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Research Article

CONGENITAL MALFORMATION EXTERNAL AUDITORY CANAL

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ABSTRACT

In contrast to anatomical variants, congenital malformations - synonymous with anomalies or dysplasia - are characterized by deviations not only from normal anatomical development but also from regular function. They can result from delayed development, abnormal embryogenesis or both due to spontaneous genetic mutations - this occurs in most congenital malformations of the outer and middle ear - genetic transmission and exogenous factors in about 10% of cases.

KEYWORDS

Congenital atresia, external auditory meatus, stenosis.

INTRODUCTION

Abnormalities of the outer ear are common and occur in approximately 5% of the general population. The most common malformations include a combined malformation of the outer and middle ear, called congenital aural atresia. 50% of ENT malformations are anomalies of the ear. The malformations of the outer ear most often affect the right ear (58-61%) and most cases (ca. 70-90%) are unilateral.

The reported prevalence varies by region of the world, from 0.83 to 17.4 per 10,000 births, and the prevalence is thought to be higher in Latin Americans, Asians and Native Americans. 1: 3,800 newborns. The incidence of malformations of the external ear has been reported from 1 in 6,000 newborns, to 1 in 6,830 newborns. Severe malformations occurred in 1: 10,000 to 1: 20,000 neonates [4], severe malformations or aplasia in 1: 17,500 neonates.

The prevalence of microtia is above 3: 10,000 according to M. Schloss . Congenital atresia of the external auditory canal occurs in 80% of patients with microtia .

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Malformations can affect the outer ear (auricle and external auditory canal), the middle ear and the inner ear, often in combination. J. Swartz and E. Faerber reported that the incidence of inner ear malformations is 11-30% in persons with external and middle ear malformations.

However, different embryogenesis of the outer/median ear and inner ear resulted in malformations of the outer and/or middle ear without malformations of the inner ear and vice versa. There are no prospective data on the incidence of middle ear malformations with or without auricular malformations . According to S. Ishimoto. auricular malformations of grade 1 (classified according to Marx) showed additional malformations of varying degrees and frequency, affecting the ossicles (6-33%), the round and oval window (6-15%), pneumatisation of the mastoid process (15%), the facial nerve passage (36%) and the external auditory canal (42%). The combined malformation of the ear, known as atresia auris congenita, including anomaly of the outer and middle ear (and rarely the inner ear: about 10% according to J. Swartz and E. Faerber), with a characteristic finding of atresia of the external auditory canal, has an incidence of 1:10,000; in 15-20% of cases bilateral malformations occurred. Males have a higher incidence than females, and atresia affects the right ear more often than the left. Unilateral atresia of the external auditory canal is much more common than bilateral atresia (3-7 times difference) .

Ear development is a continuous, complex process that begins early in embryonic life in three parts and is completed before the first decade.

Defects in the ear can be genetic or acquired. Among congenital malformations, 30% are associated with syndromes with additional malformations and/or functional loss of organs and organ systems. Examples are otofacial dysostosis (e.g. Treacher-Collins syndrome, Goldenhar syndrome), craniofacial dysostosis (e.g. Crouzon syndrome, Aper syndrome), otocervical dysostosis (e.g. Klippel-Feil syndrome, Wilderwank syndrome), otoskeletal dysostosis (e.g. Van der Huwe de Klein syndrome, Albers-Schönberg syndrome) and chromosomal syndromes such as trisomy 13 (Paetau syndrome), trisomy 18 (Edwards syndrome), trisomy 21 (Down syndrome) and 18q syndrome. Non-syndromic ear malformations show only ear anomalies without any other malformations have published a detailed list of syndromes and conditions associated with congenital ear malformations. In all genetically determined malformations (syndromic and non-syndromic), a high frequency of spontaneous genetic mutations can be assumed. Numerous studies, especially those of the inner ear, have identified various genes, transcription factors, secretion factors, growth factors, receptors, cell adhesion proteins and other molecules as responsible for ear malformations.

Congenital ear malformations with an obvious family history show an autosomal dominant inheritance of about 9%, an autosomal recessive inheritance of about 90%, and an X-linked inheritance of about 1%. Non-syndromic congenital hearing loss has a completely different distribution: autosomal dominant inheritance in about 30% of cases, autosomal recessive in about 70%, X-linked inheritance in about 2-3% and sometimes mitochondrial-linked inheritance . On the other hand, in patients with familial non-syndromic high grade microtia, Katzbach et al. reported a predominantly autosomal dominant heritability with variable penetrance. Acquired ear malformations result from exogenous damage during pregnancy.

Noxae include infections (primarily viral: confirmed for rubella, cytomegalovirus and herpes simplex virus,

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possible measles, mumps, hepatitis, polio, chickenpox, coxsackieviruses and ECHO viruses, toxoplasmosis and syphilis), chemical agents, malnutrition, radiation exposure, Rh incompatibility, hypoxia, variations in air pressure and noise exposure.

Bleeding occurring in the first half of pregnancy and metabolic disorders such as diabetes should be considered. Among chemical teratogens, drugs play a predominant role, a prime example being thalidomide, which led to a significant increase in malformations in the early 1960s. Quinine and aminoglycoside antibiotics also cause malformations. Cytostatics and drugs used in the treatment of epilepsy (e.g. diphenylhydantoin, trimetadione and valproic acid) may also be a cause. Both excessive doses of retinoic acid (retinoic acid embryopathy) and vitamin A deficiency (VAD syndrome) during pregnancy can cause ear malformations .

Many additional drugs that cause malformations have been suggested, as well as hormones, drugs, alcohol and nicotine. Environmental agents such as herbicides, mercury-containing fungicides and lead can have teratogenic effects. However, in many cases the actual cause is unknown, because not all patients with possible or suspected genetic malformations can be genetically analysed, many of the responsible genes are still unknown, and anamnestic or clinical data regarding exogenous influences may be unclear or absent. Therefore, it is not surprising that the reported proportions of genetically determined and non-genetically determined ear malformations vary widely. For example, in the combined ear malformation described as atresia auris congenita, a non-syndromic hereditary etiology without association with a syndrome was reported in 20% of cases, association with a syndrome in 10%, and an acquired nature resulting from teratogenic exposure in 20% of cases. . 0.05%; thus, it is assumed that spontaneous genetic mutations are responsible for the majority of these malformations. In contrast, the estimated contribution of exogenous factors to malformations of the outer ear (especially the auricle) is 10%.

Many classifications of ear malformations have been proposed. These classifications should facilitate a standardised clinical description of the findings and should serve as a prognostic basis for treatment interventions and their comparison. Over time, classification systems have become more detailed, especially due to modern imaging techniques such as CT and MRI. Thus, a single classification for all ear malformations seems inappropriate. Roughly speaking, it can be stated that, normally, there is a correlation between the degree of malformation of the external auditory canal and the middle ear with a corresponding conductive hearing loss. Nevertheless, normal auricles with atresia of the external auditory canal and, although rare, microtia in combination with a normal external auditory canal and a normal tympanic cavity have been reported.

Abnormal ear development leads to malformations of the external auditory canal, including membranous and/or bony atresia. If this process is incomplete, it can lead to stenosis of the cartilaginous part of the canal laterally with a more normal diameter of the bone canal and tympanic membrane medially. This condition predisposes to the formation of a cholesteatoma in the external auditory canal. Clinically, malformations of the external auditory canal can be atretic (aplastic) or hypoplastic (stenotic). Several classifications have been proposed for malformations of the external auditory canal, based on various parameters including clinical examination, radiological, surgical and histopathological findings.

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The Weerda classification of malformations of the external auditory canal includes three types:

Type A- represents a marked narrowing (stenosis) of the external auditory canal along with an intact skin layer;

Type B- represents partial development of the lateral part of the external auditory canal and atresia in the medial part;

Type C- involves complete bony atresia of the external auditory canal.

Middle ear malformations may affect the normal development of the tympanic cavity as well as the ossicles. This is due to changes in the configuration or size of the middle ear cavities as well as in the number, size, and configuration of the ossicles. There may be abnormalities of the oval window and, less frequently, of the round window. Many classifications have been described. The closely interrelated development of the external auditory canal and the middle ear has led to the classification of a combined malformation called atresia auris congenita by Altmann (32). Three degrees of severity are described:

I-type: mild deformity of the external auditory canal, normal or slightly hypoplastic tympanic cavity, deformed auditory ossicles and well ventilated mastoid process are noted;

Type II: With blind or missing external auditory canal, narrow tympanic cavity, deformed and fixed ossicles, less pneumatisation of the mastoid process;

Type III: no external auditory canal, middle ear hypoplastic, the ossicles severely deformed, and there is also inhibition of pneumatisation of the mastoid process cells.

If only a middle ear malformation is present, three degrees of severity (mild, moderate, severe) can be differentiated, similar to the changes described in the Altman classification relating to the middle ear. Congenital fixation of the stapes can occur in isolation and be the result of bone plates, aplasia or dysplasia of the annular ligament. Various other changes are regarded as malformations of the middle ear. These can only be listed: liquor-mediated middle ear fistulas, congenital cholesteatoma (congenital epidermoid), congenital dermoids and malformations of the middle ear muscles.Классификация атрезий слухового прохода по H.F. Schuknecht (1993):

Type A - atresia in the cartilaginous part of the ear canal, Grade I hearing loss;

Type B - atresia in both cartilaginous and bony parts of the ear canal, II-III degree hearing loss;

Type C - all cases of complete atresia and hypoplasia of the tympanic cavity;

Type D - complete atresia of the auditory canal with poor pneumatisation of the temporal bone, accompanied by abnormal location of the facial nerve canal and labyrinth capsule (changes detected are contraindications for hearing-improving surgery).

Bone anomalies in atresia auris congenita are predominantly characterised by fusion of the malleus and anvil, including fixation in the epitimpanal recess; bony ankylosis of the malleus neck to the atresia plate; hypoplasia of the malleus handle. The hammerhead and anvil may also be missing. A variety of malformations of the anvil and stirrup can also be found. The anvil is usually small and thin, with deformed feet, but fixation of the stirrup is rare. The anvil-stem joint may also appear fragile and may sometimes exist only as a fibrous joint. Full

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visualisation of the stirrups may be difficult due to an overlying mass on top of the bone.

Ishimoto et al. found only a very limited correlation between deformities of the auricle and malformations of the middle ear.

In addition, a number of authors R. A. Jahrsdoerfer [36], R. Siegert, T. Mayer, H. Weerda have developed various classifications and scoring systems, based on CT data of the temporal bones, for a preliminary assessment of the prognosis of the surgical outcome.

The classification of the R. Jahrsdoerfer (J system), which was proposed in 1992. The J system consists of nine anatomical structures: presence of the stapes, antevertal window, cochlear window, ventilated middle ear space, presence of malleolar-anvil joint, pneumatisation of the mastoid process, anvil-stem joint, location of the tympanal segment of the facial nerve and the normal appearance of the auricle (Table 1).

Table No. 1

R. grading system. Jahrsdoerfer

Parameters	Баллы
Stage	2
Eucharist window	1
Eardrum	1
Facial nerve	1
Malleolar-anacetabular junction	1
Pneumatisation of mastoid process	1
Anvil-stem joint	1
Window of the cochlea	1
Auricle view	1

The streak is awarded 2 points as it is considered the most important factor. The remaining eight anatomical components are given a score of 1. An overall score of J ≥6 indicates that the patient may be a candidate for canaloplasty (48) (Table 2).

Table 2

R.A. Jahrsdoerfer prognostic scale

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Prognosis for surgical treatment	Score n=10
Excellent	10
Very good	9
Good	8
Satisfactory	7
Intermediate	6
Poor	5 or less

A number of studies have confirmed the validity of System J, showing a correlation between a higher degree and better hearing results.

R. Siegert et al. used their scoring system to demonstrate that despite significant differences between the groups of malformations assessed, it is not possible to reliably predict the degree of middle ear malformation in individual cases. On this basis, they extended the classification proposed by R.A. Jahrsdoerfer using their own semi-quantitative scoring system (Table 3) to evaluate temporal bone malformations (based on CT scan) and to assess differential indications for surgical intervention, in particular to establish a prognostic basis for suitability for middle ear reconstruction. This scale includes the degree of development of structures that are considered critical to the success of middle ear reconstruction surgery.

Table 3

The R. Siegert, based on preoperative CT scans of the temporal bones

Anatomical features	Findings	Points
Outer auditory canal	Norm	2
	Soft tissue atresia	1
	Bony atresia	0
Pneumatisation of the mastoid process	Excellent	2
	Poor	1
	Absent	0

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Middle ear cavity volume	Wide	2
	Medium	1
	None	0
Middle ear cavity pneumatisation	Good	2
	Poor	1
	None	0
Facial nerve	Normal Dislocation	4
	Slight dislocation	2
	Significant dislocation	0
Vessels	Normal dislocation	2
	Slight dislocation	1
	Major dislocation	0
Malleus + anvil	Normal	2
	Dysplasia	1
	Absent	0
Hammer	Norm	4
	Dysplasia	2
	Absent	0
Vestibule window	Normal	4
	Obliterated	0
Window of the cochlea	Normal	4
	Obliterated	0
Score summation		0-28

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A high score corresponds to well-developed or normal structures. The external auditory canal, size of the tympanic cavity, configuration of the ossicles and free windows are important spatial parameters in tympanoplasty. Aeration of the mastoid process and tympanic cavity allows conclusions about the functional status of the eustachian tube. Abnormal arterial and/or facial nerve pathways do not rule out surgery, but increase the risk of complications. In addition to a spiral scan, the use of a sagittal and coronary scan or subsequent reconstruction of these slices is required in some cases for an accurate assessment. A normal auricle is usually combined with normal middle ear structures. Thus, such ears almost always receive scores close to a maximum of 28 points. In the case of malformations of the auricle of a higher degree, the score is usually significantly lower. Based on their findings, Siegert et al. formulate the following recommendations:

1. In bilateral middle ear anomalies with an external auditory canal atresia, reconstructive surgery should be started with the better hearing ear, if the patient has a score of 15 or higher;

2. In the case of a unilateral anomaly, surgical treatment is indicated with a minimum score of 20 and after the patient has been fully informed about possible complications.

3. In patients with lower scores, hearing aids only.

N.A. Mileshina (2003) developed a 26-point temporal bone CT scan in children with external auditory canal atresia. Data are recorded separately for each ear (Table 4)

Table No. 4

Assessment of computed tomography findings in congenital malformations of the temporal bone (Mileshina N.A., 2003)

CT scoring	Number of possible points right ear	Number of possible points left ear
Atresia of the external auditory canal 2/1/0		
Pneumatisation of the mastoid process 2/1/0		
Size of tympanic cavity 2/1/0		
Anvil-stem complex 2/1/0		
Anvil-stem joint 2/1/0		
Oval window 4/2/0		
Circular window 4/2/0		
Canal of the facial nerve 4/2/0		
Sigmoid sinus, jugular vein 4/2/0		

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Total points 26		

Notes: 2/1 point - mild pathology; 0 point - severe pathology; 4/2 points - normal. In the empty columns, the right and left ears should be recorded respectively. With a score of 18 or more, a meatotympanoplasty can be performed to improve hearing. In patients with external auditory canal atresia and conductive hearing loss of degrees III-IV with severe congenital pathology of the auditory ossicles, labyrinth windows, facial nerve canal, having a score of 17 or less, the hearing-improving phase of surgery will not be effective. If this patient has microtia, only plastic surgery to reconstruct the auricle is rational.

At the same time, H. M. Diab modified and adhered to the R. Jahrsdoerfer. He developed a prognostic scoring scale based on a study of the structure of the middle ear based on CT scans of the temporal bones and measurements obtained on the cadaveric temporal bones (Table 5).

Table 5

Distribution of scores based on middle and inner ear structure (H. M. Diab)

Anatomical features	Findings	Scores
Antrum prominent	Decrease of less than 30%	2
	Reduction of 30-50%	1
	Reduction greater than 50%	0
Location of sigmoid sinus	Normal presentation	2
	Preconception with covering 1/3 of the mastoid part	1
	Pre-presentation covering 2/3 of the mastoidal area	0
Location of temporomandibular joint	Slight dislocation	2
	Moderate dislocation	1
	Severe dislocation	0
Pneumatisation of the mastoid process	Less than 30% reduction	2
	Reduction of 30-50%	1

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	More than 50% reduction	0
Middle ear cavity	Wide	2
	Medium	1
	Absent	0
Facial nerve	Normal dislocation	2
	Slight dislocation	1
	Large dislocation	0
Malleus + anvil	Not detected	0
	Detected	1
Hammer	Not detected	0
	Detected	2
Vestibule window	Detected	1
	Not Determined	0
Window of the cochlea	Determined	1
	Not Determined	0
Score	Findings	17

Table 6

The prognosis for the outcome of the operation depends on the number of points scored

Anticipated outcome of the operation	Scores			
	17-14	13-11	10-8	< 8
	A	B	C	D

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Note: A) Excellent prognosis for surgical treatment; B) Good prognosis for surgical treatment; C) Satisfactory prognosis for surgical treatment; D) Surgery is not indicated.

Abnormal arterial and/or facial nerve pathways do not always rule out surgery, but increase the risk of complications. Patients with atypical facial nerve pathways and patients with severe middle ear malformations should not be considered candidates for surgery.

The bone thickness of the cranial vault can be measured, especially in the temporal and parietal regions in patients who are planning to be fitted with a bone-anchored hearing aid (BAFA).

Thus, a CT scan not only demonstrates suitability for surgery, but also identifies contraindications. The chosen classification for malformations of the outer and middle ear will allow the otosurgeon to determine the surgical treatment and post-operative prognosis for this pathology.

In general, the history plays an important role in unilateral hearing loss without an outer ear defect, in which the diagnosis is ambiguous and sometimes delayed. It should include the occurrence of hearing problems and the exclusion of other causes of hearing loss, such as infections or surgery. Isolated middle ear malformations and otosclerosis may have similar characteristics. If malformations are not suspected, a CT scan is often not performed and the diagnosis is made by a tympanotomy. In order to classify patients with ear malformations and treat them effectively, accurate knowledge and a uniform description of the defects are needed. Clinical and audiometric examination methods as well as radiological methods can be used. Accurate anatomical description of malformations using imaging procedures is necessary, especially with regard to the planning and outcome of surgical reconstructions of the outer and middle ear.

A newborn with an auricular deformity needs a detailed examination of the craniofacial structures. All such patients require a thorough examination of the skull, face and neck for configuration, symmetry, facial proportions, chewing apparatus, bite, hair and skin condition, sensory function, speech, voice and swallowing. The middle ear function should be investigated particularly carefully because the development of the outer ear usually closely correlates with the development of the middle ear. Abnormalities of the ear may be accompanied by pre-auricular fossae or tags, and partial or complete paralysis of the facial nerve. In addition to a basic examination of the ears (examination, palpation, photodocumentation), attention should be paid to any anatomical features that may increase the risk or compromise the success of middle ear surgery. Such findings include abnormalities of the Eustachian tube resulting from adenoid hyperplasia, marked septal deviation or nasal hyperplasia, and the presence of a cleft palate (even if it is merely submucosal). Ear malformations can occur in combination with syndromes; therefore, changes in the internal organs (e.g., heart and kidneys), nervous system and skeleton (e.g., cervical spine) should be excluded by a multidisciplinary team (e.g., pediatrician, neurologist and orthopedist). For genetic reasons, a preoperative evaluation of facial nerve function is necessary when planning reconstructive middle ear surgery. Congenital anomalies of the outer and middle ear are rare causes of conductive hearing loss in children. Hearing loss in patients with malformations of the external ear canal is conductive, although a small proportion of patients will also have a neurosensory component. Conductive loss is usually at a threshold of 40-60 dB, depending on ossicular deformity, ossicular mobility and the degree of pneumatisation of the

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temporal bone. Hearing loss associated with these minor malformations, including congenital ankylosis of the stirrup, persistent stirrup artery, malleolar fixation and absence of the oval window, can range from mild to severe, can be missed during newborn hearing screening and cannot be diagnosed until the child is able to undergo behavioural testing.

Diagnosing a conductive hearing loss in a child with grade II or III microtia and associated atresia of the external auditory canal is very simple - the abnormality is easily identified on physical examination and appropriate testing can be performed. Following newborn hearing screening, recommended testing protocols include otoacoustic emission testing in the normal or slightly abnormal ear, auditory brainstem response testing by air and bone conduction (ABR) in the normal ear and if the atretic ear is unilateral then testing by bone conduction, if the process is bilateral then bone conduction ABR is performed. A child with a conductive hearing loss with a normal auricle and atresia of the external auditory canal may be deceptive, but hearing loss is usually detected in newborn hearing screening or sometimes later in school screening. Careful binocular microscopic examination either in the office or under anaesthesia with ABR testing for air and bone conduction can diagnose atresia of the external auditory canal or minor malformations and conductive hearing loss.

With an estimated incidence of 1 in 10,000 to 20,000, malformations of the external auditory pathway cause a moderate to severe conductive hearing loss; the bone conduction thresholds are usually normal. About 70% of the external auditory canal atresia process is unilateral, boys are affected more often than girls, the right ear is affected more often than the left, all for unknown reasons. The diagnosis of an external auditory canal atresia is not difficult because of the accompanying microtia. However, the condition can also occur in the absence of microtia, so a careful otoscopic examination is necessary to identify the auditory canal. The search for other concomitant abnormalities, including renal, vertebral and craniofacial/structural abnormalities, should also be considered. Syndromic conditions associated with external auditory canal atresia include hemifacial microsomia/Goldenhar and Treacher-Collins syndromes.

Once the diagnosis has been established, audiometric evaluation includes air conduction and bone conduction testing (both for normal and atretic ears if unilateral) and bone conduction thresholds for the child with bilateral auditory canal malformation. By 3 months of age, a complete audiological evaluation including auditory brainstem response (ABR) testing for air and bone conduction should be performed. Even in patients with unilateral atresia of the external auditory canal, it is crucial to document the hearing status of the normal ear to make sure it can hear well. Older children can be tested behaviorally in a sound booth using techniques such as visual reinforcement audiometry (ages 2-4 years) and conditional play audiometry (ages 4-7 years). Neurosensory auditory acuity level is a method of measuring bone conduction thresholds for each individual ear in a patient with bilateral external auditory canal atresia. Hearing should be monitored at 3 to 6 month intervals during the first 2 years and annually thereafter, mainly to ensure that the ear continues to hear normally in patients with unilateral atresia of the external auditory canal. Parental monitoring of speech and language development is also a key adjunct to speech/language therapy if the child is not progressing. Normal hearing in 1 ear is sufficient for normal speech and language development; however, the development of expressive speech should be monitored closely and

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interventions such as speech therapy or bone conduction enhancement may be prescribed.

In bilateral atresia of the external auditory canal, the level of malformation and degree of hearing loss may be different for each side. Early (by 3-6 months) auditory rehabilitation in these children is of paramount importance for normal speech and language development. This rehabilitation involves the use of special bone conduction hearing aids. For optimal bone conduction, the bone oscillator is attached tightly to the mastoid process (or other area of the skull) with a strap or tape placed on the head. The benefits of amplification in a child with unilateral external auditory canal atresia are unclear, and the decision to fit a bone conduction hearing aid in this patient group is made after careful discussion with the family, otolaryngologist and audiologist. Factors to consider include the possible benefits of atretic ear stimulation, the possibility of a critical period for true binaural processing, psychosocial factors, and the lack of outcome data in children with unilateral ear atresia. For a normal hearing ear, hearing protection/preservation is critical, and an exudate in the middle ear requires close monitoring and aggressive treatment.

The temporal bone is the part of the cranial bone that forms the ear. The vital organs of hearing, such as the external auditory canal, the eardrum, the auditory ossicles, the cochlea, the semicircular canal, the otolith organ, the internal auditory canal, the facial nerve and the internal carotid artery, all cluster in this narrow space. Although a simple radiological examination of the middle and inner ear (Schüller and Stenwens method) gives some results and is still commonly performed today, it is difficult to see the structure of the temporal bone clearly. After the advent of computed tomography (CT) in the 1970s, spiral scans were developed in the second half of the 1980s, making it possible to show the temporal bone in more detail. Many patients with microtia/atresia of the NAS also have middle ear abnormalities, and a CT scan is necessary to understand the middle ear.

Currently, CT scan is the method of choice for external ear and middle ear malformations. Routine X-rays have no role in malformations of the ear. MRI allows more detailed detection of inner ear abnormalities and abnormal nerves in the inner ear canal, but cannot demonstrate abnormalities of the outer and middle ear.

Infants are sometimes referred for an outpatient CT scan at a few months of age. The timing of the CT scan varies from one healthcare facility to another. A CT scan is performed in the first months after birth for the following reasons: (1) it is the best method to identify the outer and middle ear; (2) it shows the presence or absence of inner ear abnormality and can determine the cause of hearing loss; (3) it shows the presence or absence of congenital otitis media cholesteatoma and (4) it is used to understand the severity of temporal bone anomaly early in life to confirm that reconstruction of the external auditory canal is possible and to reassure the patient's family. Thanks to recent advances in CT equipment, the radiation dose is decreasing, but the radiation exposure with CT is still tens of times higher than with plain radiography. In addition, because of their smaller body size, children have 2-5 times more exposure to an organ than adults under the same conditions. In addition, the contrast of the image in children tends to be worse because they have less fat and fewer organs than adults. Thus, the dose of radioactivity has to be increased when more contrast is required. A CT scan in children should therefore be carefully planned with the above in mind. Congenital cholesteatoma media has been reported in

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4-7% of patients with congenital stenosis or atresia of the external auditory canal. With the progression of cholesteatoma media otitis media, the surrounding bones are destroyed and inner ear disorders and facial paralysis are induced; serious symptoms associated with meningitis and brain abscesses develop when the lesion reaches the skull. It is therefore better to have a CT scan as soon as possible to rule out congenital cholesteatoma media otitis media. However, the diagnosis of otitis media cholesteatoma on CT imaging is sometimes difficult because fetal residues and exudate often persist in the middle ear cavity for 1 year of life. In addition, it may take several years before the disease becomes advanced enough to show obvious symptoms, even if it was diagnosed in infancy. MRI is useful diagnostically to help differentiate congenital cholesteatoma from other soft density exudates of the middle ear. On MRI, a cholesteatoma will appear with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.

If the patient has no auditory brainstem response or hearing acuity is progressively worsening, a CT scan should be performed as soon as possible to evaluate the inner ear. However, residual hearing acuity is not only seen in patients with unilateral microtia, in whom the normal ear has normal hearing acuity, but also in patients with bilateral microtia. Thus, CT scans performed at an early age in preparation for the use of hearing aids and at the beginning of hearing training provide little useful information. Reconstruction of the external auditory canal is often performed around the age of 10, and a CT scan is performed immediately prior to surgery to obtain information for surgery. A CT scan performed in infancy does not provide direct information for reconstruction of the outer ear. Because the skull bone continues to grow until the age of 15, the mastoid bone undergoes a different mastoidectomy than it did in infancy. CT findings in infancy, however, are useful for understanding middle ear anomalies and the potential to improve hearing acuity after reconstruction of the external auditory canal in the future. Some authors suggest performing an initial CT scan at around 3 years of age for patients without signs of otitis media, such as overt cholesteatoma media, or for patients without severe or progressive hearing loss.

The diagnosis of malformations requires the best local resolution in a CT scan. This is primarily due to the small slice thickness (0.5-1 mm), bone algorithm and high zoom (low field of view). The radiation exposure can be lower than in tumour diagnosis, which also requires a soft window, but cannot be as low as in a low-dose CT of the sinuses, because the increased image noise will not allow sufficient visualisation of small details, especially of the stapes. The specific values depend on the CT device used. For modern multidetector CTs the current limit is around 120 mAs. Because of the excellent post-processing capabilities, especially for multiplanar reconstructions, the helical method should be chosen. A prerequisite for high quality multi-planar reconstructions is a small step (0.3 mm) and for 3D reconstructions the use of soft tissue data. Often small details such as the stapes are better studied with multiple oblique reconstructions.

CT scan has a high degree of clarification when malformations of the outer and middle ear are suspected, including other causes of conductive hearing loss. Only in a few cases, the cause cannot be found. Even with today's CT technology, it is difficult to diagnose an isolated stapes fixation or every detail of abnormally formed ossicles.

Nevertheless, a CT scan is indicated in patients with suspected anomalies of the outer and middle ear and is absolutely essential for preoperative planning.

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Children with congenital stenosis of the external auditory canal are at risk for cholesteatoma due to skin entrapment within a narrow canal, especially if the canal is less than 2 mm (Cole and Jahrsdoerfer, 1990). If canal size is insufficient to assess a cholesteatoma in the clinic, a high-resolution computed tomography (CT) scan of the temporal bone is required. Cholesteatoma of the canal usually does not occur before the age of 3 years; therefore, obtaining a CT scan after this age is reasonable. A CT scan is not indicated during the neonatal period - there is no reason to expose the newborn to delivered radiation, although very little. No important clinical decisions (e.g. surgical decision) will be made during the neonatal period or in the first year of life. If a CT scan is performed during the neonatal period, a repeat CT scan will be required before surgery or if complications such as cholesteatoma are suspected. Traditional two-dimensional (2D) imaging has been widely used to image individual temporal bone structures. However, the complex multi-spatial orientation of these structures located in a compact area often makes it difficult to assess their three-dimensional (3D) orientation in the temporal bone and their complex interrelationships. Recent developments in software technology have made it possible to quickly create three-dimensional volumetric images from conventional 2D data. These three-dimensional (VR) images can be sliced in any plane and rotated in space, providing a three-dimensional view of the anatomy of the temporal bone. Microanatomical structures that are poorly visible in conventional 2D imaging can be clearly depicted using overlapping reconstructions at smaller intervals. In addition, reformatted images provide additional information about a variety of conditions including congenital malformations, vascular anomalies, inflammatory or neoplastic conditions and temporal bone injury.

Thin slices (minimum 1mm), high resolution CT scans have changed the possibilities of treating malformations of the outer and middle ear, providing precise anatomical detail of the middle and inner ear. This is important in determining eligibility and when planning surgery. If a family wants to assess their child's candidacy for surgery, a CT scan is performed when the child is old enough that sedation is not necessary (around 4-5 years).

CONCLUSIONS

In reviewing the world literature, we found that some important issues concerning etiopathogenesis, diagnosis, and preoperative examination in patients with external ear malformations have not been adequately addressed in the works of Russian and foreign otosurgeons.

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