Otology seminar

Image of Congenital Malformation of the Inner Ear

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Introduction

Embryogenesis:
- 3th wks: otic placode
- 4th wks: otocyst (otic vesicle)
- 5th wks: three portion
- 8th wks: Cochlea 1 \( \frac{1}{2} \) turns
- 10th wks: Cochlea 2 \( \frac{1}{2} \) turns
- 26-28th wks: hair cell & auditory neuron
- Birth: ossification of the otic capsule completed
4.5th weeks

endolympathic duct

utricular & saccular portion
5th weeks

- superior scc
- endolympathic duct
- saccule
- posterior scc
- cochlear duct
7th weeks

- superior scc
- endolymphatic duct
- cochlear duct
- lateral scc
- posterior scc
22th weeks
Embryology
Introduction

- A premature arrest or aberration theory
- Evidence: the resemblance of most malformed inner ears to the appearance of the inner ear during embryogenesis
- General rule: the earlier the developmental arrest the more severe the deformity and the worse the hearing will be.
Introduction
Introduction-Classification
(Jackler, 1987)

Malformation limited to the membranous labyrinth
Complete membranous labyrinthine dysplasia (Siebenmann-Bing)
Limited membranous labyrinthine dysplasia
    Cochleosaccular dysplasia (Scheibe)
    Cochlear basal turn dysplasia

Malformation of the osseous and membranous labyrinth
Complete labyrinthine aplasia (Michel)
Cochlear anomalies
    Cochlear aplasia
    Cochlear hypoplasia
    Incomplete partition (Mondini)
    Common cavity
Labyrinthine anomalies
    Semicircular canal dysplasia
    Semicircular canal aplasia
Aqueductal anomalies
    Enlargement of the vestibular aqueduct
    Enlargement of the cochlear aqueduct
Internal auditory canal abnormalities
    Narrow internal auditory canal
    Wide internal auditory canal
HRCT of Temporal bone

Axial view
HRCT of Temporal bone
Axial view
Malformation limited to the membranous labyrinth

- Over 90% of congenital deafness
- Bony labyrinth is normal; Not clinical use; based on histopathologic exam
- Classification:
  1. Complete membranous labyrinthine dysplasia (Siebenmann-Bing)
  2. Limited membranous labyrinthine dysplasia
     - Cochleosaccular dysplasia (Scheibe)
     - Cochlear basal turn dysplasia
Complete membranous labyrinthine dysplasia (Siebenmann-Bing)

- First described by Siebenmann and Bing in 1907
- Extremely rare
- Associated with cardioauditory (Jervell and Lange-Neilsen) and Usher's syndrome
Limited membranous labyrinthine dysplasia

Cochleosaccular dysplasia (Scheibe, 1892)

- Most frequent histopathologic finding in congenital deafness
- Pars inferior (cochlea and saccule) involvement; SCCs and utricle are normal
- Cochlea: organ of Corti is partial or complete missing; cochlear duct collapse; Reissner`s membrane adherent to the limbus; stria vascularis: colloidal inclusions
- Saccule: collapse
- Hearing: variable
Limited membranous labyrinthine dysplasia

Cochlear basal turn dysplasia

- Related familial high-frequency SNHL
- Hearing: from minimal symptomatic to normal hearing (compensated)
Malformations of the membranous and osseous labyrinth

- Complete labyrinthine aplasia (Michel)
- Cochlear anomalies
  1. Cochlear aplasia
  2. Cochlear hypoplasia
  3. Incomplete partition (Mondini)
  4. Common cavity
- Labyrinthine anomalies
  1. Semicircular canal dysplasia
  2. Semicircular canal aplasia
- Aqueductal anomalies
  1. Enlargement of the vestibular aqueduct
  2. Enlargement of the cochlear aqueduct
- Internal auditory canal abnormalities
  1. Narrow internal auditory canal
  2. Wide internal auditory canal
I. Complete labyrinthine aplasia (Michel aplasia)

- Severest deformity and exceedingly rare
- Arrest occurs before the formation of otic vesicle (3th weeks)
- Total deaf
- Associated with anencephaly and thalidomide exposure, external ear abnormalities
- Incidence - Over estimated; less than 1% of all congenital inner ear malformation
- Confused with labyrinthine ossification (acquired during life, a sizable and dense otic capsule)
Complete labyrinthine aplasia
(Michel aplasia)

HRCT, axial view
- Absence of any inner ear structure
- IAC: missing
- Malleus, Incus: normal
Michel dysplasia
9 y/o girl, bil. profound congenital SNHL

Axial CT scan
- Petrous bone aplasia
- Inner ear: absent
- Medial wall of middle ear: flattened
- Malleus: normal
Michel dysplasia
9 y/o girl, bil. profound congenital SNHL

Coronal CT scan
- Normal EAC and middle ear
- Malleus, incus(+), stapes(-)
- Dehiscence of medial wall of ME; internal acoustic meatus
II. Cochlear anomalies

- Cochlear aplasia
- Cochlear hypoplasia
- Incomplete partition (Mondini)
- Common cavity
Cochlear aplasia

- Cochlea is complete absent
- Arrest in the development of the cochlear bud (5th weeks)
- Rare, deaf
- Radiographically, a vestibule and SCCs (usually deformed, dilated or hypoplastic)
- D/D: labyrinthine ossification; by otic capsule bone anterior to the IAC and promontary
3D-cochlear aplasia

(1) anterior view  (2) posterior view  (3) lateral view  (4) anterolateral view
(5) posteromedial view  (6) inferior view  (7) superior view
Animation
Cochlear aplasia-Vertical rotation
Animation
Cochlear aplasia-Horizontal rotation
Cochlear aplasia

CT, axial

- Cochlea: absence
- Vestibule: dilated
- Facial n. located at the usual location of the cochlea
Cochlear aplasia, bilateral
Cochlear hypoplasia

- Arrest during 6th week
- A hypoplastic cochlea consisting of a single turn or less
- 15% of all cochlear anomalies
- Hearing is variable
- A small bud of variable length (1 to 3 mm) protrudes from the vestibule
- 50%; the vestibule is frequently enlarged with accompanying SCCs malformation
3D-cochlear hypoplasia

(1) anterior view  (2) posterior view  (3) lateral view  (4) anterolateral view

(5) posteromedial view  (6) inferior view  (7) superior view
Animation
Cochlear hypoplasia-Vertical rotation
Animation
Cochlear hypoplasia-Horizontal rotation
Cochlear hypoplasia

Axial CT

- C: small cochlear bud
- ▽: IAC; small and narrow
- ▼: facial n.; separate channel
Incomplete partition (Mondini)
Roert K. Jackler-Ch 27

- Arrest at 7th week
- Cochlea only 1.5 turn
- Most common type of cochlear malformation (>50%)
- Radiographically:
  1. Cochlear size: 5-6mm vertically (normal: 8-10mm)
  2. Absence of a scalar septum, partially or completely
- than the number of cochlear turn
Incomplete partition (Mondini)

- Histologically: correlate of Mondini dysplasia, Organ of Corti: variable
- Auditory function: variable; normal to profound deafness; 41 ears, three-tone average: 75 dB
- 20%; SCC deformities
3D-Mondini dysplasia

(1) anterior view  (2) posterior view  (3) lateral view  (4) anterolateral view
(5) posteromedial view  (6) inferior view  (7) superior view
Animation
Mondini dysplasia-Vertical rotation
Animation
Mondini dysplasia—Horizontal rotation
Incomplete partition (Mondini)
HRCT, coronal
Incomplete partition
Severe v.s. Mild
Incomplete partition
New classification

- Incomplete partition
  1. Incomplete partition type I (IP-I) cystic cochleovestibular malformation
  1. Incomplete partition type II (IP-II) Mondini malformation
Incomplete partition type I (IP-I) cystic cochleovestibular malformation

- A cystic dilated vestibule accompanied the cystic, empty cochlea
- The dimensions of the cochlea and vestibule are normal
- Arrest at 5th week
- The cribriform area between the cochlea and IAC is defective
- The vestibule was dilated in all case
- No enlarged VA (vestibular aqueduct)
Incomplete partition type I (IP-I)

Axial CT: Empty, cystic cochlea (without any partition); Dilated vestibule
Incomplete partition type II (IP-II) Mondini malformation

- Cochlea: 1.5 turn; only the middle and apical turns form a cystic cavity
- The size of the cochlea and vestibule are normal
- Arrest at 7th week
- Vestibular dilation is minimal than IP-I
- All had enlarged VA, bilateral and symmetric
Incomplete partition type II (IP-II) Mondini malformation

- 60%; enlarged IAC
- Slattery and Luxford: ganglion cell in human are found in the lower 1.5 turns of the cochlea
Incomplete partition type II (IP-II) Mondini malformation

Axial CT: dilated vestibule; EVA; confluence of middle and apical turn
Histopathological slides
## IP-I v.s. IP-II

<table>
<thead>
<tr>
<th></th>
<th>IP-I</th>
<th>IP-II</th>
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</thead>
<tbody>
<tr>
<td>Cochlea</td>
<td>Empty cyst</td>
<td>Middle &amp; apical turns form cystic cavity</td>
</tr>
<tr>
<td>Arrest</td>
<td>5th wks</td>
<td>7th wks</td>
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<tr>
<td>Modiolus</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Vestibular dilation</td>
<td>++</td>
<td>+</td>
</tr>
<tr>
<td>Enlarged VA</td>
<td>-</td>
<td>+, bilateral symmetric</td>
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<tr>
<td>Jackler et al.</td>
<td>Severe</td>
<td>Mild</td>
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<tr>
<td>Phelps et al.</td>
<td>Pseudo Mondini (meningitis)</td>
<td>Classic Mondini</td>
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</tbody>
</table>
Common cavity

- The cochlea and vestibule are confluent
- Arrest at 4th week (otocyst stage)
- Size of the cyst vary; average: 7mm vertically, 10mm horizontally
- A large common cavity had an enlarged IAC
- IAC enter the common cavity at its center
Common cavity

- Histologically: cyst cavity wall containing a primordia of the membranous labyrinth
- Hearing is usually poor
3D-common cavity

(1) anterior view
(2) posterior view
(3) lateral view
(4) anterolateral view
(5) posteromedial view
(6) inferior view
(7) superior view
Animation
Common cavity-Vertical rotation
Animation
Common cavity-Horizontal rotation
Common cavity deformity with enlarged internal auditory canal
Labyrinthine anomalies

- Semicircular canal dysplasia
- Semicircular canal aplasia
Semicircular canal dysplasia

- Dysplasia of the lateral SCC is a common type of inner ear malformation.
- 40% malformed cochlea accompanying dysplasia of the lateral SCC.
- Arrest at 6th week.
- Lateral SCC > posterior or superior SCC (earlier in embryogenesis).
Semicircular canal dysplasia

- Radiographic: a short, broad cystic space confluent with the vestibule
- **Caloric test:** most: absent or reduced; few: normal
- **Hearing:** normal or near-normal
- Associated with conductive hearing loss due to congenital stapes fixation
Semicircular canal dysplasia
3S-LSCC dysplasia
Animation
LSCC dysplasia-Vertical rotation
Animation
LSCC dysplasia-Horizontal rotation
Semicircular canal aplasia

- Only **one fourth** as common as SCC dysplasia
- Usually associated with cochlear anomalies
- Arrest **before 6th week**
Malformations of the vestibular and cochlear aqueduct

- Enlargement of the vestibular aqueduct
- Enlargement of the cochlear aqueduct
Enlargement of the vestibular aqueduct (VA)

- The most common radiographically detectable malformation of the inner ear
- The diameter of a normal VA: between 0.4 and 1.0 mm (halfway between the common crus and its external aperture)
- Enlarged VA: > 2.0mm
Enlargement of the vestibular aqueduct (VA)

- Large VA syndrome (Valvassori`s and Clemis`:): sole detectable abnormalities
- 5th week: a short, broad pouch; “J” shape (adulthood)
- Histologically: sac and aqueduct are thin-walled and lack both vascularity and rugose features
Enlargement of the vestibular aqueduct (VA)

- Bilateral
- Hearing:
  1. normal or mild impaired in children then gradually deteriorate through childhood,
  2. variable; 40% profound SNHL
  3. Sudden decrements following head trauma
Enlargement of the vestibular aqueduct (VA)

- Associated CHL due to stapes fixation; avoid stapes surgery (CSF otorrhea)
- Large VA syndrome; familial clusters
- Surgical intervention (endolymphatic sac):
  1. Shunt; postoperative hearing loss; no benefit
  2. Obliteration: muscle or fascia, no conclusion
Enlargement of the vestibular aqueduct (VA)

- Evidences of acquired deformity, ?:
  1. The bone surrounding the VA may showed signs of erosion (not stable)
  2. The existence of CSF pressure (gusher) within the inner ear (CI & stapedectomy)
  3. CT or MRI finding: a defect of the cochlear modiolus at the distal end of the IAC
3D-Enlarged Vestibular Aqueduct

(1) anterior view  (2) posterior view  (3) lateral view  (4) anterolateral view
(5) posteromedial view  (6) inferior view  (7) superior view
Animation
EVA-Vertical rotation
Animation
EVA-Horizontal rotation
EVA v.s. Normal
Enlargement of the cochlear aqueduct (CA)

- **Jackler:** nerve seen in radiographic evidence
- Misinterpretation: wide internal funnel that opens into the posterior fossa
- Medial orifice: 3-4 mm; > 10mm
- **Criteria:**
  1. > 1mm; lateral orifice
  2. > 2mm between inner and posterior fossa
Enlargement of the cochlear aqueduct (CA)

- Schuknecht and Reisser: 1400 temporal bone (29 congenital inner ear malformation): no CA > 0.2 mm at narrowest
- Clinical standpoint: <1mm; undetectable
Enlargement of the cochlear aqueduct (CA)

* Normal CA. The medial aperture typically lies inferior to the IAC and just above the jugular bulb.
Developmental anomalies of the internal auditory canal

- Narrow internal auditory canal
- Wide internal auditory canal
Narrow internal auditory canal

- A failure of eight nerve development
- Normal facial function and IAC less than 3 mm in diameter: only the facial n.
- A relative contraindication to CI
Narrow internal auditory canal
Wide internal auditory canal

- Incidental finding in healthy individuals
- Criteria: > 10 mm in diameter
- Associated with spontaneous CSF leakage and gusher during stapes surgery (hearing prognosis poor)
- Contraindicate stapedectomy
Reference

- Jackler RK. Ch 27 Congenital Malformation of the Inner Ear, Pediatric Otolaryngology, Cummings
Reference