multiparity
MICROTIA: PRESENTATION

- Unilateral in 90% of cases
- Bilateral in only 10% of cases
- Right side > Left side (60% right)
- Hearing loss normally in affected ear
  - can be bilateral or in the ear without microtia
MICROTIA: ASSOCIATED ANOMALIES

- 50% microtia cases associated with other anomalies
- Common associated anomalies:
  - Congenital aural atresia (CAA)
    - Present in almost all cases of severe microtia
      - Cholesteatoma (Associated with CAA)
  - Hemifacial microsomia
  - Acrofacial Dysostosis
MICROTIA: ASSOCIATED ANOMALIES

- Associate anomalies:
  - Goldenhar syndrome
MICROTIA: ASSOCIATED ANOMALIES

- Associated anomalies:
  - Treacher Collins Syndrome
MICROTIA: CLASSIFICATION

- Marx Classification
- Type 1:
  - Mild deformity
  - All structural components of auricle present
MICROTIA: CLASSIFICATION

- Type 2:
  - Atypical microtia
  - Some auricular structures
  - Helical changes
  - Auditory meatus patent
MICROTIA: CLASSIFICATION

- Type 3:
  - Classic microtia
  - Few auricular structures
  - CAA
  - Conchal or lobular remnant (MC is lobular)
MICROTIA: CLASSIFICATION

- Type 4:
  - Anotia
  - No auricular structures
  - CAA
MICROTIA: AUDIOLOGY

- **Hearing status**
  - Degree of microtia associated with degree of middle ear deformity
  - CHL common in microtic ear
  - Non microtic ear can have hearing loss
  - Protect normal hearing ear
    - Lower threshold for ventilation tubes
  - Bilateral hearing loss
    - Indication for bone anchored hearing aids
MICROTIA: MANAGEMENT

• Observation
  - Type 1 microtia
  - Prior to surgery: 5-8 years
  - Advantages: no risk, possibility for future reconstruction
  - Disadvantages: cosmesis, psychosocial issues
  - Main focus: hearing and speech
MICROTIA: MANAGEMENT

- Prosthesis
  - Adhesive: possible removal of microtic ear
  - Magnetic: microtic ear removed, anchors placed in a 2 step surgical procedure
  - Usage: failed or cannot have reconstruction
MICROTIA: MANAGEMENT

- **Prosthesis**
  - **Advantages:** cosmesis
  - **Disadvantages:**
    - Adhesive:
      - future reconstruction difficult
      - dislodgment
      - remove at night
      - cost
    - Magnetic:
      - no future reconstruction
      - remove at night/daily maintenance
      - cost
      - surgery
MICROTIA: MANAGEMENT

Reconstruction

- Types:
  - Autogenous rib: four step procedure
  - Medpor: porous polyethylene, more difficult procedure

- Advantages: cosmesis, low maintenance
  - Autogenous rib: lower risk for extrusion/infection
  - Medpor: no donor site morbidity

- Disadvantages: surgery, flap failure, scar
  - Autogenous rib: donor site morbidity, pneumothorax
  - Medpor: higher risk for extrusion/infection
MICROTIA: MANAGEMENT

- Reconstruction
  - Autogenous rib reconstruction stages
    - 1: Cartilage implantation
    - 2: Lobule transfer
    - 3: Creation of post auricular sulcus
    - 4: Tragus reconstruction
  - Complications:
    - Hematoma
    - Pneumothorax
MICROTIA: MANAGEMENT

- Reconstruction:
  - Autogenous
  - Medpor
SUMMARY

- Microtia is auricular failure to develop
- Embryology: branchial arches 1-2
- Etiology: mainly sporadic, possible genetic causes
- Associated anomalies: 50% of cases
  - commonly associated with CAA
SUMMARY

- Type 1: minimal deformity
- Type 2: helix deformity
- Type 3: no recognizable auricular structures
- Type 4: no auricle

Management: observation, prosthesis, reconstruction
  - Monitor hearing and speech
NEW DEVELOPMENTS

- Vacanti Mouse: biodegradable scaffolding
  - http://www.youtube.com/watch?v=LHHe_q3DNas
- Stelarc
RESOURCES


