



knowledge changing life

OVAL WINDOW CSF LEAK ASSOCIATED WITH INCOMPLETE PARTITION TYPE 1 OF THE COCHLEA: REVIEW OF REPAIR TECHNIQUES

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DISCLOSURES

I have nothing to disclose.

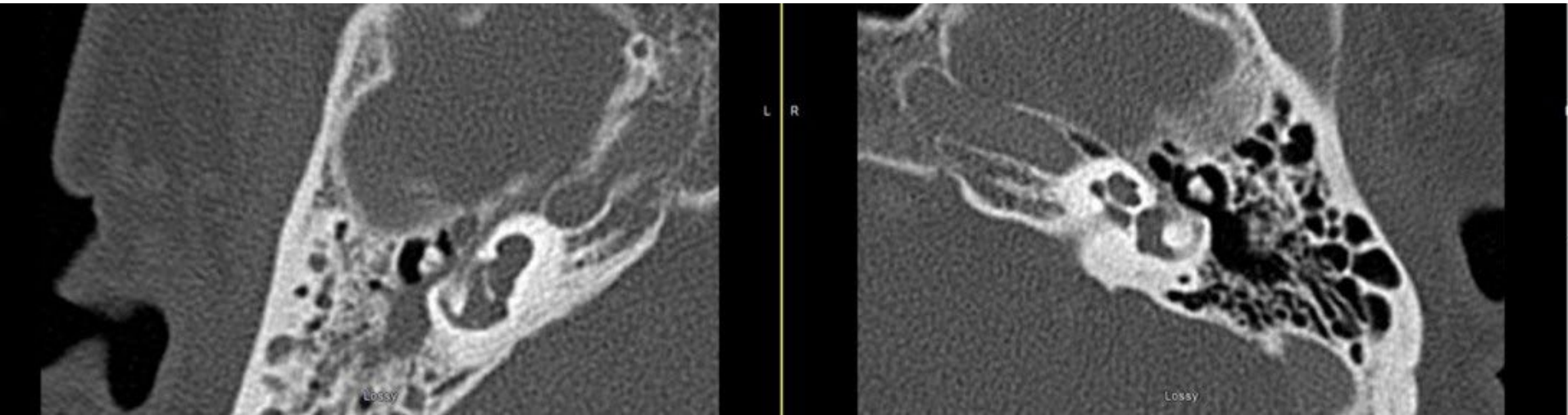
CASE REPORT

67-year-old man presented with right-sided CSF rhinorrhea of unknown duration

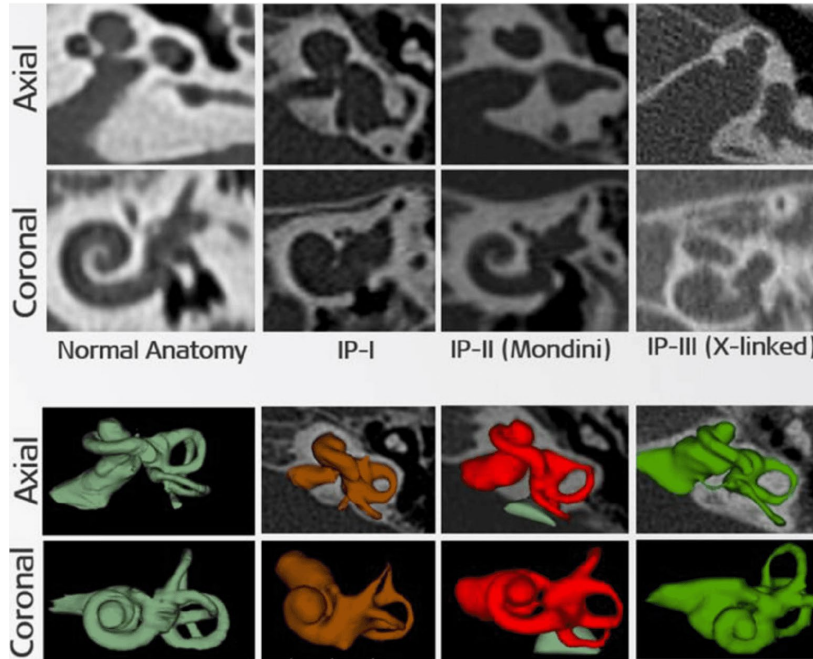
- PMH: 4 episodes meningitis (1st episode at 46), congenital profound right SNHL
- PE: bilateral tympanic membranes were unremarkable. No spontaneous or gaze evoked nystagmus was present. Audiogram demonstrated profound SNHL on the right. Clear rhinorrhea from the right naris tested positive for beta 2 transferrin.

CASE REPORT CONTINUED

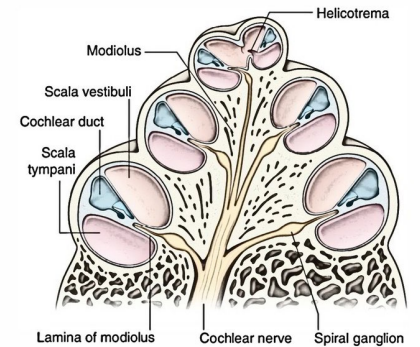
CT scan demonstrated type 1 incomplete partition of the cochlea on the right with a bony defect at the oval window



INCOMPLETE PARTITION



- Congenital anomaly of the inner ear
- cochlea is clearly separated from the vestibule and the external contour of the cochlea is nearly normal in size, but there are defects involving the modiolus and interscalar septa
- Results in SNHL
- 3 Types
 - IP-II (Mondini) is the most common of all inner ear malformations, but IP-I has a higher incidence of CSF leak/meningitis.



COMPLICATIONS OF IP

- Profound SNHL
- CSF Leak
- Recurrent meningitis
 - usually presents in childhood, our patient was asymptomatic until adulthood

CASE CONTINUED

- Surgical Repair
 - Transmastoid repair of CSF leak and fat graft obliteration of the mastoid (complete mastoidectomy was performed)
 - After opening the facial recess, mucosal adhesions were noted at the stapes superstructure with ballooning of the mucosa below the stapes and on the promontory
 - incus was mobilized and extracted through the antrum
 - A clear CSF leak was noted through the oval window between the tympanic segment of the facial nerve and stapes superstructure

CASE CONTINUED



CASE CONTINUED

- The Eustachian tube orifice was obliterated with strips of fascia wedged into place by the incus body after removal of the long process
- Small pieces of fascia, fat, and fibrin sealant were then placed into the middle ear space
- Fascia was then packed directly into the oval window between the tympanic segment of the facial nerve and the stapes superstructure at the site of the CSF leak, sealing the defect
- Fat and fibrin sealant were placed in an alternating fashion into the mastoid cavity until it was completely packed

FOLLOW UP

- 1 week
 - no rhinorrhea or vestibular upset
- 12 weeks
 - *Streptococcus pneumoniae* meningitis
 - Developed intermittent right-sided, clear rhinorrhea consistent with recurrent CSF leak

REVISION SURGERY

- Stapes superstructure and footplate were completely removed
- Vestibule was firmly packed with bone wax, perichondrium and cartilage in layers
- Eustachian tube was also repacked with cartilage and a perichondrial graft
- Middle ear space and mastoid were then packed with multiple layers of fat and fibrin sealant
 - At 6-month follow up, he had experienced no further CSF rhinorrhea or episodes of meningitis

REVIEW OF LITERATURE

- Review of literature of patients presenting with recurrent meningitis with IP Type 1
 - Systematic review
- **SEARCH PHRASE:**
 - Incomplete partition + Mondini + cochlear anomaly
- **INCLUSION CRITERIA:**
 - Must specify that defect was repaired
 - Must present with recurrent meningitis
 - What age initial presentation of recurrent meningitis was (at least over or under 18)
- 13 case reports and case series encompassing 30 total patients met the criteria

Author	Year	No. cases	Sex	Initial Presentation	Inclusion Criteria	Exclusion Criteria
Alford et al	2020	46	M	Single disease, vestibular abnormalities with recurrence	Temporal meningitis, cerebellar atrophy, hydrocephalus, IP Type 1, cochlear anomalies, and facial nerve palsy	None, None, 6 months, following surgery
Alford et al	2020	46	M	Vestibular abnormalities	Temporal meningitis	None at 12 months
Chen et al	2019	13	M	Vestibular abnormalities	Temporal meningitis and facial	None at 12 months
Chen et al	2019	18	F	Vestibular abnormalities	Temporal meningitis and facial	None at 12 months
Hong et al	2019	10	Child ages	Childhood	Childhood	Childhood
Hong et al	2019	10	Child ages	Childhood	Childhood	Childhood
Chen et al	2016	7	M	Vestibular abnormalities	Temporal meningitis	Childhood
Chen et al	2016	8	F	Vestibular abnormalities	Temporal meningitis	Childhood
Hong et al	2016	2	M	Vestibular abnormalities with recurrence, vestibular abnormalities	Childhood, Temporal meningitis and facial	Childhood, None at 12 months following surgery
Hong et al	2016	14	F	Vestibular abnormalities with recurrence, vestibular abnormalities	Childhood, Temporal meningitis and facial	Childhood, None at 12 months following surgery
Hong et al	2016	2	M	Vestibular abnormalities	Temporal meningitis and facial	None at 12 months
Hong et al	2016	5	M	Vestibular abnormalities with recurrence, vestibular abnormalities	Childhood, Temporal meningitis and facial	Childhood, None at 12 months following surgery
Hong et al	2016	4	F	Vestibular abnormalities	Temporal meningitis and facial	None at 12 months
Hong et al	2016	3	F	Vestibular abnormalities	Temporal meningitis and facial	None at 12 months
Hong et al	2016	13	M	Vestibular abnormalities	Temporal meningitis and facial	None at 12 months
Hong et al	2016	5	F	Vestibular abnormalities	Temporal meningitis and facial	None at 12 months
Hong et al	2016	4	M	Vestibular abnormalities	Temporal meningitis and facial	None at 12 months
Hong et al	2016	11	F	Vestibular abnormalities	Temporal meningitis and facial	None at 12 months
Chen et al	2014	12	F	Vestibular abnormalities	Temporal meningitis, cerebellar atrophy, hydrocephalus, IP Type 1, cochlear anomalies, and facial nerve palsy	None (temporal meningitis, cerebellar atrophy)
Chen et al	2014	11	M	Vestibular abnormalities	Temporal meningitis, cerebellar atrophy, hydrocephalus, IP Type 1, cochlear anomalies, and facial nerve palsy	None (temporal meningitis, cerebellar atrophy)
Chen et al	2014	12	F	Single disease, vestibular abnormalities with recurrence	Childhood, Temporal meningitis, cerebellar atrophy, hydrocephalus, IP Type 1, cochlear anomalies, and facial nerve palsy	Childhood, None following surgery (temporal meningitis, cerebellar atrophy)
Chen et al	2014	11	F	Vestibular abnormalities	Temporal meningitis, cerebellar atrophy, hydrocephalus, IP Type 1, cochlear anomalies, and facial nerve palsy	None (temporal meningitis, cerebellar atrophy)
Chen et al	2014	7	M	Vestibular abnormalities	Temporal meningitis, cerebellar atrophy, hydrocephalus, IP Type 1, cochlear anomalies, and facial nerve palsy	None at 9 years
Chen et al	2014	075	M	Multiple ear and function tests, vestibular evoked myogenic audiogram	Single disease	None at 9 months
Chen et al	2014	6	M	Single disease	Temporal meningitis and facial	None (temporal meningitis)
Chen et al	2013	26	M	Childhood	Childhood	Childhood
Chen et al	2011	0.5	M	Childhood	Temporal meningitis, cerebellar atrophy, hydrocephalus, IP Type 1, cochlear anomalies, and facial nerve palsy	Childhood (temporal meningitis, cerebellar atrophy)
Chen et al	2011	1.4	M	Single disease	Temporal meningitis, cerebellar atrophy, hydrocephalus, IP Type 1, cochlear anomalies, and facial nerve palsy	None at 6 months
Chen et al	2009	4.2	Child ages	Vestibular abnormalities	Temporal meningitis and facial	None at 19 months
Chen et al	2007	4.6	Child ages	Vestibular abnormalities	Temporal meningitis and facial	None at 19 months

REVIEW OF LITERATURE

- Age of onset
 - 13% (4/30) Patients presented in adulthood
 - Average age onset in patients under 18 was 6.3
- Operative techniques
 - Vestibular obliteration and Closure without obliteration (simple packing with various materials)

REVIEW OF LITERATURE

- Operative techniques continued
 - 27/30 cases described materials used in surgery

LIMITATIONS

- Small sample size
 - This review only accounts for cases up to February 2020, IP first classified in 2002
- Outcomes may be surgeon/skill dependent

DISCUSSION

- Adult presentation: was defect in oval window always there and was not previously picked up vs trauma or increased ICP contribution
 - No previous CT temporal bone
- Research supports packing to obliterate vestibule
 - Risk of recurrence high
 - Cannot comment on materials used and outcomes, additionally cannot comment on outcomes with approach

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