



## Surgical Indication of Inner Ear Malformation associated with Bacterial Meningitis

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### Abstract

Bacterial meningitis in children is a life-threatening infection of the central nervous system that is mostly associated with inner ear malformations and Cerebrospinal Fluid (CSF) leakages. With bacterial meningitis, inflammation may spread to the healthy ears and cause bilateral deafness in some cases. In this paper, two cases of congenital inner ear malformations are presented. In Case 1, inflammation of the bacterial meningitis spread to the healthy ears and cause bilateral deafness and the patient received cochlear implant surgery. In Case 2, there was no history of meningitis or any findings of CSF leakage on CT, but the patient received exploratory surgery and repair of the potential risk of CSF leakage. In recent years, inner ear malformations have been increasingly diagnosed after newborn hearing screening, without any signs of meningitis. Early identification of the specific type of inner ear malformation and determining the associated risk of meningitis are vitally important. In cases of inner ear malformations with a high risk of CSF leakage and bacterial meningitis, surgical repair of CSF leakage should be considered before bacterial meningitis develops.

**Keywords:** Cerebrospinal fluid (CSF); Bacterial meningitis; Computed tomography (CT); Newborn hearing screening (NHS)

### Introduction

Bacterial meningitis in childhood is a life-threatening infection of the central nervous system that is mostly associated with inner ear malformations and Cerebrospinal Fluid (CSF) leaks. Up to 33% of cases of repeated meningitis in children are caused by otolaryngological etiologies [1]. Bacterial meningitis is not only potentially life-threatening, but serious complications, such as brain dysfunction, can occur. In addition, inner ear inflammation associated with meningitis may cause severe bilateral hearing loss (total deafness) [2].

In recent years, inner ear malformations have been increasingly diagnosed with temporal bone Computed Tomography (CT) after Newborn Hearing Screening (NHS), without any signs of meningitis. Although it is obvious that not all cases of inner ear malformation require prophylactic surgical closure of CSF leakage to prevent bacterial meningitis, early identification of the cases at high risk of bacterial meningitis is required after NHS. In this paper, two cases of congenital inner ear malformations are presented, and surgical repair of CSF leaks is considered.

### Case Presentation

#### Case 1

A young boy, 5years and 4months old, was brought to our emergency department because he had high fever and his consciousness had become cloudy. He had severe neck stiffness and hypertonia of the upper limbs. Blood tests showed neutropenia. On cerebrospinal fluid examination, increasing number of neutrophils in the cerebrospinal fluid was observed, and the cerebrospinal fluid sugar level was decreased. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) showed brain edema. Temporal bone CT showed bilateral congenital inner ear malformations, incomplete partition Type I (IP-1) (Figure 1a and b). In addition, a soft tissue shadow was found in the tympanum and mastoid cavity that suggested a CSF leak of the right ear (Figure 1a).

In fact, this patient had already been diagnosed as having bilateral inner ear malformations

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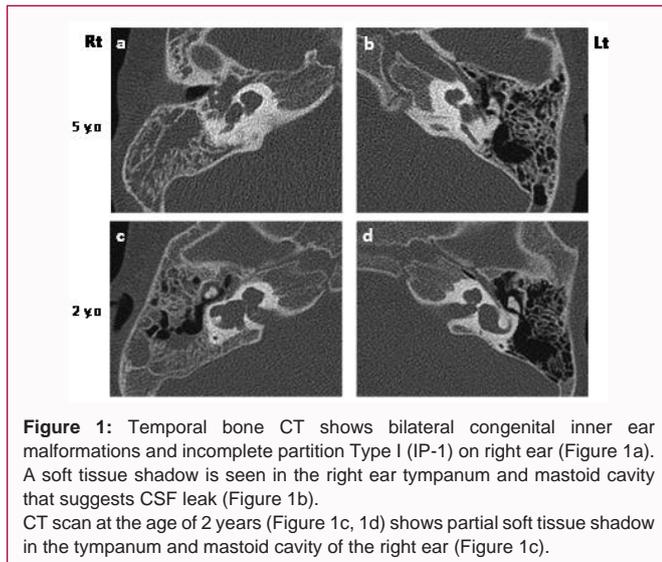
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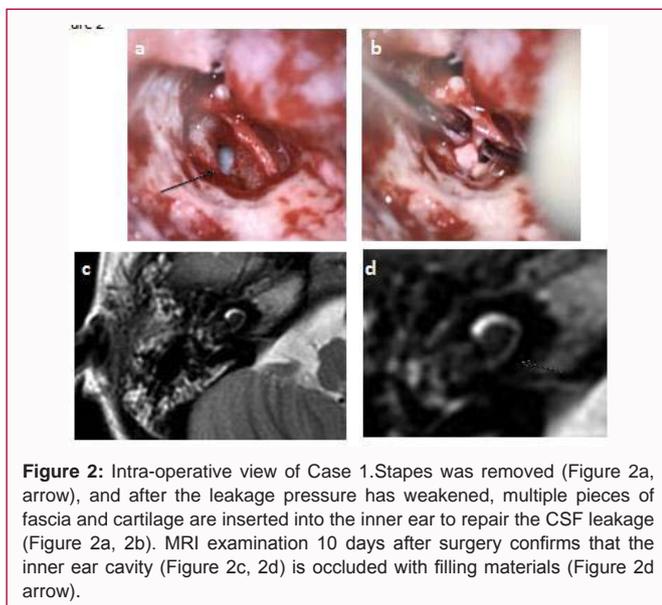
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**Figure 1:** Temporal bone CT shows bilateral congenital inner ear malformations and incomplete partition Type I (IP-1) on right ear (Figure 1a). A soft tissue shadow is seen in the right ear tympanum and mastoid cavity that suggests CSF leak (Figure 1b). CT scan at the age of 2 years (Figure 1c, 1d) shows partial soft tissue shadow in the tympanum and mastoid cavity of the right ear (Figure 1c).



**Figure 2:** Intra-operative view of Case 1. Stapes was removed (Figure 2a, arrow), and after the leakage pressure has weakened, multiple pieces of fascia and cartilage are inserted into the inner ear to repair the CSF leakage (Figure 2a, 2b). MRI examination 10 days after surgery confirms that the inner ear cavity (Figure 2c, 2d) is occluded with filling materials (Figure 2d arrow).

at the age of 2 years at another hospital, and a hearing aid had been fitted for the contralateral left ear. The patient underwent a newborn hearing test at birth and was judged as refer; he was then seen by another doctor at the age of 4 months to undergo a complete hearing test. ABR showed severe hearing loss of the right ear and intermediate hearing loss of the left ear. He started using a hearing aid for the left ear. At the age of 2 years, he underwent CT examination and was diagnosed as having bilateral congenital inner ear malformations (Figure 1c and d). Partial soft tissue shadow that suggested fluid collection was found in the tympanum and mastoid cavity of the right ear. Since he had never had a history of meningitis, watchful waiting was selected. After the diagnosis of inner ear malformations at the age of two, he developed sudden bacterial meningitis at the age of 5 years and 4 months.

The patient was immediately admitted to the ICU and given intravenous antibiotics (Ampicillin: ABPC 300 mg/kg + Ceftriaxone: CTRX 114 mg/kg). On the second day of hospitalization, he showed mild hemiparesis and facial paralysis. Since *Streptococcus pneumonia* was detected on CSF culture, ABPC was increased (400 mg/kg), and

methylprednisolone (mPSL) pulse therapy was added. Intravenous antibiotics were administered for 14 days, and the meningitis was cured. During the course of the illness, there were no complaints of otalgia or tympanic findings suggestive of acute otitis media.

A CSF leak due to the right congenital inner ear malformation was considered the source of the meningitis infection. The CSF leak was repaired via the oval window with removal of the stapes of the right ear. Under general anesthesia, atympanostomy was performed with resultant significant watery otorrhea. The liquid was sugar-positive and was considered to be CSF. A small defect at the central part of the stapes footplate was found, and CSF was discharged from the bone defect. Stapes was removed, and after the leakage pressure weakened, multiple pieces of fascia and auricular cartilage were inserted into the inner ear to repair the CSF leak (Figure 2a and 2b). Spinal fluid pressure was not lowered by lumbar spinal drainage or mannitol. It was decided to wait after removal of the whole stapes, and when the cerebrospinal fluid leak decreased, it was repaired. MRI examination 10 days after surgery confirmed that the inner ear cavity was occluded with filling materials. There was no recurrence of the CSF leak or of the meningitis after surgery (Figure 2d arrow). Although the CSF leakage was stopped by surgery, hearing loss of the opposite ear occurred due to inner ear inflammation caused by bacterial meningitis. Therefore, cochlear implant surgery on the opposite ear was performed.

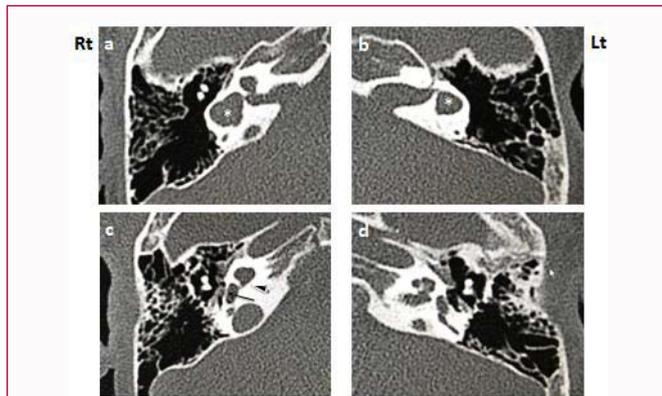
## Case 2

A young girl, 3 years and 6 months old, underwent a newborn hearing test at birth and was judged as refer; she then underwent a complete hearing test at 4 months. ABR showed severe hearing loss in her right ear. During the course, although there were no episodes of acute otitis media or meningitis, she was brought to our hospital at the age of 3 years and 6 months for a detailed examination. CT confirmed a right inner ear malformation (IP-1) (Figure 3c arrow head). Development of the right mastoid air cells was good, and they were well-ventilated. Although there was no evidence of a bone defect of the stapes footplate, a small soft tissue shadow was observed between the cruses of the stapes (Figure 3c arrow).

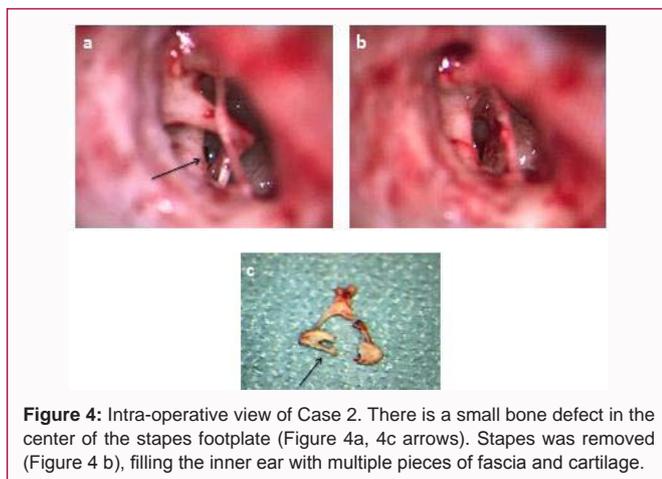
Since the right inner ear congenital anomaly was IP-1, it was explained to her parents that they should pay particular attention to the symptoms of bacterial meningitis at the onset of acute otitis media. Since the parents wanted preventive surgery if the risk of bacterial meningitis was high, it was planned to check the footplate of the stapes at surgery and perform a definitive CSF leak repair if necessary. The findings at surgery showed a membranous bulging between the cruses of the stapes. When this was cut, there was a small bone defect in the center of the stapes footplate (Figure 4a and 4c arrows). Although the CSF leakage was not observed from the oval window, definitive surgical repair was performed by filling the inner ear with removal of the stapes, similar to Case 1 (Figure 4b).

## Discussion

Case 1 was a bilateral case of hearing impairment due to inner ear malformations, while Case 2 was a unilateral case (both IP-I). Case 1 was diagnosed as having an inner ear malformation on CT at 2 years of age, and a CSF leak in the right ear was suspected. However, since there was no clinical history of bacterial meningitis, the parents were advised to follow him carefully. Because the patient developed bacterial meningitis at 5 years and 4 months, he underwent surgical repair of the CSF leak by filling the inner ear. The CSF leak stopped after surgery, but unfortunately, hearing loss of the good hearing



**Figure 3:** CT confirms right inner ear malformation (incomplete partition type 1: IP-1) (Figure 3c arrow head) and bilateral dilated vestibules (Figure 3a,3b white asterisk). Development of the right mastoid air cells is good, and they are well-ventilated. A small soft tissue shadow is observed between the cruses of the stapes (Figure 3c arrow).



**Figure 4:** Intra-operative view of Case 2. There is a small bone defect in the center of the stapes footplate (Figure 4a, 4c arrows). Stapes was removed (Figure 4 b), filling the inner ear with multiple pieces of fascia and cartilage.

ear due to bacterial meningitis was observed, and cochlear implant surgery was performed. On the other hand, in Case 2, an inner ear malformation (IP-I) was diagnosed during examination for unilateral hearing loss. There was no history of meningitis or any findings of CSF leak on CT, but the patient underwent exploratory surgery because IP-I is one of the high-risk inner ear anomalies associated with bacterial meningitis. Since there was a small bone defect in the stapes footplate, surgical repair of the potential risk of CSF leak was performed in Case 2.

Inner ear malformations may involve bone defects of the stapes footplate. Furthermore, in cases with weakness between the cochlea and internal auditory canal, CSF leakage is likely to occur from the small hole of the stapes footplate. Rao et al. [3] reported that 59% of cases of repeated bacterial meningitis in children were due to anatomical congenital malformations of the temporal bone. Savva et al. [4] reported that persistent CSF leaks from inner ear malformations were observed in 15% to 25% of repeated bacterial meningitis cases in children. In cases with bacterial meningitis, inflammation may spread to the healthy ear and cause bilateral deafness in some cases. Therefore, in cases with inner ear malformations with obvious cerebrospinal fluid leakage, surgical repair of the CSF leakage should be performed before the development of meningitis.

Children with certain congenital malformations of the inner ear, including those with Michel type, cochlear aplasia, common cavity

deformity, cochlear hypoplasia, and IP-I, have a higher incidence of spontaneous CSF leaks and resulting meningitis [5]. IP-I is one of the incomplete partition anomalies and accounts for approximately 20% of inner ear malformations [6,7]. In IP-I, there may be a defect between the cochlea and internal auditory canal, and the CSF completely fills the cochlea. IP-I may also involve a defective stapes footplate. Therefore, meningitis can occur in IP-I patients. On the other hand, there is no fragility of the site separating the internal auditory canal and the cochlear cavity in IP-II, so that the risk of cerebrospinal fluid leakage is low. Previously, there were several reports of repeated bacterial meningitis due to Mondini type inner ear malformations, but Mondini type strictly conforms to IP-II in the classification of Sennaroglu [6,7], so the risk of meningitis is low in Mondini type deformity.

Since  $\beta$ 2-transferrin is present only in the CSF, it is very useful for diagnosing CSF leaks [8]. However, when there is a copious CSF gusher, care must be taken, because a large amount of CSF otorrhea can occur into the external auditory canal even with a small tympanostomy incision. Therefore, in the case of a bulging tympanic membrane, it is better to consider tympanostomy as an examination for final confirmation in the operating room.

Muzzi et al. [9] reported in their review that the average diagnostic delay from the first episode of meningitis was  $3.44 \pm 3.41$  years, the number of meningitis episodes that occurred before the correct diagnosis and definitive surgical treatment was  $3.27 \pm 1.81$ , and certain numbers of children died from inner ear malformation-related meningitis. Newborn hearing screening programs should prompt a diagnostic work-up in the case of hearing impairment to prevent inner ear malformation-related meningitis. Early identification of the specific type of inner ear malformation and determination of the associated risk of meningitis are of vital importance. Since NHS provided an opportunity for the early identification of high-risk inner ear anomalies, temporal bone CT should be recommended to children with sensorineural hearing loss early in life. In cases with specific inner ear malformations, in our opinion, prophylactic surgical repair of CSF leakage should be performed to prevent bacterial meningitis.

## Conclusion

Bacterial meningitis in childhood is not only a life-threatening infection, but also a definitive cause of severe bilateral hearing impairment. In children with the high-risk type of congenital inner ear malformations, such as Michel type, cochlear aplasia, common cavity deformity, cochlear hypoplasia, and IP-I, especially with associated CSF leakage, surgical repair should be considered before bacterial meningitis develops.

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