



Preoperative Otolaryngology Examination

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Special Features

1. Patients with inner ear malformations may apply with recurrent meningitis which may be a fatal situation.
2. Spontaneous footplate fistula may be seen in incomplete partition I and cochlear hypoplasia II.
3. Progressive hearing loss with sudden hearing loss attacks may be due to incomplete partition type II (IP-II).

Preoperative evaluation of patients with inner ear malformations (IEM) usually represents audiological assessment such as behavioral testing, auditory brainstem response testing, otoacoustic emission testing, educational testing, and radiological testing such as computed tomography and magnetic resonance imaging. However, in this chapter we would like to focus on otolaryngological evaluation and emphasize certain clinical points which must be addressed prior to surgery.

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5.1 History Taking

Characteristics of hearing loss and recurrent meningitis are very important to think about inner ear malformations. The patient may have unilateral or bilateral hearing loss. Unilateral hearing loss at birth is a strong indication for radiological evaluation for unilateral IEM.

Family may give a history of progressive hearing loss. There may be situations where the patient passes neonatal hearing screening tests at birth but then experiences progressive hearing loss with sudden hearing loss attacks. This is suggestive of incomplete partition type II (IP-II).

History may reveal rhinorrhea after head trauma. This is a situation seen in IP-I or cochlear hypoplasia type II (CH-II) where there is a stapes footplate fistula. There may be a history of recurrent meningitis. This presents an ENT emergency.

5.2 ENT Examination

Otolaryngological examination starts with a full physical examination. Otoscopy is particularly important. Otitis media with effusion (OME) should be treated prior to surgery particularly for cochlear implantation. Persistent effusion after adequate medical therapy necessitates ventilation tube placement. Cochlear implantation in the presence of effusion complicates surgery by obscuring the anatomical structures both by

hypertrophic mucosa and bleeding. Tube placement should be done at least 3 months prior to cochlear implantation. Implantation can be done with the tubes in place. If the patient has a subtype of IEMs with a risk of gusher it is advisable to remove the tubes and allow sufficient time for the perforation to heal before CI surgery.

Another very important point is to differentiate effusion from cerebrospinal fluid in the middle ear. IEMs like IP-I and CH-II are prone to stapes footplate fistulas which may lead to CSF leakage in the middle ear and subsequent meningitis. Cases of recurrent meningitis and persistent effusion behind tympanic membrane are suggestive of IP-I or CH-II. OME is a mucosal disease and tympanic membrane shows increased vascularity. This is very useful to differentiate the patients with CSF in the middle ear where there is no increased vascularity of the tympanic membrane. It is very important to keep in mind that patients with unilateral effusion or CH-II and IP-I might have CSF in the middle ear. In these patients, stapes footplate fistula is the point of CSF leakage and must be closed either preoperatively or intraoperatively.

These patients may also present with rhinorrhea and they may be operated by neurosurgery for suspected anterior skull base defects without success. Patients with rhinorrhea and hearing loss must be investigated for IEMs and stapes footplate fistulas.

Patients may have visible signs such as unilateral cup ear or hypoplastic auricle. Sometimes unilateral facial paralysis may accompany cochlear hypoplasia cases.

IEMs may be unilateral where contralateral ear and hearing are normal, making the diagnosis more challenging. This delay may cause recurrent meningitis leading to ossification within the ipsilateral labyrinth as well as ossification of the contralateral normal hearing ear. Therefore, patient may lose the option of cochlear implant [1]. Therefore, it is an ENT emergency to notice and repair stapes footplate fistulas particularly in IP-I and CH-II.

A significant number of the patients suffer from comorbidities. These are really important for anesthesiology both for radiological evalua-

tion and surgery. Each comorbidity must be consulted with the relevant specialty. For example, a patient with CHARGE syndrome should be consulted with the department of pediatrics, ophthalmology, neurology, cardiology, orthopedics, and pediatric surgery.

Genetic consultation is also important both for etiology and family consulting.

5.3 Radiological Evaluation

A patient with fluctuating or progressive SNHL needs a radiological evaluation. If the patients benefit from hearing aids and a CI is not indicated, only MRI is sufficient for the diagnosis of IP-II. This is for the concern of avoiding radiation to the child. If the patient has profound SNHL and a CI is planned, temporal CT and MRI are necessary. This is usually done at 6 months of age and if available, cone beam CT is advantageous to minimize radiation.

5.4 Management

In case of total hearing loss there is a possibility of a diagnosis of complete labyrinthine aplasia, rudimentary otocyst, cochlear aplasia, or cochlear nerve aplasia which are definite indications for an ABI [1]. In such a situation, **radiological evaluation should be done as early as possible, not later than 6 months of age** [2].

If an IP-II is diagnosed the patient is advised to wear helmets during sports and avoid contact sports because head trauma may cause further hearing loss in IP-II. In addition, the family is informed that the patient may have attacks of sudden hearing loss. **In such a case they need to apply to a hospital immediately for steroid treatment to recover hearing.** Aim is to provide natural hearing and delay cochlear implantation as long as possible.

Patients with stapes footplate fistula need immediate middle ear exploration to check the footplate for a fistula. This is an ENT emergency to avoid further meningitis. Pneumococcal vaccination is mandatory but if the fistula is not

repaired vaccination will not provide protection against meningitis.

5.5 Conclusion

Patients with IEMs require special attention throughout the management. A meticulous preoperative evaluation prevents further problems and complications in an already complicated patient group.

References

1. Sennaroğlu L, Bajin MD. Classification and current management of inner ear malformations. *Balkan Med J.* 2017;34(5):397–411.
2. Sennaroglu L, et al. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. *Otol Neurotol.* 2011;32(2):187–91.