

Imaging in children with hearing loss

Bildgebende Diagnostik bei Schwerhörigkeit im Kindesalter

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ABSTRACT

Background Since the introduction of hearing screening in Germany in 2009, pediatric hearing disorders are detected at an early stage. Early therapy is essential for language development. Imaging plays a central role in diagnosis and therapy planning.

Method Imaging findings of the most relevant causes of pediatric hearing disorders are presented. Specific attention is given to the method used in each case – CT or MRI.

Results and Conclusions While CT is the method of choice for conductive hearing loss, a combination of CT and MRI with high-resolution T2-3D sequences has been established as the best diagnostic method for sensorineural hearing loss.

The most common causes of conductive hearing loss in childhood are chronic inflammation and cholesteatoma. Congenital malformations of the outer or middle ear are less frequent. In the case of sensorineural hearing loss, the cause is located in the inner ear and/or the cochlear nerve or the cerebrum. In these cases, congenital malformations are the most common cause.

Key Points

- CT and MRI are necessary to identify morphological causes of hearing disorders and to clarify the possibility of hearing-improving ear surgery or cochlear implantation.
- Contraindications for surgical procedures must be excluded.
- Anatomical variants that may be risk factors for surgery must be described.

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ZUSAMMENFASSUNG

Hintergrund Seit Einführung des Hörscreenings 2009 in Deutschland werden kindliche Hörstörungen frühzeitig erkannt. Eine frühe Therapie ist für die Sprachentwicklung essenziell. Die bildgebende Diagnostik spielt eine zentrale Rolle in der Diagnostik und Therapieplanung.

Methode Bildgebende Befunde der wichtigsten Ursachen einer kindlichen Hörstörung werden vorgestellt. Dabei wird gezielt auf die jeweils zur Anwendung kommende Methode – CT oder MRT – eingegangen.

Ergebnisse und Schlussfolgerungen Während bei einer Schallleitungsstörung die CT die Methode der Wahl ist und in den allermeisten Fällen auch ausreicht, hat sich bei der Schallempfindungsschwerhörigkeit eine Kombination aus CT und MRT mit hochauflösenden T2-3D-Sequenzen als bestmögliche Diagnostik etabliert. Die häufigsten Ursachen einer Schallleitungs-schwerhörigkeit im Kindesalter sind chronische Entzündungen und Cholesteatome, seltener angeborene Fehlbildungen des Außen- oder Mittelohres. Bei einer Schallempfindungsstörung liegt die Ursache im Innenohr und/oder dem Nervus cochlearis oder dem Großhirn. Hier sind angeborene Fehlbildungen die häufigste Ursache.

Introduction

Hearing screening in newborns has been required by law in Germany since January 1, 2009. Consequently, pediatric hearing disorders can be detected systematically at an early stage. Permanent or longer-term pediatric hearing impairment affects language development [1]. Therefore, it is important for these children to undergo a hearing-improving intervention in a timely manner.

Hearing loss can be classified as follows [2]:

- **Conductive hearing loss (CHL)** is the result of a problem transferring sound waves to the inner ear: The pathophysiology is usually mechanical and affects the structures of the pinna, the external auditory canal, the eardrum, and the middle ear, particularly the auditory ossicles. CHL is usually acquired.
- **Sensorineural hearing loss (SNHL)** is caused by a problem transferring nerve impulses from the inner ear to the brain: The pathophysiological cause is thus the structures of the inner ear, particularly the cochlea, and the cochlear nerve. In contrast to CHL, SNHL in children is typically congenital.
- **Combined hearing loss** is a combination of CHL and SNHL.

Diagnostic imaging

Both MRI and CT play a central role in the diagnosis of pediatric hearing disorders. Imaging is used not only to determine the etiology but also to plan the optimal treatment [3, 4].

Note

The objectives of imaging in the case of hearing loss in early childhood are [2]

- Identification of possible structural causes of the hearing loss
- Search for additional anomalies that can help to diagnose an underlying syndrome
- Evaluation of whether a hearing-improving ear surgery or cochlear implant (CI) would be promising
- Determination of anatomical risk factors in connection with surgical planning

CT is the only established imaging method for the diagnosis of CHL. In contrast, the optimal imaging modality for SNHL is a topic of discussion in the literature [5–7].

MRI and CT provide complementary information: The external auditory canal, middle ear, and the bony labyrinth including the internal auditory canal can be evaluated with CT. MRI allows direct evaluation of the cochlear nerve (CN) and the fluid compartments of the inner ear, as well as assessment of the brain structures and thus the diagnosis of cerebral comorbidities.

Note

MRI and CT together offer the greatest reliability for the diagnostic imaging of sensorineural hearing loss and should therefore be used in combination in the ideal case.

CT

A dedicated high-resolution technique is needed to achieve an appropriate level of detail regarding the petrous bone. Imaging is performed using fine collimation and a submillimeter slice thickness. Oblique coronary and oblique sagittal reconstructions are generated from the primary axial images [8]. One disadvantage of CT is the use of ionizing radiation in the pediatric population. However, performing a low-dose CT examination of the temporal bone to reduce the radiation dose can be problematic since high image quality with the best possible resolution is needed to evaluate the tiny structures [8]. However, in the case of isotropic imaging, it is usually possible to position the head of the child on an incline so that the radiosensitive ocular lenses are outside the path of primary X-ray beam.

At our facility, CT is performed as an axial spiral acquisition at 120 kV and 200 mA on a 128-slice scanner (Ingenuity, Philips Amsterdam, The Netherlands) with a collimation of 0.625 mm and a slice thickness of 0.67 mm.

MRI

While MRI provides significantly worse visualization of the bony structures of the inner ear, it is clearly superior to CT in the evaluation of the auditory nerve due to the excellent soft-tissue contrast. Optimal visualization of the CN requires a high-resolution, thin-slice, T2-weighted sequence. This sequence also provides detailed information about the fluid-filled labyrinth and can visualize relevant anomalies of the inner ear. In general, MR images are acquired on the axial plane. A 3D acquisition method allows generation of almost isometric voxels, thus allowing a diagnostically important secondary reconstruction perpendicular to the course of the CN in the region of the internal auditory canal.

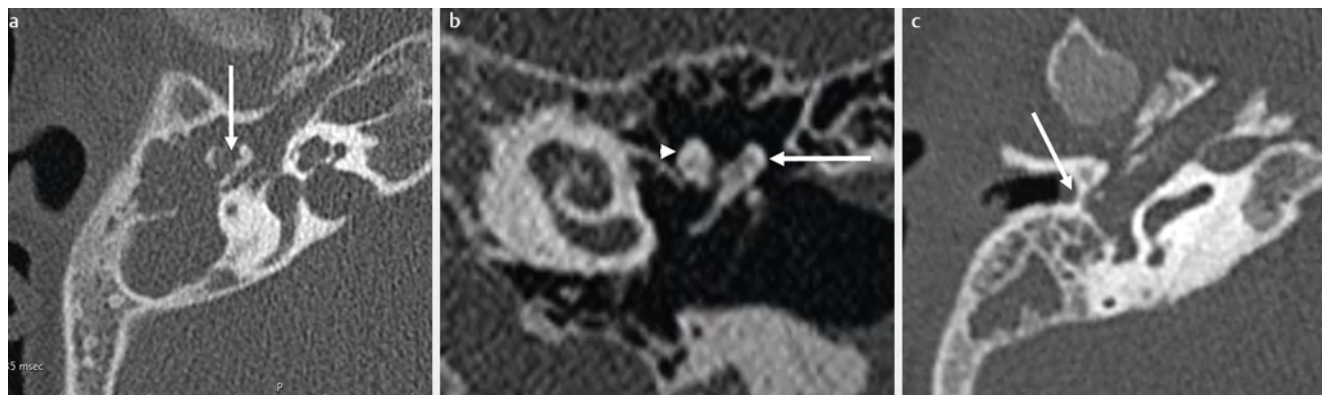
At our facility, we use 3D-SPACE (Sampling Perfection with Application optimized Contrast using different flip angle Evolution) sequences on the axial plane with $0.25 \times 0.25 \times 0.35$ (interpolated from $0.5 \times 0.5 \times 0.4$ mm) voxel size (3T, Prisma Fit; Siemens, Erlangen, Germany). Special 3D gradient echo sequences, like CISS (constructive interference in steady state), are also suitable for visualizing the structures of the ear on MRI.

In addition, to evaluate the brain and in particular to rule out malformations, at least one T2-weighted sequence of the entire cranium should be acquired. Contrast administration is only necessary in the case of justified suspicion of a schwannoma in the case of a predisposing primary disease, e. g., neurofibromatosis 2.

Due to the long duration of the examination and the necessity for vibration-free images, sedation is often needed in children up to preschool age.

Conductive hearing loss

The most common cause of acquired conductive hearing loss in infants and toddlers is acute **otitis media**. Due to fluid collections in the tympanum, the mobility of the eardrum and the auditory ossicles is limited, thus affecting the transfer of sound to the inner



► **Fig. 1** Conductive hearing loss, CT. **a** 2-year-old boy, congenital cholesteatoma: tympanic cavity and parts of the mastoid are filled with soft tissue, ossicles, tympanic cavity wall and mastoid cells are destroyed (arrow). **b** 4-year-old boy, 1 year after petrous bone fracture, coronary reconstruction: dislocation of the incudomalleolar joint- the malleus head (arrowhead) is clearly medial to the incus (arrow), the socket is empty. **c** 1-year-old boy: Hypoplasia of the external auditory canal, atresia of the tympanic membrane (arrow), non-ventilated, hypoplastic tympanic cavity.

ear. This type of CHL is characterized by intermittent mild to moderate hearing loss that is usually reversible after the infection resolves and does not require any diagnostic imaging. Imaging is only indicated in the case of clinical suspicion of a complication of otitis media, e. g., in the case of peripheral facial paresis, signs of meningitis, or symptoms of sinus vein thrombosis, and the onset of rotational vertigo or indications of acute involvement of the inner ear.

Persistent acquired CHL is an indication for cross-sectional imaging. It can be caused by chronic otitis media with perforation of the eardrum, cholesteatoma, or trauma.

A cholesteatoma is a collection of keratinized squamous epithelium and is often referred to as “skin at the wrong location”. The disruption in sound transmission can be the result of the mass effect or erosion of the ossicles [9]. In children, it is necessary to differentiate between congenital and acquired types.

Congenital cholesteatomas, which are very rare, develop from residual embryonic epithelial cells, which can be found everywhere in the temporal bone, e. g., in the middle ear, but also in the eardrum, in the external auditory canal, and in the mastoid. They can be associated with atresia of the external auditory canal but can also occur in otherwise healthy children. If the diagnosis is made too late, irreversible destruction of the auditory ossicles and the surrounding bony structures can occur.

Note

Congenital cholesteatomas can occur at any location in the petrous bone.

Due to the pathogenesis, acquired cholesteatomas can be divided into retraction pocket cholesteatomas and non-retraction pocket cholesteatomas. Retraction pocket cholesteatomas occur as a result of hypoventilation and inflammatory processes of the middle ear resulting in retraction of the eardrum in the direction of the middle ear. Non-retraction pocket cholesteatomas can occur secondary to the migration of epithelial cells from the external auditory canal through a perforated eardrum (inflammatory, post-traumatic, or iatrogenic) into the tympanum.

On CT, a cholesteatoma presents with soft tissue attenuation, typically in connection with bone erosion in the region of the ossicles, the scutum, or the tympanum (► **Fig. 1a**). However, a lack of erosion does not exclude a cholesteatoma. Therefore, reliable differentiation between a cholesteatoma and granulation tissue, effusions, and other soft-tissue lesions in a chronically inflamed middle ear or mastoid is not always possible based solely on CT.

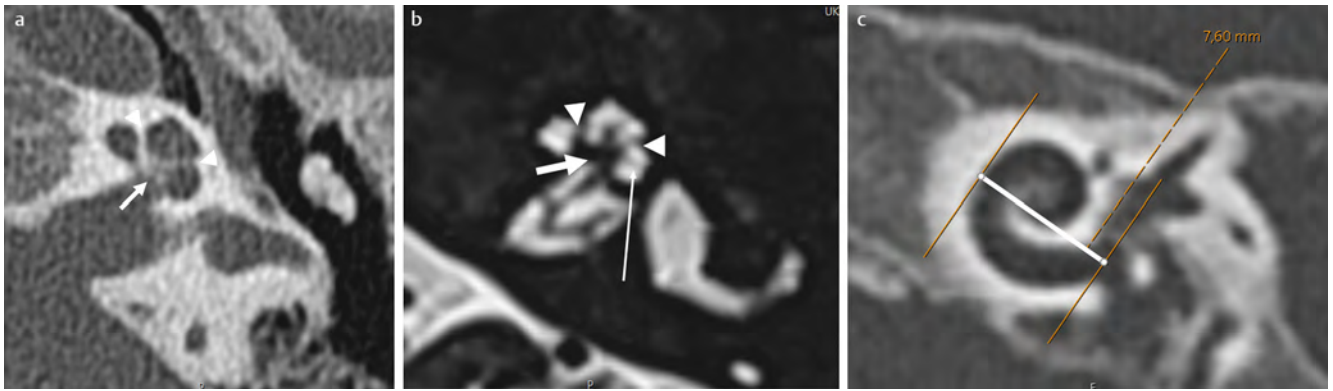
Diffusion-weighted (DWI) MRI sequences have proven to be helpful for the differentiation between cholesteatomas and non-cholesteatoma soft tissue since cholesteatomas have diffusion restriction. The specificity of diffusion restriction in cholesteatomas greater than 5 mm in size is up to 100% in the literature [10]. The typical pronounced artifacts on conventional echoplanar DWI sequences at the base of the skull can be significantly reduced by using alternative read-out strategies, like TSW-DWI, non-EPI (HASTE) DWI or RESOLVE-DWI [11, 12].

Note

On CT, cholesteatomas are seen as tissue with soft tissue attenuation, typically with bone erosion. If the latter is absent, the detection of diffusion restriction can confirm the diagnosis.

In the case of head trauma with petrous bone fracture, fractures or luxations in the region of the auditory ossicles can occur. The pathology is often initially not able to be detected due to the secretory otitis media. Therefore, in the case of persistent CHL after trauma, a follow-up CT examination should be performed.

Rare congenital causes of CHL are **malformations** of the outer and middle ear. This includes hypoplasia or atresia of the external auditory canal, the eardrum, or the tympanum and malformations of the auditory ossicles (► **Fig. 1c**). Such malformations can be isolated or can occur in combination and often as part of a syndrome. The inner ear and the internal auditory canal are typically not affected in these patients due to their different embryological origin.



► **Fig. 2** Normal anatomy and internal structure of the cochlea: Both CT **a** and MRI **b** show the modiolus (arrow) and the interscalar septum (arrowheads) well. The lamina spiralis is more visible on MRI than on CT due to the high soft-tissue contrast (long arrow). **c** The oblique sagittal reconstruction allows the CDL to be determined by measuring the distance between the center of the round window and the opposite boundary of the cochlea.

Sensorineural hearing loss

Pediatric SNHL can be unilateral or bilateral and is usually congenital. The prevalence of congenital SNHL is 1–2 per 1000 live births [13]. Unilateral SNHL is significantly more frequently associated with malformations of the inner ear than in the case of a bilateral presentation [14].

The *cochlea* is the primary hearing organ. The modiolus is a central bony pillar around which the cochlea turns 2.5 times and that can be seen as a structure with bone density on CT and as a hypoechoic structure on MRI (► **Fig. 2a, b**). The spiral lamina projects from the modiolus and divides the cochlea into an upper (scala vestibuli) and a lower (scala tympani) compartment. The scala tympani is the target site for cochlear implantation (CI).

The goal of imaging is to rule out cochlear anomalies that could limit the success of CI. Therefore, the presence of the 2.5 turns should be evaluated and the required length of the electrode must be determined. The cochlear duct length (CDL) is calculated from the CT scan using a simplified formula (► **Fig. 2c**).

Moreover, particularly after meningitis, sclerosis of the cochlea, which can be identified based on the hypointense signal on MRI and calcifications on CT, must be ruled out. Masses in the cochlea, e. g., intracochlear schwannomas, can very rarely be detected on MRI.

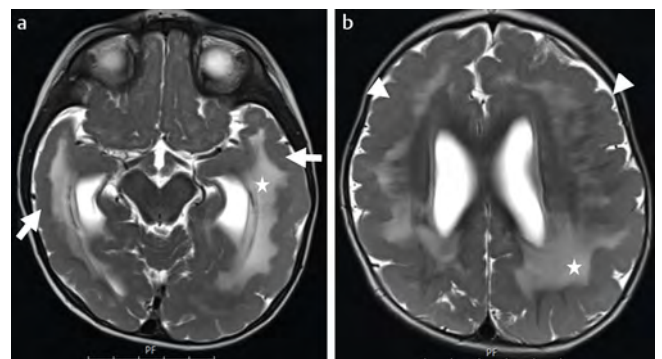
Note

The CT report should explicitly describe the morphology of the cochlea, the number of turns, and the presence of the inner septation, the spiral lamina, and the modiolus [15].

Imaging findings when diagnosing sensorineural hearing loss

Brain

Hearing disorders can be caused by brain anomalies that are often caused by a prenatal infection. These often result in structural changes in the brain that can be seen on MRI, e. g., leukoencephalopathy, calcifications, or gyration disorders (► **Fig. 3**). The diagnosis of such pathologies helps with the etiological workup of hearing loss and can also provide prognostic information regard-



► **Fig. 3** Congenital CMV infection, SNHL, 1-year-old girl, MRI T2 weighting: Severe gyration disorder with pachygyri temporal (short arrows), polymicrogyri frontal (arrowheads), and marked leukoencephalopathy (asterisk).

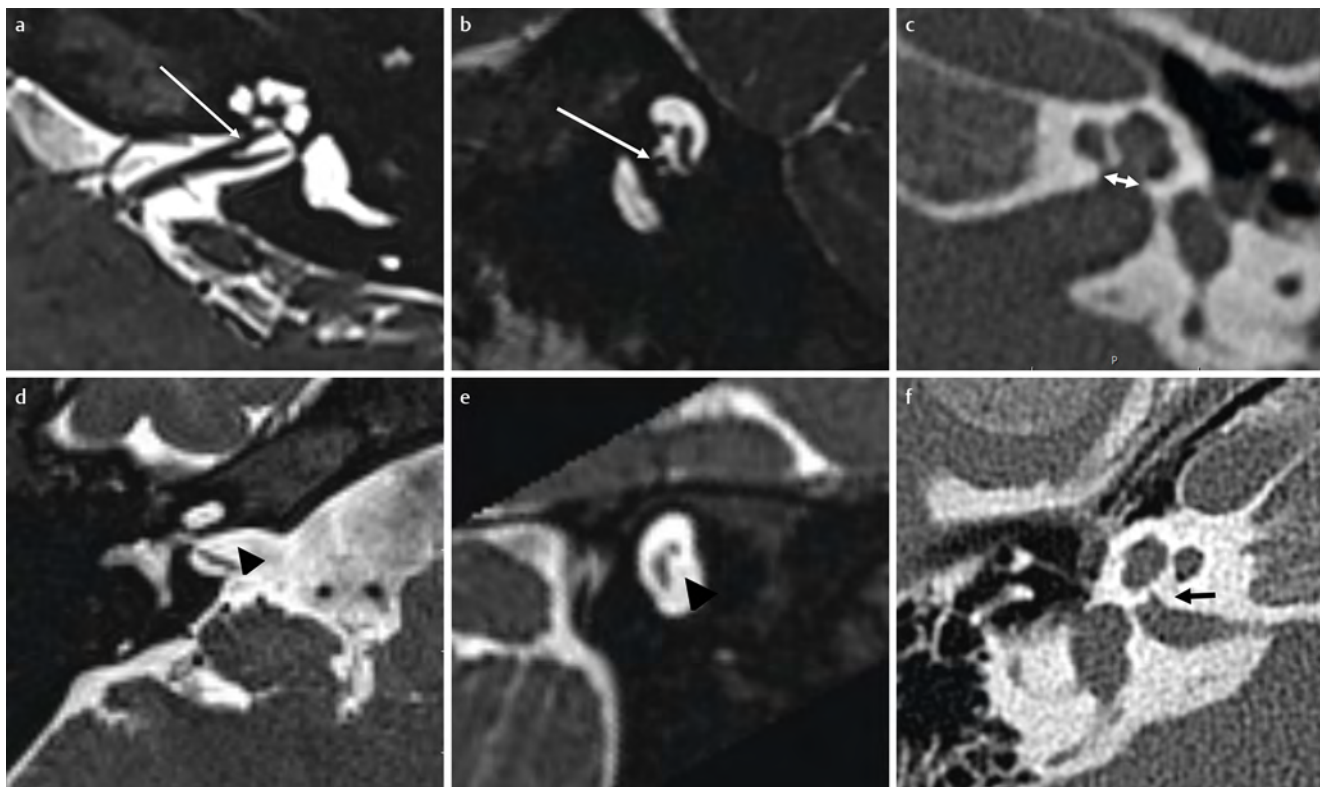
ing the success of CI since this largely depends on the subsequent cooperation of the patient, particularly during the rehabilitation phase.

Vestibulocochlear nerve

Evaluation of the vestibulocochlear nerve and especially the cochlear nerve requires particular attention since aplasia of the cochlear nerve is a contraindication for implantation of a cochlear implant. In contrast, hypoplasia is only a relative contraindication but affects the prognosis.

It is recommended to generate parasagittal reconstructions perpendicular to the course of the cochlear nerve from the thin-slice isovoxel MRI sequences. The cochlear nerve is seen in the anterior lower quadrant of the internal auditory canal (IAC). In the normal case, the vestibulocochlear nerve is approximately twice as thick as the facial nerve. After it divides into the vestibular and cochlear portions, the cochlear nerve is at least as thick as the overlying facial nerve (► **Fig. 4a, b**). If the caliber is smaller, hypoplasia must be assumed [8]. The cochlear nerve canal (CNC) can be easily visualized on CT (► **Fig. 4c**).

Aplasia of the cochlear nerve can be missed in transverse slices, particularly in the case of a hypoplastic internal auditory canal



► **Fig. 4** Top row: Inconspicuous cochlear nerve on the left (arrows) **a** MRI CISS sequence transverse, **b** reconstruction perpendicular to the internal auditory canal, **c** CT shows a normal width of the CNC (>1.9 mm, double arrow). Bottom row: Aplasia of the right cochlear nerve: It does not show up on MRI in the transverse slice **d**, nor in the oblique sagittal reconstruction **e** (arrowheads). **f** CT shows a bony occlusion of the CNC (black arrow) = "isolated cochlea", additionally hypoplasia of the IAC.

(IAC). In parasagittal reconstructions, this can be diagnosed with great certainty by visualizing only 3 instead of 4 nerve structures (► **Fig. 4d, e**).

However, the finding is not always so definitive. A narrow IAC or artifacts, e. g., due to movement or dental braces, can limit the ability to evaluate MRI images. CT can also be helpful in the evaluation of cochlear nerve deficiency (CND): If the CNC is narrower than 1.9 mm at the widest location, cochlear nerve deficiency is highly probable [16]. If the CNC is completely occluded by a bony protrusion, it is referred to as an isolated cochlea and is always associated with a CND (► **Fig. 4f**).

Internal auditory canal

Hypoplasia of the IAC can also be indicative of a cochlear nerve deficiency, but the correlation is significantly less pronounced than in the CNC [16, 17] so that this finding is less relevant in practice. Aplasia can occur, for example, as part of Michels syndrome (see below).

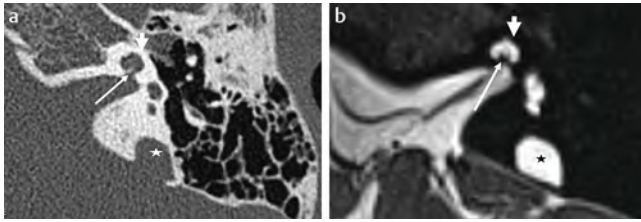
Inner ear

The inner ear begins developing already in the third week of pregnancy. In the eighth week, the cochlea is fully developed. The vestibulum is fully developed in the 11th week and the semicircular canals in the 19th to the 22nd week. The bony labyrinth is fully developed in the 23rd week [18].

The earlier a problem occurs in the embryonic period the more severe the deformity, resulting in varying degrees of differentiation of the labyrinth.

According to Sennaroglu [19], inner ear malformations can be classified as follows:

- Aplasia of the labyrinth (Michel deformity): This is the most severe type of inner ear malformation and is the result of a developmental problem in the third gestational week. It is characterized by the complete absence of the inner ear structures and the cochlear nerve. The IAC is hypoplastic or aplastic. Additional anomalies in the region of the middle ear and the base of the skull and relating to the course of vessels are often seen.
- Cochlear aplasia: This anomaly occurs approximately in the fourth gestational week. The cochlea is completely absent, and the region appears sclerotic. The vestibulum and semicircular canals can be hypoplastic, dilated, or normal. The finding must be differentiated from inflammatory ossification based on the absence of the promontorium.
- Common cavity: If the problem occurs at the start of the fifth gestational week, there is no differentiation between the vestibulum and the cochlea. They form a common cavity.
- Incomplete partition type I (IP-I): In the case of a developmental problem at the end of the fifth gestational week, the vestibulum and cochlea are normal in size and differentiated from one another, but the internal architecture is absent. The cochlea is an empty cyst and there is no identifiable modiolus.



► **Fig. 5** 12-year-old girl with branchio-oto-renal syndrome and progressive mixed hearing loss: Incomplete partition type II (Mondini malformation), **a** CT, **b** MRI. Lack of differentiation between middle and apical convolutions (short arrows), preserved modiolus (thin arrows). Massively dilated aquaeductus vestibuli (asterisk) in the sense of an LVAS.

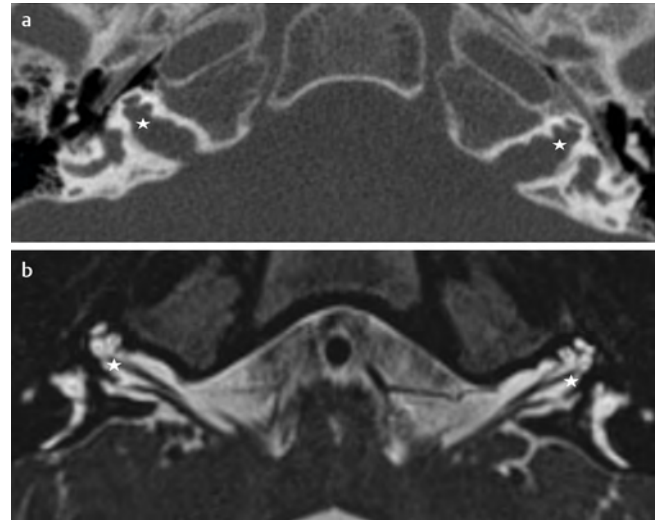
- Incomplete partition type II (IP-II, Mondini malformation): If the problem occurs in the seventh gestational week, the basal turn and the modiolus are present. However, the middle and apical turns cannot be differentiated from one another and the interscalar septum and spiral lamina are absent. The vestibulum and vestibular aqueduct are often dilated. IP-II is the most common form of cochlear malformation (50%) [20] and is often seen as part of complex malformations (► **Fig. 5**). In contrast to the malformations described so far, IP-II is not an absolute contraindication to CI.

X-linked deafness (perilymphatic gusher phenomenon) is classified as incomplete partition type III (IP-III). As a result of an atypical connection between the internal auditory canal and the basal turn of the cochlea, the intracranial pressure is transferred to the endolymph. Imaging shows an absent modiolus and a bulbous dilation of the fundus of the IAC that has a broad connection to the basal turn of the cochlea. The cochlea has a corkscrew-like appearance (► **Fig. 6**). Clinical signs of the gusher phenomenon include pronounced hearing impairment in the low-frequency range and hearing loss at a very young age (also see Section 6).

Dysplasia and aplasia of the equilibrium organ do not play an immediate role in hearing loss but often occur as part of complex malformation syndromes or inner ear malformations, e. g., in CHARGE syndrome, and should trigger an intensive search for malformations in the region of the cochlea.

Large vestibular aqueduct syndrome (LVAS) refers to dilation of the bony aqueduct (visible on CT) or the endolymphatic duct (visible on MRI). The latter arises from the posterior vestibulum and runs through the petrous bone to the endolymphatic sac, which ends in the epidural space of the posterior cranial fossa.

Normally, the aqueduct is not wider than the posterior semicircular canal and has a diameter < 1.5 mm [21]. It is considered dilated when the middle third is ≥ 1 mm on axial slices or the opening measures ≥ 2 mm [22, 23] (► **Fig. 5**). A dilated aqueduct can be an isolated occurrence, but in most cases (approx. 85%) it is combined with other malformation or occurs as part of a malformation syndrome [20].



► **Fig. 6** 5-year-old boy with X-linked hearing loss **a** CT, **b** MRI: Incomplete partition type III: The basal cochlear turn has a wide connection to the internal auditory canal (asterisk), resulting in a “corkscrew-like” image impression of the cochlea. The modiolus is missing.

LVAS manifests as sensorineural hearing loss or mixed, typically fluctuating and/or progressive, hearing loss in childhood. It is caused by a transfer of intracranial pressure to the labyrinth resulting in damage to the hair cells. Hearing can still be normal at birth. The first symptoms often appear suddenly, either spontaneously or in connection with mild head trauma [20]. There is a significant correlation between the extent of dilation of the aqueduct and the degree of hearing loss [24]. Physical activity resulting in significant fluctuations in pressure, e. g., jumping on a trampoline and headers in soccer, should therefore be avoided.

There were various attempts in the past to take into account the complexity of inner ear malformations and develop a feasible classification system [19]. The most recent classification is the INCAV system with the letters indicating the various parts of the inner ear: Internal auditory canal (I), cochlear Nerve (N), Cochlea (C), vestibular Aqueduct (A), and Vestibulum (V) [25] (► **Table 1**). As a result of this structured approach, malformations of the inner ear structures can be viewed individually, with a category and a number for categorization being assigned to every ear structure. Zero “0” means that the inner ear component is normal. Higher numbers indicate the severity of the malformation. This classification is performed individually for each ear. Thus, I0N0C0A0V0 indicates a normal finding. I3, N3, and C6 are absolute contraindications for CI. These patients can be easily identified at a glance. The classification can be used for MRI and CT. On CT a narrow CNC as a surrogate for a CND is classified as “CX”.

► **Table 1** INCAV system for classifying inner ear malformations according to Adibelli et al. 2017 [25].

INCAV classification (MRI)	0	1	2	3	4	5	6
Internal auditory canal (I)	Normal	Dilated	Narrow	Atresia			
Cochlear nerve (N)	Normal	Thickened	Hypoplasia	Aplasia			
Cochlea (C)	Normal	IP-II	IP-III	Hypoplasia	IP-I	Common cavity	Aplasia
Vestibular aqueduct (A)	Normal	Dilated					
Vestibulum (V)	Normal	Malformation of the semicircular canal	Dilated	Hypoplasia		Common cavity	Aplasia

External auditory canal, tympanum, and mastoid

CT evaluation of the bony structures is decisive for surgical CI planning also in the case of SNHL. A partial mastoidectomy is performed in order to place the CI electrode. Therefore, prior evaluation of the pneumatization of the mastoid is important. Visualization of the tympanic course of the facial nerve is important since anomalies in this region can result in serious surgical complications as a result of injury to the facial nerve. Multiplanar reconstructions perpendicular to the tympanic course of the nerve are suitable for this. The caudal bony boundary of the facial canal can be effectively evaluated (► **Fig. 7**).

It continues to be important to identify anomalies and bony protrusions into the round window through which the electrode is inserted into the cochlea in the case of CI.

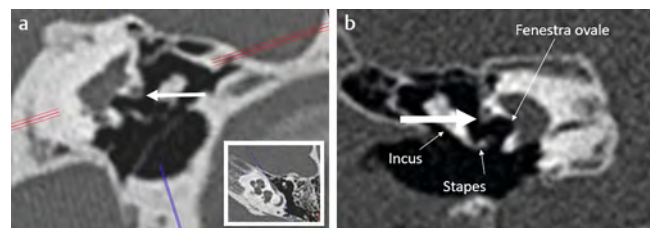
Anatomical variants that are important for surgical planning

Some anatomical variants are extremely important for the planning of cochlear implantation and must be mentioned in the report. These are, for example, an atypical course of the carotid artery, a high-riding or dehiscent jugular bulb, or a protrusion of the sigmoid sinus into the mastoid (► **Fig. 8a**).

An atypical connection between the basal cochlear turn and the internal auditory canal can occur not only in connection with X-linked hearing loss but also as an isolated anatomical variant. During surgery, this can result in a massive loss of spinal fluid in terms of a gusher phenomenon, particularly in the case of opening of the footplate, e. g., in the case of suspected otosclerosis. In spite of sufficient surgical closure, sensorineural hearing loss can occur. Persistence of the CSF leak results in deafness and poses a risk of meningitis.

Since this anomaly is not always visible on axial CT scans, Varadarajan et al. [26] recommend oblique multiplanar reconstructions along an axis between the apical cochlear turn and the center of the IAC (► **Fig. 8b, c**).

Hearing-improving surgical procedures in addition to CI are available. These include various implantable hearing systems that use sound conduction via bone (BoneBridge, OSIA, BAHA). An interdisciplinary conference with phoniatrians, pediatric audiologists, surgically active ENT physicians, and specialized radiologists has become established for selecting the suitable surgical method, surgical planning, and subsequent rehabilitation and for



► **Fig. 7** Tympanic course of the facial nerve: An oblique coronal reconstruction perpendicular to the course of the nerve allows the assessment of the bone cover **a**: Normal findings. The facial canal is covered by bones in the caudal direction (arrow), **b**: The nerve hangs far into the tympanic cavity (thick arrow), the bony cover is missing.

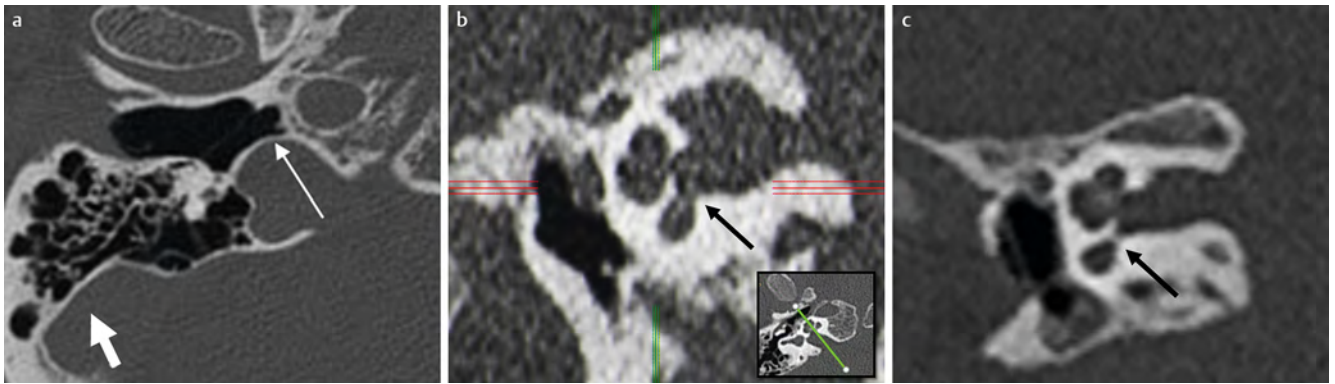
ensuring mutual understanding between the individual disciplines. These interdisciplinary conferences are now required by quality management when certifying CI centers.

Summary

High-resolution CT and MRI play an important role in the evaluation of hearing loss in children. CT is the method of choice for the evaluation of bony anomalies in the region of the middle ear and the labyrinth. It is important for diagnosis and surgical planning with regard to both conductive hearing loss and sensorineural hearing loss. MRI provides information about the membranous labyrinth, the vestibulocochlear nerve, and the brain, which is essential for the diagnosis and treatment of sensorineural hearing loss. MRI and CT in combination provide the highest possible diagnostic accuracy.

The goal of imaging is to visualize the anatomy and to identify malformations causing the hearing loss. At the same time, contraindications for CI must be ruled out and anatomical variants that could lead to surgical complications must be identified.

An understanding of the embryogenesis and anatomy of the inner ear in connection with standardized classification systems for inner ear malformations is important for describing and interpreting CT and MRI images. An interdisciplinary case conference with phoniatrians, pediatric audiologists, surgically active ENT physicians, and specialized radiologists has become established



► **Fig. 8** Variations of anatomy important for surgical planning, CT, **a**: Protrusion of the sigmoid sinus into the petrous bone (short arrow) and of the bulbous venae jugularis into the tympanic cavity (long arrow), **b**: Atypical connection between the basal turn and the internal auditory canal visible only in the oblique reconstruction (small image) (arrow), **c**: for comparison, normal findings – complete bony separation of the basal turn and the IAC (arrow).

for selecting the suitable type of treatment and discussing surgically relevant findings and is the standard at certified CI centers.

Conflict of Interest

The authors declare that they have no conflict of interest.

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