Common Ear Diseases

8

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8.1 Congenital Malformation of Ear

Congenital ear malformations include auricular malformation, external ear canal atresia, middle ear malformation and inner ear malformation.

Congenital external ear malformation and congenital middle ear malformations are both common ear abnormalities. Unilateral ear malformation is more common, while 25–30% of the patients have bilateral malformation. Congenital external ear and middle malformation are usually divided into microtia/atresia and middle ear malformation. Patients with congenital external ear malformation usually have two physiological defects, i.e. facial defects and hearing impairment. Plastic surgery and hearing improvements are urgent wishes of the patients and their families.

8.1.1 Congenital Auricular Malformation

Congenital auricular malformation is caused by abnormal development of the first and the second branchial arch. Genetic factors are the main causes of this abnormal development. Half of the malformations in the ear, nose, and throat region affect the ear. Malformations of the external ear (pinna or auricle with external auditory canal [EAC]) are collectively termed microtia. Microtia is a congenital anomaly that ranges in severity from mild structural abnormalities to complete occuring absence of the external ear (anotia). Microtia occurs more frequently in males (2-3:1), is predominantly unilateral (70–90%), and more often involves the right ear. The reported prevalence varies from 0.83 to 17.4 per 10,000 births. Microtia may be genetic (with family history, spontaneous mutations) or acquired. Malformations of the external ear can also involve the middle ear and/or inner ear. Microtia may be an isolated birth defect, but associated anomalies or syndromes are described in 20–60% of cases, depending on the study design. These generally fit within the oculo-auriculo-vertebral spectrum; defects are located most frequently in the facial skeleton, facial soft tissues, heart, and vertebral column, or comprise a syndrome. Diagnostic audiological investigation of microtia includes clinical examination, audiologic testing, genetic analysis and, especially in higher grade malformations with EAC deformities, computed tomography (CT) or cone-beam CT for the planning of surgery and rehabilitation procedures, including implantation of hearing aids.

Clinical Manifestation

Auricular malformation includes anotia, protruding/prominent ear (also called Dumbo ears), monkey ear, accessory auricle, macrotia and microtia. Anotia and microtia are often accompanied with atresia, external ear canal stenosis or middle ear malformation.

A German doctor Max has proposed the following classification system:

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- Type I: Auricular malformation is slight, auricular size is slightly smaller than normal ear, and each part of auricular structure can be clearly distinguished;
- Type II: Auricular size is 1/2~1/3 of normal size, auricular structure is partially preserved;
- Type III: Severe malformation of auricle, usually like a peanut (Fig. 8.1).

Diagnosis

Inquire if there are similar cases in the family, the history of other diseases and drug history of mother during pregnancy. Although the diagnosis could be made according to physical examination, it is still necessary to make a comprehensive examination for accurate anatomical description of malformation of middle ear, inner ear and facial nerve. Examination of auditory function, ear X-ray and CT scan are helpful to identify the auditory function, ear canal, mastoid, ossicular chain and inner ear.

Treatment

When patients ask for auricular plastic surgery, the surgery can be performed according to their individual conditions. It is usually done at around six years old before accepting education. Some surgeons advise surgery after 10 years old because thoracic growth at that time is not influenced if limb cartilage is removed to reconstruct the auricle. Artificial materials, such as silicone stent and high-density porous polyethylene (Medpore) can be used to reconstruct auricle. It has been reported that the artificial auricle made with biological silicone by 3D printing technology is used to reconstruct the auricle. To improve hearing of patients with bilateral severe malformation of ear canal atresia, canalplasty and tympanoplasty should be performed before entering school. Bone-anchored hearing aid (BAHA) could be used after birth to improve hearing and speech development as early as possible, BAHA is available, and bone bridge implant is optional.

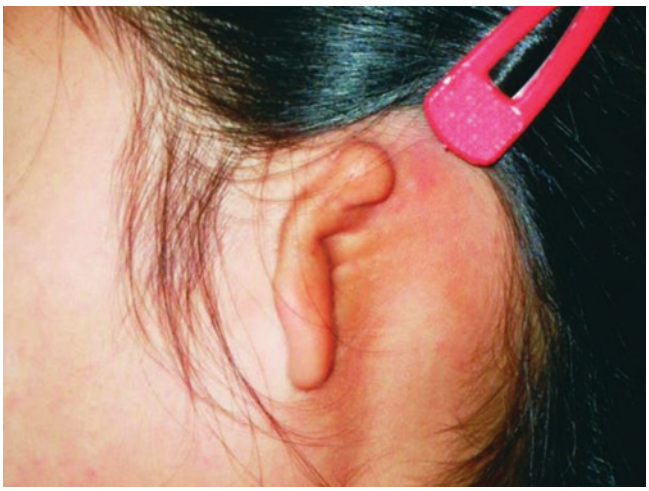


Fig. 8.1 Congenital auricular malformation

8.1.2 Congenital Malformation of External and Middle Ear

8.1.2.1 External Auditory Canal Malformation

It is classified as external ear canal stenosis and atresia including cartilaginous and bony atresia (Fig. 8.2).

8.1.2.2 Middle Ear Malformation

It includes malformation of tympanic cavity, tympanum antrum, mastoid and Eustachian tube. The most common malformations are ossicular chain and facial nerve malformations.

Ossicular Chain Malformation

Ossicular chain is a lever structure which includes malleus, incus and stapes. Stapes transmits sound by direct connection with vestibular window, so almost all the auditory reconstruction surgeries focus on the stapes.

Classification of ossicular chain malformation by Okano in 2003 is as follows:

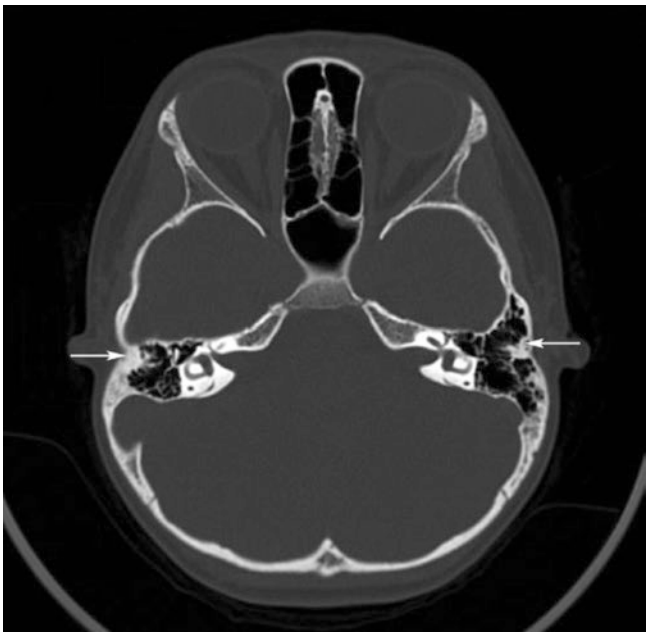


Fig. 8.2 Bilateral atresia of external ear canal (arrow) CT scan

- Type I: discontinuity between the incus and the stapes with mobile stapes;
- Type II: congenital fixation of the stapes with ossicular deformity;
- Type III: congenital fixation of the malleus and deformity of the incus with mobile stapes.

Okano classification is a relatively simple and practical one.

Facial Nerve Malformation

The proportion of facial nerve malformation in ear malformation is high and correlates with severity of ear malformation. At 1994, Tongjia Leng analysed 250 cases with congenital external and middle ear malformation of which 26.7% were facial nerve malformation and 59.2% were chordal tympanic nerve malformation. The most common malformation of facial nerve is; bony defect of Eustachian tube and cover of vestibular window by Eustachian tube. The most common malformation of chorda tympani is absence of nerve. Care must be taken on bony defect of Eustachian tube. Though bony defect of Fallopian tube in middle ear may not be showed on CT scan, it can be judged by some indirect signs such as Eustachian tube abnormalities at geniculate ganglion, defect of vertical segment, defect of vestibular window and pyramidal process.

8.1.2.3 Diagnosis and Treatment

Diagnosis can be made based on physical examination of auricle and external ear canal, moderate and severe conductive hearing loss, and CT scan showing malformations of external ear and middle ear.

Classification of Ear Malformations

De la Cruz Ear Malformation Classification (Table 8.1)

Table 8.1 De La Cruz ear malformation classification

Minor malformation	Severe malformation
Normal pneumatization of mastoid	Poor pneumatization of mastoid
Normal oval window and footplate of stapes	Defect or malformation of oval window and footplate of stapes
Good relationship between facial nerve and footplate of stapes	Abnormal facial nerve course
Normal inner ear	Abnormal inner ear

Jahrsdoerfer Grading System (Only for Bony Atresia (Table 8.2)

Table 8.2 Jahrsdoerfer grading system

Parameters	Scores
Stapes present	2
Oval window present	1
Middle ear space	1
Facial nerve position	1
Malleus-incus complex	1
Mastoid pneumatized	1
Incus-stapes connected	1
Round window present	1
Appearance external ear canal	1
Maximum total score	10

Table 8.3 Evaluation of patients with congenital middle ear malformation for reconstruction surgery by Jahrsdoerfer grading system [1]

Scores	Operation indication
10	Excellent
9	Very good
8	Good
7	Fair
6	Marginal
<5	Poor

Firstly, bone conduction threshold should be confirmed as normal by audiometry or ABR, and inner ear should be identified as normal by imaging examination.

Treatment Regimen

If inner ear development is well identified by auditory tests and CT scan, both feature and hearing can be improved by combination of pinnaplasty, canalplasty and tympanoplasty. Such surgery is challenging because of high incidence of facial nerve malformation, temporal bone abnormality, hypoplasia of tympanum and mastoid in patients with external and middle ear malformation. If a doctor chooses patients with wrong indications or does not know well about the anatomy of temporal bone of patient with ear malformation; facial paralysis, sensorineural hearing loss and other severe complications can occur. Therefore, appropriate preoperative evaluation is essential.

Preoperative evaluation of patients with bony atresia is usually based on Jahrsdoerfer grading system (Table 8.3). Generally, patients with 6 scores or more are indicated for surgery, patients with 8 scores or more may regain good hearing following canalplasty and tympanoplasty (air-bone gap <25 dB). In patients with 5 scores or less, surgical risk may outweigh the benefit.

Patient with a score of 4 or more can be treated with vibrant soundbridge (VSB), and those with a score of 3 or less are not indicated to take surgeries. VSB is a device implanted in the middle ear, which is applied for mixed hearing loss, conductive hearing loss and sensorineural hearing loss. Kiefer

et al. reported that combined VSB implant and pinnaplasty could be applied for bilateral ear malformation. Although VSB has many advantages over tympanoplasty such as: reconstruction of the external ear canal is not needed, no postoperative complications like atresia and infection in external ear canal, no recurrent conductive deafness, small external volume and easier to bury in hair; it is an expensive implanted device for hearing loss. Nevertheless, VBS implantation cannot take place of classical tympanoplasty which is the best choice for patients with well-developed temporal bone.

Patients with 5 scores or less should implant Bone-anchored hearing aid (BAHA). Patients younger than 3 years old are suggested to wear soft band BAHA but not titanium-nail implantation which is indicated for patients older than 3 years old. The thickness of bone cortex for implant should be more than 4 mm. Titanium nail should be anchored as one-stage operation. Canalplasty is indicated for congenital external and middle ear malformation, especially for those with canal stenosis with a diameter of less than 2 mm. Poorly developed mastoid is not indicated for auditory reconstruction to avoid acquired cholesteatoma in the canal.

The results of hearing improvement following auditory reconstruction vary from patients. The poorer long-term result than short term result may be related to factors such as lateralization of tympanic membrane, repeated stenosis and infection of canal (18.3–31%), refixation of ossicular chain, sensorineural hearing loss etc.

8.1.3 Congenital Inner Ear Malformations

Congenital inner ear malformation is a rare disease with an incidence of 1/2000–1/6000. It is subdivided according to the malformation sites into cochlear malformation, vestibular malformation, semicircular canal malformation, bony labyrinth malformation, internal auditory canal malformation, inner ear nerve malformation, blood vessel malformation etc. It includes malformations like hypoplasia, absence, deformation, translocation, stenosis etc. Common inner ear malformations are as follows.

1. Michel malformation
   Also known as Complete labyrinthine aplasia of inner ear. Complete absence of auditory and vestibular function (Fig. 8.3).
2. Mondini malformation
   Severe cochlear hypoplasia with a normal basal turn and a cystic fuse of middle and apical turn (Fig. 8.4).
3. Common Cavity malformation
   In common cavity malformation, developmental arrest occurs at the fourth week of gestation and is defined as a single cavity that represents the undifferentiated cochlea and vestibule (Fig. 8.5).

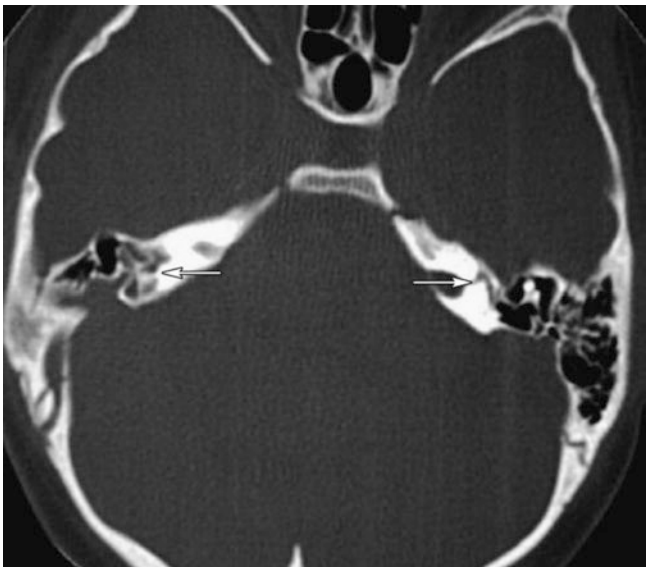


Fig. 8.3 CT scan of Michel malformation of bilateral inner ear (arrow)

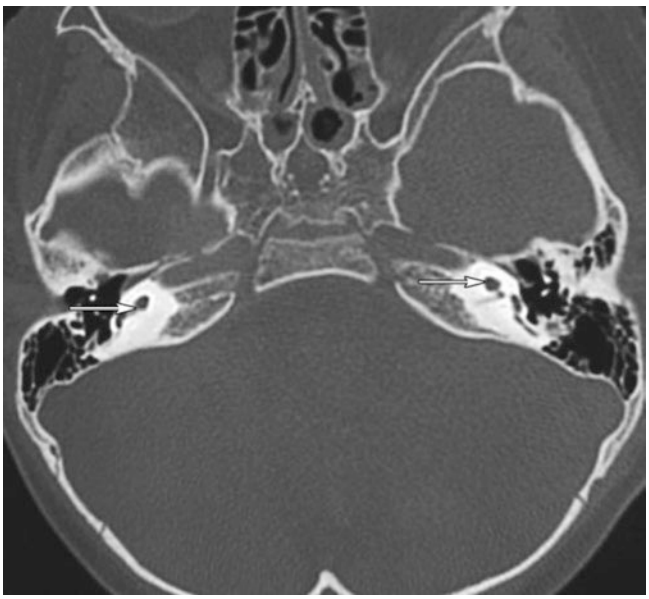


Fig. 8.4 Mondini malformation of bilateral inner ear (arrow) CT scan

4. Enlarged vestibular aqueduct
   The width in the middle of the long axis of the aqueduct exceeds 1.5 mm. It may be related to SLC26A4 gene mutation (Fig. 8.6).

If the patients with external and middle ear malformation have severe inner ear malformation and thus profound sensorineural hearing loss, care must be taken to perform external and middle ear surgery except for auricular plastic surgery for patients' psychological and social need. Patients with indications choose to have cochlear implantation.

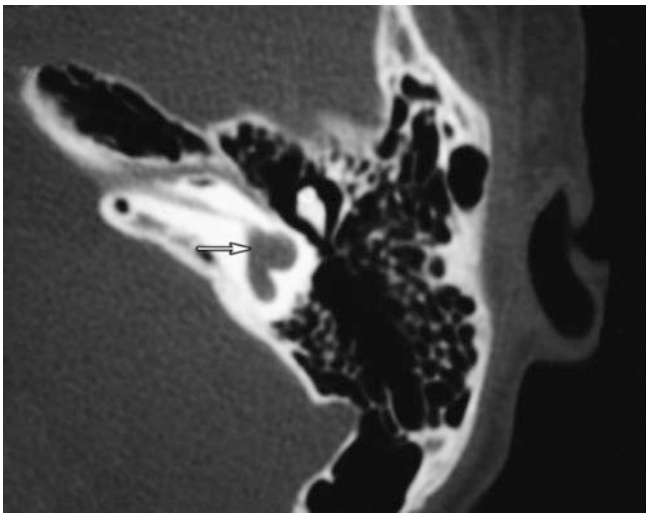


Fig. 8.5 Common cavity malformation (arrow) CT scan

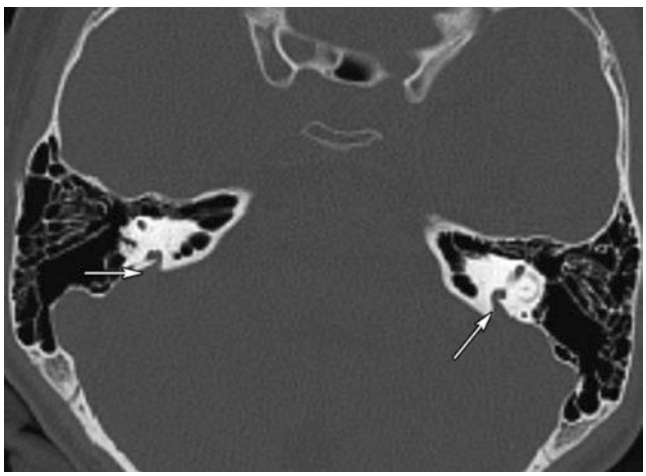


Fig. 8.6 CT scan of bilateral aqueduct vestibule expansion (arrow)

8.1.4 Congenital Pre-auricular Fistula

Congenital pre-auricular fistula is a common otological disease, which occurs due to abnormal integration of hillock node at the first and second brachial arch or poor closure of the first brachial arch at embryonic stage.

Clinical Manifestations

1. Symptoms

   Usually asymptomatic, or local swelling, pain or festering following infection.
2. Signs

   Fistula is usually unilateral. Generally, the orificium fistula is located before helix crus, and the other end is blind. A little white sticky and caseous secretion may be extracted from the orificium while being pressed.

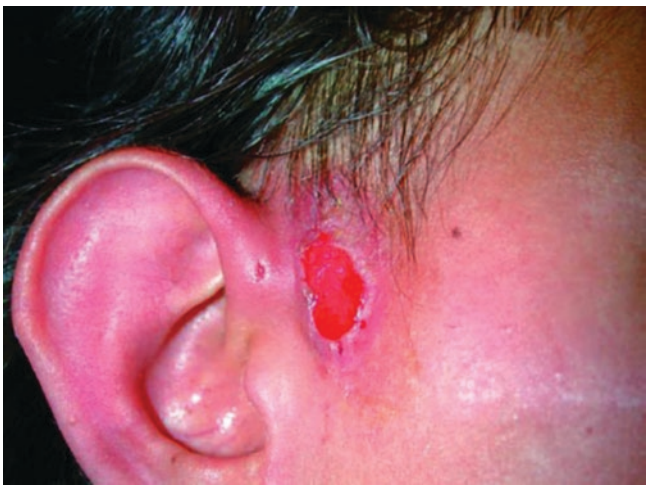


Fig. 8.7 Congenital preauricular fistula

Repeated infection may lead to formation of cyst or abscess which may transform into purulent fistula or scar once it ruptures (Fig. 8.7).

Therapy

1. No treatment is needed if there is no infection. Systemic antibiotics for acute infection. If abscess forms, incision and drainage are needed followed by surgical resection once infection is controlled.
2. Surgical indications
   (a) A history of recurrent or persistent infection;
   (b) No previous infection, but local itch and exudation of secretion;
   (c) No symptoms, but surgery required by patient.

Injecting a little methylene blue solution into the fistula before operation and guiding with a probe during surgery is helpful to remove the fistula completely.

8.2 Trauma of Ear

8.2.1 Trauma of External Ear

Clinical Manifestation

1. Local symptom
   (a) Auricular hematoma

   It usually occurs after ear contusion. External force may break the blood vessel of auricle, and thus lead to blood deposit between cartilage and perichondrium. Soft semicircle and reddish subcutaneous mass may appear on auricle skin. Patient may have no obvious symptoms except local pain. Infection or gradual organization and calcification may occur if hematoma is not treated promptly.

(b) Hemorrhage

Anterior superficial temporal artery and posterior auricular artery are the main blood supply of auricle which forms a rich anastomosis. Injury to these arteries may result in severe bleeding which could be paused temporarily by direct pressure.

2. General symptoms

Patients may feel dizzy and usually don't have general symptoms following simple auricular trauma. Patients with trauma of other organs may have changes of vital signs such as blood pressure, pulse and respiration, and symptoms of corresponding organs.

Examination

Firstly, pay attention to the patient's vital signs. Secondly check the injured area and the surrounding areas. The patient's vital signs must be checked before the injured site and other organs are examined.

Examination and management may be performed at the same time for serious injuries. In unconscious patient or seriously injured patient; doctors have to make a primary diagnosis based on experience with careful examination of the patient (Fig. 8.8).

Diagnosis

Diagnosis can made based on the traumatic history and physical examination. Severity of injury must be assessed preliminarily, including depth and contamination of wound, vital signs, complications and so. Injury of middle ear and inner ear may result in hearing loss, tympanic membrane perforation, hemotympanum, vertigo, or facial paralysis. Temporal bone must be taken into consideration.

Fig. 8.8 Auricular trauma

Treatment

1. The management of auricular hematoma

The auricular hematoma is not easy to be absorbed due to few subcutaneous tissue and poor blood supply of auricle. The accumulated blood can be extracted with a thick needle and dressed with pressure. If it does not heal following repeated operations, it can be incised parallel to the helix so that the hematoma can be exuded, or the blood clot can be removed. Pressure dressing and prevention from infection are essential.

2. Management of the external ear canal

Prevention from infection is the primary objective. The ear canal must be disinfected strictly and must not be irrigated with any liquid. The dust, cerumen and debris can be removed with suction, curettage and cotton swab. The ear canal should be kept as dry as possible. Nail purple oil is not recommended because it may disturb examining the wound. Sterilized antibiotic gauze or iodoform gauze can be used to pack the canal to prevent from infection and stenosis if necessary. If too much granulation tissue blocks the canal, it can be removed with curettage followed by canalplasty once the infection is under control.

3. General Management

Systemic Management

Adequate broad-spectrum antibiotics are recommended for prevention of infection. Tetanus immunoglobulin is required for deep wound.

8.2.2 Tympanic Trauma Tympanic Membrane Trauma

Clinical Manifestations

Sudden ear pain, immediate hearing loss accompanied by tinnitus, a small amount of bleeding in the external ear canal and vertigo. Occasionally, they may present with dizziness, nausea and mixed hearing impairment.

Examination

Tympanic membrane has many irregular shapes or fissure shaped perforation. There can be a little bloodstain within the external ear canal or blood scab. Small amount of blood can be found around the edge of the Tresses (Fig. 8.9). When there is plenty of bleeding or aqueous humor outflow, it suggests cerebrospinal fluid otorrhea caused by the fracture of the temporal bone or skull base. Conductive or mixed hearing damage can result from audition examination.

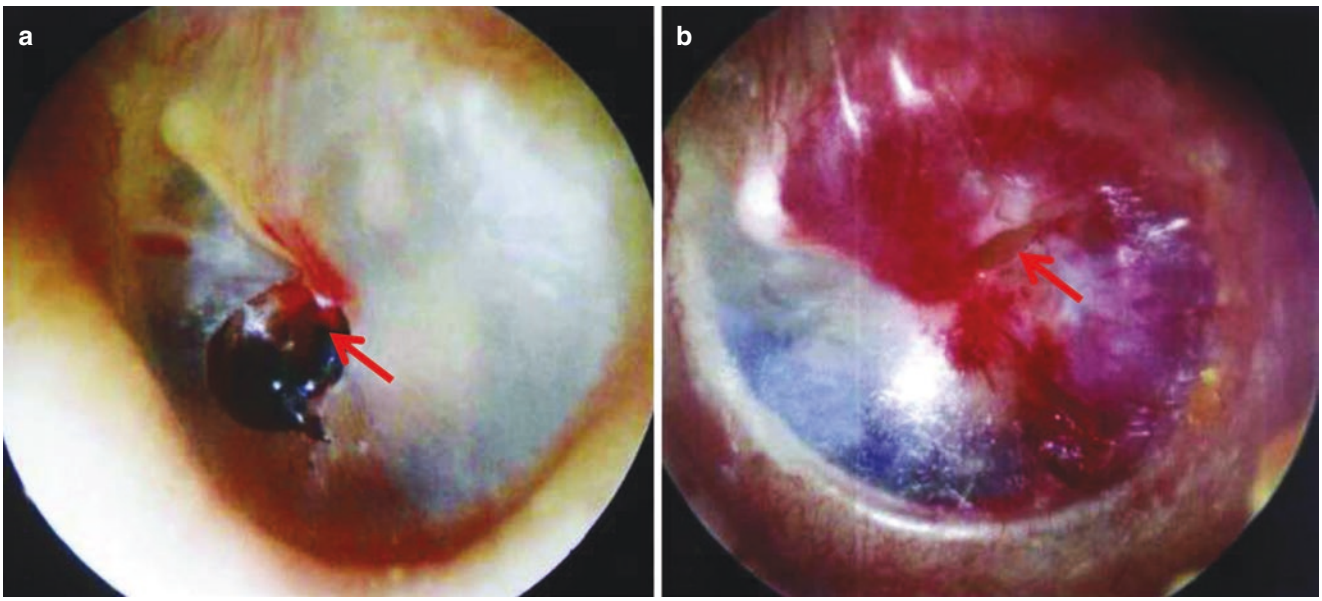


Fig. 8.9 Tympana perforation. (**a**) Tympanum perforation, little bloodstain. (**b**) Tympanum fissure perforation

Treatment

1. Clear foreign matter in the external acoustic canal and block it with sterilized cotton ball.
2. Avoid catching a cold. Don't violently blow your nose.
3. Irrigating or dripping medicine to the external ear canal is forbidden. Most traumatic perforations can self-heal after about 3–4 weeks. If the perforation is too large to heal spontaneously myringoplasty should be considered.

8.2.3 Temporal Bone Fracture

Classification

1. Longitudinal fracture

   Longitudinal fractures comprise 80% of all temporal bone fractures. They are frequently caused by a lateral force over the mastoid or temporal squama, also usually produced by temporal or parietal blows. The fracture line parallels the long axis of the petrous pyramid. It starts in the pars squamosa (mastoid or external auditory canal), as seen in the image above (Fig. 8.10 the temporal bone transverse fracture), and extends through the posterosuperior bony external canal, continues across the roof of the middle ear space anterior to the labyrinth, and ends anteromedially in the middle cranial fossa in close proximity to the foramen lacerum and ovale. The most common course of the fracture is anterior and extralabyrinthine; however, although rare, intralabyrinthine extension is possible. Again, bilateral temporal bone fractures are present in 8–29% of all fractures, according to the medical literature.

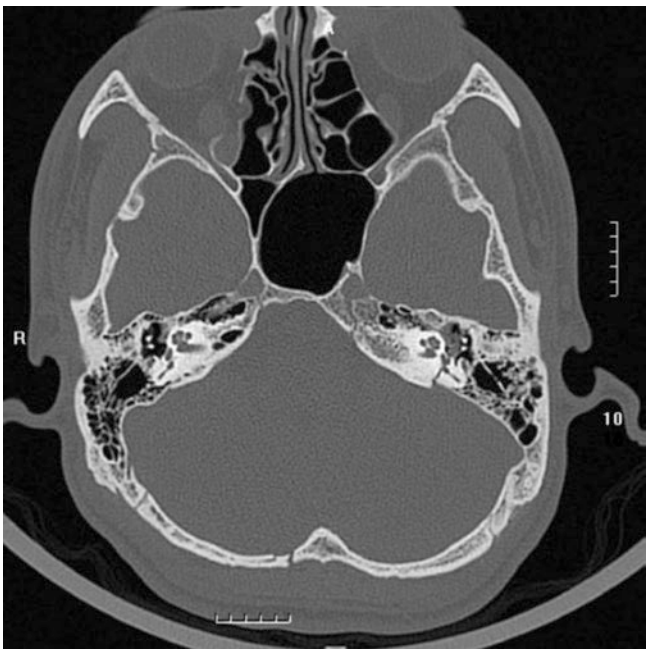


Fig. 8.10 The left temporal bone transverse fracture CT scan

Signs and symptoms include bleeding into the ear canal from skin and tympanic membrane laceration, hemotympanum, external auditory canal fractures, ossicular chain disruption that produces conductive hearing loss, and facial nerve paralysis. Twenty percent of longitudinal fractures injure the facial nerve and cause paralysis. The injury site is usually the horizontal segment of the nerve distal to the geniculate ganglion. CSF otorhinorrhea is common but usually temporary. Sensorineural

hearing loss may occur as a result of concussive damage. Vertigo occurs but is not related to the severity of the fracture.

2. Transverse fracture

   Transverse fractures comprise 20% of all temporal bone fractures. They are usually caused by a frontal or parietal blow but may result from an occipital blow. The fracture line runs at a right angle to the long axis of the petrous pyramid and starts in the middle cranial fossa (close to the foramen lacerum and spinosum). It then crosses the petrous pyramid transversely and ends at the foramen magnum. It may also extend through the internal auditory canal and injure the nerves directly. Cochlear and vestibular structures are usually destroyed, producing a profound sensorineural hearing loss and severe ablative vertigo. The intensity of the vertigo will decrease after 7–10 days and then continues to decrease steadily over the following 1–2 months, leaving only an unsteady feeling that lasts approximately 3–6 months, until compensation finally occurs. Intense nystagmus (third degree) is present since the initial fracture, with the fast component beating away from the fracture site. The nystagmus is easily seen by the naked eye. Nystagmus also decreases progressively in intensity (third degree, second degree, first degree) and then finally disappears.

   Rarely, a mixed hearing loss may occur. Facial nerve injury occurs in 50% of transverse fractures. The injury site is anywhere from the internal auditory canal to the horizontal segment distal to the geniculate ganglion. Pneumolabyrinth may be noted.

   Histopathology reveals hair cell loss, ganglion cell loss, and supporting cell loss. In rare cases, labyrinthitis ossificans occurs secondary to the trauma or subsequent infection. This must be kept in mind when cochlear implant is considered after a temporal bone fracture.

3. Mixed fracture

   These patterns, which extend both longitudinally and transversely, are common. According to some authors, these patterns occur more often than isolated transverse or longitudinal fractures. A range of 62–90% of temporal bone fractures were designed as a mixed pattern in medical literature.

4. Petrous apex fracture

   Rarely-seen. It can damage the second-sixth cranial nerves leading to corresponding eye symptoms and trigeminal neuralgia or facial sensation disorder. The petrous apex fractures can damage the internal carotid artery, leading to the fatal bleeding.

   The temporal bone fractures can be accompanied by cerebrospinal fluid leak. Cerebrospinal fluid leakage is light red in the early stage, gradually turns into crystal and the test result is sugary liquid (glycosuria test paper can be used).

Treatment

1. Keep airway clear and perform tracheotomy if necessary.
2. Control hemorrhage and infuses fluid or transfuses blood in time to prevent hemorrhagic shock and maintain the normal function of circulation.
3. Antibiotics should be applied in time and the ear should be sterilized to prevent infection of the encephalic infection or ear infection. If there is cerebrospinal fluid leak, take the head-up or half supine posture; most cerebrospinal fluid leakage can stop spontaneously. If leaking doesn't stop over 2–3 weeks, the dural defect can be repaired through the ear canal to control the cerebrospinal fluid leakage.
4. The peripheral facial paralysis caused by the temporal bone fracture should be operatively decompressed. Perforation of tympanic membrane, loss of acoustic chain, conduction deafness or facial paralysis and other malady should be repaired by tympanoplasty or facial nerve operation.

8.3 Disease of External Ear

8.3.1 Furuncle of External Auditory Canal

It is also called local otitis externa, mainly refers to acute suppurative lesions of the cartilaginous skin which occurs in the external ear canal as Fig. 8.11. Pathogenic bacterium is mainly the staphylococcus aureus and pseudomonas aeruginosa.

Clinical Manifestation

Symptoms and Signs

Patients have pain and drainage. Sometimes, a foul-smelling discharge and hearing loss occur if the canal becomes swollen or filled with purulent debris. Exquisite tenderness accompanies traction of the pinna or pressure over the tragus. Otoscopic examination is painful and difficult to conduct. It shows the ear canal to be red, swollen, and littered with moist, purulent debris and desquamated epithelium.

Otomycosis is more pruritic than painful, and patients also complain of aural fullness. Otomycosis caused by *A. niger* usually manifests with grayish black or yellow dots (fungal conidiophores) surrounded by a cottonlike material (fungal hyphae). Infection caused by *C. albicans* does not show any visible fungi but usually contains a thickened, creamy white exudate, which can be accompanied by spores that have a velvety appearance.

Furuncles cause severe pain and may drain sanguineous, purulent material. They appear as a focal, erythematous swelling (pimple) (Fig. 8.11).

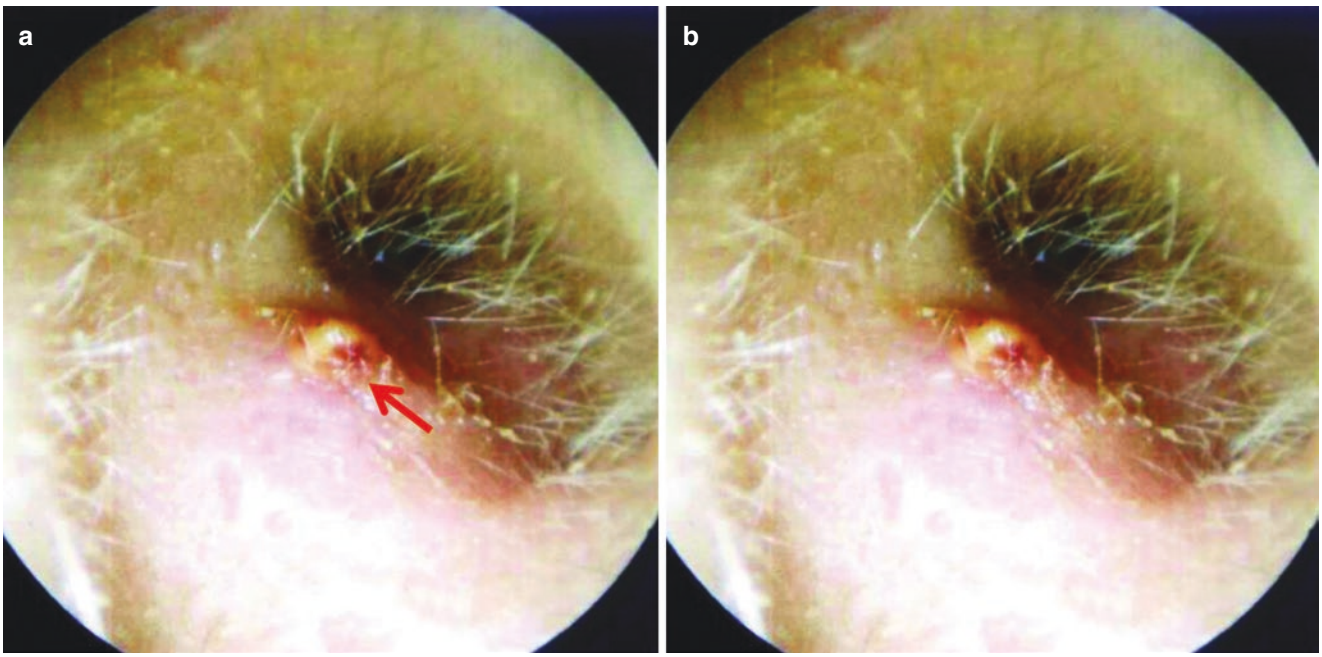


Fig. 8.11 Furuncle of external auditory canal

Treatment

1. Local therapy

   When the furuncle is not mature, sterilize the area locally with ethanol, apply the detumescence gauze dipped with ichthammol on the furuncle, and use antibiotics when necessary. In the presence of purulent bolt, incise with sterile scalpel after ethanol disinfection, remove the abscess, apply the antibacterial gauze with erythromycin on and replace it every day until the lesion is involuted. When the abscess is fluctuated, incise along the external ear canal after the ethanol sterilization to drainage.
2. Systemic therapy

   Patients with fever or systemic disease may be treated with oral or intravenous antibiotics according to local or systemic antimicrobial susceptibility tests result and do analgesic therapy when there is severe pain.

8.3.2 External Ear Canal Cholesteatoma

Clinical Manifestations

Often occur in adults. Uninfected people have no symptoms. Patients with Severe cholesteatoma showed ear coagulation sensation; tinnitus and secondary infections have ear pain, headache and smelly secretion in the external ear canal.

Specific Examination

1. The deep section of the external ear canal is blocked with white or yellow cholesteatoma, and the canal surface is covered with multilayer flaky substance (Fig. 8.12).
2. After removing the related-bigger cholesteatoma, there could appear bone destruction and absorption at the external ear canal, obviously expanded bony part of the external ear canal.
3. Entire tympanum membrane can be congested and invaginated.

The big external ear canal cholesteatoma invades the mastoid to damage its sclerotin (Fig. 8.13), complicating with the cholesteatoma's type middle ear mastoitis, causing peripheral facial paralysis.

Diagnosis

Pathogenic exam result of cholesteatoma could confirm the diagnosis.

Treatment

1. In the absence of concurrent infection, cholesteatoma is easier to be taken out and removed like cerumen.
2. In the presence of infection, pay attention to control infection. Partially remove or all of cholesteatoma.

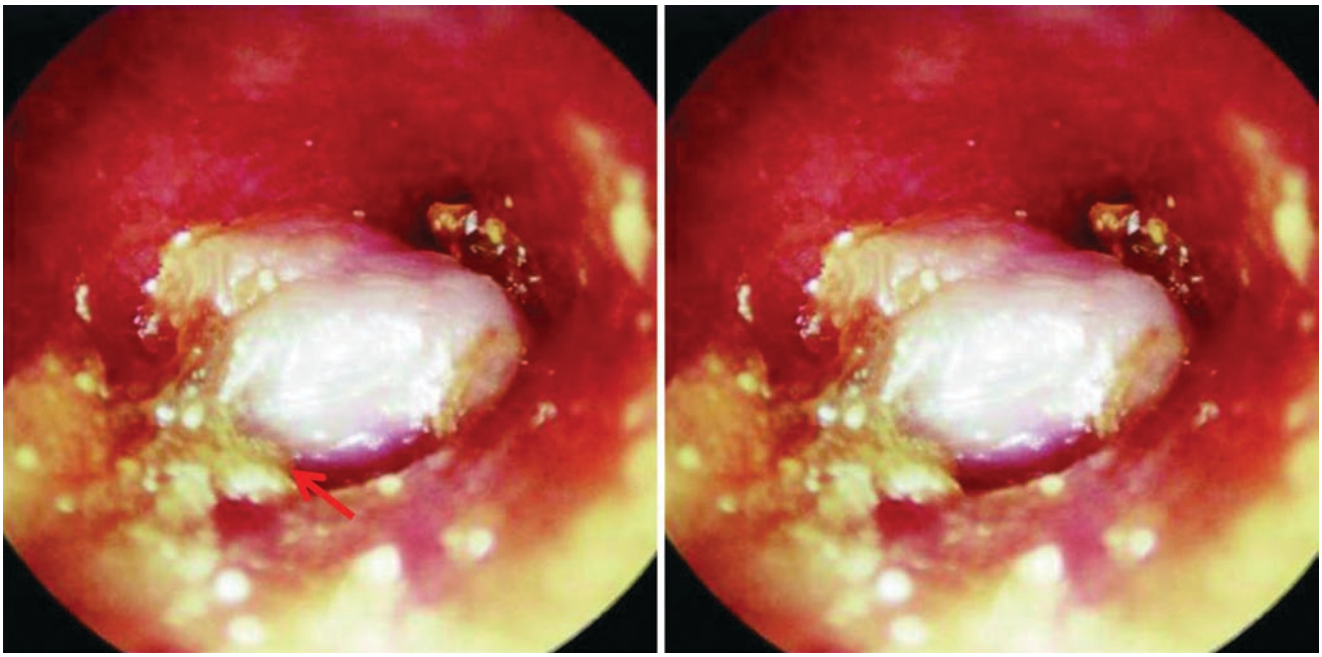


Fig. 8.12 The external ear canal cholesteatoma

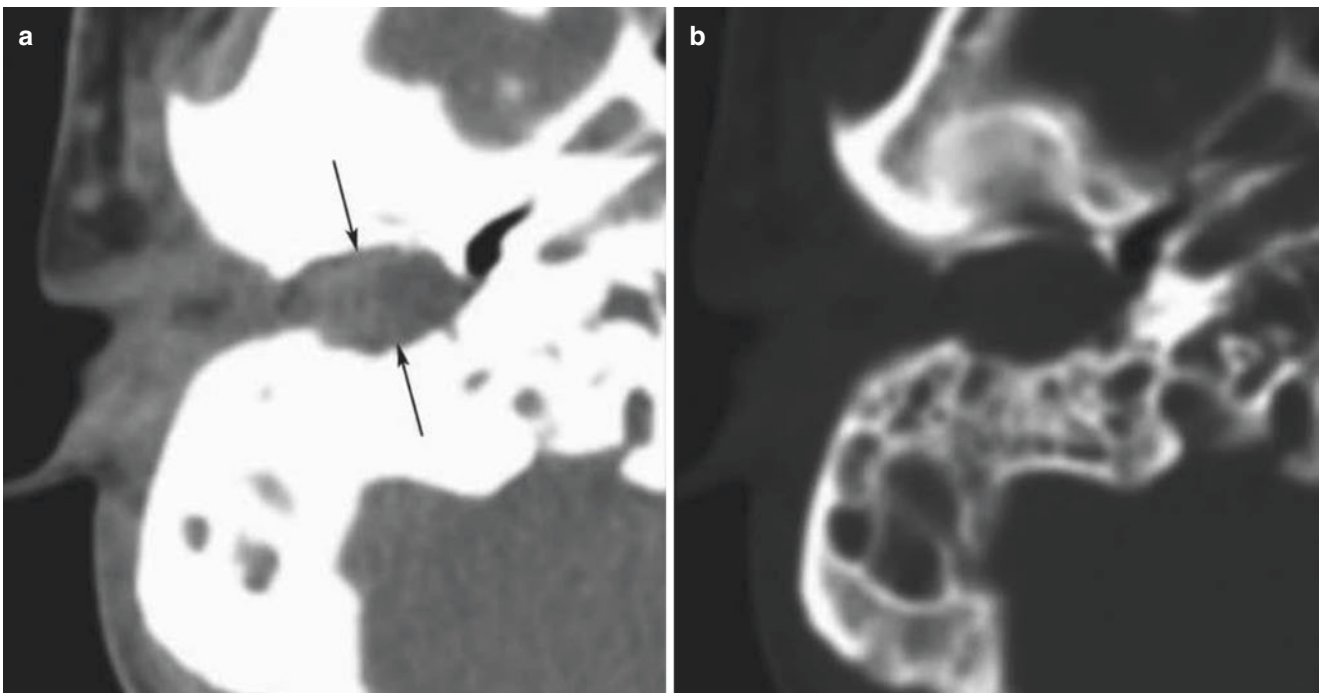


Fig. 8.13 Axial CT scan: external ear canal cholesteatoma. (**a**) Soft tissue fenestra, manifesting soft tissue density shadow filled in right external ear canal (arrow). (**b**) Bone fenestra, manifesting bone destruction of the right external ear canal, harden bone edge and slightly enlarged external ear canal; unwell gasified mastoid and increased mastoid sinuses density

3. In patients with severe infection, the chlosteatoma is to be removed under general anesthesia and operating microscope. Control the infection by applying broad-spectrum antibiotics. Follow-up, observe and remove the residual or regenerative cholesteatoma after surgery.
4. Patients with the external auditory meatus cholesteatoma which has invaded the mastoid should be treated by radical mastectomy or modified mastectomy.

8.4 Diseases of Middle Ear

Middle ear disease is common in otorhinolaryngology, head and neck surgery, with symptoms such as ear discomfort, pain, swelling sensation, occlusion sensation, otorrhea, hearing loss, tinnitus, dizziness, etc. Trauma, infection or pressure change caused by obstruction of eustachian tube in the middle ear are most causes of the disease. Fever, malaise and other symptoms are associated with infective middle ear disease. Otitis media is the most common one among middle ear disease.

Classification criterion of otitis media in 2012 by the academy of otolaryngology of Chinese Medical Association is as follows.

1. Secretory otitis media (SOM)
2. Suppurative otitis media
   (a) Acute suppurative otitis media
   (b) Chronic suppurative otitis media: resting stage; active stage.
3. Middle ear cholesteatoma
4. Special otitis media
   (a) Tuberculous otitis media
   (b) HIV positive otitis media
   (c) Mesotoxic otitis media
   (d) Fungal otitis media
   (e) Necrotizing otitis media
   (f) Radioactive otitis media
   (g) Aero-otitis media [2]

8.4.1 Secretory Otitis Media

Clinical manifestation

1. Hearing loss

   Most of patients with acute secretory otitis media have the history of upper respiratory tract infection, with gradual hearing loss, and louder speech.
2. Otalgia

   Acute secretory otitis media may cause slight otalgia at the onset, and it develops while secondary infection to patients with chronic SOM.
3. Inner ear occlusion sensation

   Adult patients always complain of occlusion or distention sensation and can be relieved temporarily by pressing the tragus.
4. Tinnitus

   Not heavy generally, but there could be a "crackling" sound. While the head moves, yawning or blowing the nose, gurgling appears.
5. Acoustic immittance

   The acoustic immittance test has great value to diagnose. The flat type (type B) is the typical curve of the disease, sometimes-high vacuum type (type C). All the acoustic reflexes disappeared.

Examination

1. Tympanic membrane

   Acute SOM will cause hyperemia at pars flaccid or entire tympanic membrane, manifestation as shortened, deformed or disappeared cones light, shifted-to-posterior and obvious malleus's axillary process. The tympanic membrane is yellowish, orange-red glossy or amber when the tympanic effusion happens. For the chronic SOM, the color is grayish blue or milky white as shown in Fig. 8.14. There is hairy line on tympanic membrane, and when the head position changes, it's still parallel to the ground. The bubbles could be seen through the tympanum membrane and may increase while doing eustachian tube insufflation.

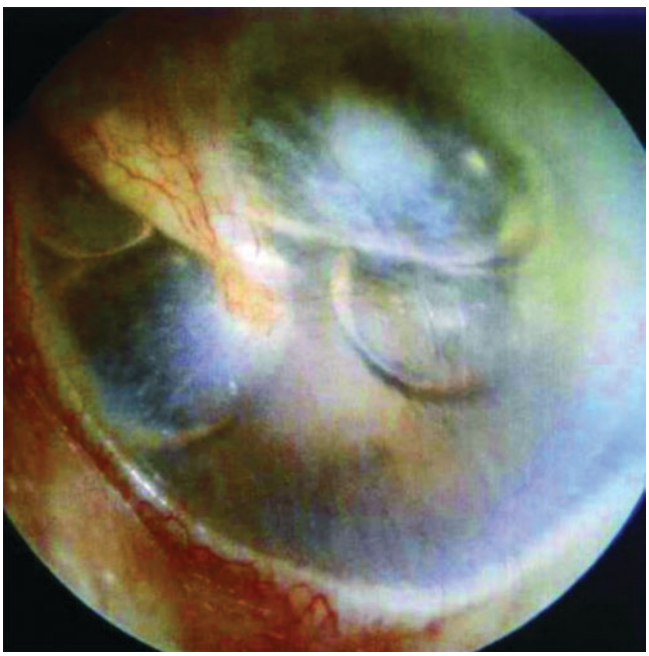


Fig. 8.14 Secretory otitis media, manifesting fluid flat tympanum hydrous and inside bubbles

2. Pneumatic otoscope examination

   It manifests the tympanum membrane movement limitedly.

3. Audition test

   Tuning fork test and pure tone threshold test show conductive deafness. In severe cases, hearing loss of about 40 dBHL can be found, mostly low frequency, and could be improved after eliminating the effusion. Acoustic immittance figure is mostly flat type (Type B). Vacuum type (Type C) suspects the dysfunction of eustachian tube, including cavum tympanum hydrops appearing in part of them. Severe patients could reach 100 dBHL at ABR test and can't be used to diagnose nerve deafness.

4. Imaging diagnosis

   Ossa temporal CT scan: different degrees of hydrops occur in the pneumatic cavity of middle-ear system. Most of CT values are under 40 Hu.

   Mastoid MR Examination: the mastoid, mastoid cells without any signals, becomes hypersensitive and has high specificity diagnosis value (Figs. 8.15 and 8.16).

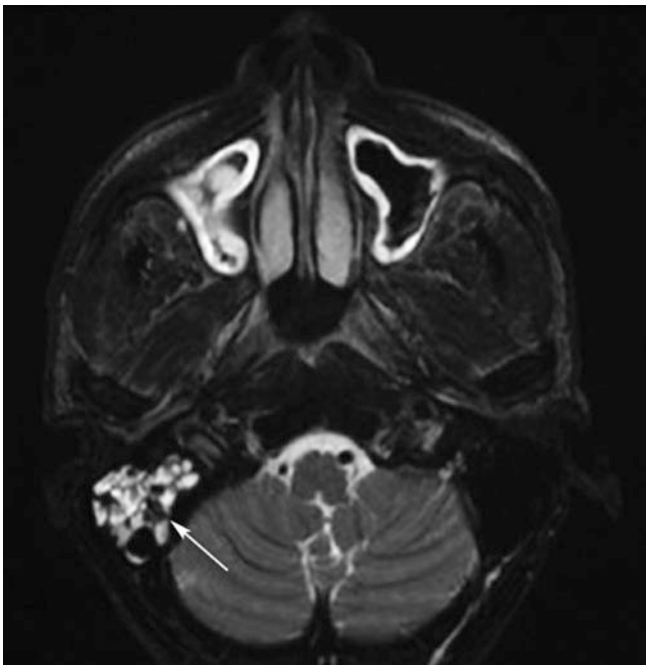


Fig. 8.16 Bilateral mastoid plain MR scan (T_2WI). Well-gasified left nidus Vespa without signal manifestation, right mastoid nidus Vespa filled with T_2WI-high-signal fluid (arrow)

Differentiate Diagnosis

Tympanotomy could help make a definite diagnosis.

Treatment

The first choice for the patients is 3-month non-surgical therapy grasping the surgical indications strictly. Etiology therapy, improving the middle ear ventilation drainage and removing the tympanic effusion are the therapeutic principles of the disease.

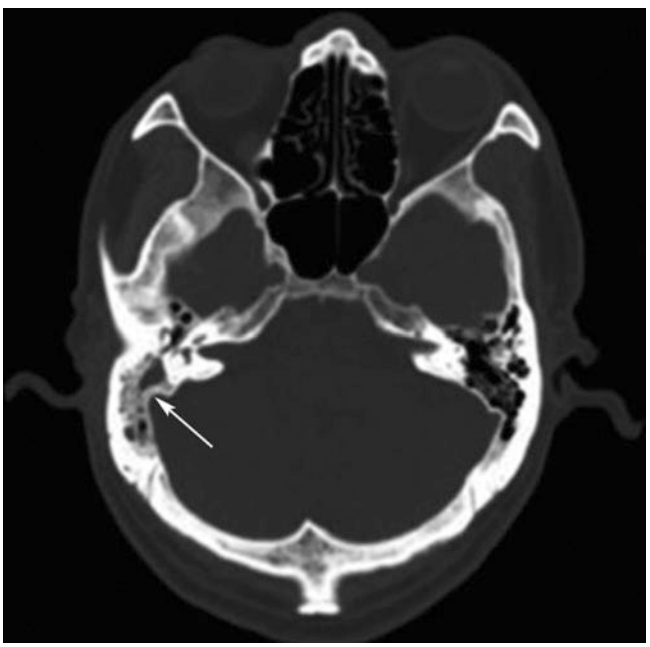


Fig. 8.15 Bilateral mastoid plain CT scan (bone fenestra). Well-gasified left nidus Vespa, right mastoid nidus Vespa filled with slightly high-density fluid (arrow)

1. Non-surgical therapy
   (a) Antibiotics: select the appropriate antibiotics according to the severity of the lesion at acute phase.
   (b) Keep the nasal cavity and the eustachian tube unobstructed: inhale intranasal with 1% ephedrine solution and antibiotics contain hormones alternately, 3–4 times each day.
   (c) Promote cilium movement and excretion function: dilute mucin beneficial to ciliary excrete, lower superficial tension of eustachian tube mucosa and the opening pressure of the eustachian tube.
   (d) Takes glucocorticoid drug, dexamethasone, prednisone orally as adjuvant therapy.
2. Surgical therapy
   (a) Eustachian tube insufflation: Valsalva maneuver. Wave's ball method outraging method at chronic phase
   (b) Tympanic puncture and drainage: local anesthesia for adult patients and general anesthesia for infants.
   (c) Tympanotomy: Considered for patients with mucous effusion which cannot be cleaned by puncture. A radial or arc-shaped incision is made in the interior and lower quadrants of tympanic membrane with tympanic knife. Mucosa of tympanic wall should not be injured. After tympanic incision, fluid cavity should be absorbed.
   (d) Tympanoplasty: If the effusion is too viscous to be discharged and the eustachian tube function is difficult to return to normal in a short time after head radiotherapy, tympanoplasty can be considered (Fig. 8.17).

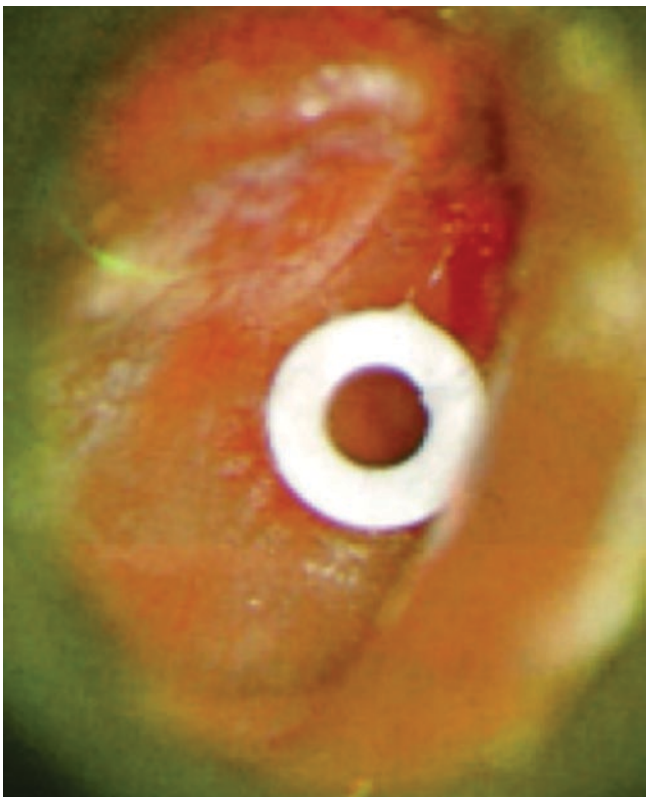


Fig. 8.17 Tympanoplasty

(e) Long-term repeated attacks, suspecting granulation tissue formation or other irreversible lesion formed in the middle ear mastoid cavity or ossicular ossicles destruction, early simple mastoidectomy, epitympanotomy, upper tympanostomy or posterior tympanostomy should be performed to clear the lesion.
(f) Active treatment of nasopharyngeal or nasal diseases.
(g) Balloon angioplasty: In the early twenty-first century, Europeans developed a kind of balloon dilation catheter to treat sinusitis. Ockermanm (2010) and it was first applied to the eustachian tube by imputing the saccule at the Eustachian tube orifice to expand the tube under guidance of nasal endoscopy on a cadaveric head model. A slight crack at eustachian tube cartilage was found but no injury to the bone which confirms the feasibility and security of the balloon angioplasty. Subsequently, he applied it in the clinic, and found that the function of eustachian tube was significantly improved after surgery without any obvious complications. The results of 2-year follow-up showed that the balloon dilatation had a good long-term effect.

Secretory otitis media is closely related to dysfunction of eustachian tube, and dysfunction of eustachian tube may be related to dysfunction of tensor veli palatinis and decrease of surfactant in eustachian tube. Balloon dilatation of eustachian tube can cause small tear in the submucosa of eustachian tube, thinning the submucosa and enlarging the lumen. Moreover, the teared tissue is repaired by fresh scar tissue, i.e. collagen fibers are compressed and filled by fibroblasts, neovascularization and inflammatory cells, instead of collagen fibers, which are regenerated and repaired. Therefore, it is not easy to restenosis. After enlarging the eustachian tube lumen, it can improve drainage and restore ciliary function. It may also redistribute the active substances on the surface of eustachian tube mucosa and help to restore its function.

Surgical methods: After general anesthesia, the nasal cavity was contracted by 1% adrenaline for 5 min. The nasal cavity and nasopharynx were examined by 0 nasal endoscopy. The tip of 70 or 30 or 45 duct was placed at the nasopharyngeal orifice of the eustachian tube, and the guide wire was introduced into the eustachian Pump pressure, swell balloon, slowly add to 10 atmospheric pressure, maintain 2 min, pump decompression, suction to negative pressure, exit balloon and guide wire.

If the nasal cavity can not be inserted into both nasal endoscopy and balloon catheter implant due to anatomical reasons, nasal endoscopy can be inserted through oropharyngeal approach, and balloon catheter can be inserted through nasal cavity.

8.4.2 Acute Suppurative Otitis Media

Clinical Manifestations

1. Systemic symptoms are chills and fever. Children have severe systemic symptoms, such as high fever, convulsions, vomiting and diarrhea. Once the tympanic membrane is perforated, the body temperature gradually decreases and the symptoms of the whole body are obviously alleviated
2. Otalgia: deep ear throbbing pain or tingling, can radiate to the ipsilateral head or teeth, the pain is intense, causing restlessness, unable to sleep. Children cry endlessly, turn their head and neck, grasp their ears with their hands and refuse to eat. After tympanic membrane perforation and purulence, the earache is relieved.
3. Tinnitus: deafness, stuffy hearing, hearing loss gradually. When the earache is severe, deafness is often neglected, occasionally accompanied by vertigo, and deafness is alleviated after tympanic membrane perforation.
4. Otorrhea: After tympanic membrane perforation, there are mucopurulent efflux, which can be blood and water samples at first, and then become purulent.

Examination

1. Otoscopy

   Otoscope examines tympanic membrane congestion, which is at first congestion of the flabby malleolus stalk, then radial congestion of the tense part, and finally diffuse congestion and bulging outward. Finally, tympanic membrane perforation, because of small perforation, pus pulsatile overflow, visible flashing pulsatile light, known as "beacon sign", necrotic can see large tympanic membrane perforation (Fig. 8.18).
2. Ear palpation

   Mild tenderness appears at the tympanum antrum site of mastoid.
3. Audition examination

   Conductive deafness often.
4. Hemogram

   Quantity of WBC and neutrophils increases, and the hemogram gradually becomes normal after tympanum tresses.

Treatment

The principle is anti-infection, drainage and removing roots.

1. Systemic therapy
   (a) Apply the antibiotics early at full dose:

   After doing tympanic membrane tresis, take the abscess for fine culture and drug sensitization. Choose the appropriate antibiotics due to the reference result, and stop the pills after keeping treating for couple days after the symptoms are gone.

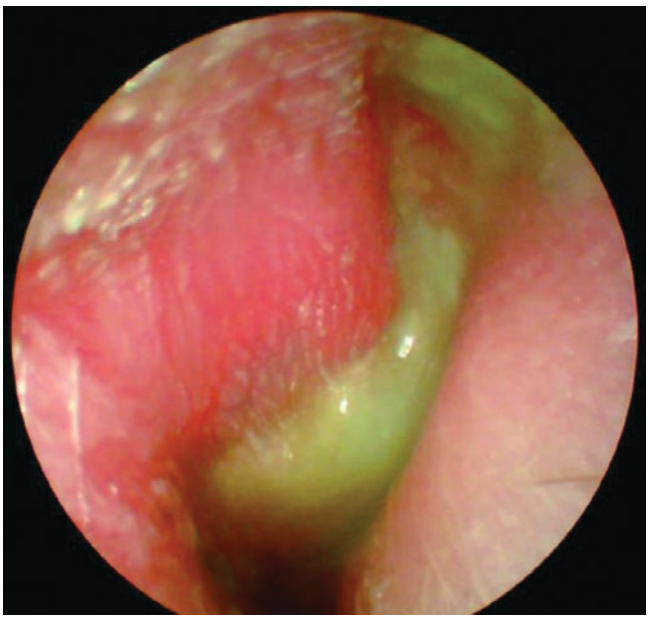


Fig. 8.18 Acute suppurative otitis media under microscopy

   (b) Rest enough, adjust the diet and keep bowels open. The severe patient should pay attention to the support therapy like applying the glucocorticoids, etc. Ask the pediatrician to cooperate while necessary.
2. Local therapy
   (a) Ear inhalation

   Use 2% phenols to do ear inhalation before the tympanic membrane tresis; after perforating, use 3% peroxide to clear the external ear canal and inhalation antibiotics ear drops.
   (b) Tympanic membrane incision therapies: accurate tympanic membrane dissection can be unobstructed drainage, is conducive to the rapid dissipation of inflammation, so that systemic and local symptoms are alleviated.
   (c) Short-term uses of nasal congestion drugs: such as 1% ephedrine nasal drops, reduce the swelling of nasopharyngeal mucosa; help restore Eustachian tube function.
3. Etiology therapy

   Active therapy of chronic diseases of the nose and pharynx.

8.4.3 Chronic Suppurative Otitis Media

Chronic suppurative otitis media is chronic suppurative inflammation at middle ear mucosa, bone periosteum or bone cortex, its clinical features are long-term intra-auricular pus, tympanic membrane perforation, hearing loss, etc.

Clinical Manifestation

1. Intra-auricular discharge the overflow can be discontinuous or persistent. Secretions can be sticky or thin, sometimes with blood. Amount of secretions differ, and discharge would increase when water flows into ear.
2. Hearing loss: Hearing loss degrees differ.
3. Tinnitus: Some patients may be with paroxysmal or persistent tinnitus.

Examination

1. Tympanic membranous perforation

   It would locate in dense part, and size and shape of perforation usually differ, which can be manifested as central small perforation, kidney shaped perforation or large perforation, but there are residual edges in the tympanic membrane, and there is no destruction of the tympanic ring. Smooth tympanum antrum mucosa or slight tympanum antrum edema would be observed through proliferation (Fig. 8.19). Ossicular chain is

usually complete or only partial malleus handle is necrotic.

2. Hearing test generally result manifest as mild conductive hearing loss.
3. Mastoid X-ray imaging or Temporal CT scan

   Mastoid may manifest as gasified type or barrier type, soft tissue shadow would be seen in middle ear. No cortex damage would be noticed (Fig. 8.20).

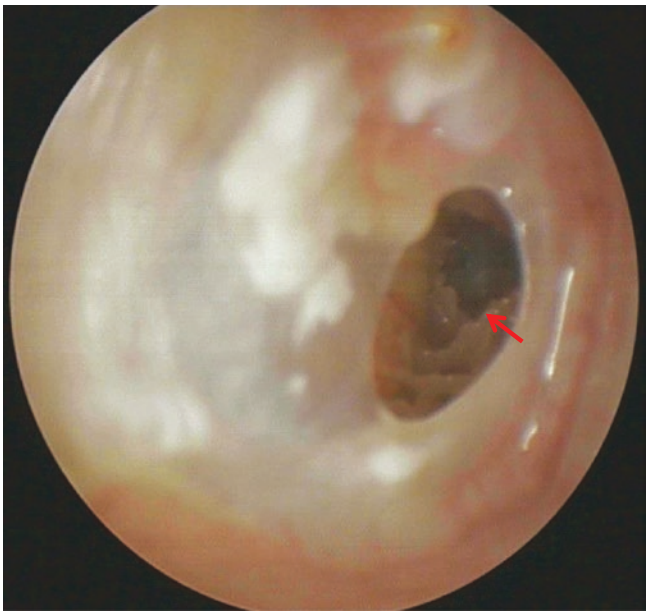


Fig. 8.19 Tympanic membranous perforation

Treatment

If drainage is unblocked unblocked, local medication can be used, such as 0.3% ofloxacin ear drops, 2.5% chloramphenicol glycerin ear drops and 3% boric acid ear drops. 3% hydrogen peroxide solution can be used to wash ears before medication. The principle of treatment is to prevent recurrence and to restore hearing by tympanoplasty after inflammation control. In inflammatory stage, antibiotics should be selected reasonably according to the results of bacterial culture and susceptibility test. When the local purulent secretion is large, it should be cleaned with 3% hydrogen peroxide solution, cleaned or sucked up by an aspirator, and then dripped with antibiotic ear drops, such as 0.3% ofloxacin solution and 0.5% chlortetracycline solution. After inflammation control, tympanoplasty can be considered to reconstruct hearing.

8.4.4 Cholesteatoma of the Middle Ear

Cholesteatoma of the middle ear is a cystic structure located in the middle ear. It is composed of stratified squamous epithelium and contains cholesterol crystals, exfoliated skin cells, keratins and bacteria. It is not a true tumor. According to the pathogenesis, it is usually divided into acquired primary cholesteatoma and acquired lipoma. Cholesteatoma can be secondary to chronic suppurative otitis media. Chronic suppurative otitis media can also be secondary to bacterial infection of cholesteatoma. Therefore, this disease is called chronic otitis media with cholesteatoma.

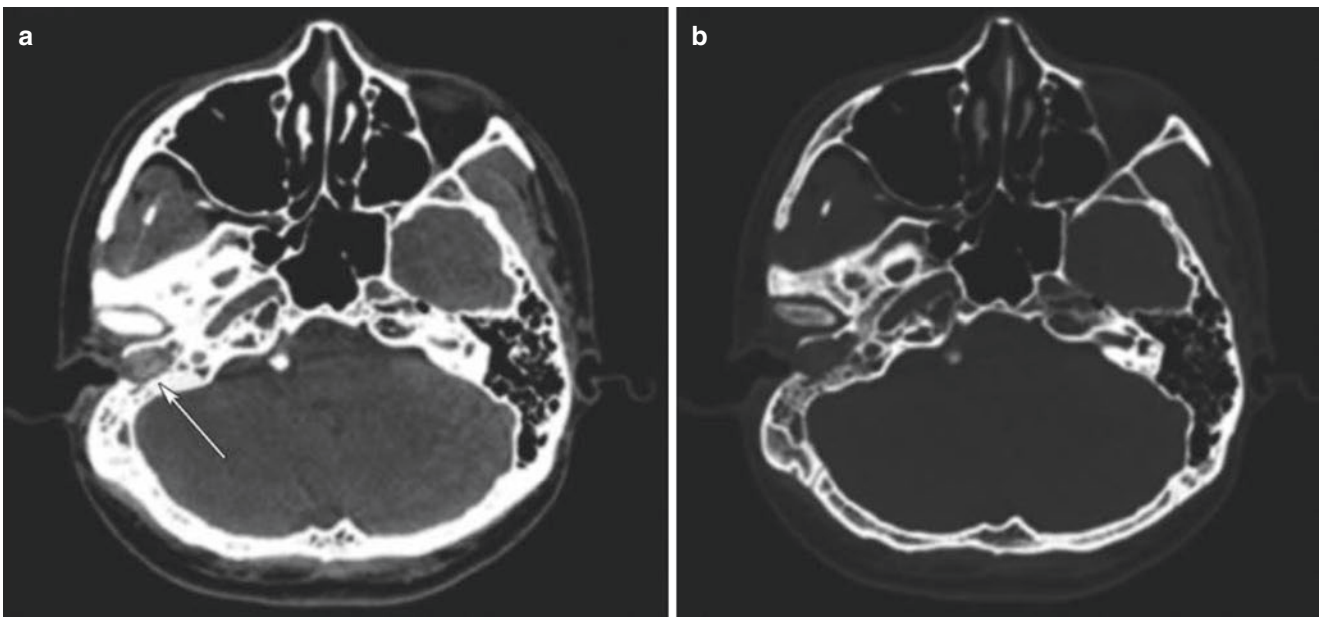


Fig. 8.20 Temporal axial CT scan of right chronic middle ear mastoid otitis. (**a**) Soft tissue window CT plain scan; (**b**) bone window CT plain scan. Well-gasified left mastoid, no abnormality; small right mastoid nidus vespa, within high density shadow, middle ear cavity and mastoid sinus enlarges and little sclerotin damage within soft tissue density shadow stuff (arrow)

Clinical Manifestation

1. Acquired primary cholesteatoma is rare. No Otorrhea or tympanic membrane perforation can be found before infection. Acquired cholesteatoma is characterized by long-term otorrhea, pyotympanic membrane perforation and hearing loss
2. Long-term persistent or intermittent otorrhea. Stench exists.
3. Complication: Cholesteatoma of the middle ear may cause intracranial and extracranial complications, such as subperiosteal abscess, peripheral facial paralysis, labyrinthine inflammation, epidural abscess, thrombophlebitis of the sigmoid sinus, meningitis, brain abscess and even hernia, because of its strong bone destruction characteristics. Serious cases may cause death
4. There are marginal perforations in the relaxation or tension. From the perforation, there are grayish-white scaly or bean dregs-like substances in the tympanic chamber with strong odor (Fig. 8.21)

Examination

1. Audiological test

   It is usually a severe conductive hearing loss, such as lesions and the cochlea. Mixed hearing loss.
2. CT examination

   For patients with clinically suspected cholesteatoma high-resolution temporal bone CT scan, to accurately understand the cholesteatoma scope, ossicular's change. The destruction of the facial nerve canal, semicircular canal, tympanic cavity, etc. Which is typically characterized by soft tissue shadow in the middle ear, homogeneous density, dense and sharp border, and often accompanied by bone destruction (Figs. 8.22 and 8.23).

Treatment

Once the middle ear cholesteatoma is diagnosed, it should be operated as soon as possible. There are many kinds of surgical methods, mainly open and close (wall-closing) two types. Endoscopy can be used for wall-closure surgery to see areas that are difficult to be seen under the operating microscope, such as the upper tympanic chamber, tympanic sinus, eustachian tube and so on, which is expected to greatly reduce the incidence of residual lesions. After cholesteatoma is com-

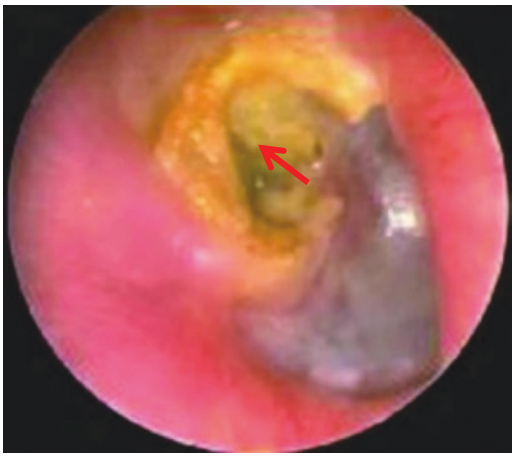


Fig. 8.21 Middle ear cholesteatoma endoscopy

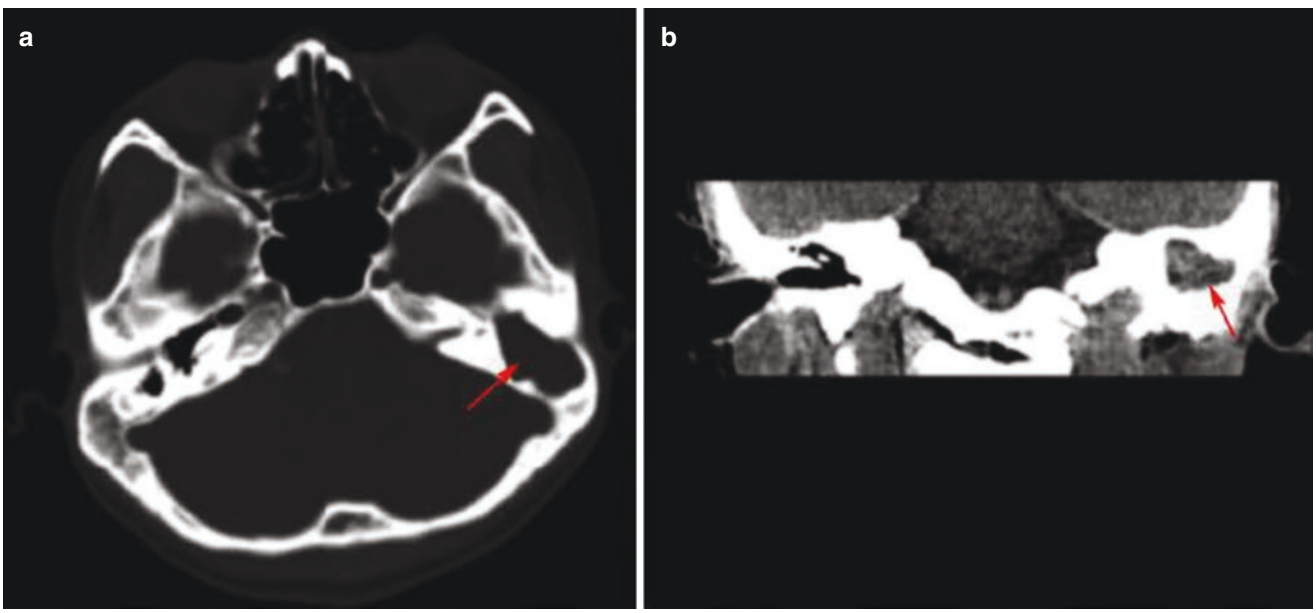


Fig. 8.22 Left mastoid cholesteatoma's temporal CT scan manifestation. (**a**) Bone window axis. (**b**) Soft tissue window coronary reconstruction. Left mastoid is sclerosis type, where left mastoid sinus and aditus extend and large sclerotin damage zone could be seen, within soft tissue density shadow stuff (arrow) and its peripheral sclerotin has hyperplasia sclerosis

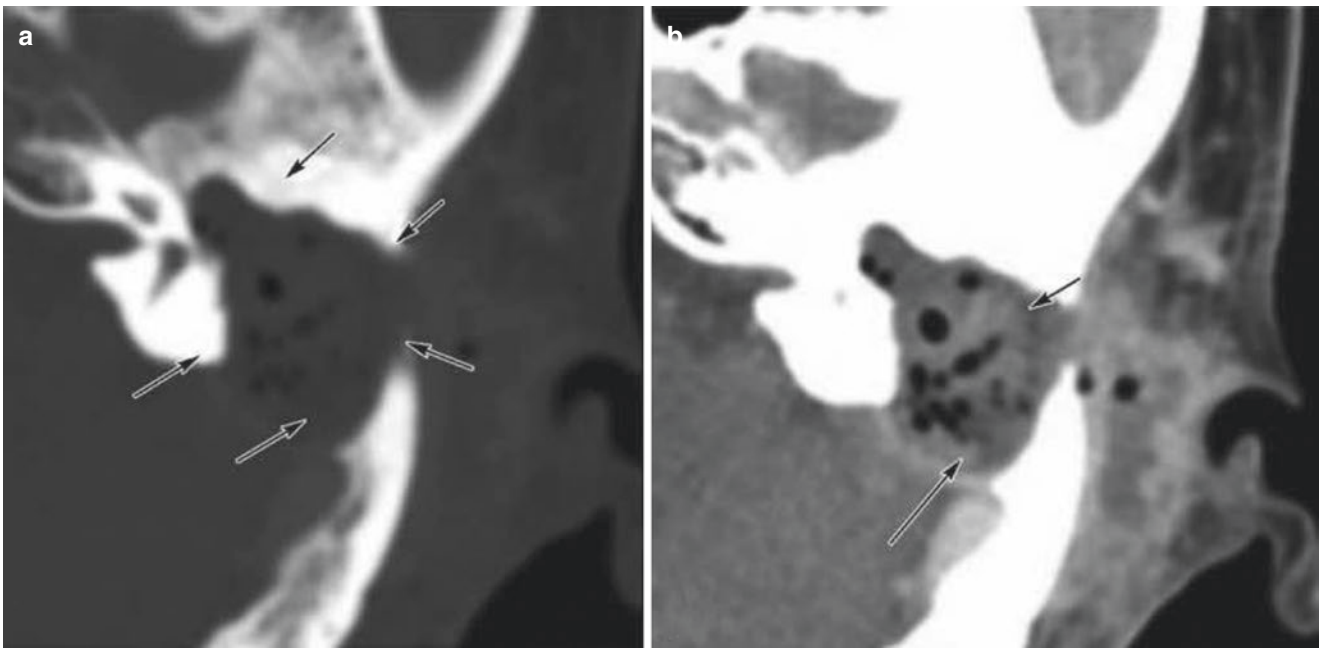


Fig. 8.23 Temporal axis CT plain scan imaging manifest large cholesteatoma. (**a**) Bone window says left mastoid sinus enlarges, sinus wall sclerotin damage and sclerotin hyperplasia sclerosis (arrow); (**b**) enhanced scanning says non-enhance mastoid intra-sinus soft tissues, sinus cavity and its adjacent soft tissues communicate and constitute a sinus canal (short arrow),internal wall sclerotin damage and involved endo-cranium (long arrow)

pletely removed, ossicular chain reconstruction and tympanoplasty can be performed as appropriate to preserve or improve hearing.

8.4.5 Sequelae of Otitis Media

1. Atensive/Adhesive Otitis Media;
2. Tympanosclerosis;
3. Cholesterol granuloma of the middle ear;
4. Occult otitis media.

8.5 Otosclerosis

Clinical Manifestation

1. Hearing loss

   There is no cause for gradual hearing loss in both ears. Hearing loss is slow and gradually aggravated. Excessive fatigue, excessive tobacco and alcohol, and after pregnancy and childbirth can cause hearing loss significantly aggravated. Affect patients' social activities.
2. Tinnitus

   Tinnitus intermittent or persistent bass tinnitus. Most occur simultaneously with deafness.
3. Willis mishearing

   It is better for the patient to have a better hearing in the noisy environment than in the quiet environment. This phenomenon is called Willis's wrong listening or Willis's auditory perversion.
4. Vertigo

   A few patients experienced mild transient vertigo after head movement.

Examination

1. Signs

   The outer ear canal is more spacious, the tympanic membrane is normal, and the activity is good. Sometimes, at upper quadrant of tympanic membrane, a red area is seen, which is the manifestation of hyperemia of the promontory mucosa. This phenomenon is called Schwartz sign, which is one of the characteristics of clinical otosclerosis.
2. Audiometry
   (a) Tuning fork test RT 256 Hz negative, 512 Hz positive showed early hearing damage; 256 Hz and 512 Hz were negative, suggesting that hearing damage aggravated. Gelle test, a test of the mobility of the ossicles, is negative.
   (b) Pure tone audiometry Bone conduction hearing curve is V shape under 1000 Hz or 2000 Hz region decreased said Kaladze notch, suggesting that the stapes floor be fixed, will be eliminated after a successful operation.

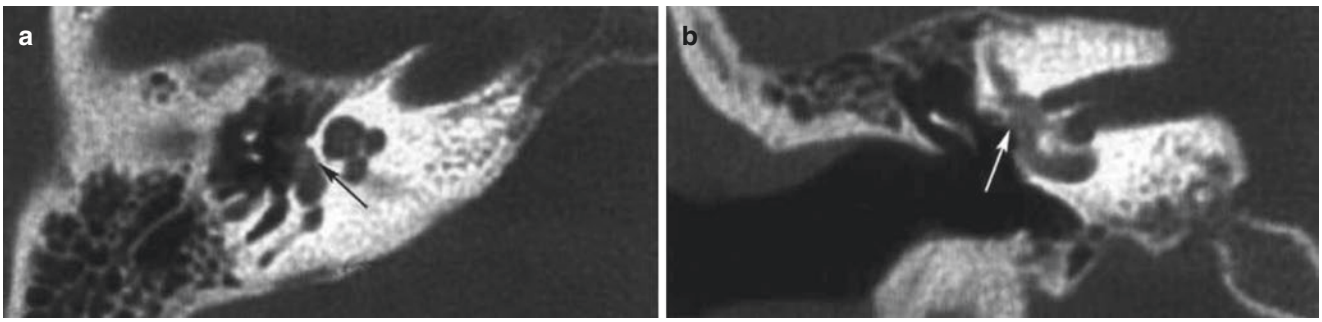


Fig. 8.24 Otic sclerosis HRCT imaging. (**a**) Axis imaging, manifestation thickened stapes basal plate (arrow); (**b**) Coronary reconstruction imaging, manifesting bone labyrinth sclerotin absorption, density reduction and point calcified lesion (arrow)

(c) Acoustic impedance test: The tympanic curve showed A type, As type or biphasic curve, and the reflex threshold of stapes muscle increased or disappeared.
(d) Auditory brainstem response test: I wave and V wave latency prolongation or threshold increases.

3. Imaging examination

Temporal bone X-ray showed no lesions at middle ear and mastoid. High resolution spiral CT scanning of the temporal bone revealed thickened stapes floor, and sclerosis of the vestibular window, cochlear window, bony labyrinth and inner ear tract as Fig. 8.24.

Treatment

1. Surgical method: Stapes surgery and inner ear fenestration were selected according to the condition. Stapes surgery includes stapes swing and stapes resection. Fenestration of stapes floor and implantation of artificial auditory ossicles. Stapedectomy can use hand drill, drill and laser.
2. Medical therapy: Sodium fluoride therapy and chondroitin sulfate therapy.
3. Hearing aids can be selected for those who are not suitable for or unwilling to undergo surgery, and cochlear implants can be performed for those with severe bilateral deafness.

8.6 Disease of Facial Nerve

8.6.1 Facial Nerve Anatomy

1. Facial nerve composition

Facial nerve is composed of motor nerve, parasympathetic nerve, taste fiber and sensory nerve fiber.

(a) The motor nerve originates from the motor nucleus at the lower part of the pons, and innervates all facial expression muscles except the levator palpebrae superioris, buccal and stylohyoid muscles of stapes.
(b) Parasympathetic nerve originates from the superior salivary nucleus and distributes in lacrimal gland, submandibular gland and sublingual gland respectively through the sphenopalatine ganglion and the mandibular ganglion.
(c) The gustatory fibers mainly connect the anterior 2/3 gustatory receptors of the tongue and pass through the genicular ganglion to the nucleus tractus solitarius.
(d) The sensory fibers are mainly the skin sensation of the auricle and part of the external auditory canal.

2. The course of facial nerve

According to route of facial nerve motor nerve fibers, courses can be divided into the following sections.

(a) Superior nuclear segment: It originates from facial nerve center at cerebral cortex and descends towards facial nerve motor neuclei.
(b) Nuclear segment: It is course of facial nerve in pons.
(c) Cerebellopontine angle segment: It is the segment from subpons edge to inner ear aditus.
(d) Inner ear canal segments: It is the segment from inner ear aditus to inner ear canal basement.
(e) Labyrinthine segment: It is the segment from inner ear canal basement to geniculate ganglion.
(f) Tympanic segment: It is the course at tympanic inner membrane, from geniculate ganglion to pyramidal eminence. It divides into greater superior petrosal nerve which ends at lacrimal gland.
(g) Mastoid segment: It is the segment from pyramidal eminence to stylomastoid foramen, during which it divides stapes muscle and tympanic chord nerve.
(h) External temporal segment: It is the segment which ends at pathetic muscle through parotid gland after crossing stylomastoid foramen.

8.6.2 Diagnosis of Peripheral Facial Paralysis

Peripheral facial paralysis is caused by facial nerve nucleus or lesions below the nucleus.

Clinical Manifestation

The main manifestations are facial muscle disorders of voluntary movement, including the disappearance of frontal lines, inability to frown, raise eyebrows, incomplete closure of eyes, shallow nasolabial sulcus, droop and tilt to the opposite side, more obvious when speaking and showing teeth, leakage of breath when whistling, and easy outflow along the mouth corner when drinking water. There may be a loss of taste, tear or saliva secretion. The face is stiff and expressionless when facial paralysis is complete on both sides.

Etiological diagnosis:

1. The causes of peripheral facial paralysis can be preliminarily understood by detailed inquiry and medical history collection. These causes include congenital facial nerve deformities, bacterial and viral infections such as otitis media, herpes zoster, temporal bone fractures caused by trauma, and facial neuritis.
2. Physical examination

   Detailed physical examinations, including static and motor examinations, otolaryngology and nervous system examinations, and general examinations. Examination can provide valuable clues for diagnosis. House-Brackmann classification is usually used to assess the degree of facial paralysis (Table 8.4).
3. Imaging examination

   CT, MRI and ultrasonography.
4. Localization diagnosis
   (a) Lacrimal gland secretion test
   (b) Stapes reflex
   (c) Taste test
   (d) Saliva secretion test
5. Electrophysiological diagnosis
   (a) Electromyography
   (b) Nerve electrogram
   (c) Nerve excitability test

8.6.3 Common Peripheral Facial Paralyses

1. Bell's Palsy

   Bell's palsy is an acute peripheral facial paralysis of unknown origin, also known as idiopathic facial paralysis. It can occur at any age, but more people are 20–40 years old.

   The exact cause is unknown, but it may be related to ischemia caused by vasospasm, viral infectious immune response, heredity and other factors. The clinical manifestations were sudden and rapidly aggravated peripheral complete or incomplete facial paralysis. There may be a history of cold wind or viral infection. Early stage may be accompanied by side ear or subauricular pain, a small number of facial and tongue numbness, facial tactile abnormalities. The mastoid process may have tenderness, the posterior part of the tympanic membrane may have slight congestion, but it will disappear in a few days. To diagnose this disease, other diseases causing peripheral facial paralysis should be excluded.

   Treatment is divided into non-surgical treatment and surgical treatment. Non-surgical treatments include glucocorticoids, vasodilators, vitamins, physiotherapy, acupuncture and moxibustion, etc. Pay attention to cornea protection. Surgical treatment is mainly facial nerve decompression, but its indications, timing and scope of decompression are still controversial.
2. Auricular herpes zoster

   Herpes zoster virus infection of facial nerve induced peripheral facial paralysis, also known as Hunt syndrome, was reported by Ramsay hunt for the first time in 1910. The clinical feature is ear herpes accompanied by peripheral facial paralysis. First, common in the early stage of severe earache, the hyperemia, cluster appeared herpes conchae and around after herpes broken exudate (Fig. 8.25). Peripheral facial paralysis occurs later, can be incomplete facial paralysis, severe cases can be complete

Table 8.4 House- Brakeman facial nerve functional recovery assessment criteria

Damage degree	Level	Definition
Normal	I	Bilateral symmetry, normal function of each district
Mild facial paralysis (just perceptible)	II	Slight facial nerve movement inability, complete eye closure under slight force. Slight facial asymmetry while strongly smiling, slight synkinesis, no facial spasm
Moderate facial paralysis (significant difference)	III	Obvious inability of facial nerve movement, no facial appearance loss, possible eyebow raisement disability, complete eye closure while under force, powerful mouth movement while forcing but asymmetric, obvious synkinesis and spasm
Moderate and severe dysfunction	IV	Obviously inable facial muscle movement, appearance loss, eyebow raisement disability, unable to completely close eyes even while under force, asymmetric mouth movement, obvious synkinesis and spasm
Severe facial paralysis	V	Slight facial movement, palpebral closure disability, only slight quarrel movement at mouth corner, synkinesis and spasm disappeared
Complete facial paralysis	VI	Facial muscle movement disability, tension loss, no synkinesis, no spasm

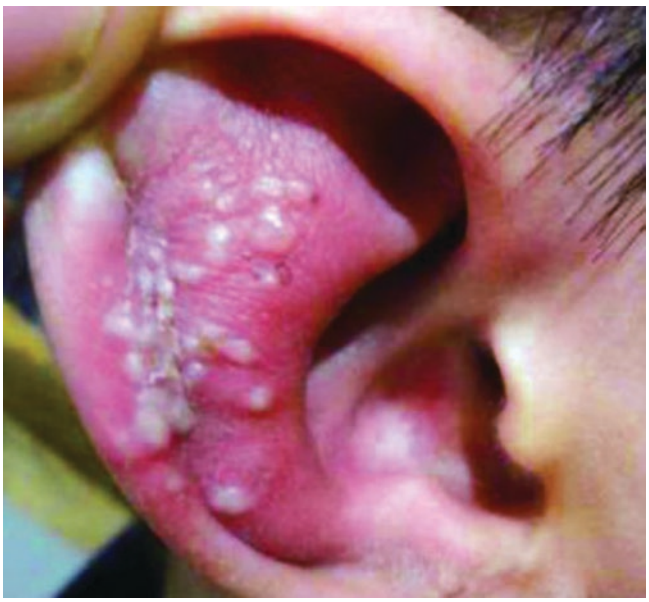


Fig. 8.25 Auricular herps zoster

facial paralysis. If the virus invaded the cochlear nerve, vestibular nerve, trigeminal nerve, can appear ear, deafness, tinnitus, vertigo, facial pain. Other patients may have other neurological symptoms and signs. The therapy principle is similar to Bell facial paralysis.

8.7 Malignant Tumor of Temporal Bone

Temporal Bone Malignant Tumors

It accounts for 1/5000–1/20,000 of otological cases, and squamous cell carcinoma (SCC) is the most common. Squamous cell carcinoma accounted for 60–80% of the external auditory canal, middle ear and mastoid process, followed by adenocarcinoma, cystadenocarcinoma and basal cell carcinoma (Fig. 8.26), and melanoma was rare (Moffat et al.) summarized six histological types of squamous cell carcinoma: highly differentiated, moderately differentiated, poorly differentiated, clear cell morphology, spindle and verrucous squamous cell carcinoma. The incidence of squamous cell carcinoma of the temporal bone is 1/1,000,000,000–6/1,000,000,000, of which 60–70% occur in the auricle, 20–30% in the external auditory canal, and 10% in the middle ear and mastoid process. Age is more than 50–60 years old, there is no gender difference. The vast majority of auricles are basal cell carcinomas. The following is limited to squamous cell carcinoma of the temporal bone.

Etiology

The main cause is exposure to ultraviolet or excessive radiation, such as radiotherapy for nasopharyngeal carcinoma, especially in people with delicate skin. In 18 cases of temporal bone tumors, 39% (7/18) were radiation-related tumors (after radiotherapy for nasopharyngeal carcinoma): 5 squamous cell carcinomas and 2 sarcomas. For uncovered sites, such as squamous cell carcinoma of the external auditory canal, it may be related to genes. In addition, the correlation between chemical agents, such as chlorine-containing disinfectants, has also been reported. Chronic suppurative otitis media mentioned in previous literature is a significant pathogenic factor, which has not been confirmed yet. Most middle ear cancers with chronic otitis media are infected by human papillomavirus (HPV). About 50% of patients had a long history of otorrhea before using antibiotics.

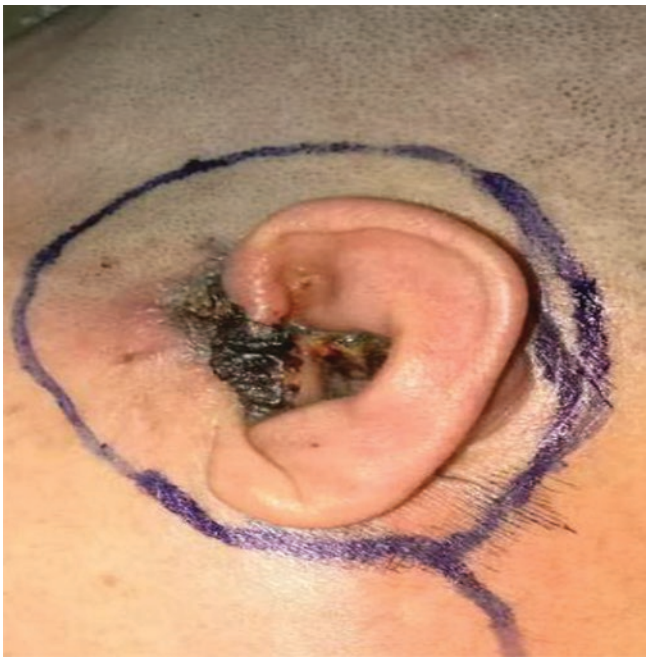


Fig. 8.26 Basal cell carcinoma of external ear

Clinical Manifestations

The first symptoms are mostly otorrhea, and intraauricular hemorrhage or hemorrhagic secretions are more common. Hearing loss is conductive in the early stage. Tumors can invade along bone walls or existing vascular and nerve pathways, and cochlear damage can lead to sensorineural deafness. With the growth of tumors, there are earache, dizziness and facial paralysis. Diffusion from the external auditory canal to the temporomandibular joint, parotid gland or directly through the weak external auditory canal bone wall, petrous scales bone suture or cartilage notch of the external auditory canal invaded the infratemporal fossa, causing difficulty in opening the mouth, external auditory canal, preauricular mass, etc. (Fig. 8.27); invasion of the fifth, sixth, fifth, eighth, nineteenth, eighth, eighth, eighth, eighth, fifth, fifth, fifth, fifth and fifth cranial nerves could cause corresponding

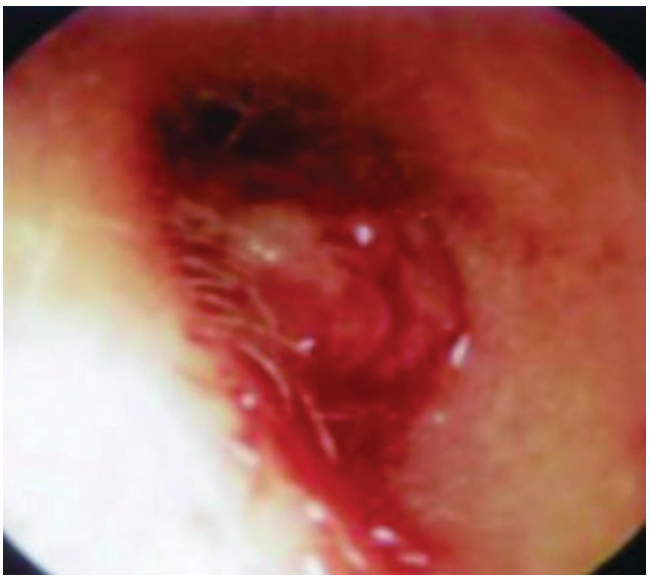


Fig. 8.27 Soft tissue mass with slight enhancement in the left external auditory canal

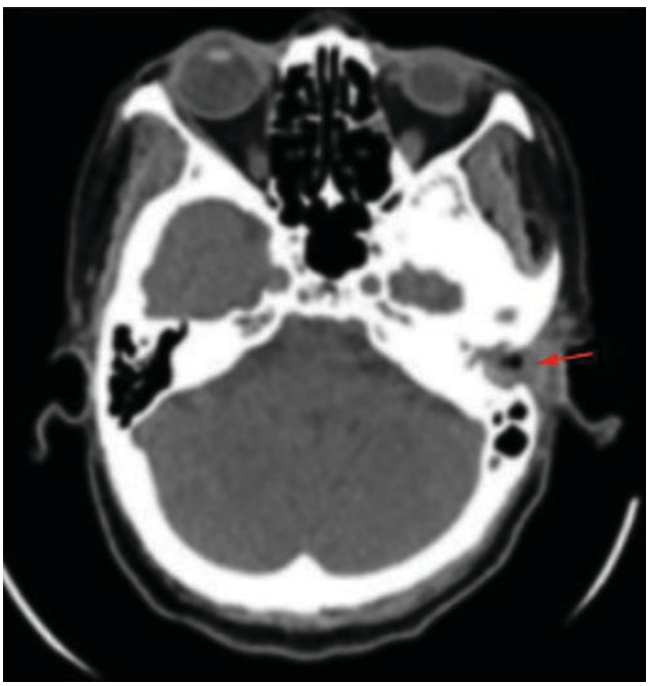


Fig. 8.28 Axis CT of temporal bone demonstrates carcinoma of the left auditory meatus (arrow), with slight bone destruction of the external auditory

symptoms; invasion of the jugular foramen, causing internal Jugular Artery massive hemorrhage of the internal carotid artery; attacks the tympanic cap, dura mater and temporal lobe upwards, causing headache and meningitis. Late stage may have cervical lymph node enlargement and distant metastasis (Fig. 8.28).

Surgery and Reconstruction

1. Surgical methods for temporal bone cancer are limited T1 stage of external auditory canal, especially for the posterior wall of external auditory canal, which can be resected sleeve-like. Previously, the tumors were resected completely together with the skin of external auditory canal, but now they are only between simple tumor resection and extended partial temporal bone resection (LTBR).
   (a) Lesions confined to the external auditory canal: Most skull base surgeons advocate LTBR for patients with stage T1 and T2 who refuse or cannot be enlarged for medical reasons: removal of cartilage and bone of the external auditory canal, tympanic membrane and malleolus, incus, and other adjacent tissues. Surgical procedure: The external auditory canal was closed into a dead space and mastoidectomy was performed to enlarge the facial nerve recess. The upper tympanic cavity was enlarged as far as possible. The tympanic bone was ground down to the tympanic ring. The temporomandibular joint was dissected from the Tragus Cartilage and the anterior external auditory canal bone. The anvil-stapes joint was separated and the whole cylindrical tissue was removed. The operation can enlarge the superficial parotid gland, temporomandibular joint and condyle. LTBR is more conducive to complete resection of tumors than conventional mastoidectomy. Many studies have shown that the survival rate of routine mastoidectomy plus post-operative radiotherapy is similar to that of LTBR (Fig. 8.29).
2. Tumors beyond the external auditory canal: tumors originating from the middle ear and mastoid process, regardless of their size, belong to stage T3 or T4. They should be treated as follows: (1) small patch resection; (2) subtotal temporal bone resection (STBR); (3) total temporal bone resection (TTBR), also known as extended temporal bone resection.

1. *TTBR procedure:*

   These involve a complete removal of external auditory meatus, auricle and surrounding skin. Parotid gland, facial nerve, ascending ramus of mandible, zygomatic arch, temporomandibular joint, pterygoid muscle and foramen ovale were resected to protect trigeminal nerve. The internal carotid artery and the external carotid artery were separated in the upper neck. The internal carotid artery was completely resected with the cerebral nerves (IX, X, XI) and the medial jugular vein. The posterior vertebral artery was separated from the communicating branch of the neck 1 and the deep side of the carotid muscle to remove the stylomastoid foramen and expose the carotid artery into the cranial orifice. The middle cranial

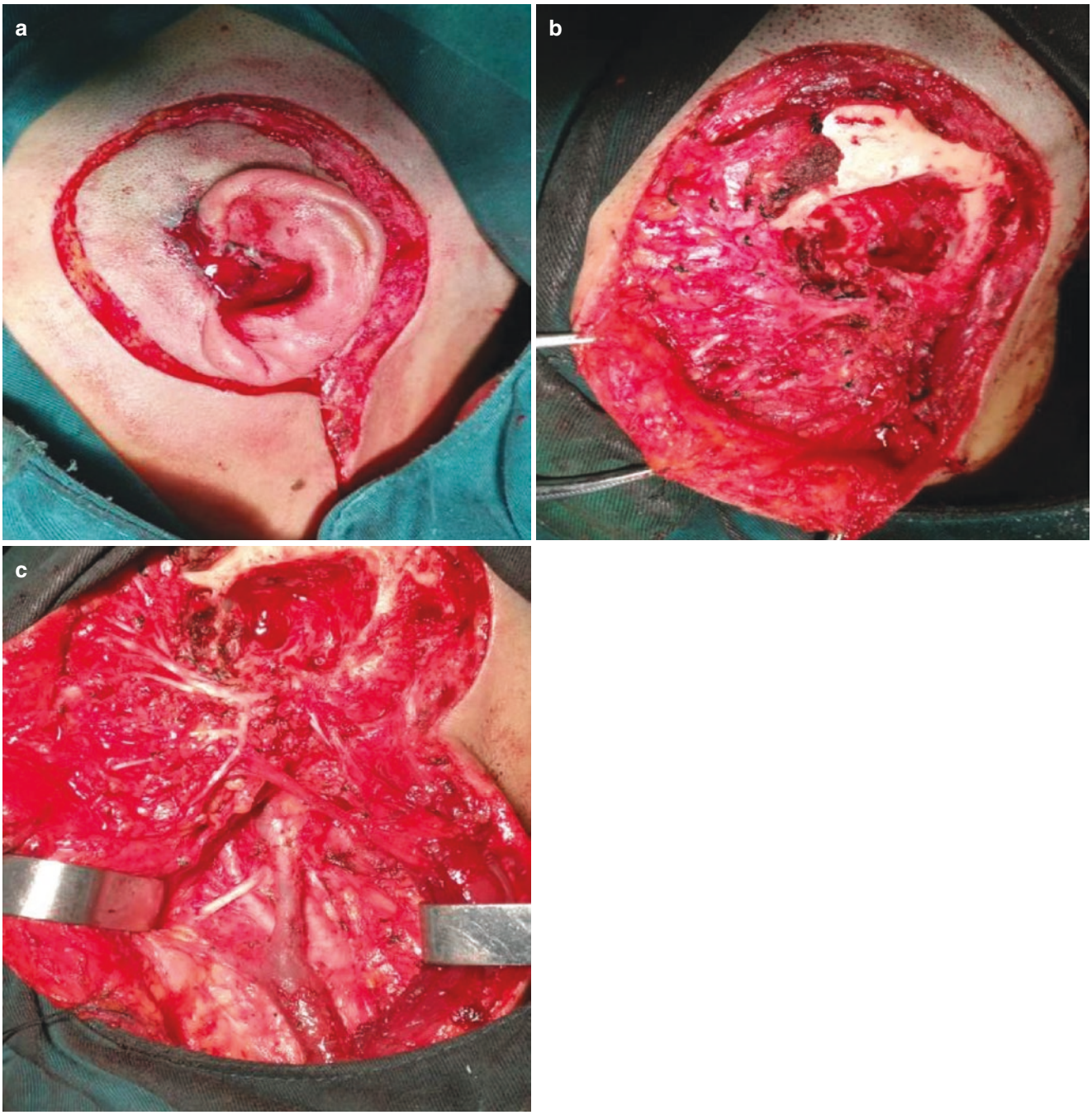


Fig. 8.29 (**a**) Total auriculectomy. (**b**) Superfical parotidectomy and Lateral temporal bone resection. (**c**) Neck dissection

fossa was resected to expose the trigeminal nerve and the carotid artery in the petrous region. The dura mater of the middle cranial fossa and the transverse sinus were ligated behind the sigmoid sinus. The clival bone adjacent to the petrous apex and the carotid artery canal was resected forward. Carotid artery was excised and transplanted. Suboccipital incision was performed to direct the jugular bulb and foramen magnum. The dura mater was resected backward to sigmoid sinus, through communicating sinus to tentorium cerebellum, and forward to superior petrosal sinus. In the sacrificial group, 6 cranial nerves were removed to release the temporal bone. If the extent of resection is large, it must be carefully separated and resected layer by layer, especially in occipital condyle, clivus, carotid artery or cavernous sinus. Note that basal venous plexus hemorrhage is more frequent.

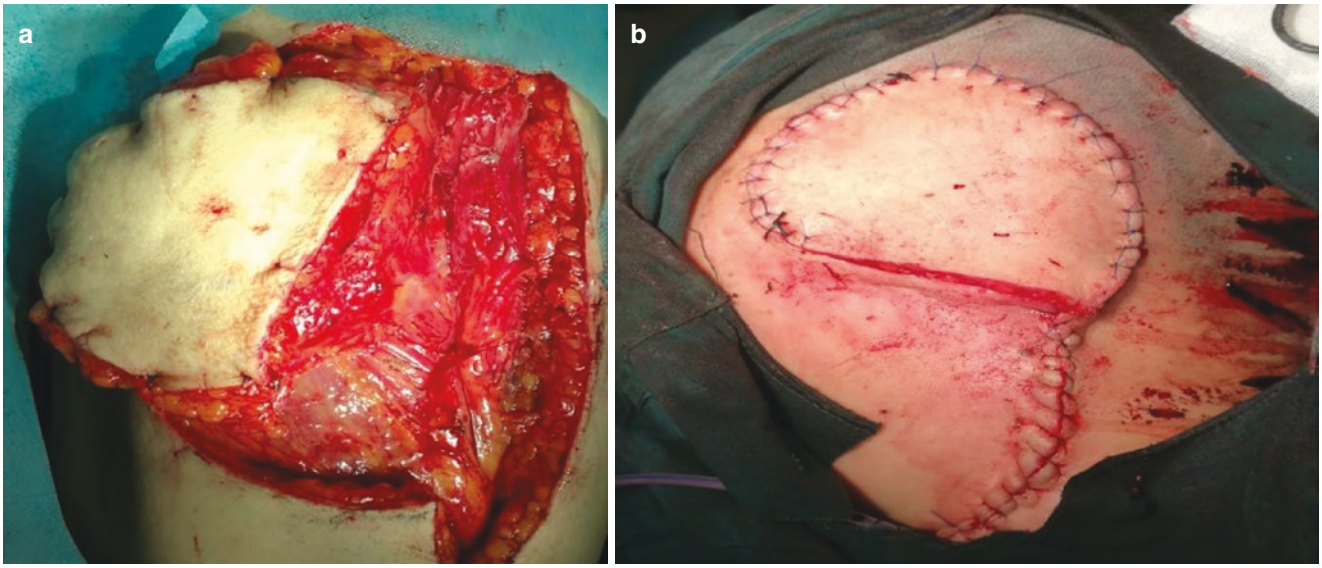


Fig. 8.30 (**a**) Anterolateral thigh (ALT) flap. (**b**) Defect following reconstruction with an anterolateral thigh (ALT) flap

2. *Reconstruction*

   Mostly local musculocutaneous flaps, such as temporalis, trapezius, pectoralis or platysma cervicalis, were mostly used for reconstruction, and the surface was covered with skin or subcutaneous mucosal grafts. Temporal myocutaneous flaps are the most commonly used. It is believed that microvascular flaps can repair large meningeal defects or carotid artery exposure. The dura mater can be repaired with artificial meninges or fascia lata. Nerve transplantation can reconstruct facial nerve or other brain nerves, such as nerve pairs IX and IX. Some scholars believe that hypoglossal nerve should not be used to repair facial nerve, because the loss of hypoglossal nerve function will aggravate the loss of glossopharyngeal nerve and vagus nerve symptoms (Fig. 8.30).

8.8 Implantable Artificial Hearing Device

With the rapid development of a multidisciplinary clinical audiology, ear microsurgery technology, bio electronic technology and biomedical engineering technology, a variety of artificial implantable hearing device constantly available, has been widely used in clinical therapy for hearing loss, different types and different degrees of loss of patients. Implantable artificial hearing device comprises a semi implantable bone anchored hearing aid (BAHA), vibrant implantable Sound bridge (VSB), cochlear implant (CI), bone bridge, auditory brainstem implant (ABI) and auditory midbrain implant system implant (AMI), etc.

8.8.1 Bone Anchored Hearing Aids

Bone anchored hearing aid (BAHA) is a semi-implantable hearing device through bone conduction mode, which is constituted with the fixture, the titanium abutment and sound processor, among which the sound processor receives and amplifies the sound and converted to sound vibration the base and the titanium implant, and the bone conduction sound vibration directly to the cochlea, the inner ear and the auditory nerve auditory stimuli produced (Figs. 8.31 and 8.32). For patients with unilateral sensorineural hearing loss, BAHA is used to transfer the sound from the affected side to the contralateral cochlea, which can reduce the negative effect of the head shadow effect. It can expand the sound field and improve the speech recognition and comprehension ability of the patients in the noisy environment.

Indications

Patients with unilateral or bilateral conductive or mixed hearing loss due to various causes have partial hearing loss, expect to improve language communication ability, unable or unwilling to choose hearing reconstruction surgery, and unwilling or unable to wear ear canal hearing aids. Patients with unilateral sensorineural hearing loss due to various reasons are unwilling or unable to wear ear canal hearing aids. Children should be about 3 years old and have sufficient thickness of the skull to implant titanium screws, young people can wear BAHA soft band (BAHA soft land) first.

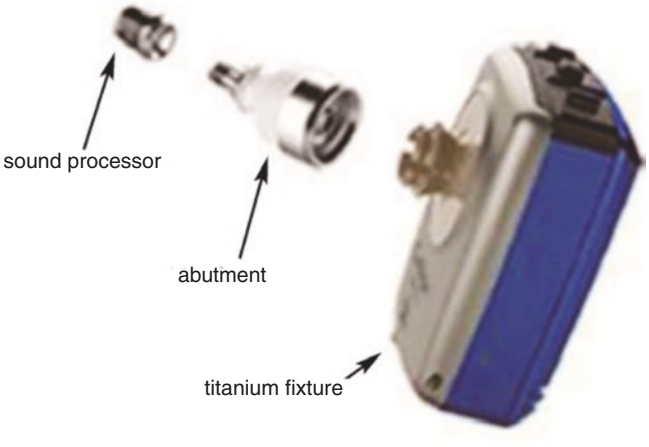


Fig. 8.31 BAHA structural diagram

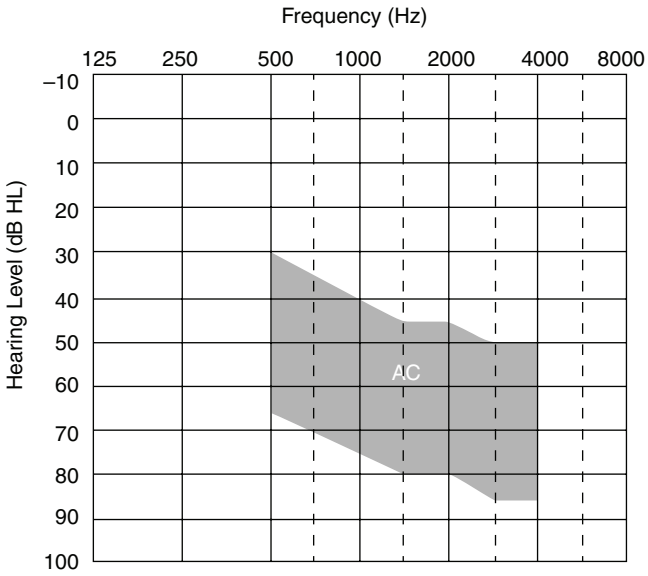


Fig. 8.32 VSB implant sensorineural deafness gas-conductive threshold range

Contraindication

1. Skull thickness is not enough or the bone soft, prone to implant instability.
2. Patients with posterior cochlear lesions.
3. Persons with severe mental retardation.
4. Patients with severe mental illness.
5. Poor overall condition, cannot tolerate surgery.

The relative contraindications of BAHA implantation include osteogenesis imperfecta, Paget's disease, severe osteoporosis, etc.

Operation Methods

BAHA implantation can be performed under local anesthesia or general anesthesia, with simple operation and high safety. It can be completed in two phases, and can be completed in one phase.

1. One-stage operation
   (a) the BAHA model is use to locate the insertion point behind the ear, from the mastoid bone region at the midpoint of the external ear canal 5.0~5.5 cm. A U shaped flap with a diameter of about 2.5 cm is placed in front of the pedicle to expose the mastoid periosteum.
   (b) drill a deep 3~4 mm round hole and screw in the titanium screw.
   (c) reposition the flap and suture the incision.
   (d) after 3~4 months (about 6 months), titanium screws and bone tissue achieved good osseointegration and undergo the next stage operation.
2. Two-stage operation
   (a) Place the flap along the original incision and remove the subcutaneous tissue around the screw
   (b) drill a round hole on the top of the screw with a disposable drill.
   (c) the base is fixed to the screw by this hole.
   (d) reduction flap.
   (e) The patients were followed up for 6 weeks. After the local healing, the sound processor was installed on the base.

Complication

The most common complications are skin and soft tissue infections. Patients with diabetes, immune suppression or long-term immunosuppressive drugs often require a longer bone fusion time, and the incidence of wound infection is relatively high.

8.8.2 Vibriant Acoustic Bridge

The Vibrant Sound Bridge (VSB) is a kind of artificial semi implantable middle ear hearing device developed according to the principle of electromagnetic induction. It was manufactured by MED-EL company in Austria. It won the European CE, American FDA and China food and Drug Administration certification respectively. At present used in many hospitals to carry out surgical implantation of VSB. Its biggest advantage that it does not destroy the normal anatomic structure of the middle ear, and retain the integrity of ossicular chain. VSB consists of two parts: audio processor (AP) and vibrant ossicular reconstructive

prosthesis (VORP). Floating MS transducer (FMT) can be surgically fixed to the ossicular chain, cochlear window or vestibular window.

Indication

1. In patients with moderate or severe sensorineural hearing loss, the threshold of air conduction is within the shadow range at Fig. 8.32. The effect of traditional hearing aid is not satisfied or unwilling to wear hearing aids, and the whole frequency hearing loss is higher than that of low frequency.
2. Mild, moderate or severe conductive deafness or mixed hearing loss in adults, including the etiology of otosclerosis, chronic suppurative otitis media (including middle ear cholesteatoma), congenital external ear atresia, congenital middle ear deformity and other bone conduction threshold in the shadow range at Fig. 8.33.
3. Air bone gap less than 10 dB, some scholars' think that can be less than 15 dB.
4. Speech recognition rate is above 50%.
5. The past 2 years the hearing fluctuation is less than or equal to 15 dB.
6. There is no abnormality in the skin of the implant site.
7. Normal developments, normal brain function, the correct expectations.

Contraindication

1. Posterior cochlear deafness or central hearing loss.
2. Active period of middle ear infection.
3. Chronic effusion of the middle ear.
4. Perforation of tympanic membrane with repeated infection of middle ear and high expectation value.

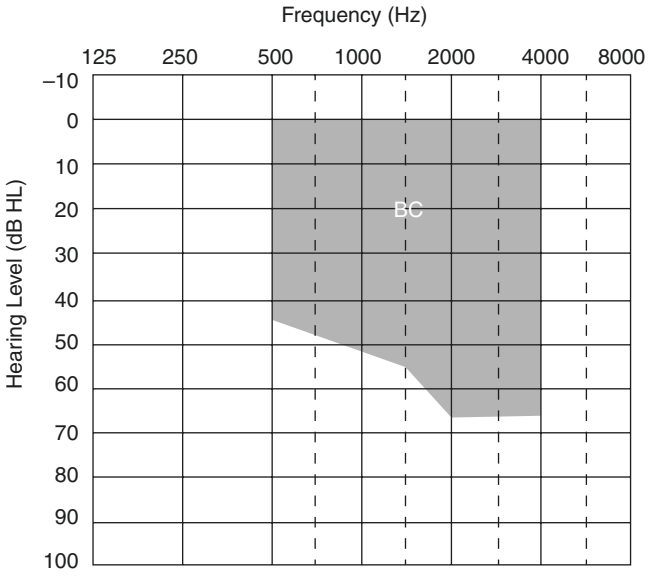


Fig. 8.33 VSB implant conductive or mixed deafness bone-conductive threshold range

Surgical Method

The surgical approach using postauricular incision, can use S or C shaped incision for mastoidectomy, the facial recess opened in the ear tympanic, skull surface implant bed receiving coil and the modem, and then by the facial recess into the wire and MT in the middle ear cavity through small titanium clips on FMT FMT will be fixed in the long process of incus, or directly to the package of FMT fascia placed vertically in the round window membrane in vitro auditory processor in the ear and the corresponding position of the receiving coil. The posterior ear incision can also be applied for VSB implantation through the external ear canal approach.

Complication

VSB implantation may occur under the scalp hematoma, wound healing, facial nerve, chorda tympani nerve, ossicular chain (long limb of incus necrosis) or inner ear damage, cause taste changes, facial paralysis, hearing loss was not significantly improved or aggravated, tinnitus, vertigo, labyrinthitis, very small due to device failure again operation. SB implantation is not recommended for MRI examination at present.

8.8.3 Cochlear Implant

Cochlear implant (CI) is a biomedical device that helps hearing impaired people engineering their hearing and speech communication skills. This Engineering device has been used in clinic since 1970, and has a history of more than 40 years. With the development of related science and technology, products are constantly updated and replaced. The number of electrodes ranges from single-conductor to 24-conductor. The types of electrodes are straight, curved, soft and ultra-soft. Speech processing technology is constantly upgrading. At present, hundreds of thousands of people around the world have received CI implantation and benefited from it. In 2003, China first formulated the Guidelines for Cochlear Implantation. In 2006 and 2013, the Guidelines were revised comprehensively to provide guidance for clinicians, hearing and speech rehabilitation workers, and to further standardize cochlear implantation in China and improve overall treatment.

Indication

Patient selection criteria: cochlear implants are mainly used for the therapy of severe or profound sensorineural hearing loss.

1. Selection criteria for patients with pre-lingual deafness
   (a) implantation age is usually 12 months ~6 years old. The younger the implantation age, the better the effect.

(b) If 3-6 months wearing of hearing aids does not effect, Children with severe or profound hearingloss at both ears should be considered to take direct cochlear implantation.
(c) No operative contraindication.
(d) The guardians and (or) implanted patients have the correct understanding and proper expectation of cochlear implantation.
(e) they have the condition of hearing and speech rehabilitation education.

2. Selection criteria for patients with postlingual deafness
(a) Postlingual deafness patients of all ages.
(b) Biaural severe or profound sensorineural hearing loss, relying on hearing aids can not carry out normal auditory and speech communication.
(c) No operative contraindication.
(d) The implanter himself and/or his guardian have a correct understanding of cochlear implantation and appropriate expectations.

[Contraindication]

- Absolute contraindication

  Severe deformities of the inner ear, such as Michel deformity cochlear deformity, absence or interruption of the auditory nerve, acute suppurative inflammation of the middle ear and mastoid.
- Relative contraindication

  Frequent seizures can not be controlled; severe mental, intellectual, behavioral and psychological disorders, can not match the hearing and speech training.

Guidelines for the clinical practice of cochlear implantation in special conditions.

1. White matter lesions

   If white matter lesions is shown on MRI, intelligence, neurological signs and MRI should be reviewed. If there is no setback of mental and motor development, no deformity of outer hearing and speech system, no positive pyramidal signs or signs of change at neurological examination, but only high signal lesion at cerebral white matter on MRI (like DWI), observe inspections above with the interval of 6 months. If the high signal lesion does not expand on MRI, cochlear implantation could be considered.
2. Auditory neuropathy (auditory neuropathy spectrum disorder)

   It is a kind of special sensorineural hearing loss, which is caused by the dysfunction of inner hair cells, auditory nerve synapses and (or) auditory nerve itself. Audiology has its typical characteristics, manifestations of otoacoustic emission (OAE) and (or) cochlear micro phonics (CM) and normal auditory brain response (ABR) thousands of missing or severely abnormal. At present, cochlear implant is effective for most patients with auditory neuropathy, but it may be ineffective or ineffective for some patients, and the patient and/or guardian risk must be informed before the operation.
3. Bilateral cochlear implantation

   Bilateral implantation can improve the acoustic source localization, speech understanding under quiet and background noise, and help to obtain more natural sound perception, and promote the development of hearing, speech and music appreciation ability. Bilateral simultaneous implantation or sequential implantation can be selected, and two times of sequential implantation, the shorter the operation interval, and the more conducive to postoperative speech rehabilitation.
4. Cochlear implantation for residual hearing

   Patients with residual hearing loss, especially those with high frequency hearing loss, are suitable for electrode implantation with residual hearing preservation. Postoperative acoustic and electrical stimulation mode can be selected, but the risk of loss or loss of residual hearing in patients and/or guardians should be told before the operation.
5. Artificial cochlea implantation in the abnormal-inner-ear-structure

   Abnormal inner ear structure related to artificial cochlea implanting includes common cavity deformity, undeveloped cochlea, ossified cochlea, cochlea stenosis, etc. The artificial cochlea could be applied to most of the patients with any symptoms above. Organize the preoperative case discussion, handle cautiously during surgery and it is suggested to monitor facial nerve. There will be difference at personal effect after surgery.
6. Artificial cochlea implantation in the chronic-otitis-media accompanying with tympanic membrane tresses.

   If the inflammation is under control, the patients should be offered one-stage or phased operation. One-stage operation is radically treating the lesion (or mastoid cavity autologous tissue padding or the ankylotia), repairing the tympanum membrane and implanting the artificial cochlea simultaneously; phased operation is clearing the lesion, repairing the cavum tympanic membrane tresses or blocking the external ear canal firstly and implanting the artificial cochlea about 3–6 months after the former surgery.

Pre-operation Evaluation

1. Medical history taking

   Knowing the etiologic agents by asking the medical history. The point of ear medical history should be on the etiology and process of hearing loss, which could be concluded from the history of the audition, titinus and dizziness, the ototoxicity drug exposure, noise exposure,

systemic acute and chronic inflammation, past otology medicine, family hearing loss, wearing hearing aid, development factors (systemic or local developmental deformity, intelligence development, etc) and other pathogenesis (epilepsy, mental condition, etc.) and to the hearing-loss infants, there should also be the history of the maternal fetation, the parturition, the infant growth, and speech growth. Besides, to all the patients, the doctors should ask about the speech-language ability (lamprophonia, understanding ability, presentation ability, etc) and desire to improve communicating.

2. Systemic or ear examinations

   It includes systemic examination, the examination of the auricular, the external ear canal, the cavum tympanumic membrane, etc.

3. Audition and vestibular function examination

   (a) exam items

   - Pure tone audiometry

     It includes the threshold value of the air and bone conduction. The infants under 6 years old could be applied the behavioral audiometry, including observing, visual reinforcement audiometry and play audiometry.
   - Acoustic immittance

     It includes the cavum tympanum imaging and the stapes muscle reflex.
   - Auditory evoked potential:

     It includes the ABR, 40 Hz, audition event related potential or auditory steady-state response (ASSR) and cochlear micro phonics exam.
   - oto acoustic emission (OAE)

     Do oto acoustic emission or temporary evoked oto acoustic emission (TEOAE) to the deformity.
   - Speech audiometry

     It can be divided into speech recognition rate test and speech recognition threshold test. Choose the accurate speech test material, open and (or) closed type (Appendix A).
   - Aided effect evaluation

     Aided audition threshold test and (or) speech recognition test after optimization match.
   - Auricular function exam (patients with the dizziness history and able to cooperate)
   - Bone promontory stimulation test (if necessary)

   (b) audition inclusion criteria

   - The pre-lingually deafened

     It's needed to do both the subjective and objective comprehensive audition evaluation.
     - objective auditory assess: click ABR reaction threshold >90 dB nHL, 40 Hz audition event-related potential reaction threshold (under 1000 Hz) > 100 dB HL. Audition steady-state response threshold (not less than 2000 Hz frequency) > 90 dB nHL; f ailed at oto acoustic emission of both ears (except for the neurotic)
     - subjective auditory assess: average threshold of single ear's behavioral audiometry>80 dB HL, aided audition threshold (over 2000 Hz frequency) > 50 dB HL; aided speech recognition rate (closed type disyllable) ≤70%, conclude that the patients can't cooperate can't get benefit from the hearing aid.
   - the post-lingually deafened

     Patients with average auditory threshold of ears' pure tone air conduction over 80 dB HL has profound hearing loss; patients have severe hearing loss if the open-phrase recognition rate of its good-hearing ear is less than 70%.

     Remnant audition: patients with good audition to the low frequency, whose audition threshold over 2000 Hz >80 dB HL and communication hearing-aid cannot help, could be implanted the artificial cochlea; To the patients whose remnant audition cannot be detected, it should be informed of to the patients themselves or their guardians that the bad hearing restoration risk after surgery.

4. Imaging evaluation

   As regular, CT scan the thin layer of the temporal, do the inner ear MRI and the skull MRI, 3-dimensionally reconstruct cochlea if necessary.

Operation-Related Request

1. Request to the physicians

   The physicians should have abundant middle-ear-mastoid microsurgery experience, been professionally and systemically trained at artificial cochlea operation and done 20 artificial cochlea implantation cases personally under guidance of experienced physicians at least.

2. Request to the operation room and basic equipment

   The room should be in good asepsis condition and contain the operating microscope, otology drill and relevant device.

3. Pre operative preparation

   Preoperative conversation should be done by the operation doctor and the audiologist, letting the patients or their guardians know about all the risk and complications possibly occur during the surgery, know the benefit and risk the patients could get from implanting the artificial cochlea and sign their name on the informed consent (Appendix B).

4. Operation procedure and methods

   As regular, operate through the mastoid crypt entrance access cochlea fenestra or cochlea fenestra entrance access at post auricle incision. The detail should be according to the relevant request of all types of the artificial cochlea implanting device.

5. Interoperation monitoring

   Do the electrode immittance test and electricity-evoked nerve reaction test due to the artificial cochlea device to know the integrity of electrode and auditory nerve reaction to electric stimulation.
6. Disposal after operation

   Take imaging (skull X-ray imaging) after operation to eliminate the electrode's location and the rest exams are is like other common oto operation.
7. Surgery complications

   Common complications include the cavum tympanic membrane tresses, the external ear canal injury, abnormal smelling sense, dizziness, titinus, facial muscle tics and pain, infection, scalp hematoma, cerebrospinal fluid leakage, facial nerve paralysis, meningitis, intracranial hematoma, shifting and exposure of the implantation service, electrode prolapsed, split or necrotic flap, etc. The complications should be dealt due to the relevant conditions.
8. Switching-on and debugging

   Turn the device on about 1–4 weeks after surgery usually, debug 1–2 times during the following 1-month and arrange the time due to the patients' condition. Lengthen the debugging interval properly after the audition stabilizes and finally debug the device once a year. Switching-on and debugging methods should be done according to the technical requirements. If the contralateral ear benefits from the hearing aid, it is advised to fit hearing aids.

Audition Speech Restoration After Implanting

The patients who implant the artificial cochlea have to be trained by scientific audition speech restoration training to promote their development of the diagnosis, expression and speech application ability. The guardians or patriarchs of infant patients implanted the artificial cochlea should master vital audio speech restoration information and skills under professional guidance at qualified rehabilitation institution, practice actively, try to be supporters, guides and accompanists of all the hearing-impairment children restoration education process to maximum the restoration effect. Adult with artificial cochlea implant should accept the guidelines of the audition adaption training and speech recognition training due to suggestions of doctors.

Restoration evaluation at the artificial cochlear implant includes the assessment of the implantation sound field, speech audition ability and language ability. The 'flying love, a Chinese audiometry software and the International simplest Chinese speech test set for adults with cochlear implantation developed by Xin Xi's group (from China), containing the most latently developed Chinese AzBio sentences, Chinese CNC mono-syllabary and Chinese BKB-SIN test, reflect the mandarin recognition assessment system for adult cochlear implant initially develops, which is enough to filtrate cases before operation and finish post-operation long-term restoration assessment. To this infant cochlear implant whose speech language ability can't finish the audition, language and speech test above, investigators should interview the teachers of patriarchs close to the infants to finish the evaluation paper. Suggested papers are meaningful auditory integration scale (MAIS), infant-toddler MAIS (IE-MAIS), patriarchs/parents evaluate ability of communication hearing (PEACH), teacher EACH (TEACH), meaningful using speech scale (MUSS) and Mandarin context development of infants (MCDI). To the long-term efficacy of big samples, investigators should evaluate the audition sense with result of the categories of audition performance (CAP) and speech expression ability with result of speech intelligible response (SIR). To evaluate the life quality of pre- and post-implanting artificial cochlea, Nijmegen cochlear implant questionnaire (NCIQ) is suggested.

8.8.4 Bone Bridge

Bone Bridge is the latest active cross-flap bone conductive audition implant device applied in clinic, including an implant and a head-mounted audition processor. The implant is made with receiving coil, a modem and a bone conductive floating mass transducer (Fig. 8.34). The implant has MRI compatibility and is able to do the 1.5 T MRI. Compared to the BAHA, there is no implant exposed to obviously decrease the wound infection incidence and no need to do follow-up skin care, what makes it a trend for the bone bridge to replace the BAHA. Bone bridge has been applied in clinic over 5 years, but it just breaks the ice in China.

Clinical Indications

It includes the conductive, the mixed and the unilateral sensory deafness of adults and infants over 5 years old caused by multiple reasons. It also contains the ankylotia, ineffective tymossiculoplasty (weak bone conduction of gas >30 dB HL), otosclerosis, tympanosclerosis, abrupt deafness and acoustic neuroma.

Operation Method

According to the preoperative CT scan result and position of patients, arranging proper bone bed and insert the implant after placing the implant behind the mastoid cavity or sigmoid sinus, fix the implant with two bone cortical screw (should we replace a professional word of 'bone cortical') and place the receiving coil on the skull. Switch on and debug when the operation area heals and the flap detumescence.

Complications

The bone cortical screw loosening occurs only in clinic aboard. There are few reports because at the beginning stage the bone bridges are used in China.

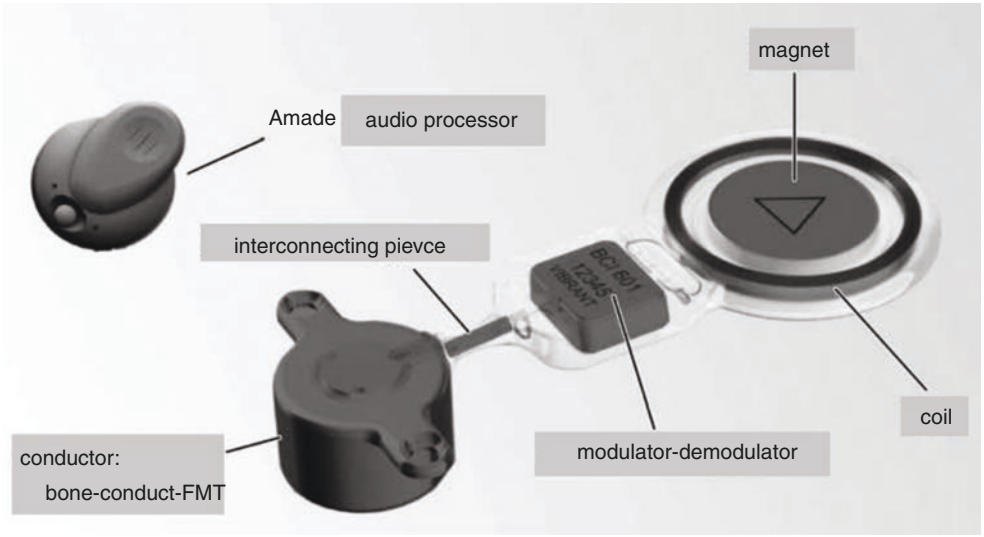


Fig. 8.34 Contents of bone bridge

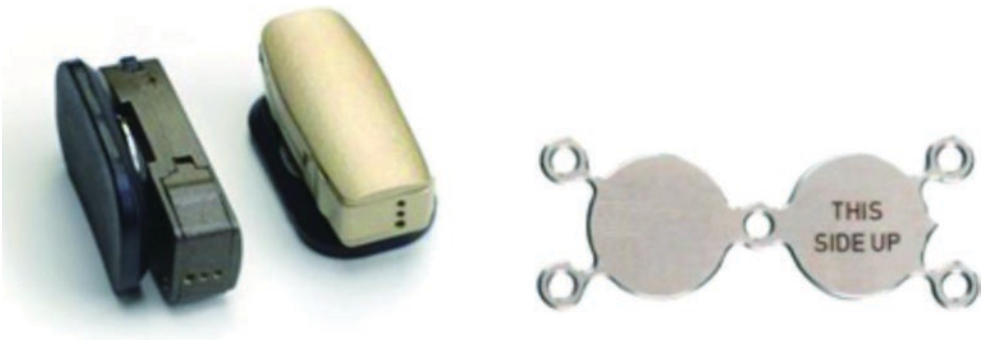


Fig. 8.35 Sound processing and implant of implant bone conductive hearing-aid system

8.8.5 Implant Bone Conductive Hearing-Aid System

The latest China-made bone conductive hearing-aid system consists the sound processing and the implant (Fig. 8.35). The implant is packed with titanium enclosure and is the thinnest (only 2.6 mm) and smallest in size compared to other same-type productions. The implant is fixed with 5 titanium screws on subcutaneous mastoid, absorbing magnetically with the sound processing outside the skin to achieve transmitting the sound. The production has advantages like easy to operate, causing little injury, without exposure in vitro and others. Difference between topic system and Bone Bridge is that the energy converter in topic system is in the sound processing in vitro.

Clinic Indications

1. Conductive or mixed hearing loss but still can benefit from the amplified sound.

   The pure acoustic bone conductive threshold of the affected ear tested under 0.5 kHz, 1 kHz, 2 kHz and 4 kHz isn't more than 45 dB HL.

2. Bilateral symmetrical conductive or mixed hearing loss.

   Difference between the pure acoustic bone conductive threshold of both ears under 0.5 kHz, 1 kHz, 2 kHz and 4 kHz isn't more than 10 dB HL or average threshold at every frequency is not more than 15 dB HL.

3. Deep unilateral sensorineural hearing loss and cannot apply the air conductive contralateral routing signals hearing aid. The pure acoustic air conductive threshold of health ear at 0.5 kHz, 1 kHz, 2 kHz and 4 kHz isn't more than 20 dB HL.

8.8.6 Auditory Brainstem Implant

Auditory brainstem implant (BMI) has similar form as the artificial cochlea. The electrode array is inserted into crypt outwards the fourth encephalcoele by surgery and straightly stimulate the auditory neurons in the brainstem cochlear nuclear complex cross the cochlea and auditory nerve to produce audition. There are already over 1000 patients implanted. ABI is mainly produced by the Australian Cochlear Company and Austrian MED-EL Company.

Clinical Indications

NF-2 patients' nerve connection between the bilateral cochlear nuclear and cochlear spiral ganglions interrupts after removing the tumor and the artificial cochlea don't effect.

Patients can't get benefit from the artificial cochlea because of the cochlea or acoustic nerve injury, loss, maldevelopment, dysneuria, ossified cochlea, severe deformity and other symptoms, caused by varieties of etiologies like trauma, congenital deformity, auditory, neuropathy, meningitis, otosclerosis.

No age limit. Both the adults and the infants could be in. There is already an infant patient who has implanted the BMI.

Operation Methods

ABI implanting could be done by the translabyrinthine approach, retro-sigmiod approach or retro-sigmoid-inner ear canal approach and other operation methods. The translabyrinth approach is the most common one. An ideal method should entirely remove the tumor and also completely expose and accurately locate the implanting field to avoid from damage to the important blood vessels, nerve and functional zone.

Complications

Severe complications include death, cerebellar injury, persistent facial paralysis, meningitis, cranial nerve injury, cranial hydrops, pseudo-meningocele etc. Less severe complications contain the cerebrospinal fluid leak, temporary cranial hydrops, traumatic hematoma, slight infection, disequilibrium, perilous-implant infection, flap infections, temporary facial paralysis, temporary phonation or swollen difficulties, headache, flat problems, and non auditory response.

Post Implanting Debugging and Effect

The ABI implant switches on and debugs mostly six weeks after operation. First switching-on should be done under electrocardiogram monitoring to prevent the vital signs' change evoked by stimulating brainstem formation. Do regular postoperative debugging to fit personal requirements of patients. Debugging includes ensuring the threshold, the maximum comfort threshold, non auditory sensation response intensity of each electrode, tone-sensation grading evaluation, ordering the electrode pairs due to the tone, programing ABI language processor and eliminating speech manifestations.

All in all general effect of ABI is worse than the artificial cochlea according to data present.

8.8.7 Auditory Midbrain Implant

Auditory midbrain implant (AMI) has similar formation as the ABI. Place the electrode into the inferior colliculus or its central core to stimulate to produce the audition. There aren't many applied cases, so we need do further relevant research. Austrian MED-EL Company and Australian Cochlear Company produce the implant.

8.9 Sudden Deafness

Dekleyn firstly reports it at 1944. Most of the scholars relate its pathogenesis to the blood circulation disorder, the virus infection, the labyrinth fenestrated membrane rupture, the trauma, the poisoning, etc. Those whose etiology isn't explicit could be called the idiopathic abrupt deafness. It is defined by the Chinese Medical Association Otorhinolaryngology Head and Neck Surgery branch (2015) as the sensorineural hearing loss abruptly occurs inward 72 h with unclear pathogenesis, during which audition loses not less than 20 dB HL between two adjacent frequencies. Definition from the U.S. guideline in 2012 is the sensorineural deafness rapidly develops without clear pathogenesis inward 72 h, whose hearing loss at three successive frequencies isn't less than 30 dB. As research result from the Chinese Abrupt Deafness Research Center shows that the median onset age is 41-year-old, there is no obvious imbalance gender ratio, the left deafness onsets more than the right. The bilateral abrupt deafness incidence is low, about 1.7–4.9% of all cases, which is only 2.3% at researches of Chinese Research Center.

Clinical Manifestation

1. Suddenly happened hearing loss
2. Tinnitus (about 90%)
3. Ear stuffy-fullness feeling (about 50%)
4. Dizziness or giddiness (about 30%)
5. Paracusia acris or weak hearing
6. Periodic paraesthesia (common in anakusis)
7. Some patients develop mental disorders like anxiety, somnipathy and influence the living quality.

Diagnosis

Diagnosis gists from Chinese Medical Association Otorhinolaryngology Head and Neck Surgery branch at 2015:

1. The sensorineural hearing loss abruptly occurs inward 72 h with, during whom audition loses not less than 20 dB HL between two adjacent frequency, most of whom is unilateral and minority of whom is bilateral occurred or occurs at both ears by step.
2. Unclear pathogenesis (including the systemic or local factors)
3. May be accompanying with the tinnitus, the ear stuffy-fullness sensation, and the periodic paraesthesia etc.

4. May be accompanying with the dizziness, vomiting and nausea.

In 2012, the U.S. Guideline recommended diagnosing despite other neurologic diseases, examine the auditory, despite the retro-cochlear lesions and intensely deprecate the CT scan and other Regular lab examinations.

Treatment

Because the pathogenesis is unknown, there is no unified pattern to treat. Most of the therapy methods are experience-depend. Main measurements include the drug therapy, including the vasodilator dextran-40, calcium antagonist nimodipine, antihistamine methionine betahistine, glucocorticoid (oral or intra-tympanic injection), anticoagulation batroxobin, heparin, etc., and other methods like hyperbaric oxygen (HBO).

- Therapy guidelines In 2015 Chinese Medical Association Otorhinolaryngology Head and Neck Surgery branch point out that typing the abrupt deafness according to the auditory curve could guide the therapy and prognosis, the improving-inner-ear-microcirculation drug and the glucocorticoid effect on all types of abrupt deafness, proper combined drugs are more effective than the single drug, the effect order at all types should be low frequency decline type (best) > flat decline type (better) > high frequency decline type and anacusia type (worst).

1. Basic treatment advice
   (a) Abrupt deafness acute onset stage (inwards 3 weeks):
   Main change is the inner ear vasculopathy. Suggest applying the glucocortocid plus hemarheology therapy (including the hemo-dilution, improving the flow and decreasing the viscosity or fibrinogen, which is done by drugs like the ginkgol biloba extract (GBE), batroxobin, etc.
   (b) Application of glucocorticoid:
   Oral: prednisone 1 mg/kg per day (top 60 mg), taken at a drought in the morning, thrice daily, stay on it for 2 more days if it effects, no need to back-titrate. Withdrawal straightly if no response. The hormone could also be administrated by vein. Reckoning analysis due to the prednisone dosage, it should contain methylprednisolone 40 mg or dexamethasone 10 mg, same courses as oral hormone. The hormone therapy is priorly suggested to systemic injection. The local injection is for the rescue therapy, including the intra-tympanic or retro-auricle injection. The intra-tympanic injection should apply the dexamethasone 5 mg or methylprednisolone 20 mg, quaque omni die (QOD), 4–5 times. Retro-auricle injection should apply the methylprednisolone 20–40 mg or dexamethasone 5–10 mg, qod, 4–5 times. If the patients difficult in subsequent visiting, it could be applied the diprospan 2 mg (1 ml), retro auricle injection once. About these patients have hypertension history, diabetes history and other medical histories, the glucocorticoid or local administration due to the condition under close attention to blood pressure and blood glucose monitoring could be used to them with their permission.
   (c) Abrupt deafness could make secondary nerve injury, which can be cured by taking the nutritive psychotropic drugs (such as the mecobalamine, nerve nutrition factors, etc.) and antioxidants (such as the lipoid acid, the ginkgo biloba extract, etc.) at and after acute staging.
   (d) Same type drugs aren't suggested to be combined utilized.
   (e) It is still debatable about the hyperbaric oxygenation response, so the hyperbaric oxygenation isn't the best therapeutic regime. If the regular treat response isn't good, the rescuing measure should be considered.
   (f) The drug withdrawal could be advanced if audition restoration rates isn't good during the courses and delayed due to the condition. To these patients who don't get good efficacy, the hearing aid, the artificial cochlea or other auditory assistance devices could be applied according to the hearing loss degree.
2. Recommended grading therapy proposal
   The anacusia type, the high frequency decline type and the flat decline type have low curing rate, so the patients with any of them should be active treated as soon as possible.
   (a) Low-frequency decline type
   - It may be caused by the membranous labyrinth hydrops, so there should be salt-restriction. The patients should not be transfused too much transfusion and better no saline solution;
   - patients with average hearing loss <30 dB have high self-curing rate, could orally take drugs, including the glucocorticoid, the methionine betahistine, the improving venous-return drug (such as the aescuven forte) etc. and could also be considered to intra-tympanic or retro-auricular administration of the glucocorticoid (such as the methyprednisolone, the dexamathesone, the diprospan, etc.); patients with average hearing loss ≥30 dB could be venously injected the ginkgol biloba extract (GBE) + the glucocorticoid; minority of patients, getting no efficacy with regimen but exacerbating the ear stuffy-fullness sensation, can be treated with lowering-fibrinogen drug (such as the batroxobin) and other improving-venous-reflux drugs.

(b) High-frequency decline type
- improving microcirculation drugs (such as the ginkgol biloba extract (GBE)) + glucocorticoid;
- the ion channel blockers (such as the lidocaine) have good efficacy to decrease the high-tone tinnitus;
- the nutrition neurotic drugs could be considered (such as the mechobalamine, etc)

(c) All-frequency hearing loss (including the flat decline type and the anacuria type)
- decreasing-fibrinogen drug (such as the betroxobin);
- the glucocorticoid;
- the improving-inner-ear-microcirculation drugs (such as the ginkgol biloba extract (GBE).

The combined medication therapy is suggested.

The U.S. Guideline recommends that the intratympanic injection should be the rescuing therapy when other therapies failed and the initial hormone therapy (including the oral and intratympanic injection) and the hyperbaric therapies could be applied appropriately. No commendation to other drug therapy regimens.

Recommended efficacy grading by the Chinese Medical Association Otolaryngology Head and Neck surgery branch (2015):

1. *Recovery* The damaged frequency audition threshold reinstated to the normal, the health-ear level or the quality premorbid.
2. *High-efficacy* Average audition at the damaged frequency increases over 30 dB.
3. *Efficient* Average audition at the damaged frequency increases 15–30 dB.
4. *Inefficient* Average audition at the damaged frequency increases less than 15 dB [3].

8.10 Benign Paroxysmal Positional Vertigo

The benign paroxysmal positional vertigo (BPPV) is the temporary dizziness concomitant with nystagmus evoked when head moved to a specific location. Because most of the BPPV is self-limited diseases, curing after several days or months, it is 'benign'. It was firstly reported by Barany. BPPV is the most common peripheral vertigo disease, occupying 1/4 of vertigo diseases and whose morbidity is 10.7/100,000–64/100,000.

Etiology

The etiology of the BPPV isn't ensured. It can be idiopathic and also secondary to factors following:

1. When the age-related change or anaplasis occurred red in the labyrinth, the maculae utriculi degenerate, the otolith detaches and drops into the semi buccal tube (often in the posterior, occasionally in the lateral and the anterior).
2. Trauma

   Slight head trauma or head accelerating novation like the whiplash injury. The otolith dropping into the semi buccal tube can also occur after the stapes surgery?
3. Ear disease

   The mid-ear mastoid inflammation like the various labyrinthitis, the chronic suppurative otitis media, the Ménière disease catalase, the perilymphatic fistula, etc.
4. insufficient blood supply to inner ear

   It is caused by the arteriosclerosis and the hypertension, the capsule colloid membrane pinch outs and the otolith drops into the semibuccal tube.

Pathogenesis

There are many theories about the BPPV's pathogenesis, whose trendency is the cupulolithiasis theory and the canalithiasis theory (Fig. 8.36)

1. Cupulolithiasis theory

   It is reported by Schukne in 1969, the basophil granules after the utriculus otolith degeneration drop into the semi buccal tube (mainly the posterior canal) edge crest, what evokes the concentration difference between the endolymph and the edge crest to cause the ratio change and finally changes the ridge crest gravity and the linear acceleration into sensitivity (Fig. 8.36). But theory, if only the semi buccal cal tube keeps vertical to the ground, the gravity-sensitive ridge crest skewing should also be still and cause continuous vertigo and nystagmus. But the dizziness or the nystagmus in this disease could only last temporarily in few seconds, short time, so it's not enough to explain the short-term and the fatiguability of the nystagmus. Motility found that there are only 28% of the ridge crest otolith drops at the posterior canal, 21% at the lateral and 13% at the anterior in the normal ossa temporal which defends the cupulolithiasis theory.
2. Canalithiasis theory

   As research results show, the degenerated otolith crushing isn't attached to the ampullae edge crest but floating in the endolymph of the semi buccal tube. When the head moves to the stimulating area, the tube is vertical to the ground and the otolith moves by the opposite ositsmpullae direction and drops in the lowest sites of the semi buccal tube, what guide the endolymph to move the ampullae edge crest to opposite-utriculus direction, finally stimulate the ampullae edage sensory hair cells and evoke vertigo and nystagmus.

   In fact, a main difference between the cupulolithiasis theory the canalithiasis theory is that the otolith sedimentation attach is attached to the edge crest or floating in the smi buccal tube. If there are plenty of the otolith granules, there could be both simultaneously.

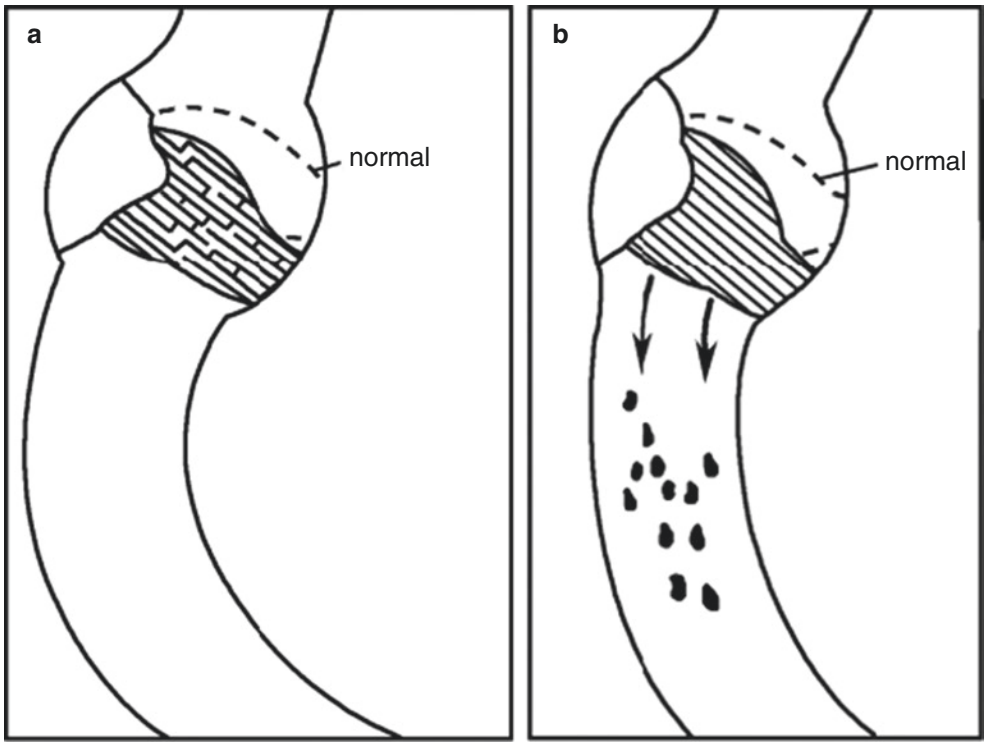


Fig. 8.36 Pathogenesis pattern graph of the cupulolithiasis theory and the canalithiasis theory

Pathology and Pathophysiology

The views differs at that the substance existing on the semi buccal tube and the ridge crest is the otolith or the others. Welling (1997), Pames (1992) and others find that those granules floating in the semi buccal tube is basophilia, considering it as the translocating otolith. Motility (1992) found that there is basophil existing on 22% of the ridge crest in 566-ossae temporal, often being seen at the lateral and anterior sites. He considered it common that sedimentation occurs in the semi buccal after death, and the sedimentation maybe not the otolith, but also can be the cell debris like the macrophage, the WBC and possibly the crushing developed from the labyrinth micro hemorrhage. Besides, because of the trauma, the mid-ear surgery and the inflammation, there could be WBC and endo-membrane debris in the endolymph aggregating in the semi buccal tube to cause same reaction as the otolith translocation and finally evokes the BPPV.

Clinical Manifestation

Typical onset manifestation is that the severe rotational vertigo occurs while heading up or turning over but disappears soon, repeat action could invoke the vertigo again but accompanying with the tinnitus and no hearing loss. According to the locations the change occurs, the manifestations are different.

1. Posterior canal BPPV (PC-BPPV)

   Happen abruptly and often be evoked while the sudden prone, head moves toward one profile or stretching the neck, sudden acceleration or deceleration while in vehicles and crouching 3–6 s (incubation period) after suddenly moving head or stimulating the site, the patients sense the temporary rotating nystagmus and fatiguable. Accompanying with the vertigo, there is the top heavy sensation or floating sense, unsteadily. The period could last for several hours, couple days even over 1 year at the long-term. There is also vomiting and nausea sensation while being attacked, but without any hearing disorders, tinnitus etc. and none central nerve manifestations and signs. Comfortable during the incubation stage.
2. Horizontal canal BPPV (HC-BPPV)

   Vertigo happens in short time too, and often occurs while turning over. While the patients turn over to the affected side, the vertigo or the nystagmus is severe, but when the head moves vertically like raising-head-up or erection after stooping, there isn't any uncomforted. It is a short-term manifestation, lasting several days or about 1 month. Compared to the PCBPPV, the incubation stage is short as 2–3 s but the lasting may be longer. The fatigue feeling may be occurring or may not.
3. Anterior canal BPPV (Slo EC-BPPV)

   With low morbidity and may be ensured due to the vertical substance's direction at rotational vertigo.

Examination

1. Dix-Hallpike positioning nystagmus test

   The most common test method is at the post-canal and the anterior canal. The procedures are ensured by the nystagmus direction: (1) Sitting position, the investigator

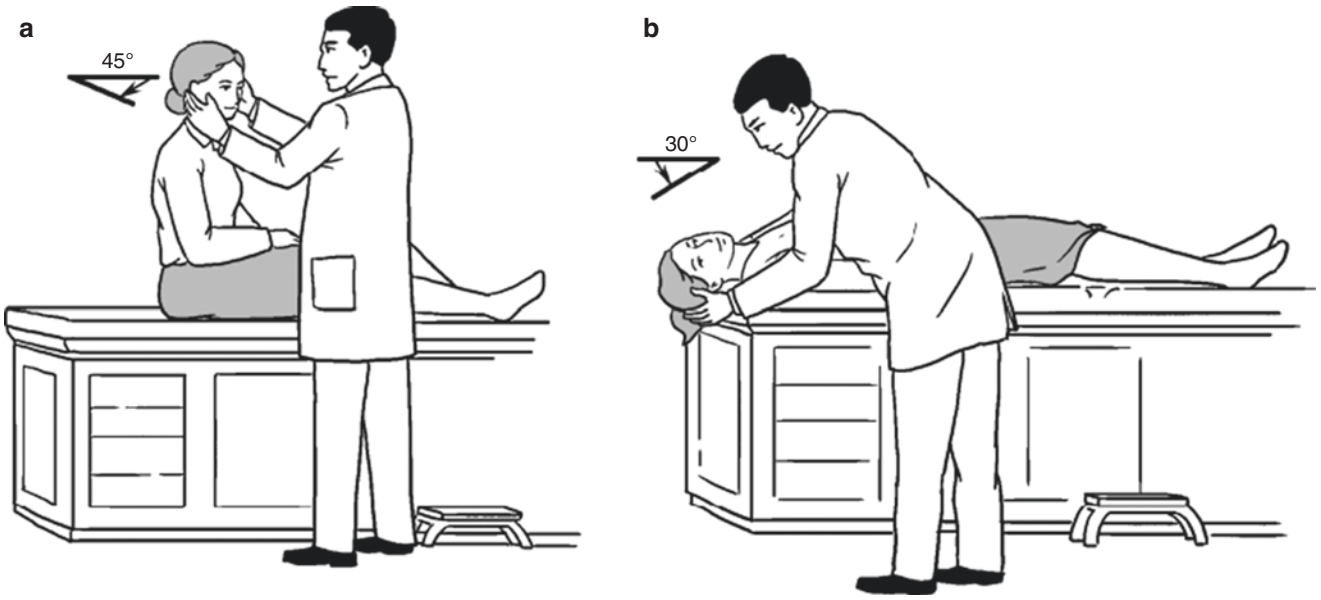


Fig. 8.37 Dix-Hallpike test. (**a**) Sitting positioning detroverse the head by 45°; (**b**) The investigator support and move the head to the supine suspension position, 30° to the horizontal plane

support and dextroverse the patients' heads by 45°with both hands, (2) the investigators support, rapidly move the head to the supine suspension position with both hands, whose angle of the horizontal plane is 30° and 45° to the vertical plane, stay for 30 s and resume into the sitting position after investigation the vertigo and nystagmus conditions (Fig. 8.37). Investigate the other side by the same way. Typical PC-BPPVs always causes temporary vertigo and vertical rotatively nystagmus after 5–15 s' incubation and it lasts for less than 30 s, with fatiguability.

2. Roll test

   Horizontal position. The investigators support and rapidly detroverse the head to left or right by 90° by both hands and investigate the vertigo and nystagmus conditions (Fig. 8.38). The typical HC-BPPV rapidly causes severe rotative vertigo and toward-earth nystagmus after several seconds' incubation and it lasts for over 30 s, without fatiguability.

3. Audiometry

   There is usually no abnormal audiological manifestation, except for that the canalithiasis is secondary to other inner-ear disease.

4. Nystagmus electro-graph examination

   Most of the results are normal. If the BPPV is secondary to the some inner-ear disease, the relative vestibular function could change.

5. Imaging examination

   The neck X-ray imaging or MRI, the temporal CT scan and others could help differentiate diagnosis.

Diagnosis and Differential Diagnosis

1. Diagnosis gist
   (a) The temporary vertigo history when the head moves to a specific position.
   (b) Positioning nystagmus test could appear dame nystagmus characteristics and has incubation stage (<30 s).
   (c) Ascertain the BPPV type due to the history and Examination result (Table 8.1) [4].
2. Differentiate diagnosis

   The BPPV should be differentially diagnosed from the central vertigo, the vestibular neuritis, the Ménière disease, the cerebrovascular diseases and other dazzling diseases (Table 8.2).

Treatment

1. General therapy

   Escape position could evoke vertigo and rest in bed while vertigo occurs, avoiding the head novation, rapid retroversion, etc. Pay attention to the psycho-therapy, eliminating the psycho-burden.

2. Medical therapy
   (a) Drug therapy: choose the antidinic drugs due to the condition to inhibit the vestibular nerve excitement, alleviating the vertigo, controlling the nauseating, vomiting and other autonomic manifestations.
   (b) Positioning therapy: The Brandt-Daroff positioning therapy method. Close the eyes stand and twist toward one side till the occipital touches the Examination table. Keep the position till the vertigo

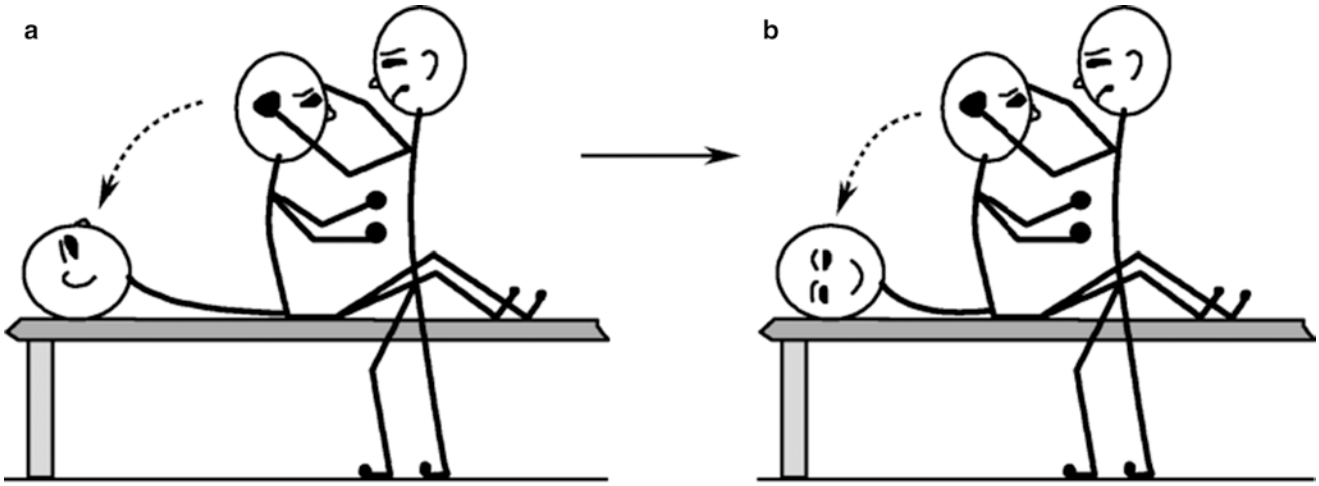


Fig. 8.38 Roll test. (**a**) At the supine positioning and keep the head in middle. (**b**) The head rapidly detroverse to right or left by 90°

Table 8.1 Differential diagnosis of positioning vertigo

	Central vertigo	BPPV	Alcoholic vertigo	Cervical vertigo
Vertigo				
Incubation	No	2–20 s	No	Yes
Lasting time	Continuous	2–40 s	Lasting while staying still	Short-term
Nystagmus				
Direction	Unsteady	Towards inferior ear		Steady
Occurring position	Plenty	One	Plenty	One
Fatigued	×	√	×	√
Property	Vertical or oblique	Rotating or horizontal	Rotation or horizontal	Horizontal

Table 8.2 Differentiate points on BPPV

Differentiate points	PC-BPPV	HC-BPPV	SC-BPPV
Evoking test	Dix-Hallpike test	Roll test	Dix-Hallpike test
Lasting time	<30s	>30s	<30s
Incubation stage	5–15 s	<3 s	5–15 s
Fatiguability	√	×	√

goes and sit up. Twist to another side after 30 s. Repeat as above till the vertigo symptoms disappear.

(c) Otolith repositioning maneuvers: choose different positional maneuvers according to the different BPPV types.

- The PC-BPPV: Epley or Semont repositioning maneuvers (Fig. 8.39)

Surgical therapy: to intractable BPPV, the patients should do the ampullae neurectomy, the vestibular neurotomy, the semi buccal occlusion, etc. if it influences the living quality of patients even after inefficient traditional therapy [5].

Prognosis

BPPV has high self-healing nature. The PC-BPPV can be self-healed after about 39–47 days, and the HC-BPPV's is 16–19 days. Symptoms of 30% of the patients last over 1 year. The recurrence rate after 2 years is about 20%, and 55% after 8 years. The recurrent patients could do the repositioning maneuvers again.

8.11 Ménière Disease

Ménière disease is a kind of inner ear disease with specific endolymphatic hydrops, manifesting as recurrent rotating vertigo, fluctuant sensory hearing loss, tinnitus and (or) aural fullness sensation. Its pathogenic change is endolymphatic hydrops (Fig. 8.40).

Clinical Manifestation

1. Clinic symptoms:

The pathological base of the meniere disease is the hydro labyrinth. Its main clinical manifestations are shown in the following:

(a) Recurrent rotating vertigo, lasting for 20 min even several hours, occurs twice at least;often accompanying with the nausea, the vomiting balance disorder but no consciousness loss; may accompanying with the horizontal or horizontally rotating nystagmus.
(b) At least once sensory neuro deafness at pure tone test.
(c) Indirect or persisting tinnitus, the ear stuffy-fullness sense
(d) Despite other diseases could cause vertigo.

2. Clinic staging

The clinic staging is according to average audition threshold under the pure-tone test investigation and the speech recognition rate (Table 8.3):

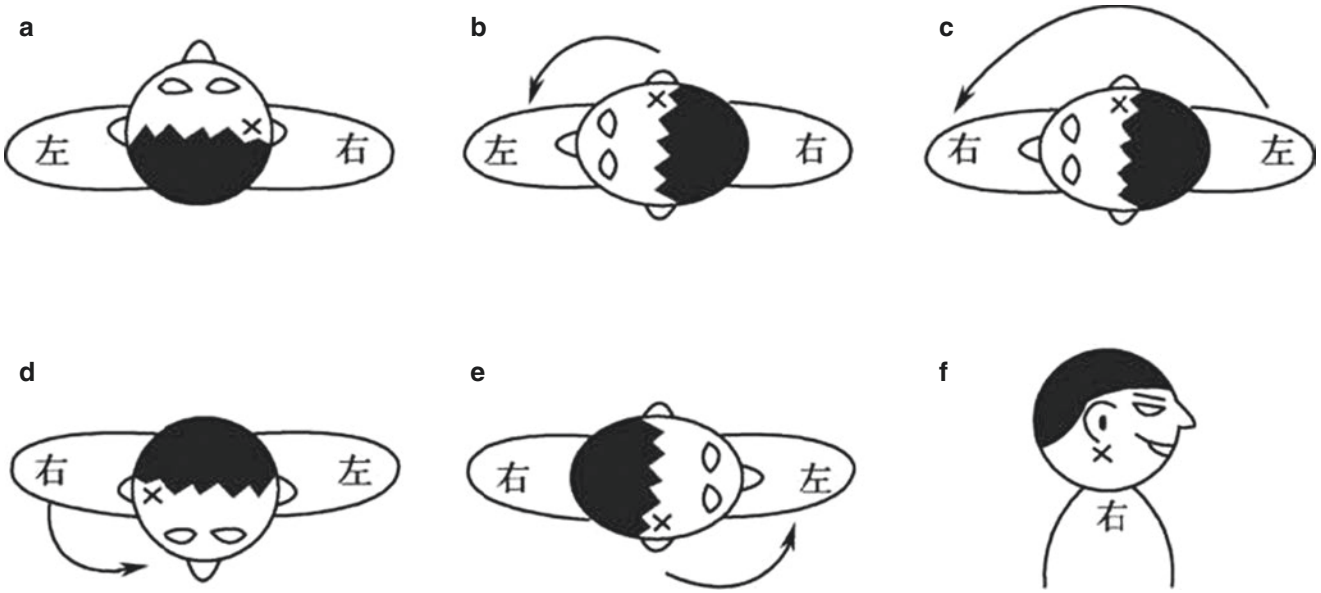


Fig. 8.39 HC-BPPV positioning maneuvers

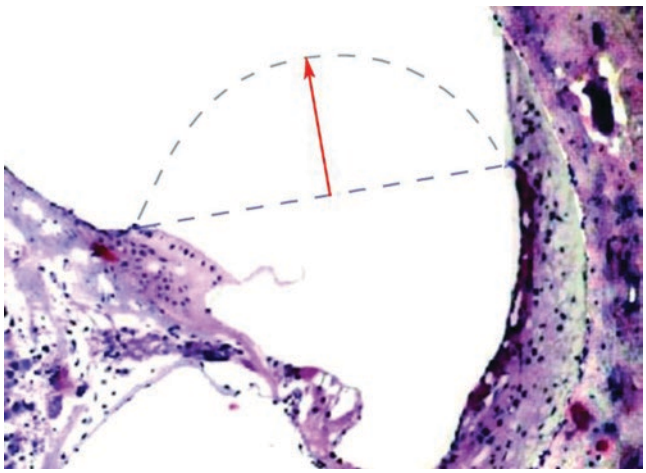


Fig. 8.40 Diagram of endolymphatic hydrops in guinea pig cochlea

Table 8.3 The meniere's clinic staging

Stages	Average threshold of pure-tone (dB)	Speech-recognition rate (%)
I	≤25	≥70
II	26–40	≥50
III	41–70	≥50
IV	>70	<50

The prophase: indirect normal audition or slight low-frequency hearing loss.

The mid-phase: indirect low frequency and high-frequency hearing loss.

The late stage: over moderate-severe all-frequency hearing loss, no audition undulation.

The pure-tone threshold applies the average value of threshold at the 250 Hz, 500 Hz, 1000 Hz and 2000 Hz, and it is the worst auditory range before therapy.

Examination

1. Enquire the medical history and the vertigo recurrence history
2. Examination
   (a) Otoscopy
   Normal tympanic membrane
   (b) Vestibular function Examination
   While the vertigo occurs, the nystagmus and the balance disorder appear. The indirect idiopathic and evoked vestibular function examinations are often normal. The evoked vestibular function examination at the effected side declines or completely disappears.
   (c) Audition
   Sensory deafness. Upward-sloping audition curve at the early stage, flat or decline curve at the later stage. The audition has the undulation and the recruitment phenomenon.
   (d) Glycerinum test
   After taking the 50% glycerinum by 2.4–3 ml/kg, the 250–1000 Hz auditions improved to positive, ≥15 dB.
   (e) Imaging examination
   The inner ear and the cerebellopontine angle CT scan or MRI Examination help differentiate diagnosis the Ménière disease. The temporal CT scan occasionally manifests the badly gasified peripheral aqueduct vestibuli, with short and straight lumen. Some

patients manifest the straightened and slimmed aqueduct vestibuli under membranous labyrinth MRI. The intra tympanic injection of the gutamide dilution through the auripuncture is recently applied to prepare the three dimensional fluid attenuated inversion recovery MRI (3D-FLAIR MRI) 24 h later, which is used to distinguish the intra-, the peri-lymph gap boundary, manifest the hydro labyrinth condition and is possible to offer accurate proof to differentiate diagnose the Ménière disease.

Diagnosis Gist

1. Paroxysmal vertigo occurs not less than 2 times, lasting 20 min to several hours and often accompanies with the automatic nerve system dysfunction and the disequilibrium, without consciousness loss.
2. Undulating hearing loss that the low-frequency audition loses at the early period and gradually the audition loss worsen along with the disease processing. At least do one sensory neurologic hearing loss at pure-tone test. There could be the recruitment phenomenon.
3. often accompanying with the tinnitus and (or) the ear fullness feeling.
4. Automatic nystagmus may happen.
5. Despite the vertigo caused by other diseases, such as the benign paroxysmal positioning vertigo, the labyrinthitis, the vestibular neuritis, the drug toxic vertigo, the paroxysmal deafness, vertebrobasilar artery insufficiency and the intracranial space-occupying lesion.
6. The suspicious diagnosis (unconfirmed the Ménière disease): (1) only for once vertigo occurs, the pure-tone test result is the sensory neurologic hearing loss, often accompanying with the tinnitus and the ear fullness feeling; (2) not for less than twice vertigo occurs, lasting 20 min to several hours. Normal audition, without tinnitus and the ear fullness feeling; (3) undulating low-frequency sensory neuro-hearing loss, accompanying with the recruitment phenomenon, no obvious vertigo occurrence. Diagnosis fit any one of three above is a suspicious diagnosis.

Treatment

1. Drug therapy

   Drugs act at controlling the acute onset treatment and long-term dealing especially the undulating stage therapy of vertigo. The ideal criteria of the drug therapy is to achieve aims following: (1) eliminating the vertigo; (2) build a new sensory equilibrium function with rapid and complete vestibular compensation; (3) alleviating the nausea, vomiting and other symptoms at automatic neurodysfunction. Treat as regular emergency therapy while onset to alleviate the vertigo, nausea and the vomiting.

   (a) Benzodiazepines

   Diazepam

   Is the mostly common used in the benzodiazepine which has a better vestibular sedation function and is helpful to treat the vertigo, vomiting and could against the anxiety simultaneously.

   (b) Antiemetic drugs

   They have the sedation, the anti-cholinergic and the antiemetic function, including the dimenhydrinate, the metoclopramide, the phenergan, the anisodamine, the scopolamine, the atropine, etc.

   (c) Vasodilator

   Such as the betahistine, the radix salvia miltiorrhiza, the niacin, etc.

   (d) Diuretics

   Such as the hydrochlorothiazide, the chlorealidone and the chlorobenzene pteridine. Supply potassium while taking the thiazide.

   (e) Ca2+ antagonists: intermittent period available flunarizine and nimodipine.

   (f) The middle ear administration: Cochlear window film has a semi permeable effect of intratympanic injection of drugs through the penetration into the inner ear and therapeutic effects of antagonists: intermittent period available flunarizine and nimodipine currently used gentamicin or steroids to intra tympanic injection therapy [6]. The injection of gentamicin in the tympanum can destroy the secretory function of the vestibule dark cells to alleviate the membrane labyrinth and water accumulation. But because gentamicin has the risk of hearing loss, it is more suitable for patients with severe hearing loss. Steroid injection into tympanic cavity not only increases cochlear blood flow, but also inhibits immune mediated inflammatory response, and avoids the influence of amino glycoside drugs on hearing. Commonly used drugs with dexamethasone and methylprednisolone.

2. Surgical therapy

   It is suitable for frequent seizures, heavy symptoms, longer course of disease, ineffective drug therapy, and a significant impact on work and life. It is divided into destructive surgery and non-destructive surgery. The former mainly aims at the mechanism of water accumulation and the preservation of auditory function, the latter is mainly the innervation of the vestibule of ear, and the function of hearing is not necessarily preserved. For the pure tone audiometry speech recognition it; 70 dBHL and 20% were the best choice for hearing preservation surgery. Only one ear with hearing should avoid surgical therapy.

   (a) nondestructive surgeries

   - endolymphatic sac operation: endolymphatic sac decompression, endolymphatic bursa mastoid

drainage and endolymphatic sac subarachnoid drainage. The effective rate of all kinds of endolymphatic sac operations is about 75% [7].

- cryotherapy: semicircular canal window can reduce the canal within the lymph flow, reduce its sensitivity and selectivity is chosen to permanently reduce the vestibular function, has the dual value can control the vertigo and hearing preservation.
- three semicircular canal occlusion surgery

  It may be effective intreating vertigo in Ménière disease, mild postoperative reaction, vestibular compensation set up fast and more complete compensation.

(b) destructive surgeries

- labyrinthectomy: this method can only be used for hearing loss. Including chemical labyrinthectomy and physical labyrinthectomy. The former mainly uses amino glycoside drugs, such as streptomycin injection tympanic (Fig. 8.41), and also reports on the general use of drugs. But at present, there is no unified standard for the method, dosage, and schedule of the drug delivery. The latter mainly uses surgical methods to remove labyrinthine, which is suitable for single ear disease, long-term or recurrent symptoms, and severe hearing loss.
- Vestibular neurectomy: can effectively eliminate the serious symptoms of vertigo patients and to preserve function, but not eliminate tinnitus and ear fullness. It is possible to use the middle cranial fossa path, the posterior sigmoid sinus path and the posterior route of the labyrinth.
- Others: chordal tympanotomy (chorda tympanectomy), cervical sympathectomy (cervical sympathectomy), decompression through vestibular window such as balloon angiotomy (sacculotomy) and cochlear balloon stoma (cochleosacculotomy).

3. Meniett therapy

   In recent years, the inner ear therapy instrument (Meinert) developed in the United States can obviously relieve vertigo and improve hearing. Meniett therapy through the tympanostomy tube, will continuously low pressure pulse transmission to the middle ear cavity and its function in the cochlear window. The ear has not compressibility, low pulse energy produced a movement of perilymph, causing the endolymph inward longitudinal flow lymphatic and endolymphatic sac and local circulation and in the film lost in absorption and absorption, reducing natural endolymph, improving hydrolabyrinth, therapy of Ménière disease (Fig. 8.42) [8, 9].

Therapeutic Evaluation

1. Vertigo assessment

   The number of vertigo episodes in 18~24 months after therapy was compared with the number of vertigo attacks 6 months before therapy. Press: the score = number of vertigo episodes 18–24 months after therapy/number of vertigo attack 6 months before therapy by 100. It is divided into 5 levels, that is,

   - A level: 0 (complete control, unintelligible as “cure”); level
   - B level: 1~40 (basic control);
   - C level: 41~80 (partial control);
   - D level: 81~120 (uncontrolled);
   - E level: >120 (aggravated).

2. Hearing assessment: In the 6 months before therapy is the worst time for 250 Hz, 500 Hz, 1000 Hz, 2000 Hz and 3000 Hz to average a corresponding frequency threshold for 18–24 months after therapy, the minus deviation of the mean of the assessed.

   - A level: the improvement of >30 dB or the frequency threshold <20 dBHL;
   - B level: improve 15~30 dB;

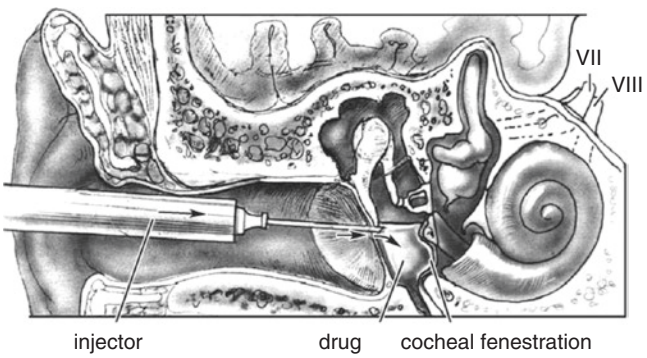


Fig. 8.41 Chemical labyrinth excision therapy

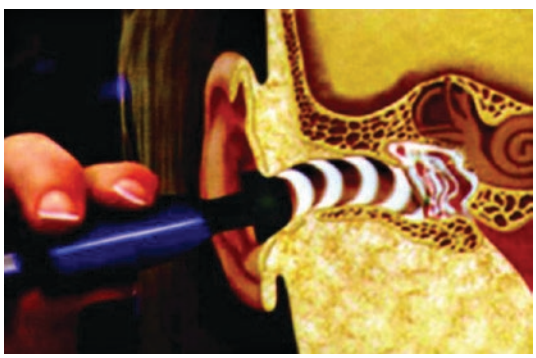


Fig. 8.42 Meinett therapy

- C level: improve 0~14 dB (invalid)
- D level: improve <0 (deterioration).

If the diagnosis is bilateral Meynier's disease, it should be evaluated separately.

3. Ability evaluation: The incidence of 18–24 months after therapy was compared with the 6 months before therapy. The score was equal to 18–24 months after therapy/the first half of the first half of the therapy day. It is divided into 5 levels, that is
   - A level: 0 (complete control);
   - B level: 1~40 (basic control);
   - C level: 41~80 (partial control);
   - D level: 81~120 (uncontrolled)
   - E level: >120 (aggravated).
   - Appendix: the day of onset: the sum of the day of activity limited/the sum of the days of observation. Limited day of activity refers to the days of 3 and 4 of the day's activities.
   - Activity score:
   - 0 points: any activity is not affected;
   - 1 point: mild activity;
   - 2 points: moderate activity, but no activity limitation;
   - 3 points: activities limited, unable to work, must rest at home;
   - 4 points: activities are severely restricted, all day rest or most activities cannot be accomplished [10, 11].

8.12 Acoustic Neuroma

Acoustic neuroma originates from the vestibular nerve (VIII brain nerve pair) the nerve cells of benign schwannoma. Originates from the nervous statoacusticus canal segment, or the inner ear nerve sheath crossing at the start of the inner ear or bottom, is one of the most common intracranial tumors, accounting for 7–12% of intracranial tumors, accounting for the cerebellopontine angle tumors 80–95%. More common in adults, the peak of the peak in 30~50 years old, less than 20 years old, children's single acoustic neuroma rare, no significant gender differences. The incidence at left ear and right ear is similar, and occasionally bilateral. Bilateral acoustic neuromas usually indicate neurofibromatosis type 2 (neurofibromatosis2), also known as central neurofibromatosis (central neurofibromatosis) or multiple bilateral acoustic neurofibromatosis (bilateral acoustic neurofibromatosis), is a kind of euchromosome dominant genetic disease.

Clinical Manifestations

The symptoms of acoustic neuromas are mainly related to the size and location of tumor and the degree of compression to peripheral nerves, blood vessels and brain tissue. Main symptoms and signs:

1. Cochlear and vestibule symptoms: Showed unilateral tinnitus and deafness, of unilateral hearing loss, a minority of patients with sudden hearing loss as the first symptom, tinnitus for high pitched, cicadas or whistle, dizziness, vertigo and unstable sense of attack [12].
2. Headache: the frontal occipital pain is accompanied by discomfort in the large orifice of the lateral occipital bone.
3. Cerebellar ataxia: Unstable walking, horizontal tremor of the eyeball, and dysfunctional motor function.
4. Cerebellopontine angle syndrome: Symptoms and signs of nerve, facial nerve, trigeminal nerve and posterior group of brain nerve disorders, cerebellar damage and brain stem compression were the symptoms of adjacent brain nerve damage. As the disease side pain, facial twitching, facial hypesthesia, facial paralysis and eating cough, hoarseness, pharyngeal reflex disappeared or decreased; ipsilateral corneal reflexes diminish or disappear.
5. Symptoms of increased intracranial pressure Including optic disc edema, headache aggravation, nausea and vomiting, diplopia, etc.

Specialist Examination

1. Check the audiological examination: It includes pure tone audiometry, speech recognition rate, acoustic conductivity, distortion product otoacoustic emission, cochlear electrogram, ABR latent Dynasty, etc. The pure tone audiometry showed different degrees of sensorineural deafness, the rate of speech recognition was significantly reduced, and the ABR examination showed the post cochlear lesion.
2. Vestibular function examination: the function of the semicircular canal is diminished or lost.
3. Peripheral facial paralysis: if there is facial paralysis, the electro facial electrogram should be examined.
4. Imaging examination
   (a) X-ray photography of the inner ear canal: enlargement of inner ear canal and destruction of otolith ridge.
   (b) axial bone CT: showed enlargement of inner ear canal (Fig. 8.43). There were ISO density or low-density lesions and obstructive hydrocephalus in cerebellopontine angle area.
   (c) Cranial MRI: MRI is the most sensitive and effective method at present. It shows that T1 weighted images of cerebellopontine angle area show low signal or equal signal. T_2 weighted images show high signal occupying lesions, and enhanced scan lesions are significantly enhanced (Figs. 8.44 and 8.45).
   (d) Cerebral angiography: can understand the blood supply of tumor and mediator embolism reduces intra operative bleeding.

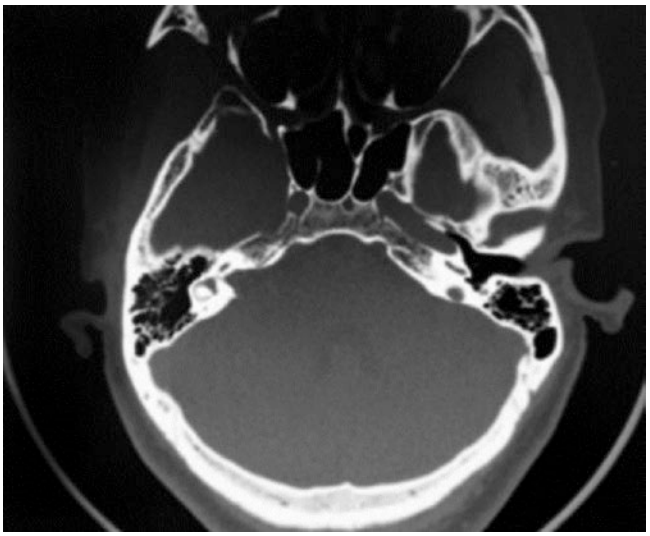


Fig. 8.43 Axial temporal CT of right acoustic neuroma: right inner ear canal enlargement

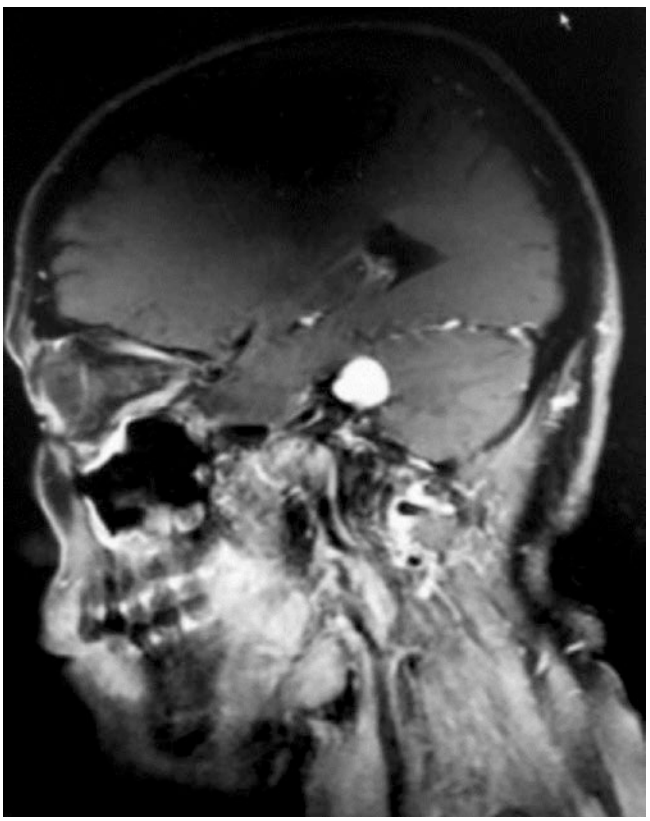


Fig. 8.44 Weighted sagittal MRI T_1WI shows: acoustic neuroma

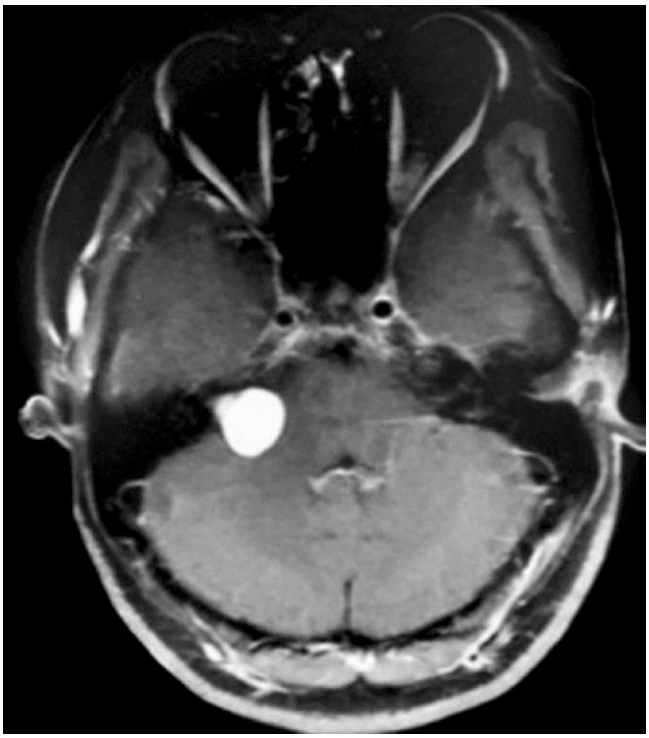


Fig. 8.45 Weighted MRI: right acoustic neuroma

Diagnosis and Differential Diagnosis

1. Diagnosis

   According to the results of patients with symptoms, signs, audiological examination and characteristic imaging (temporal CT and brain MRI) can be diagnosed.

2. Differential diagnosis

   This disease and facial neuroma, cerebellopontine angle meningioma, congenital cholesteatoma, arachnoid cysts, glioma, neuron inflammation, sudden deafness, and Ménière disease identification.

Treatment

This disease is a slow but life-threatening benign tumor of the brain. There is no drug to inhibit the growth of the tumor. Surgical resection is the first accepted therapy. Small tumors by microsurgical resection, application of facial nerve and auditory evoked potential monitoring technology, function and auditory function to maximize the overall nerve.

1. Operation the approaches to acoustic neuroma surgery include:
   (a) the middle cranial fossa approach: suitable for confined to the inner ear in acoustic neuroma, and retain the residual hearing of patients.
   (b) by the way of backward Road: applicable to the main body in the cerebellopontine angle, medium size (diameter 2.5 cm) tumor resection, which is conducive to the preservation of residual hearing, not to hurt the lost structure.
   (c) labyrinthine approaches: it is suitable for severe hearing loss, facial nerve function is normal, and the tumor originates from the inner ear canal protruding

in the cerebellopontine angle pool, which is good for preserving the facial nerve function.

(d) retrosigmoid or suboccipital approach: this approach is suitable for large auditory neuromas located mainly at the cerebellopontine angle.

2. Stereotactic radiotherapy (α knife, γ knife) Suitable for surgical contraindication, no increased intracranial pressure, tumor diameter <2 cm, unwilling to accept operation, can also be the first large tumor resection and (or) ventricular shunt relieve intracranial hypertension after α knife or γ knife therapy.
3. MRI continuous observation of tumor growth: it is suitable for patients with asymptomatic acoustic neuroma, aged (>70 years), unsuitable for operation or unwillingness to operate.

8.13 Genetic Diagnosis and Prenatal Diagnosis of Hereditary Deafness

Deafness is the most common and high incidence of deafness in human beings, with about 60% of the cases are hereditary deafness. There are about 78 million carriers of mutuated deafness gene in China with the overall carrying rate of 6%, 80% of children are born to hearing parents, and a large number of parents with delayed-onset hearing loss, mostly due to a defect in a gene polymorphism and its cause of deafness caused by environmental factors increase susceptibility to disease [13]. Hereditary deafness is mostly sensorineural deafness, which cannot be diagnosed according to clinical manifestations, and there is no effective drug therapy. Gene screening for deafness patients can, to a certain extent, clarify the cause of deafness, and guide patients and their relatives to take precautions in medication and life, prevent birth defects, control the risk of drug-induced deafness, intervene or delay the occurrence and development of deafness.

1. Genetic deafness genes could lead to the hereditary deafness, about 70% are non-syndromic deafness (non syndromic hearing loss, NSHL), and the rest about 30% are syndrome type deafness (syndromic hearing loss, SHL). The inheritance of NSHL could be expressed in a variety of genetic methods, including autosomal dominant (related chromosomal loci named DFNA), autosomal recessive (related to chromosomal loci named DFNB), X-linked NSHL (genetic linkage related chromosomal loci named DFNX), Y-linked NSHL (genetic linkage related chromosomal loci named DFNY) and maternal inheritance. Among them, 75~80% are autosomal recessive, 10~20% are autosomal dominant, X-linked NSHL is less than 2%, and a Y-linked Chinese deaf family has been reported now. Autosomal recessive deafness is mainly manifested in pre-lingual deafness, while autosomal dominant hereditary deafness is mainly manifested as post-lingual deafness [14, 15].

   In 1999, Chinese academics Xia Jiahui, etc. reported 2 unrelated autosomal dominant Chinese deafness families. A heterozygous mutation of *GJB3* was found in two Chinese deaf families, and the *GJB3* gene was cloned. As of March 2015, there were 144 loci of non syndromic hearing loss in the world, including 31 autosomal dominant genes, 56 autosomal recessive genes, and 5 X-linked NSHL genes. At present, the target of deafness gene screening has been widely carried out in China. It is mainly focused on screening of *GJB2*, *SLC26A4* and mitochondrial *12sRNA A1555G* mutations [16].
2. Diagnostic techniques and applied strategies for genetic hearing loss gene screening

   (a) Screening and diagnostic techniques for deafness genes

   There are a variety of diagnostic techniques for deafness gene screening, which are simply summarized as two kinds of indirect and direct methods.

   - Direct method

     It is the sequence analysis of the target gene directly to identify whether there is a pathogenic mutation. It includes direct sequencing, gene chip, mass spectrometric analysis and SNP typing, each with advantages and disadvantages. The purpose of direct sequencing is to determine whether the specific genes of the subject are defective. It needs to make sure that a gene is a pathogenic gene of a disease, and known mutations, and take the peripheral blood or other tissue samples (including paraffin sections) of patients (or pro-band), to detect whether there is a genetic mutation of the gene by testing. This is the most clinically practical genetic deafness gene diagnosis strategy. The most commonly used method of mutation analysis is PCR-direct sequencing, and GeneChip and the next-generation of genome wide sequencing is also widely used in the detection of cloned deafness genes [17]. For the deaf families or the sporadic patients with unclear genotype, hot spot mutation screening is the most popular method. Gene mutation screening is based on a variety of experimental technology (PCR-HELP, fluorescent PCR, IMLDR, etc.) [18].
   - Indirect method

     Chromosome haplotype analysis, is closely linked to genes or genetic markers which are often passed to offspring based on inspection and therefore the adjacent DNA is passed to the offspring, can indirectly determine the causative gene is passed to the offspring, the purpose is clear the subject whether there from parental genetic dis-

ease gene. Genetic markers for accurate positioning information which needs to clear disease genes and the disease gene is closely linked to the relationship, the samples of patients (or pro-band) and their family members in peripheral blood or other tissue specimens (including paraffin), genetic heterogeneity of the ring shadow, DNA recombination the influence of. The linkage analysis uses a variety of DNA polymorphic loci widely used in the genome, especially gene mutation sites or adjacent polymorphic loci as markers. FFLP, SSCP, AnT and other techniques can be used for linkage analysis. This method is often used in the location and cloning of a new pathogenic gene for hereditary deafness.

Due to genetic deafness with genetic heterogeneity is very strong, most of the existing detection technology for mutation to, used for screening, has no clinical specificity, and detection of hereditary deafness are standard genetic deafness gene screening and diagnosis at home and abroad and there is no unified standard [19], at present. The team led by Professor Feng Yong of Xiangya Hospital, Central South University, has filed a diagnosis of genetic deafness phenotype of the strategy based on the hope from the current hot gene screening and has gradually achieved the purpose of gene diagnosis. Figure 8.46 is a genetic diagnosis strategy for genetic deafness developed by the Xiangya Hospital of Central South University.

(b) Clinical application strategies

- Candidate gene detection based on clinical phenotype: the application range is phenotypic—genotyping of sporadic patients or deaf people. According to the database and literature reports, we have drawn the phenotype genotype spectrum of genetic deafness genes [20]. Based on genotype and phenotype characteristics of various hereditary deafness we have worked out effective gene diagnosis strategy for hereditary hearing loss, part of which is shown in Fig. 8.47 [21].
- Hot spot mutation screening

Applied range of deafness or phenotypic and genotypic undefined sporadic patients. For patients with unclear candidate gene screening, according to different clinical needs, Professor Feng Yong's

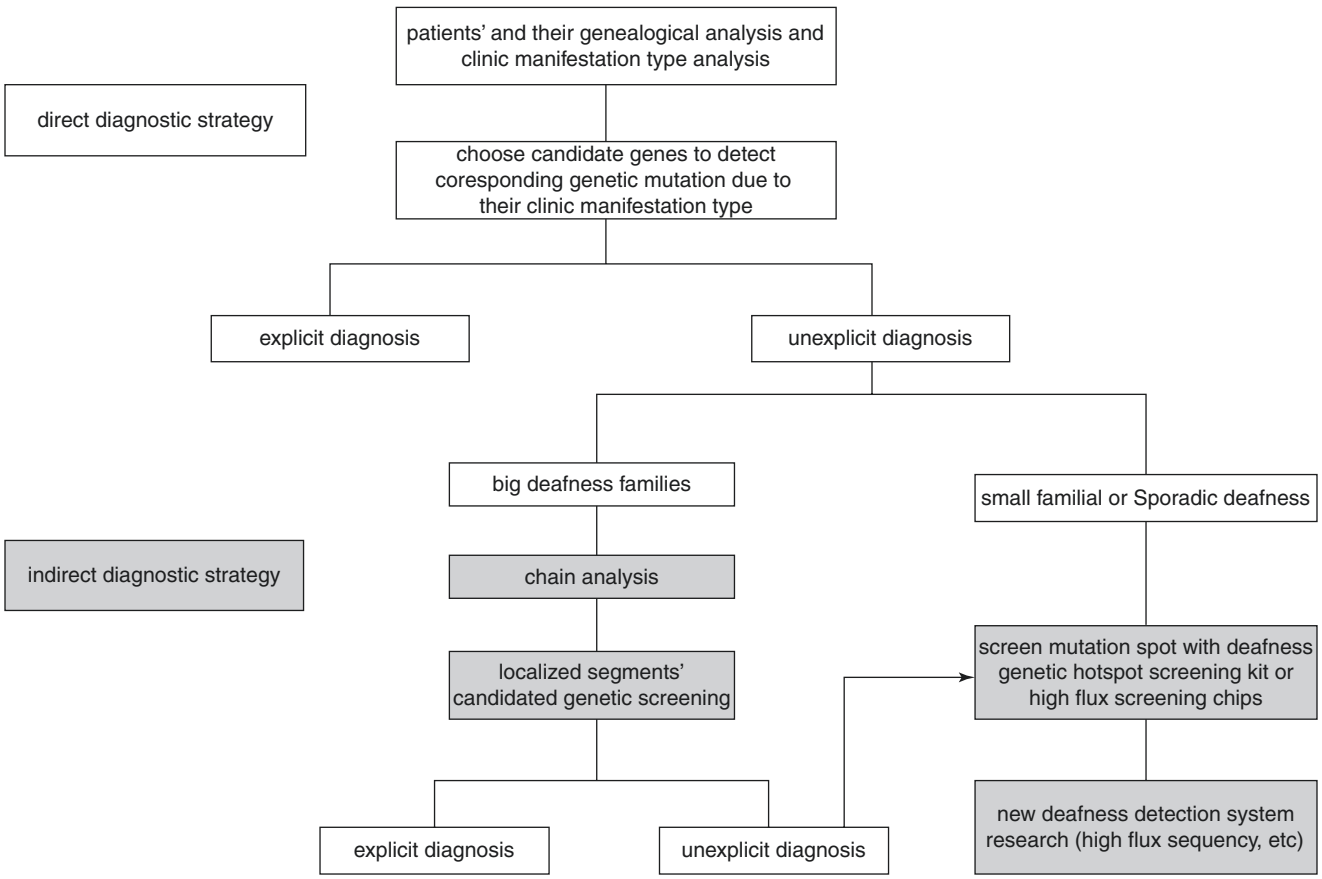


Fig. 8.46 Xiangya hospital's genetic deafness's molecular diagnostic strategy figure

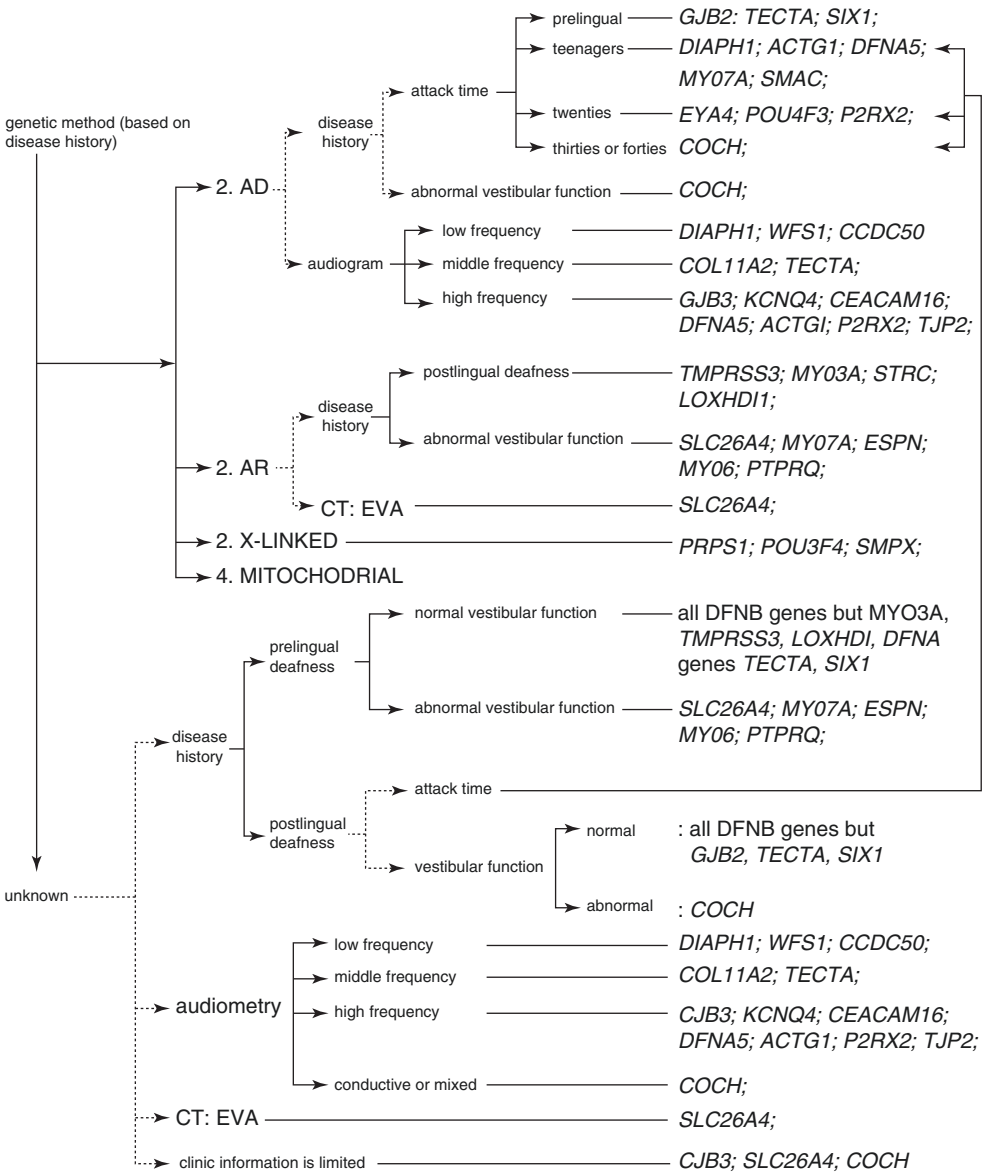


Fig. 8.47 Xiangya hospital's genetic deafness genetic diagnosis strategy

team of Xiangya Hospital, Central South University has designed various hot spot mutation kits. They are based on restriction fragment length polymorphism analysis (RFLP, which can detect 3 hot spots), fluorescent PCR Technology (detectable 14 hot spot mutations), and a new multiple band ligase Technology (IMLDR, which can detect 32 hotspot mutations) [22].

- high-throughput deafness gene diagnosis chips and copy number variation (CNVs) analysis:

Application scope is deafness gene mutuation screening in many hot spots where unclearly diagnosed patients could furthur expand the use of mutations screening, to establish a high throughput genetic deafness gene chip combined with mutation screening copy number variation for the currently known deafness mutation screening. For instance, Xiangya Hospital of Central South University [23] and Illumina company Goldengate384 chip project can detect 240 internationally recognized deafness causing mutations (including 77 dominant and 163 recessive mutations, also included 144 SNPs, of known deafness related high-throughput screening and deafness related mutation pedigree preliminary exclusion positioning. Copy number variation refers to the wide variation of human genome from deletion of 1000p (base pair) to millions of BP, insertion, repetition and complex multiple loci. The current molecular diagnosis of deafness is mainly focused on point mutation detection. There are reported copy number vari-

ants associated with deafness (WS, Usher syndrome, Teacher- Collins- Franceschetti syndrome, etc.), and CNVs has been detected in related genes. Therefore, the CNVs detection is included in the routine molecular diagnosis as a supplementary means for the detection of the clinical deafness gene, and the simultaneous detection of SNP/CNVs is realized [24].

(c) high throughput sequencing

The scope of application is to screen the genetic deafness patients without hot spot mutation, and it is also the future research direction of gene diagnosis. High throughput sequencing is based on the next-generation sequencing platform, which uses target acquisition technology to conduct high-throughput sequencing of deafness related tens or even hundreds of genes, which greatly improves detection efficiency and detection range [25]. It is suitable for finding new genes for deafness and screening for deaf patients without hot spots. Domestic and foreign scholars have cloned 8 deafness related genes (TPRN, GPSM2, CEACAM6, SMPX, HSD/7B4, HARS2, MASPI and DNMT1) using the second generation sequencing technology [26]. In addition, the development of this technology can further detect system based on the model, such as fragment enrichment technology to efficiently capture the target gene, reduce the cost of high-throughput sequencing, such as the Professor Feng Yong's team developed based on next-generation high-throughput sequencing technology "Waardenburg syndrome" and "large vestibular aqueduct syndrome" mutation detection kit.

3. Prenatal diagnosis

Prenatal diagnosis is a genetic diagnosis of a fetus with a risk of hereditary disease in the uterus before birth. By amniocentesis and chorionic villus sampling techniques, genetic and biochemical examination of amniotic fluid, amniotic fluid cells and villi were analyzed, the analysis and diagnosis of fetal chromosome and gene, is the effective way to prevent the birth of children with genetic diseases. Since the end of 1980s, PCR technology has been applied to prenatal gene diagnosis. With the development of this technology, people have received more and more attention and welcome. As far as it is concerned, it has become the most commonly used technique for genetic diagnosis of genetic disease. Various mutation gene detection methods based on this technique have become the main means of genetic disease gene diagnosis.

On the technical level, prenatal diagnosis is divided into invasive prenatal diagnosis and noninvasive prenatal diagnosis. The commonly used noninvasive prenatal diagnosis include amniocentesis, chorionic villus sampling, umbilical cord puncture, umbilical blood sampling, fetal tissue biopsy, pre-implantation genetic diagnosis; noninvasive prenatal diagnosis, including the detection of fetal cells in maternal peripheral blood by cervical exfoliated fetal trophoblast cells detection. According to the classification of the applicable stage of pre-implantation genetic diagnosis for in vitro fertilization embryo before implantation; often used in early pregnancy chorionic villus sampling (10~12 weeks), early amniocentesis (12~14 weeks), the detection of fetal cells in maternal peripheral blood (10 weeks, 15 weeks of gestation; mid pregnancy often uses the best) amniocentesis (16–18 weeks of gestation, umbilical blood sampling best) (after 16 weeks). These prenatal diagnosis methods have their own advantages and disadvantages. Amniocentesis is the most mature and widely accepted prenatal diagnosis technology because of its accuracy in diagnosis and low risk for pregnant women.

The prenatal diagnosis of hereditary deafness is currently used for members of the hereditary deafness family, which has been diagnosed by genetic diagnosis. When mother is pregnant, the fetal DNA is extracted through the above way, and then the prenatal diagnosis is made according to the genetic diagnosis of hereditary hearing loss. According to the results of prenatal diagnosis, early intervention can be done to avoid the birth of the new hereditary deafness [27].

4. The intervention system of hereditary deafness

Due to the lack of effective therapy, the three best interventions for hereditary hearing loss based on accurate gene diagnosis is the best choice. According to the result of gene diagnosis, we can intervene in deaf people of different ages. A level of intervention, specifically for the prenatal diagnosis of hereditary deafness and positive results of fetal prenatal intervention to reduce the incidence of grade two; early intervention, hearing screening and genetic screening of newborns has, according to the hearing status by effective early intervention (BAHA or cochlear hearing aids, etc.), reconstruction of hearing and speech function [28]; level three intervention, language of deaf patients, provide guidance on marriage according to audiological examination results by effective intervention (hearing aid fitting, BAHA or cochlear implantation) to establish the hearing, improve the quality of life of patients [29].

Due to the high genetic heterogeneity of genetic deafness and the fact that many pathogenic genes are not cloned, the genetic deafness detection carried out before is mostly limited to known genes or mutation sites. There are some limitations in clinical application. However, with the deepening of clinical phenotypic research on genetic deafness, the progress of molecular biology detection technology and the development of bioinformatics, the diagnosis technology of genetic deafness will continue to improve, and will eventually be widely applied in

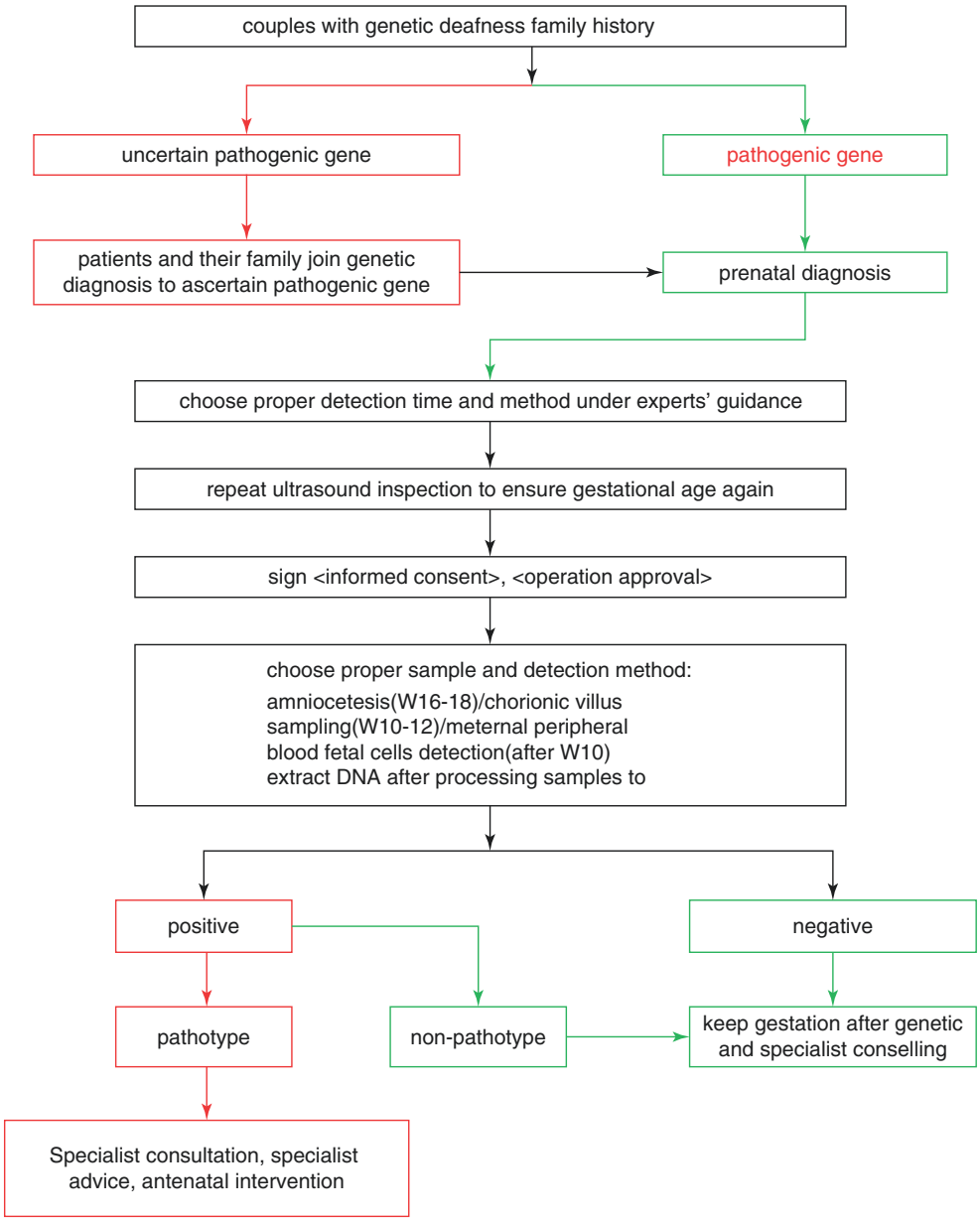


Fig. 8.48 Prenatal genetic deafness diagnostic consultation and strategy flow

clinical practice and the prenatal diagnostic consultation and strategy flow of the genetic deafness could be made due to the information at Fig. 8.48.

8.14 Neonatal Hearing Screening

Newborn hearing screening can early detect congenital hearing loss, early intervention and early rehabilitation can make it deaf but not dumb and return to the mainstream society. However, universal newborn hearing screening is a systematic project, including screening, diagnosis, intervention, rehabilitation, follow-up and quality assessment, etc., which could only be accomplished by cooperation and adjustment among all the projects [30].

8.14.1 Summary of Newborn Hearing Screening

At 1993, the U.S. National Institutes of Health (NIH) recommended to carry out the universal newborn hearing screening. At 2000, American Joint Committee on infant hearing (JCIH) published the report, 'principles and guidelines for early hearing detection and intervention of the situation' [31], the principles and guidelines for newborn hearing screening became clear [31]. At 2007, 'situation report about principles and guidelines on early hearing detection and intervention project' [32] published by the United States JCH updated 8 structures of target hearing loss, including the definition, initial hearing screening and second hearing screening, the diagnostic audi-

tory evaluation, the medical evaluation, the early stage intervention, surveillance and screening in the medical home, the communication and information infrastructure, and strengthened the monitoring to the auditory neuropathy [32].

At 1990s, China carried out the universal newborn hearing screening project (UNHS) and developed under the support of the government. Since 2004, when the former ministry of health promulgated the 'technical gists of newborn hearing screening', UNHS had gradually spreaded at some of Chinese provinces and municipalities. At 2010, they promulagated the 2010 version of the gists to detaledly formulate the process and the quality of the newborn hearing screening. Till June, 2014, after the UNHS was widely carried out at Tibet, there are 32 provinces, municipalities and autonomous regions in total where it developped. According to incomplete statistics, recently the UNHS has been carried out at 77% of China. At some municipalities and sites in China, the goverment helps expanded the newborn hearing screening projects by researching at combining screening of the newborn auditory and the gene [33].

8.14.2 The Principle of UNHS

The current screening strategy for newborns in China is universal screening. The universal hearing screening refers to the application of electrophysiological detection technology to the hearing screening of all live newborns. UNHS is a systematic project, including screening, diagnosis, intervention, rehabilitation, tracking and quality assessment. Throughout the process, multi-disciplinary cooperation is always running as a vital role on accomplishment of UNHS.

The main principles of UNHS would be followed

1. General screening. All newborns in normal delivery room and NICU should receive hearing screening during hospitalization. Those who failed to pass the procedure should be rescreened at 42 days after delivery (NICU neonates should directly enter the diagnostic procedure).
2. Accept diagnosis in 3 months. To all infants who don't pass the secondary screening, auditory and medical evaluation should be performed in 3 months to ensure the existing of hearing loss.
3. Accept intervention in 6 months. All infants who have been diagnosed with permanent hearing loss should receive early intervention within 6 months old [34].

8.14.3 Newborn Hearing Screening Technique

Otoacoustic emissions (OAE) and automatic auditory brainstem response (AABR) are commonly techniques used during UNHS.

8.14.3.1 Otoacoustic Emissions

Otoacoustic emissions (OAE) is a kind of audio energy generated from the ear, "the cochlea, the auditory ossicle chain and the tympanic membrane conduction", which are applied to the external ear canal. It reflects the function of the outer hair cells of the cochlea, but it is easy to be influenced by the function of the external middle ear. Usually, hearing loss exceeds 40 dB HL which OAE cannot record. The otoacoustic emissions technologies commonly used include the transient evoked otoacoustic emission (TEOAE) and distortion product otoacoustic emission (DPOAE). The reason why otoacoustic emission (OAE) is used for hearing screening is that it has many advantages, such as fast, accurate, objective, sensitive, noninvasive, simple and stable. But OAE also has some limitations: It can only reflect the function of the cochlear outer hair cells, the simple use of OAE screening, easily missed diagnosis of auditory neuropathy; susceptible factors of ear, ear residual fetal fat and amniotic fluid of incoming and outgoing stimulus response caused by otoacoustic emission attenuation, Create the illusion of not throughing [35, 36].

8.14.3.2 Automatic Auditory Brainstem Response

AABR is an electrophysiological detection technology based on ABR. It can reflect the function from cochlea to auditory brainstem, and is relatively less affected by external and middle ear factors. Combination use with OAE can detect lesions behind the cochlea and reduce the false negative of hearing screening. However, AABR is not sensitive to low frequency hearing loss and mild hearing loss [37]. The results of hearing screening were recored as "pass" (through) or "refer" (not pass), but not "normal" or "abnormal" [38].

8.14.4 The process of UNHS [39]

1. Normal birth neonates are screened by two-stage screening: the initial screening was completed within 48 h after birth to the discharge, and rescreening month on both ears should be carried out within 42 days. Those who have not passed the screening should be referred to the children diagnosis centers for further diagnosis at the age of 3 months
2. Infants in the neonatal intensive care unit (NICU) were screened through automatic auditory brainstem response (AABR) before discharge, and if the newborns refer at initial screening, they should be transferred to children hearing diagnostic centers appointed by the provincial health administrative department 3 months after birth.
3. Neonates with high risk factors for hearing loss should be followed up once a year till 3. If they suspected to perform hearing loss during follow-up, they should be treated for hearing diagnosis in time.

High risk factors for hearing loss [40]:

1. The neonatal intensive care unit (NICU) was hospitalized for more than 5 days.
2. Family history of children with permanent hearing impairment.
3. Intrauterine infection caused by cytomegalovirus, rubella virus, herpes virus, syphilis, or toxoplasmosis disease.
4. Crania facial morphologic deformity, including auricle and ear canal deformity, etc.
5. The birth weight was less than 1500 g.
6. Hyperbilirubinemia meets the demand for change of blood.
7. Viral or bacterial meningitis.
8. Asphyxia neonatorum (Apgar score of 1 min, 0–4 or 5 min 0~6).
9. Premature infants with respiratory distress syndrome.
10. Oxygen in vitro membrane.
11. Mechanical ventilation was more than 48 h.
12. Pregnant women had used ototoxic drugs, loop diuretics or alcohol and drug abuse
13. In clinic or suspected to be associated with hearing disorder syndrome or hereditary diseases.

8.14.5 Audiological Assessment and Diagnosis

ENT doctors have the responsibility and obligation mechanism, diagnosis and therapy of combined screening institutions and ask parents to pay more attention to the early hearing diagnosis of infant, who play important role in the universal newborn hearing screening system. The diagnostic principles and procedures are as follows:

1. Newborns without passing should be diagnosed within 3 months.
2. Newborns in NICU who refer UNHS should be directly referred to the children hearing diagnostic centers for diagnosis and follow-up.
3. The hearing diagnosis should be intersected according to the test results to determine the degree and nature of hearing impairment. Children suspected of having other defects or systemic diseases were instructed to go to related departments, suspected of hearing impairment due to genetic factors, and genetic counseling for conditional health care institutions.
4. Diagnosis process
   (a) Collection of medical history.
   (b) Otolaryngology examination.
   (c) Hearing tests should include the contents of electrophysiological and pediatric behavioral audiometry, including acoustic impedance (including 1000 Hz detection), otoacoustic emissions (OAE), auditory brainstem response (ABR) and behavioral audiometry [41].
   (d) Auxiliary examination, if necessary, relevant imaging and laboratory auxiliary examination [1].

8.14.6 Intervention

The diagnosis of children with permanent hearing impairment should be carried out in clinical.

8.14.7 Follow Up

1. The screening agency was responsible for the follow-up and rescreening after initial screening. The patients who refer UNHS should be referred to the children hearing diagnostic centers.
2. The diagnosis and treatment facility should be responsible for the follow-up, diagnosis for hearing impaired children every six months (at least one time).
3. The requirements and procedures of follow-up work should be developed and included in the routine of maternal and child health care. The maternal and child health care institutions should assist the diagnosis and therapy institutions to complete the follow-up of the children diagnosed, and collect all the data of registration and preservation, instruct the community health service center to finish that well.

8.14.8 Rehabilitation

1. For children who use artificial hearing devices, professional hearing and speech rehabilitation training should be carried out. Check and debug regularly.
2. Parents or guardians who instruct children with hearing impairment are put on record to the relevant departments of the resident and the disabled in order to receive family rehabilitation guidance.

8.14.9 Quality Control

The health administrative department should organize the formulation of the assessment and evaluation plan, supervise and inspect the screening institutions and hearing impaired diagnosis institutions regularly, control the quality of every link of newborn hearing screening, find out problems and take effective measures timely.

The newborn hearing screening center or the medical institution designated by the health administrative department to undertake the hearing impairment diagnosis and therapy work should establish the newborn hearing screening database, and carry out the information management work of UNHS [42].

8.15 Hearing Aids

8.15.1 Hearing Aids Development History

Hearing aids (hearing aid) are small expansion device for hearing loss and compensation for hearing loss. Its development has gone through seven times: set the sound cylinder device, carbon, electron tubes, transistors, integrated circuits, microprocessors and digital hearing aids era.

Today's hearing aids has entered the full digital signal processing interface generation, including compression and amplification, noise reduction technology, digital feedback control technology and direction of technology, the application of these technologies, it can meet the hearing impaired. With the continuous development of digital chip technology, hearing aid technology include the latest simulation of complex wide dynamic range compression system, net noise system, dual stabilizer digital feedback control technology, intelligent system, reality adaptive directional conversion system, open ear technology, real-time data analysis system and fitting software and hardware support system, can provide users with clear, comfortable and natural listening experience. From the appearance and development of BTE, cassette, glasses, hairpin, pen, wireless and other shapes for different patients, more beautiful appearance effect. We believe that in the future, the volume of the hearing aids will become smaller and smaller, and the function will become more and more perfect.

8.15.2 Hearing Aids Categories

There are many ways to classify hearing aids.

1. According to their shape

   Can be divided into 5 categories: cassette, glasses, behind-the-ear type, in the ear canal type, and bone-anchored hearing aid. The glasses type BTE hearing aids, also known as ear level hearing aid, hearing aid ear level hearing aid receiving sound manner than other types of more close to the physiological state.

   (a) box hearing aid: The box hearing aid appeared earlier and larger. It looked like a miniature radio. It wore a conductor to transmit the voice output signal to the earphone before wearing it. More ordinary transistor components are used, so the price is low and the background noise is high. For the fingers gone: because the box hearing aids often friction with clothing, the sound of friction often becomes noise (Fig. 8.49).

   (b) glasses hearing aids: Only a body worn to the development process of ear level aid in transition products, microphone and receiver capable of leg in different, the signal of (contralateral routing of signal, CROS), are also in the same mirror on the legs (Fig. 8.50).

   (c) BTE: BTE hearing aids is now the most widely used, the shape of slender, rely on a hard plastic ear hook bent into a semicircular hang on the ear, can use the skin or hair color shell to hide, the amplified sound through the ear hook through a plastic the incoming sound hole in the eardrum tube (Fig. 8.51).

   (d) in the ear canal hearing aids (ITE-HA)

   It includes the ITE-HA (Fig. 8.52) and the in-the-canal HA (ITC-HA). The ITE-HA contains the gen-

Fig. 8.49 Box hearing aid

Fig. 8.50 The eyeglass hearing aids (EG-HA)

Fig. 8.51 Behind-the-ear hearing aids (BTE-HA)

Fig. 8.52 ITE-HA

eral and the partial HA. The ITC-HA has the complete in-canal (CIC) hearing aid (Fig. 8.53). The ITE has an exquisite configuration and is custom-made. Input the aid into the cavum conchae or channel conceals without any tubes or wires. The output power of the ITE isn't high, so it is only helpful to the mild and moderate deafness.

Fig. 8.53 ITC

Fig. 8.54 BA-HA

(e) bone anchored hearing aids (BA-HA)

It is a surgical implanting system to treat the hearing loss, conducting the sound through the temporal bone but not the mid-ear (Fig. 8.54). The full introduction is as before (Figs. 8.31 and 8.32). The U.S FDA ratified that the BAHA could be applied on treating the conductive and mixed deafness in 1996 and on unilateral sensorineural deafness in 2002. The BAHA is used to treat the otitis media, the congenital ankylotia and the unilateral deafness who can't use the regular hearing-aids.

2. Classification due to dynamic frequency response

It can divided into two types according to the dynamic frequency response characteristics. After people measure the static frequency response of the hearing aids with several kinds of equipments, they focus more on its dynamic characteristics because that the sound we hear is in dynamic change at the frequency and the strength.

(a) Fixed frequency response hearing aids (FFR-HA)

Most of the hearing aid in market is the FFR-HA. Its frequency response is determined while being produced. The intonation button could only adjust a certain degree of the frequency response. Once the

optional Personnels set the parameters of the hearing aids, the frequency response is certain no matter what kind of acoustic condition the patient is in.

(b) Level dependent frequency response hearing aid (LEFD-HA)

The typical TILL type hearing aids is applied with the K-Amp circuit, but most of the programmable hearing aids with wide dynamic range compression circuit are more matching the LDFD-HA characteristics.

3. Classification due to the effecting area

It is divided into the group hearing aids and the personal hearing aids. The personal ones fit specific personnel and the group ones are always applied to the audio-visual education, the outdoors education and especially the deaf-infants rehabilitation or school education. According to the specific function, it could be classified into three types.

(a) Fixed wired collective hearing aid (FWC-HA)

Similar to the audio spoken language classroom system: there are a master for the teacher and an extension for each student, both having a microphone and an earphone. The master computer could attach to the recorder or other assisting facilities. The information can also be shared between the master and the extension and the extensions, which causes the free discussion possible. With the advance group hearing aids, the teacher can adjust the earphone volumes according to each one's condition without any distance limit and let all of them could hear clear and moderate voice no matter where they are sitting. All above offer benefit to the spoken language teaching and improve the deaf's linguistic competence. But a coin has two sides. The disadvantages are the position limit and the possible discrepant frequency response compared to their own.

(b) Frequency modulation hearing aid (FM-HA)

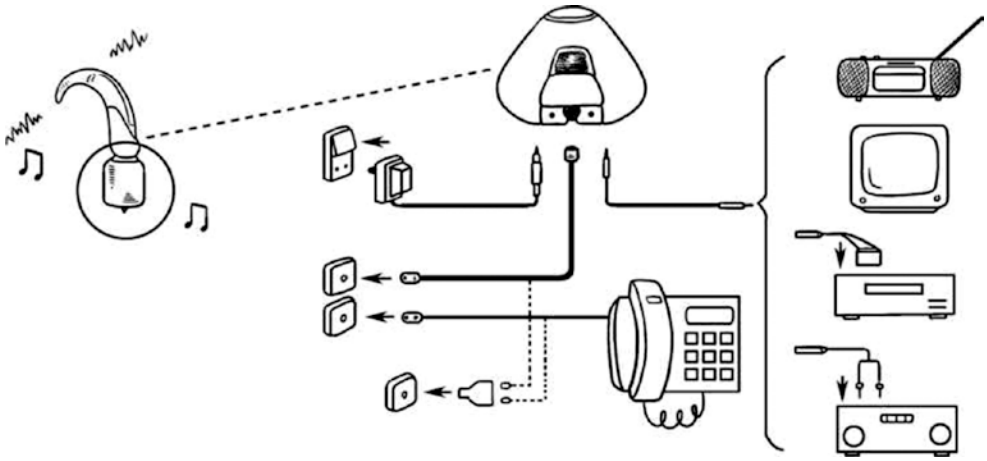
The sound source passes through the frequency modulation producer (microphone-like) to be shared and accepted by one or more frequency modulation hearing aids. The FAMHA worn by the deafness not only could accept the demodulated sound signals by the 'demodulation' units, but also it can be used as a common hearing aid because of its same fitting parameters as the common ones (Fig. 8.55). The FMHA is convenient for the deafness by expanding their action area into a 100 radius and for the outdoor education of the deaf infants. The one-to-one hearing aids fit those deaf infants who have accepted the listening and speech exercise and been able to take class with normal infants in school. The teachers wear a microphone, so the deaf could hear the teachers clearly wherever they sits. Some family set the frequency modulation producer beside the TV louder to help the deaf hear the TV audio.

(c) Closed-circuit electromagnetic induction collective hearing system (CEMIC-HS)

Also called the Induction loop system (ILS). It includes the main console (the amplifier, the frequency modulation components), the coil early arranged in the indoors such as the classrooms and the family houses and the personal hearing aids with T gear (Fig. 8.56). The trans-audience, the amplifier and the frequency modulation units could emission the electromagnetic voice of the recorder, the radio, the TV and the teachers to the area the coils encircle. The deaf could turn the hearing aid to the T (tele-coil) gear to accept the distinct voice without the distance and the number of people limit through the electromagnetic induction principle in the coil-arranged indoors room. The result is directly related to the coil setting and the T gear telecoil sensitivity.

Fig. 8.55 FM-HA

Fig. 8.56 CEMIC-HS/ILS



8.15.3 Hearing Aids Fitting

'Fitting' means that the user chooses the proper hearing aid under guideline of the professionals. Similar to the glasses, the hearing aids are also a kind of the 'fitted' auxiliary equipments for human physiologic functions.

Firstly, the hearing loss of each person varies. Cite the presbycusis examples, different deafness types, different hearing loss degrees, presence of the tinnitus, sensitivity to the voice level, audition recognition ability, etc. cause different audition difference. Different hearing loss needs different hearing aids. Actually, even to the deaf at the same property and level of the hearing disorders, the personal feeling may differ. These differences may be relative to the habits, the work environments, the subjective tolerance capacity, etc. The feeling difference cause different requirements to the hearing aids.

Secondly, there is quantity of the hearing aids brands and models, which cause that the fitting, could only be asked to be done by the professionals. Such as how the patients with severe hearing loss but sensitive to the high intensity sound choose the hearing aids? They would in-tolerate the voice while focusing on the volume up; they would not hear the voice clearly while focusing on the high intensity sound compression. Balancing the conflict is one of the main missions the professional fitting should do.

The perfect aided effect could only be achieved when the audition conditions and the chosen hearing aids' coefficient completely tally. The hearing disorders type decides the hearing aids property, and the 'fitting 'is the bridge between them. Most of the unfitted hearing aids can't achieve aided effects, possibly do harm to the remnant hearing of the wearer and even cause severer hearing loss. Due to experience above, the hearing aids should be adequately fitted by the professional fitters, then it works.

The hearing aids are the type II medical equipments in China, so there is severe licensing system on the industry service.

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