HỘI TAI MŨI HỌNG VIỆT NAM

VIETNAMESE SOCIETY OF OTORHINOLARYNGOLOGY

COCHLEAR IMPLANTATION IN ANATOMY CONGENITAL MALFORMATION PATIENTS AT ENT HOSPITAL HO CHI MINH CITY

MINH LE TRAN QUANG, PhD, MD
THAI KHUU MINH, MD

OUTLINE

1. Introduction
2. Methods
3. Results - Discussion
4. Conclusion

INTRODUCTION

- Prelingual sensorineural hearing loss (SNHL) deprives children of integration into normal life: education, employment and psychosocial status.

- Cochlear implantation are currently a treatment approved worldwide for these patients .

INTRODUCTION

- Anatomy congenital malformations contributes 20% of pediatric SNHL cases.
- Cochlear implantation in these children is challenging and postoperative outcomes vary across individuals.
- Anatomy congenital malformations include: inner ear malformations and cochlear nerve deficiency.

Sennaroğlu L, Bajin MD. Classification and Current Management of Inner Ear Malformations. Balkan medical journal. Sep 29 2017; 34(5):397-411. doi:10.4274/balkanmedj.2017.0367.

INTRODUCTION

- Classification of inner ear malformations - Sennaroglu (2017): 8 groups
  - Complete Labyrinthine Aplasia (Michel Deformity)
  - Rudimentary Otocyst
  - Cochlear Aplasia
  - Common Cavity
  - Cochlear Hypoplasia
  - Incomplete Partition of the Cochlear
  - Enlarged Vestibular Aqueduct
  - Cochlear Aperture Abnormalities

Sennaroğlu L, Bajin MD. Classification and Current Management of Inner Ear Malformations. Balkan medical journal. Sep 29 2017; 34(5):397-411. doi:10.4274/balkanmedj.2017.0367.

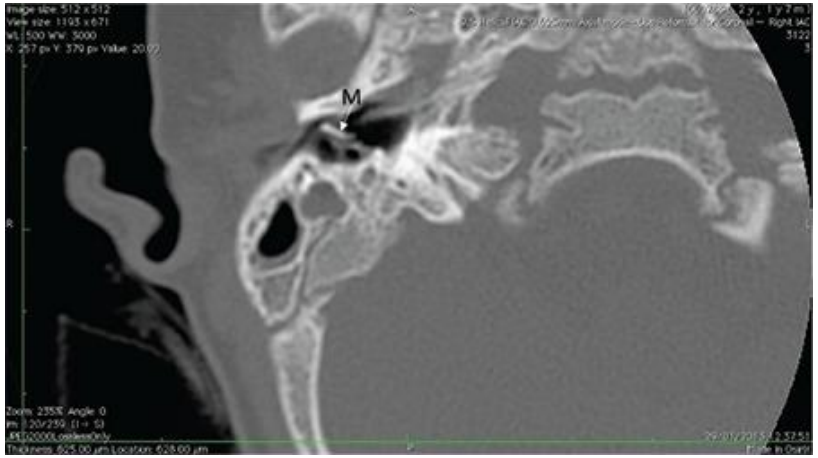
INTRODUCTION

- Cochlear nerve deficiency includes:
  - Hypoplasia: There is a separate CN but the size is less than the contralateral normal CN or ipsilateral normal facial nerve
  - Aplasia: There is no nerve in the anteroinferior part of the internal auditory canal

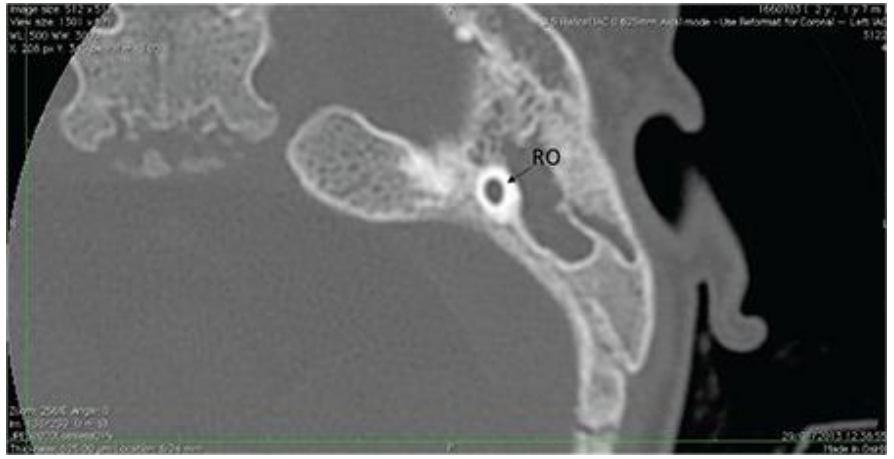
Iseli C, Adunka O, Buchman C. Cochlear Nerve Deficiency. Pediatric Cochlear Implantation. 2016:227-235.

INTRODUCTION

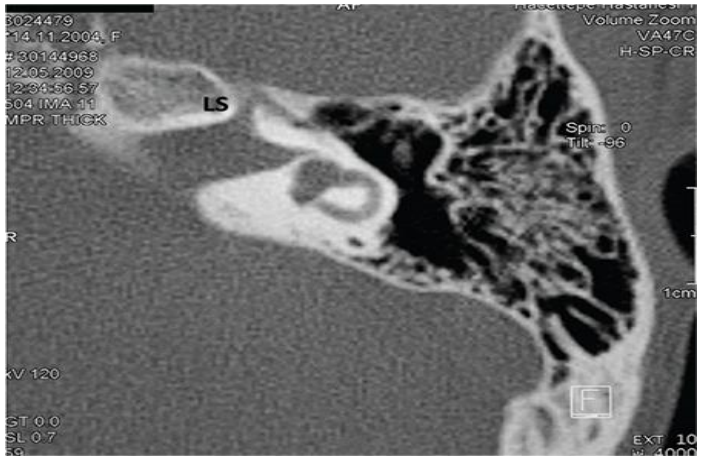
The objective of this research was to present and analyze the imaging, audiological features and cochlear implantation outcomes in anatomy congenital malformation patients.



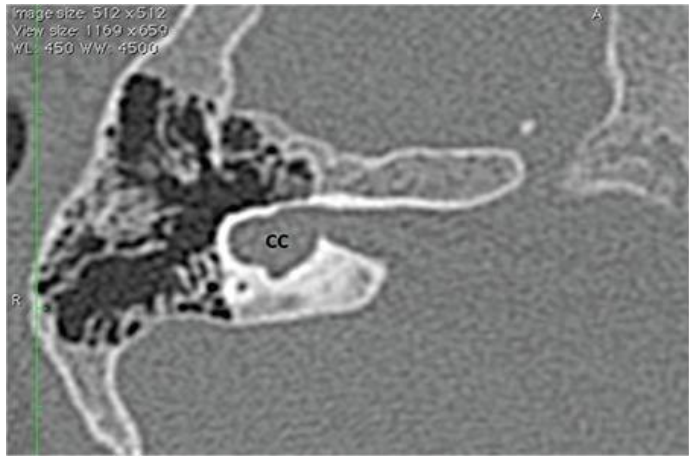
Complete Labyrinthine Aplasia (Michel Deformity)



Rudimentary Otocyst

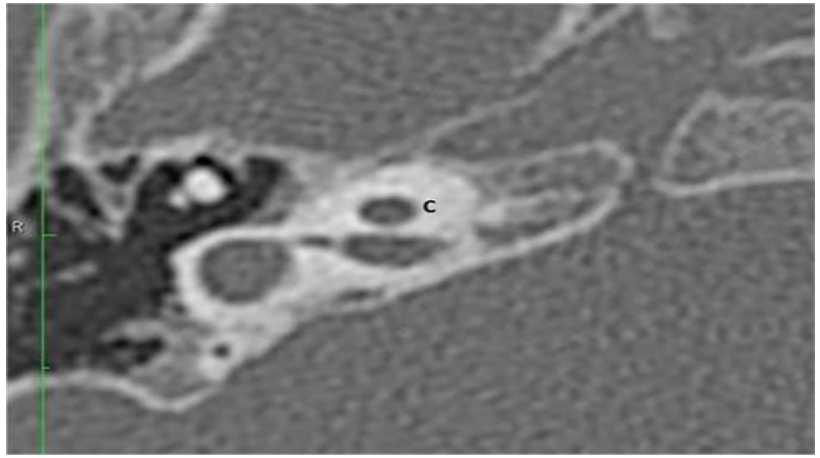


Cochlear Aplasia

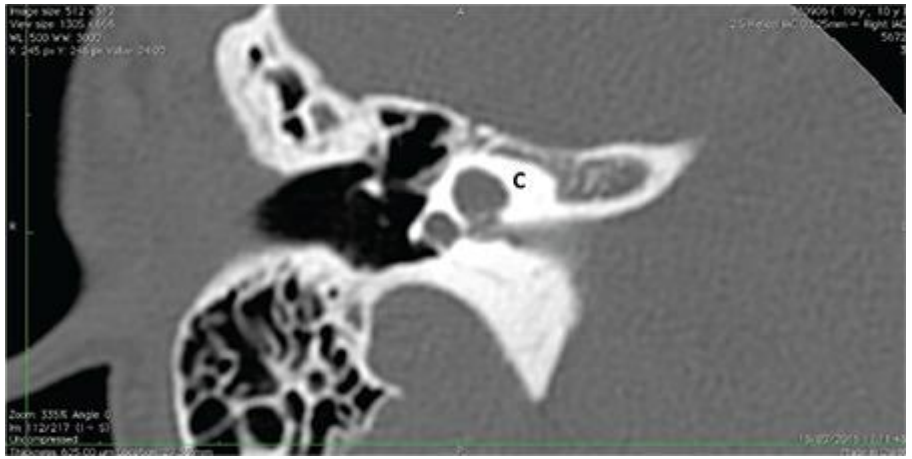


Common Cavity

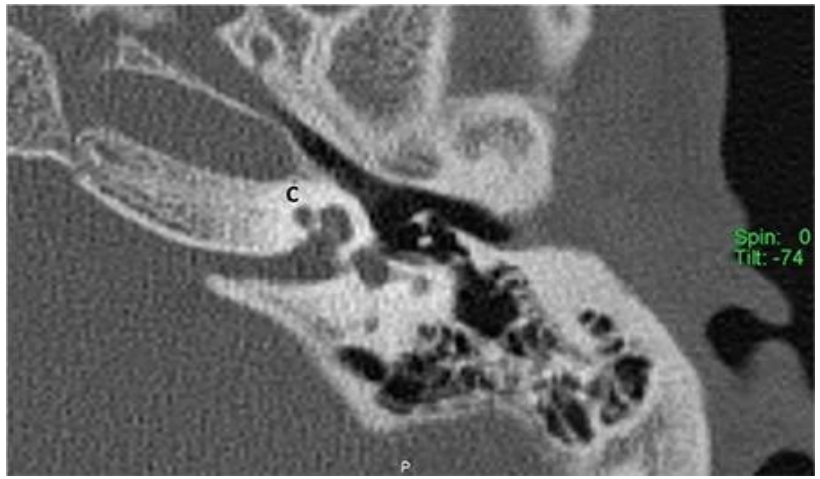
Sennaroğlu L, Bajin MD. Classification and Current Management of Inner Ear Malformations. Balkan medical journal. Sep 29 2017; 34(5):397-411. doi:10.4274/balkanmedj.2017.0367.



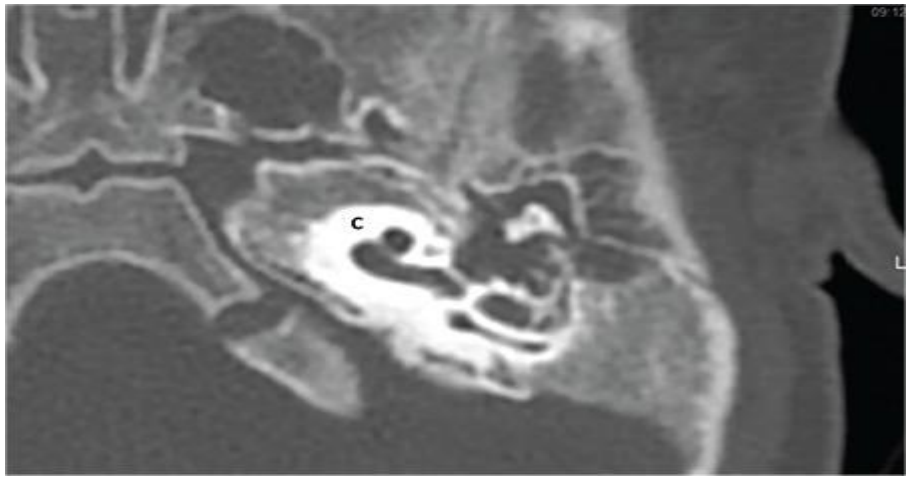
Cochlear Hypoplasia type I



Cochlear Hypoplasia type II



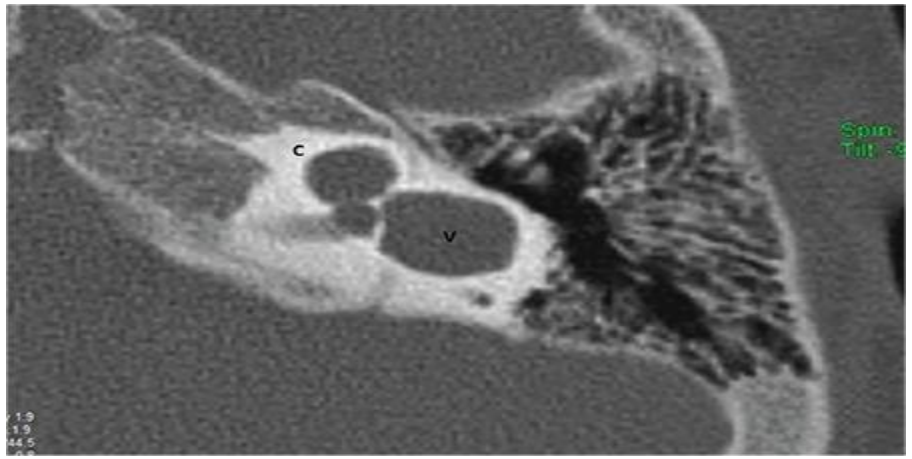
Cochlear Hypoplasia type III



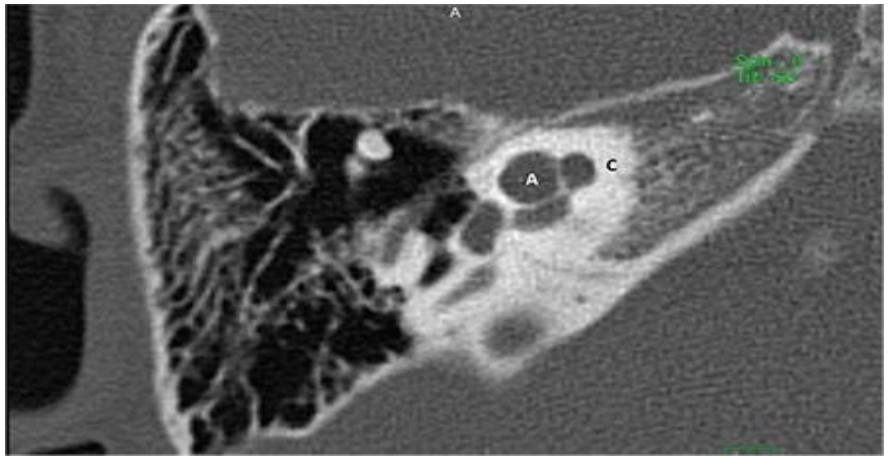
Cochlear Hypoplasia type IV

Cochlear Hypoplasia

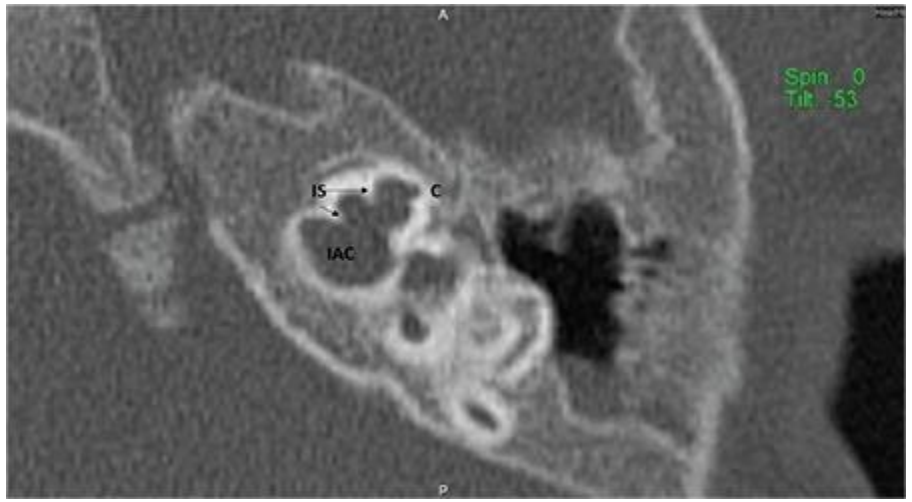
Sennaroğlu L, Bajin MD. Classification and Current Management of Inner Ear Malformations. Balkan medical journal. Sep 29 2017; 34(5):397-411. doi:10.4274/balkanmedj.2017.0367.



IP - I



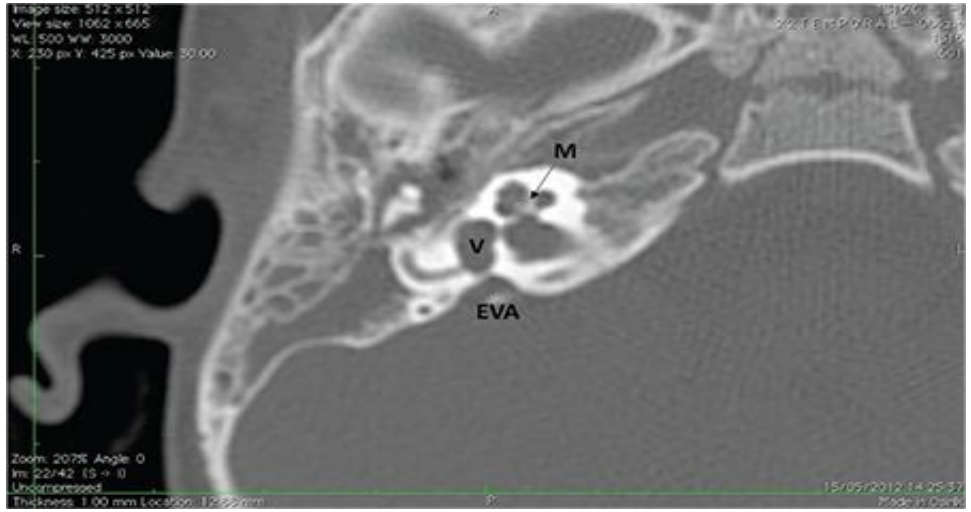
IP - II



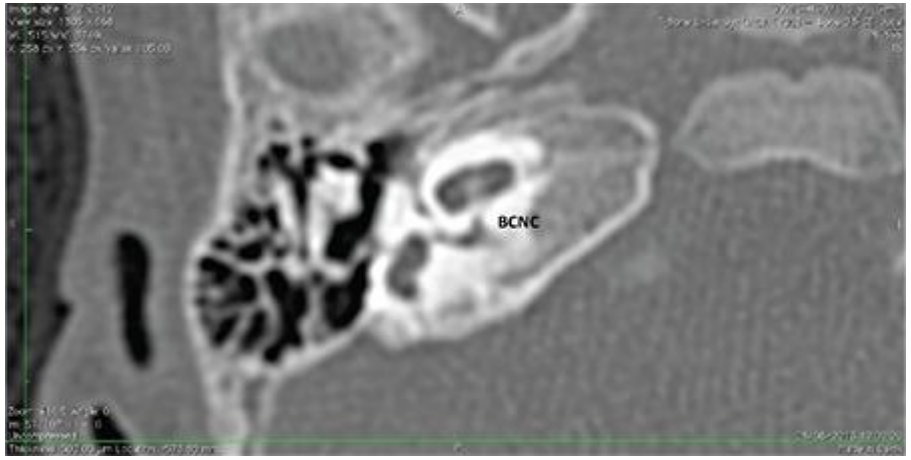
IP - III

Incomplete Partition of the Cochlear (IP)

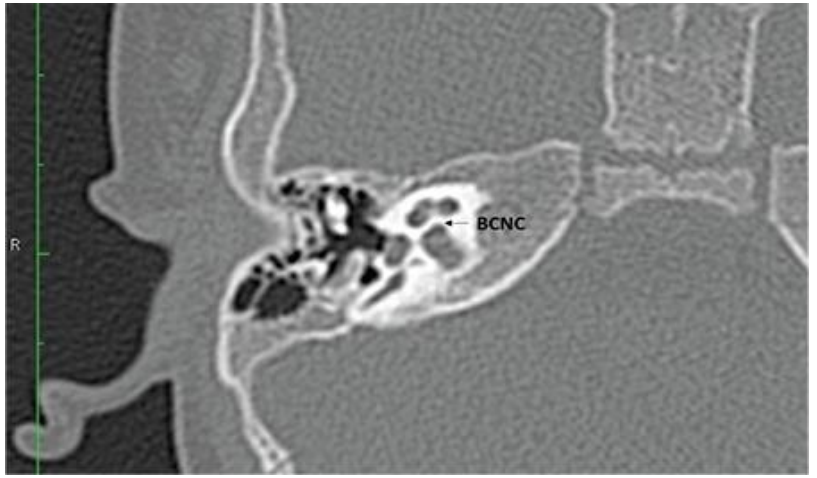
Sennaroğlu L, Bajin MD. Classification and Current Management of Inner Ear Malformations. Balkan medical journal. Sep 29 2017; 34(5):397-411. doi:10.4274/balkanmedj.2017.0367.



Enlarged Vestibular Aqueduct

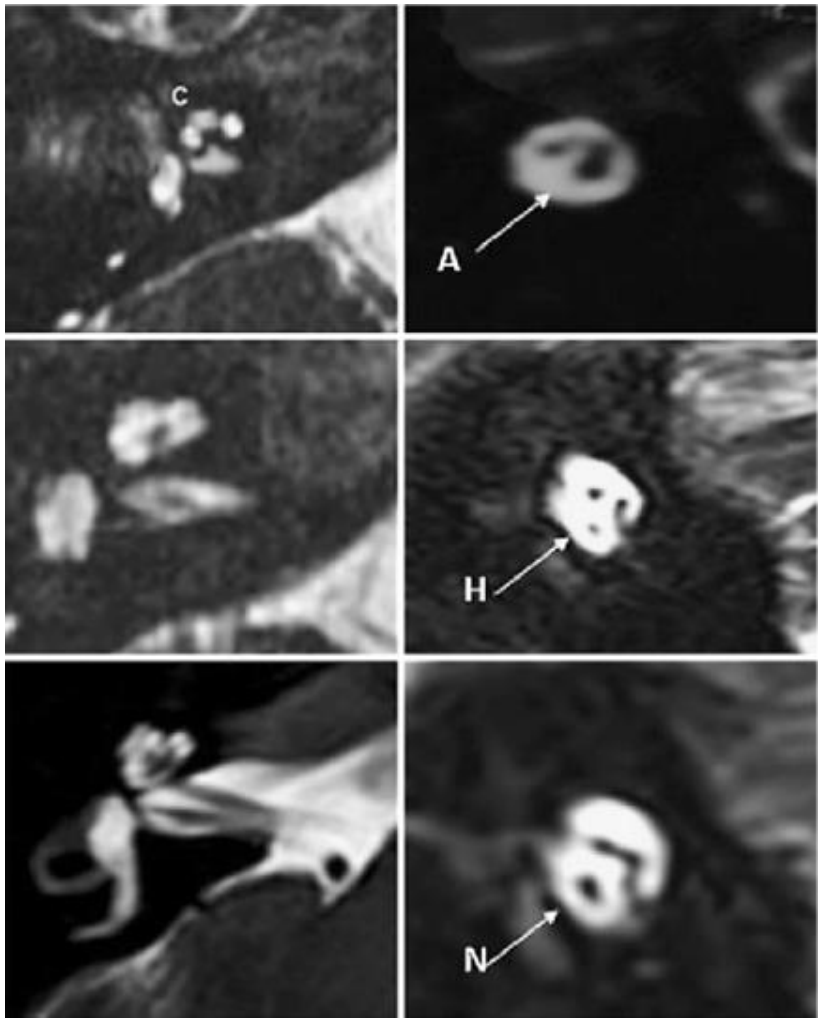


Cochlear Aperture Hypoplasia



Cochlear Aperture Aplasia

Sennaroğlu L, Bajin MD. Classification and Current Management of Inner Ear Malformations. Balkan medical journal. Sep 29 2017; 34(5):397-411. doi:10.4274/balkanmedj.2017.0367.



A. Cochlear nerve aplasia

H. Cochlear nerve hypoplasia

N. Normal cochlear nerve

Iseli C, Adunka O, Buchman C. Cochlear Nerve Deficiency. Pediatric Cochlear Implantation. 2016:227-235.

METHODS

- **Methods**

Cases series study

- **Subject of the study**

49 patients with anatomy congenital malformation underwent cochlear implantation at ENT hospital from 2010 to 2024.

METHODS

- **Inclusion criteria**
  - Anatomy congenital malformation underwent cochlear implantation.
  - Patients or parents/guardians of children agree to participate in study.
- **Exclusion criteria**
  - Anatomy malformations contraindication of CI: Complete Labyrinthine Aplasia (Michel Deformity), Rudimentary Otocyst, Cochlear Aplasia
  - Lost to follow up postoperative.
  - Insufficience auditory – verbal training postoperative.

METHODS

Procedure Steps:

- **Preoperative Consultation:** A multidisciplinary meeting including the surgeon, audiologist, and radiologist => Diagnosis, selection of surgical approach, and choice of electrode type
- **Surgical Procedure**
  - Cochlear implant surgery
  - Verification of electrode position post-surgery using a CT scan
- **Postoperative Language Training:** Conducted by audiologists and speech-language therapists
  - External Device Fitting: 4–6 weeks post-surgery once the scalp and incision site have healed.
  - Device Activation and Mapping: 3 times within the first 1–2 months, every 3 months during the first postoperative year
  - Evaluate language rehabilitation outcomes using the CAP (Categories of Auditory Performance) scale at 1year post-surgery.

RESULTS - DISCUSSION

General characteristics

Characteristics	n	%
Gender		
Male	28	57,1
Female	21	42,9
Age at surgery	5,1 ± 4,6	
Residence		
Ho Chi Minh city	16	32,7
Other provinces	33	67,3

RESULTS - DISCUSSION

Level of hearing loss

Level of hearing loss	n	%
Severe hearing loss	2	4,1
Profound hearing loss	47	95,9
Total	49	100

RESULTS - DISCUSSION

Anatomy congenital malformations

Characteristics	n	%
Inner ear malformation	15	30,6
Cochlear nerve deficiency	15	30,6
Inner ear malformation + Cochlear nerve deficiency	19	38,8
Total	49	100

The otocyst gives rise to the cochlea, vestibule, semi-circular canals and endolymphatic sac as well as the vestibulocochlear ganglion. It is therefore plausible that developmental arrest occurring early in the embryonic life would likely lead to CND associated with other congenital inner ear malformations

RESULTS - DISCUSSION

Inner ear malformations

Classification	n	%
Common cavity	3	6,1
Cochlear hypoplasia type 1	1	2,0
Cochlear hypoplasia type 2	1	2,0
Cochlear hypoplasia type 3	8	16,4
Cochlear hypoplasia type 4	2	4,1
Incomplete partition type 1 (IP-1)	1	2,0
Incomplete partition type 2 (IP-2)	13	26,5
Enlarged Vestibular Aqueduct	5	10,3
Normal inner ear	15	30,6
Total	49	100

RESULTS - DISCUSSION

- Incomplete partition type II (IP-II) is the most common anomaly, accounting for 26.5%, followed by cochlear hypoplasia type 3, which accounts for 16.4%, and enlarged vestibular aqueduct at 10.3%.
- These findings are consistent with the results of Ozkan's study, where IP-II had the highest prevalence at 29.9%, cochlear hypoplasia at 18.9%, and enlarged vestibular aqueduct at 10.2%.

Ozkan HB, Cicek Cinar B, Yucel E, Sennaroglu G, Sennaroglu L. Audiological Performance in Children with Inner Ear Malformations Before and After Cochlear Implantation: A Cohort Study of 274 Patients. Clinical otolaryngology : official journal of ENT-UK ; official journal of Netherlands Society for Oto-Rhino-Laryngology & Cervico-Facial Surgery. Jan 2021; 46(1):154-160. doi:10.1111/coa.13625.

RESULTS - DISCUSSION

- According to Sennaroglu, IP-II occurs in approximately 25% of inner ear anomalies.
- This malformation was initially described by Carlo Mondini, and when accompanied by vestibular and vestibular aqueduct enlargement, it constitutes the triad known as Mondini malformation.

Sennaroglu L. Histopathology of inner ear malformations: Do we have enough evidence to explain pathophysiology? Cochlear implants international. 2016; 17(1):3-20. doi:10.1179/1754762815y.0000000016.

RESULTS - DISCUSSION

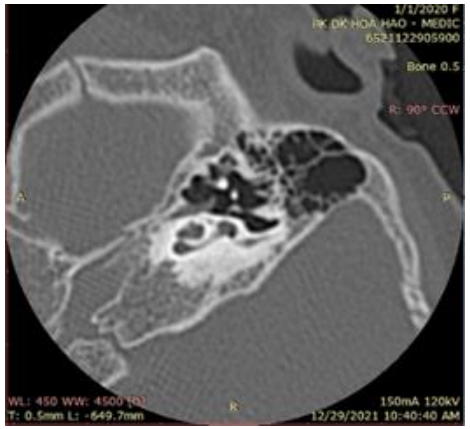
Cochlear nerve deficiency

Cochlear nerve status	n	%
CN hypoplasia	21	42,9
CN aplasia	13	26,5
Normal cochlear nerve	15	30,6
Total	49	100

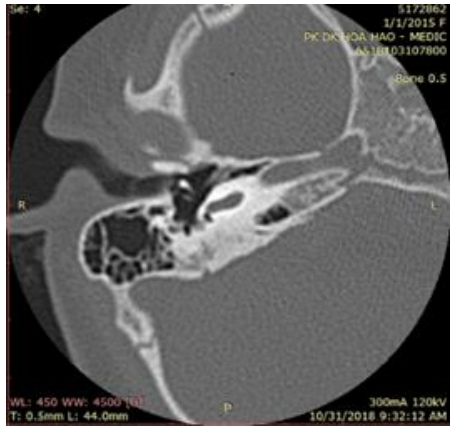
RESULTS - DISCUSSION

- The majority of patients in the study exhibited cochlear nerve hypoplasia on MRI, accounting for 42.9%. The incidence of cochlear nerve aplasia on MRI was 26.5%.
- The reported rates of cochlear nerve hypoplasia and aplasia on MRI vary across studies: Vincenti's study reported a hypoplasia rate of 60% and aplasia rate of 40%, while Birman's study reported a hypoplasia rate of 28.1% and aplasia rate of 71.9%.

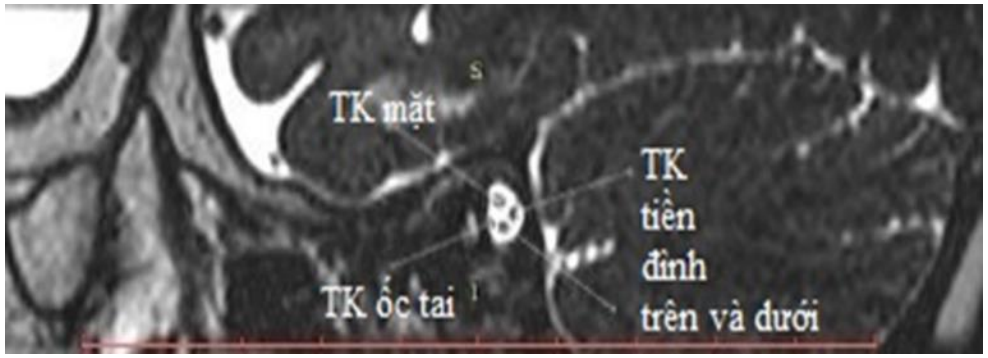
1. Vincenti V, Ormitti F, Ventura E, Guida M, Piccinini A, Pasanisi E. Cochlear implantation in children with cochlear nerve deficiency. International journal of pediatric otorhinolaryngology. Jun 2014; 78(6):912-7. doi:10.1016/j.ijporl.2014.03.003
2. Birman CS, Powell HR, Gibson WP, Elliott EJ. Cochlear Implant Outcomes in Cochlea Nerve Aplasia and Hypoplasia. Otology & neurotology : official publication of the American Otological Society, American Neurotology Society [and] European Academy of Otology and Neurotology. Jun 2016; 37(5):438-45. doi:10.1097/mao.0000000000000997



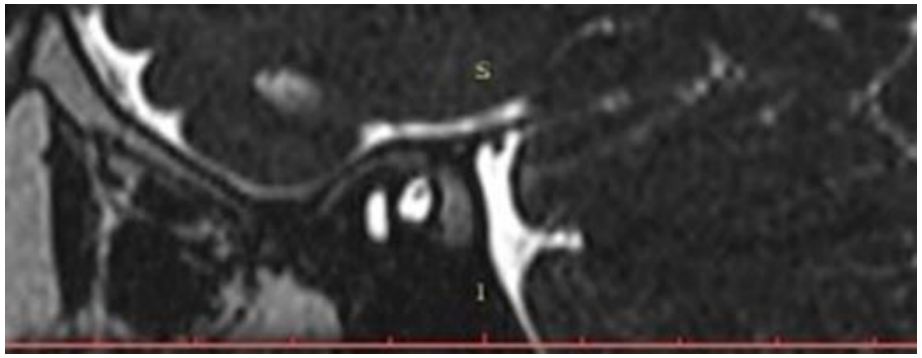
IP-2



CH-3



CN hypoplasia



CN aplasia

RESULTS - DISCUSSION

Insertion techniques

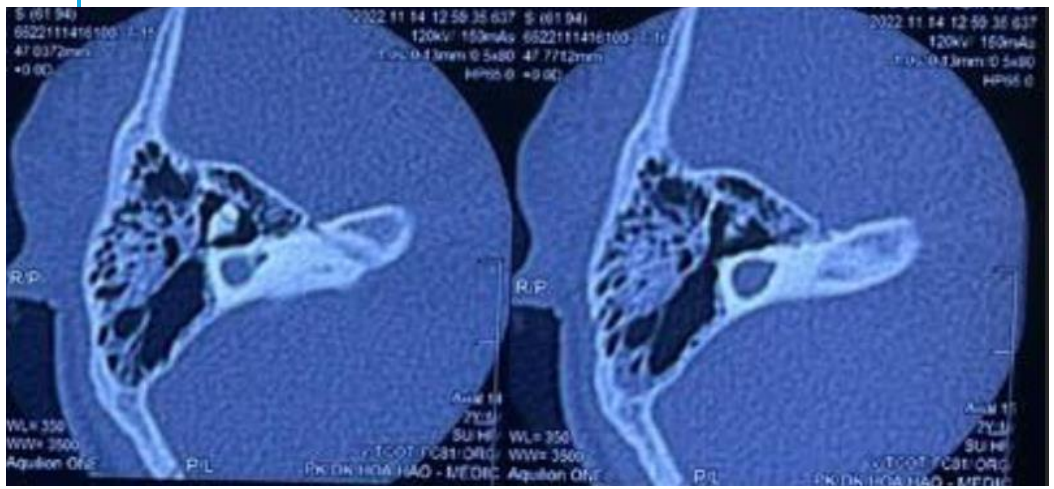
Insertion techniques	n	%
Round window	39	79,6
Cochleostomy	7	14,3
Labyrinthotomy	2	4,1
Oval window	1	2,0
Total	49	100

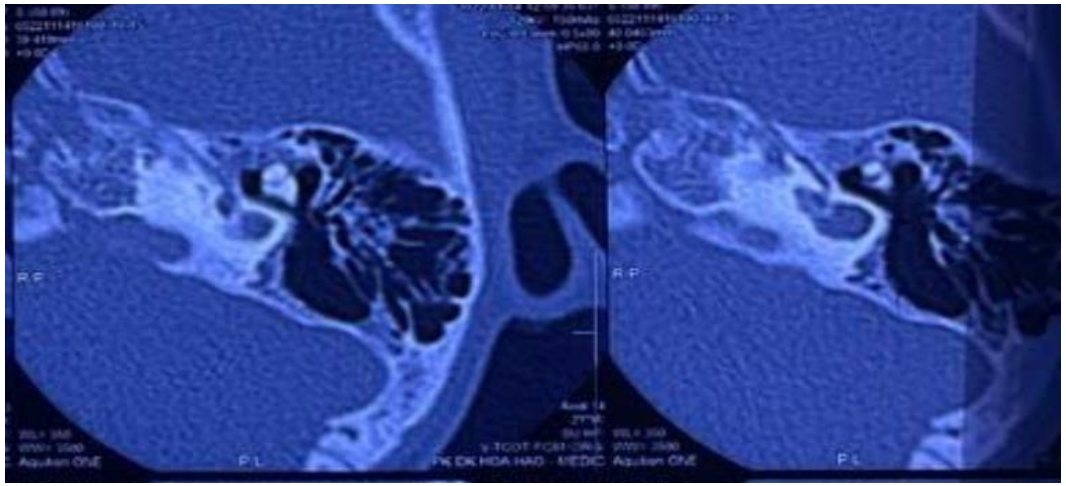
RESULTS - DISCUSSION

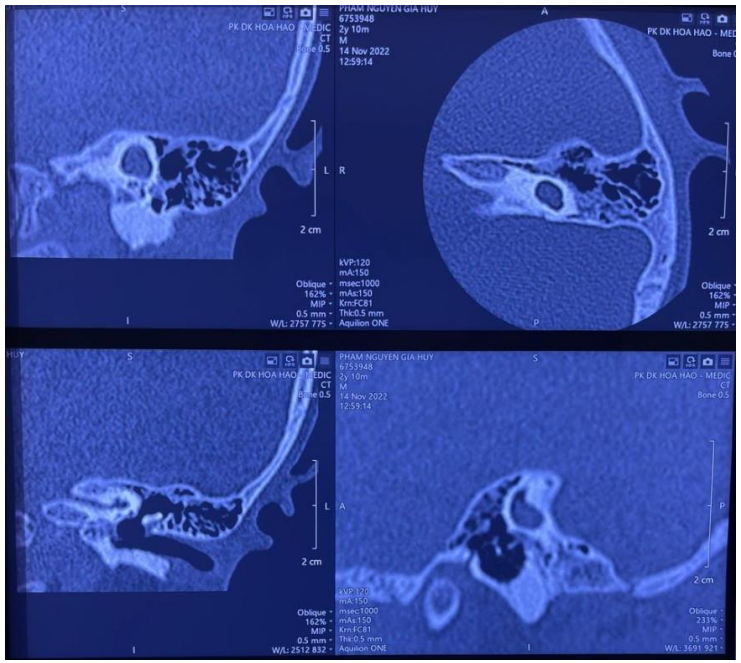
- In 79.6% of cases, the electrode was inserted through the round window membrane.
- In 7 cases (14.3%), the round window could not be located after posterior tympanotomy. In these instances, we proceeded with cochleostomy for electrode placement by identifying the promontory and drilling approximately 3mm anterior to the oval window in a downward direction.

RESULTS - DISCUSSION

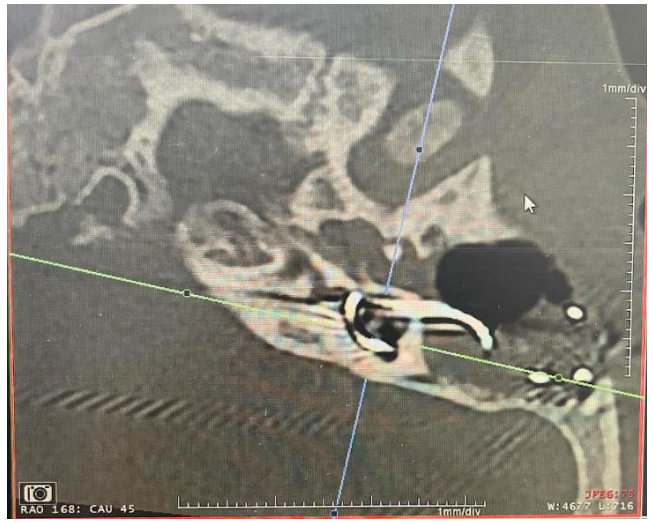
- In our study, there were three cases of common cavity cochlear malformation, where structures such as the round window membrane or round window ridge could not be identified during surgery:
  - 1 case, we were able to identify the stapes, which was subsequently removed, and the electrode was inserted through the oval window.
  - In the other 2 cases, we performed a labyrinthotomy into the common cavity and placed the electrode at the location of the lateral semicircular canal.

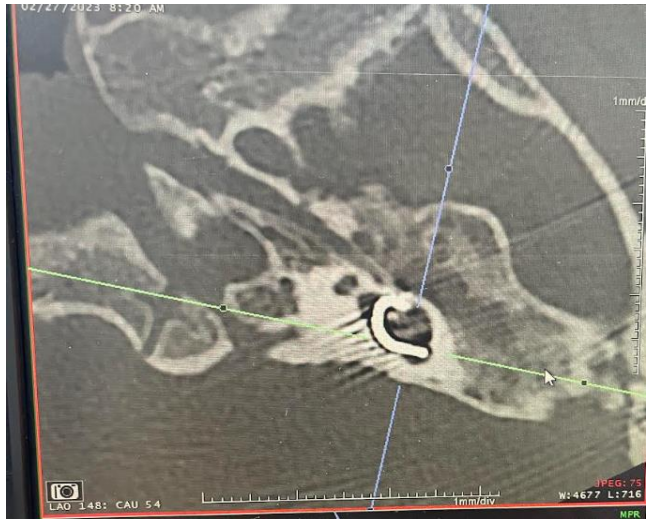


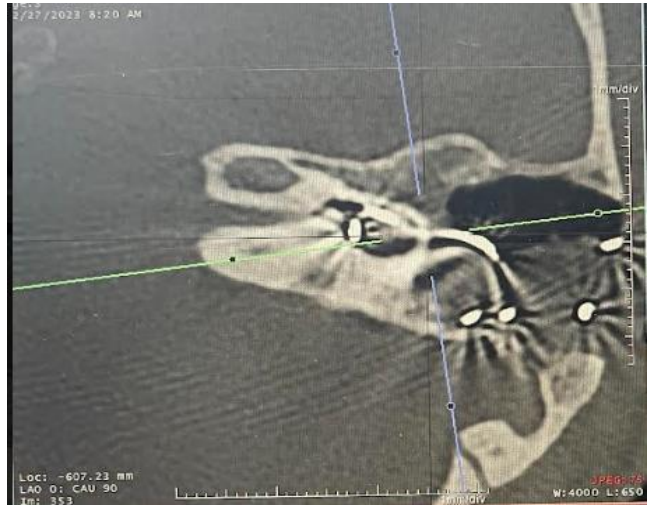


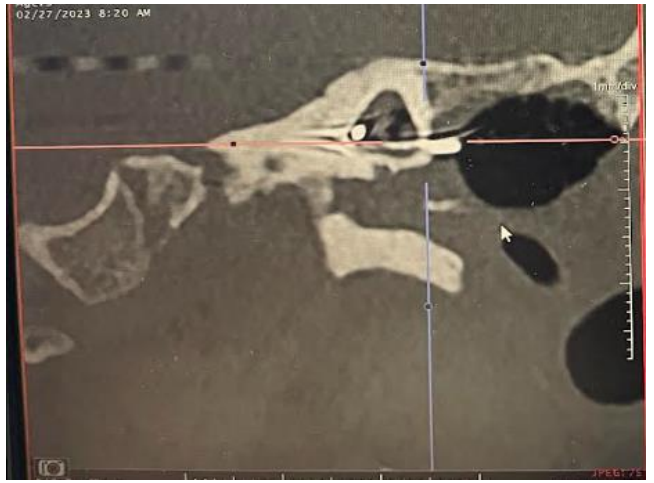


PATIENT P.N.G.H, 3 YEARS OLD
BILATERAL COMMON CAVITY

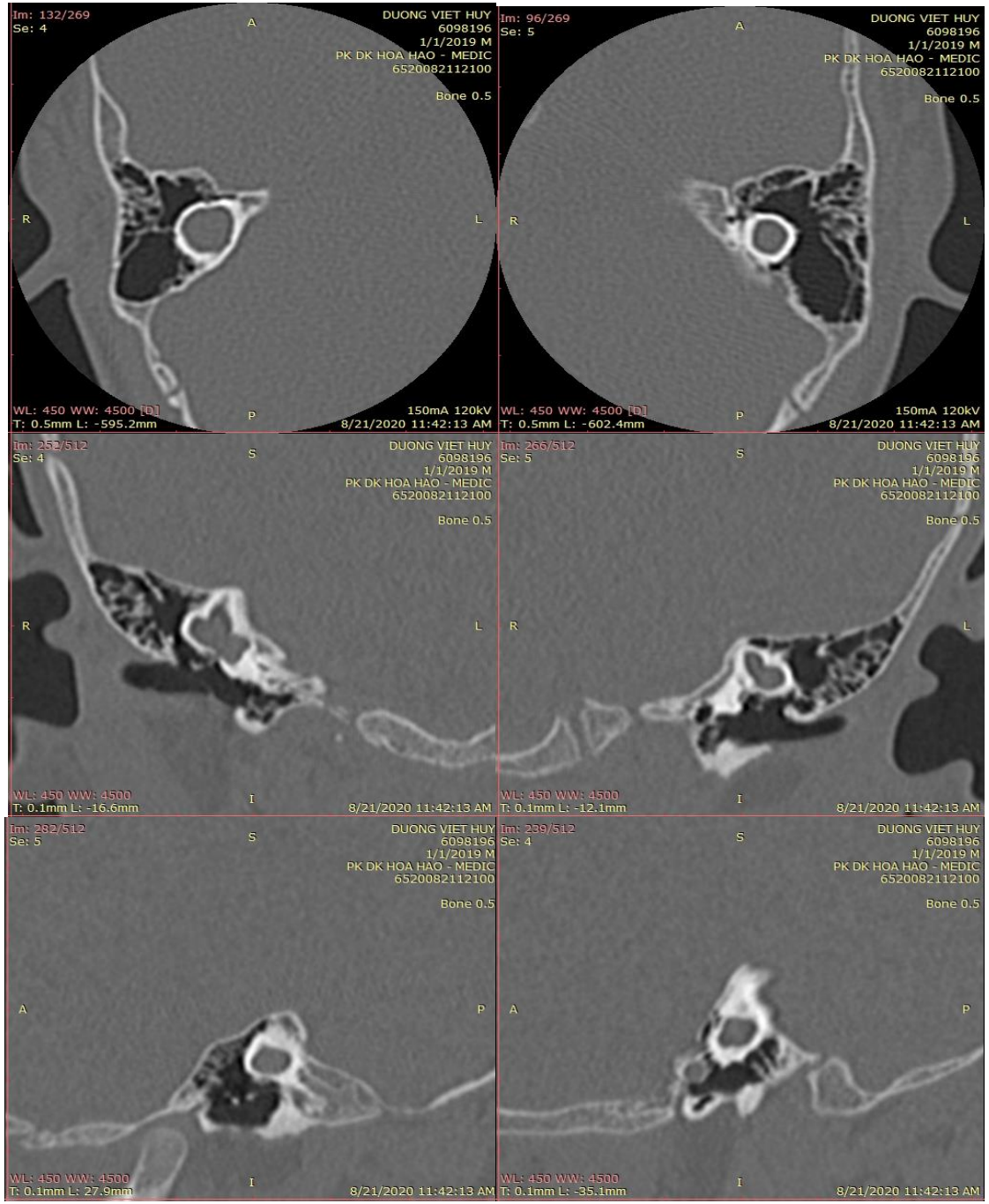




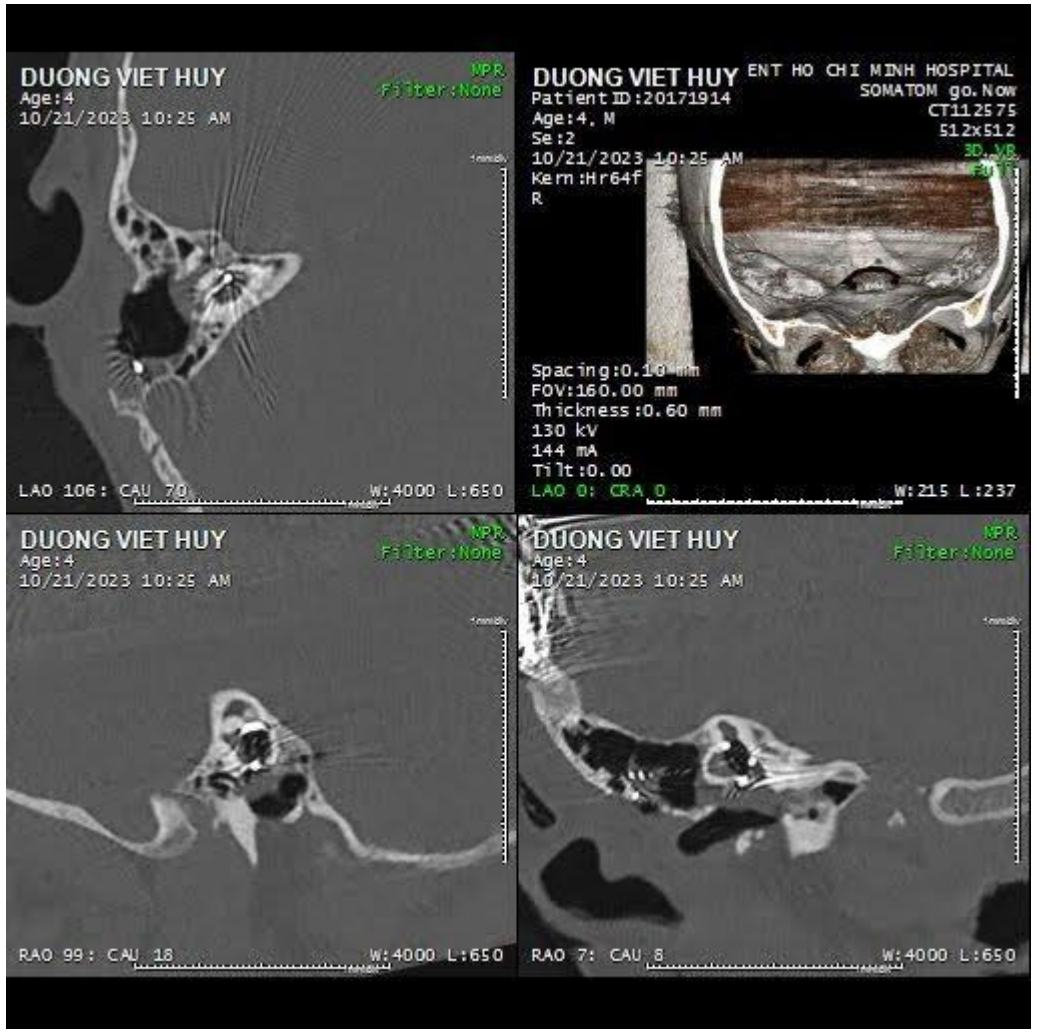




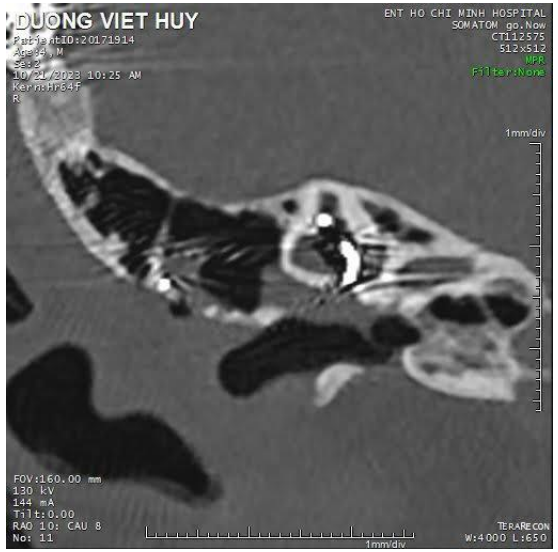
PATIENT P.N.G.H, 3 YEARS OLD POSTOPERATIVE CT-SCAN INSERTION TECHNIQUE: **LABYRINTHOTOMY**



PATIENT D.V.H, 2 YEARS OLD
BILATERAL COMMON CAVITY



PATIENT D.V.H, 2 YEARS OLD
POSTOPERATIVE CT-SCAN
INSERTION TECHNIQUE: **OVAL WINDOW**

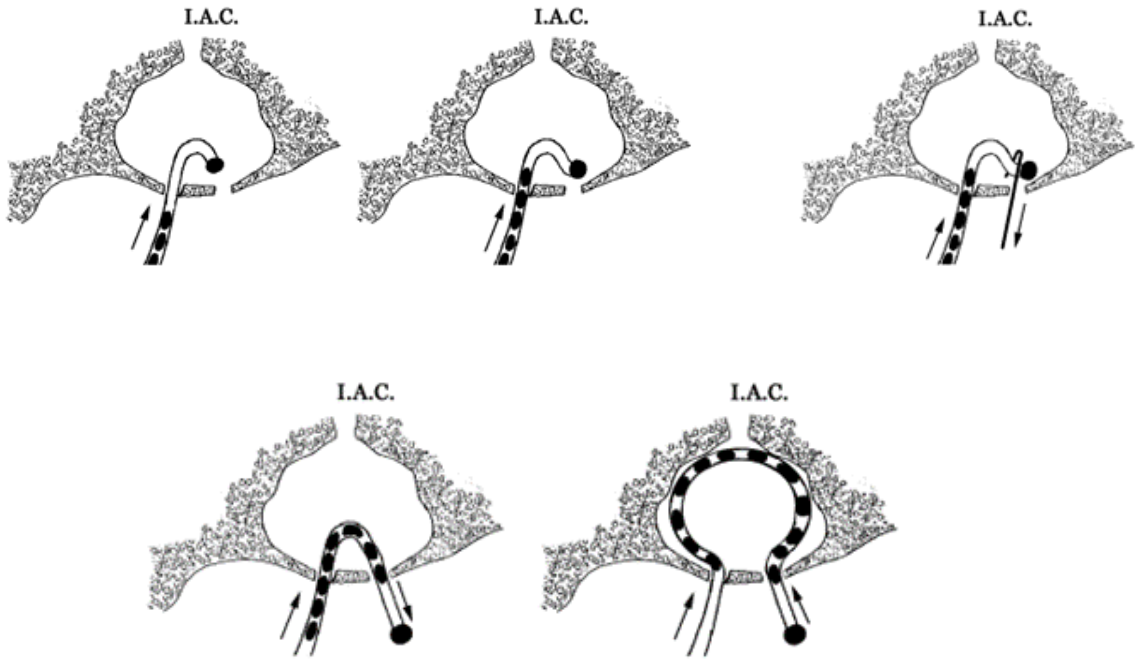


RESULTS - DISCUSSION

- Electrode placement in the common cavity is still under investigation.
- According to Sennaroglu, the common surgical approach is via a labyrinthotomy, typically at the location of the lateral semicircular canal, using a straight electrode.
- Beltrame described a specialized electrode designed for common cavity malformations, featuring an inactive tip. The common cavity is accessed by creating two openings at the location of the lateral semicircular canal, approximately 3-4 mm apart. The inactive tip of the electrode is inserted into the upper opening and pulled through the lower opening, with the remaining part of the electrode positioned in the common cavity, running along its inner wall.

1. Sennaroğlu L, Bajin MD. Classification and Current Management of Inner Ear Malformations. Balkan medical journal. Sep 29 2017; 34(5):397-411. doi:10.4274/balkanmedj.2017.0367.
2. Beltrame MA, Frau GN, Shanks M, Robinson P, Anderson I. Double posterior labyrinthotomy technique: results in three Med-El patients with common cavity. Otology & neurotology : official publication of the American Otological Society, American Neurotology Society [and] European Academy of Otology and Neurotology. Mar 2005; 26(2):177-82. doi:10.1097/00129492-200503000-00008

RESULTS - DISCUSSION



Double posterior labyrinthotomy technique - Beltrame

Beltrame MA, Frau GN, Shanks M, Robinson P, Anderson I. Double posterior labyrinthotomy technique: results in three Med-El patients with common cavity. Otology & neurotology : official publication of the American Otological Society, American Neurotology Society [and] European Academy of Otology and Neurotology. Mar 2005; 26(2):177-82. doi:10.1097/00129492-200503000-00008

RESULTS - DISCUSSION

Type of electrode

Type of electrode	n	%
Straight electrode	23	46,9
Curve electrode	26	53,1
Total	49	100

RESULTS - DISCUSSION

- According to Sennaroglu, certain types of inner ear anomalies, such as cochlear hypoplasia and incomplete partition type I, require the use of straight, short, and thin electrodes due to abnormalities in the number of cochlear turns and the size of the cochlea in these malformations.
- For common cavity deformities, the size of the cavity can vary significantly, making it necessary to assess the appropriate electrode length preoperatively. This length can be calculated using the formula $2\pi r$, where r represents the radius of the common cavity.

Sennaroğlu L, Bajin MD. Classification and Current Management of Inner Ear Malformations. Balkan medical journal. Sep 29 2017; 34(5):397-411. doi:10.4274/balkanmedj.2017.0367.

RESULTS - DISCUSSION

COMPLICATIONS

Complications	n	%
CSF leak	5	10,3
Facial paralysis	3	6,1
No complication	41	83,6
Total	49	100

RESULTS - DISCUSSION

- There were five cases of cerebrospinal fluid (CSF) leakage (10.3%), which were managed intraoperatively by sealing the cochleostomy with temporalis fascia.
- These patients were closely monitored postoperatively, and no cases of meningitis or recurrent CSF leakage were observed.
- There were 3 cases of postoperative facial paralysis (6.1%), all of which fully recovered within 6 months after surgery.

RESULTS - DISCUSSION

- The rate of cerebrospinal fluid (CSF) leakage in our study (10.3%) is higher than that reported by Melo (7.6%).
- This discrepancy may be due to differences in the types of malformations between the 2 studies. In our study, there were malformations, such as the common cavity and incomplete partition type I, which are particularly prone to CSF leakage during surgery.

Melo AS, Martins J, Silva J, Quadros J, Paiva A. Cochlear implantation in children with anomalous cochleovestibular anatomy. Auris, nasus, larynx. Oct 2017; 44(5):509-516. doi:10.1016/j.anl.2017.02.003.

RESULTS - DISCUSSION

CAP score 1 year postoperative

Malformations	n	CAP score
Inner ear malformations	15	5,7 ± 0,7
Cochlear nerve deficiency	15	5,3 ± 1,6
Inner ear malformations + Cochlear nerve deficiency	19	4,1 ± 1,6

RESULTS - DISCUSSION

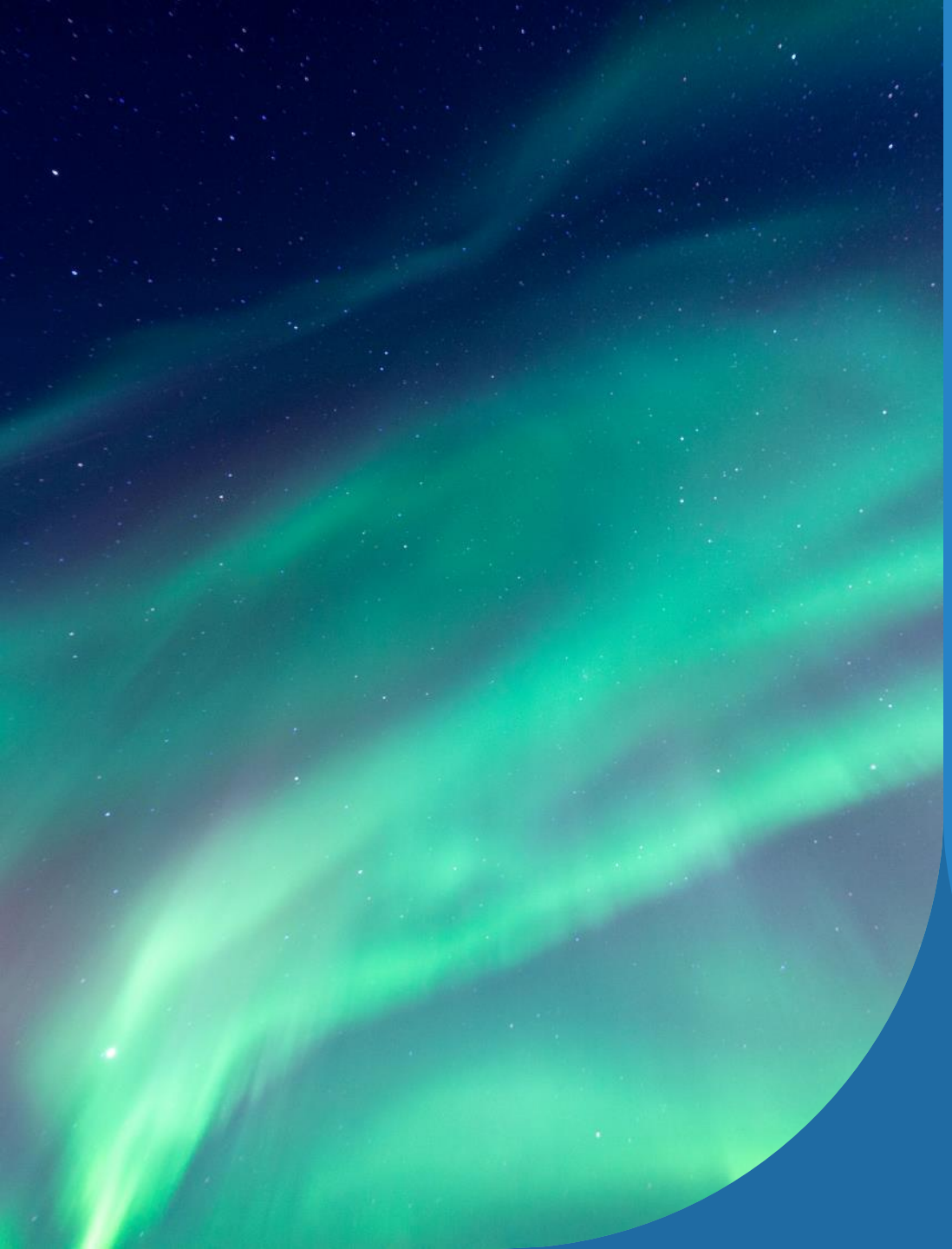
- The average score on the Categories of Auditory Performance (CAP) scale was 4.9 ± 1.5 at one year postoperatively.
- The mean CAP score in patients with only 1 type of anatomical anomaly was significantly higher than in those with 2 types of anatomical anomalies ($p < 0.05$)

=> This suggests that patients with multiple anatomical abnormalities have a poorer prognosis for language development compared to those with fewer anatomical abnormalities

CONCLUSION

- Cochlear implantation in cases of congenital anatomical abnormalities presents numerous challenges and difficulties

- However, with careful preoperative imaging studies to assess the anatomical anomalies, along with the selection of appropriate electrodes and surgical approach, CI can be performed successfully, safely, and effectively in these cases.



THANK YOU FOR YOUR ATTENTION!