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ABBREVIATION KEY:

EAC = external auditory canal.

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ABSTRACT

Congenital aural atresia occurs in approximately 1 in 10,000–20,000 births and may be surgically repaired if the middle ear malformation is limited in character. External auditory canal atresia is difficult to repair surgically, with significant risks and complications. Surgical candidacy in congenital aural atresia is based on multiple factors, central to which are the anatomy of the temporal bone and audiometric findings. High-resolution multidetector CT is the imaging technique of choice for anatomy delineation, although there are some specific indications for MR imaging in presurgical assessment. Various CT grading systems have been developed to determine surgical candidacy and are described in this review. The radiologist's understanding and precise evaluation of important anatomic structures is critical in the assessment of surgical candidacy. This review serves to familiarize the radiologist with various abnormalities seen in congenital aural atresia.

Learning Objectives: Recognize important CT-based classification systems used to determine surgical candidacy and contraindications to surgery for external auditory canal atresia repair, recognize the embryologic origin, and assess the EAC and ossicles in congenital aural atresia.

INTRODUCTION

Congenital aural atresia occurs in 1 in 10,000–20,000 births and is bilateral in approximately 30% of cases. Male individuals are affected slightly more than female individuals, and it occurs more frequently in the right ear.¹ Congenital aural atresia may occur in combination with a variety of malformations of the middle and external ear secondary to their complex, intertwined embryologic origins.² Malformations of the pinna, for example, microtia, are found in three-fourths of those patients with external auditory canal (EAC) atresia.² Repair of microtia may be performed

with or without repair of EAC atresia. Repair of EAC atresia may be entertained if sensorineural hearing is preserved³ and the middle ear malformation is limited in character. The decision of whether or not to repair an EAC atresia may be complicated and is primarily based on the radiographic appearance of the external, middle, and inner ear as well as the anatomy of closely related structures, including the facial nerve, skull base, and temporomandibular joint. Even children with significant hearing loss can receive excellent auditory rehabilitation with either a bone-anchored or bone-conducting hearing aid.^{1,4} Most

patients, however, believe that cosmesis is not very good with the Bone Anchored Hearing Aid (BAHA), and frequent wound care may be required. Thus, surgery should be offered to appropriately selected children who are likely to receive an excellent outcome, which makes preoperative radiographic evaluation even more important.

EMBRYOLOGY

The embryologic origin of the inner, middle, and external ears is complex and involves all 3 germ layer origins. Discussion of this developmental process is beneficial for understanding the associated middle ear anomalies that may accompany EAC atresia.

Development of the Inner Ear

The membranous labyrinth develops in the 4th-25th weeks of life and originates from the neuroectoderm of the otic placode. The eighth cranial nerve develops parallel to the membranous labyrinth. Development of the osseous labyrinth (which involves ossification of the labyrinthine capsule) occurs at the 16th or 17th week.⁵

Development of the Middle Ear

The body of the malleus and incus develop from the mesenchyme of the first branchial arch, with the second branchial apparatus giving rise to the handle of the malleus and the lenticular process of the incus. The stapes has a dual origin, with the suprastructure that arise from the second branchial arch and the vestibular surface of the foot plate, and the annular ligament that arises from the otic capsule. The development of the ossicles commences in the eighth week, and ossification is nearly complete by the seventh month of gestation.⁵

How the EAC Develops

The EAC arises from deepening of the first branchial groove in the sixth week of gestation, with development of the lateral one-third of the EAC (cartilaginous portion). A solid block of epithelial cells then develops at the medial portion of the groove. After complete differentiation of the inner, middle, and outer ear in the 26th week, the medial two-thirds (bony portion) of the developing EAC canalizes from the medial to lateral direction to fuse with the lateral cartilaginous portion. Failure of proper canalization of the medial two-thirds leads to atresia of the EAC.³ The floor and the sides of the bony EAC arise from the tympanic bone, which develops at approximately 12 weeks of gestation.³ Failure of canalization also results in deformity of this bone with bony overgrowth, which becomes the atresia plate (although the atresia plate may also arise from the squamosal temporal bone).³ The pinna arises from the ectoderm of the first and second branchial arches and begins at the 40th-45th day, and is fully formed by the fourth month of gestation.⁵

CLASSIFICATION OF CONGENITAL AURAL ATRESIA

The diagnosis of congenital aural dysplasia is made by clinical examination and imaging. External auditory canal atresia may be fibrous or bony. The bony atretic plate borders the tympanic cavity and may vary in thickness and pneumatization. Auricular and EAC malformations were classified by Weerda³:

- Type A EAC stenosis entails marked narrowing of the EAC with an intact skin layer.
- Type B EAC stenosis demonstrates partial development of the EAC with a medial bony atresia plate.
- Type C EAC malformation consists of complete bony EAC atresia.

A correlation between the degree of EAC atresia and the severity of microtia has been established.^{6,7}

SURGICAL APPROACHES

Two different surgical approaches for EAC atresia repair have been described: transmastoid and the anterior approach. Both approaches include mobilization of the ossicles attached to the atretic plate, which creates a tympanic membrane graft from temporalis fascia, and then attaches the neotympanum to the ossicles or stapes prosthesis.^{1,8} In the transmastoid approach, the mastoid air cells are removed to access the middle ear, and the atretic plate is then dissected.⁸ The disadvantage of this procedure is the formation of a large mastoid defect, which may be prone to infection.⁸ In the anterior approach, the surgeon will drill a canal through the atretic plate starting immediately posterior to the glenoid fossa and anterior to the mastoid cells, approaching the epitympanum inferior to the tegmen. Regardless of the technique, care must be taken to avoid injury to the ossicles or facial nerve.^{1,8} Due to the common association of congenital aural atresia and various temporal bone anomalies, such as an aberrant facial nerve, deformity of ossicles, defect of the oval window, and a lack of mastoid pneumatization, presurgical planning with high-resolution CT is necessary for surgical success.²

PATIENT SELECTION FOR SURGERY

Surgical candidacy in congenital aural atresia is based on multiple factors, central to which are the anatomy of the temporal bone and audiometric findings.⁹ Audiometric findings should confirm a significant conductive hearing impairment. There, however, is a 10%-47% incidence of inner ear anomalies associated with microtia.³ High-resolution multidetector CT is the imaging technique of choice for anatomy delineation.¹⁰⁻¹⁴ CT and MR imaging may be used in conjunction, particularly when planning surgery for congenital cholesteatoma, infection, complication from previous surgery, or labyrinthine fistula.^{15,16}

At our institution, the imaging protocol on our Siemens (Somatom 64 section scanner; Siemens, Malvern, Pennsylvania) CT scanner entails helical axial 1-mm section thick-

Table 1:– Jahrsdoerfer grading system of candidacy for surgery of congenital aural atresia

Parameter	Points
Stapes present	2
Oval window open	1
Middle ear space	1
Facial nerve	1
Malleus-incus complex	1
Mastoid pneumatized	1
Incus-stapes connection	1
Round window	1
Appearance of the external ear	1
Total available points	10

Table 2:– Interpretation of Jahrsdoerfer rating for surgery of congenital aural atresia

Rating (n = 10)	Type of Candidate
10	Excellent
9	Very good
8	Good
7	Fair
6	Marginal
≤5	Poor

ness image acquisition with a 230 FOV, 0.8 pitch, CARE Dose, CARE kV set at 120, and 200 quality mAs. A coned down axial of 0.6 mm and coronal 1-mm section thickness images are supplied in the bone algorithm.

The Jahrsdoerfer grading system¹⁷ was developed to determine surgical candidacy and to predict which patients would have more favorable hearing outcomes. The system is widely used and is based on high-resolution CT imaging findings (Table 1). Jahrsdoerfer et al¹⁷ reported a success rate of 80% in patients with a presurgical score of ≥ 8 in patients with unilateral atresia. In addition, patients with a score of ≤ 5 were considered unsuitable for surgery (Table 2).¹⁷ Shonka et al⁹ reported similar success with a Jahrsdoerfer grading score of ≥ 7 . Dedhia et al¹⁸ recently expanded Jahrsdoerfer's scoring system and highlight the importance of the malleus incus complex size and position, tegmen mastoideum location, and incudostapedial joint angle. The group also expanded the impact of the facial nerve on the grading system. Their new modified scoring system is based on a 14-point scale.¹⁸

Yellon⁴ recently expanded on work by Jahrsdoerfer¹⁷ by rationalizing surgical decision-making in the post-BAHA era. Yellon⁴ recommends BAHA for children with a Jahrsdoerfer score of ≤ 5 . Children with a Jahrsdoerfer score of ≥ 6 can be offered surgery or BAHA, depending on the family's preference.⁴

Siebert et al¹⁹ expanded on an anatomic point system based on CT findings to include the configuration of structures

considered crucial for successful middle ear reconstruction (Table 3).²⁰ Siebert et al¹⁹ recommended surgical or conservative therapy, depending on the CT score and whether the malformation is unilateral or bilateral:

- In bilateral malformations, middle ear reconstruction of the better hearing ear, if the score is ≥ 15
- In unilateral malformations, middle ear reconstruction, if the score is ≥ 20 , after a frank discussion with the patient
- In patients with lower score, hearing aids only
- Contraindications to surgery include the following: low tegmen position, risk of injury to the facial nerve due to a significantly abnormal location and position, small size and poor middle ear aeration, absence of the stapes bone, and poor status of the oval window.⁹ A recently described Active Middle Ear Implant Score by Frenzel et al,²¹ emphasizes precise CT-based assessment of the oval window, round window, middle ear pneumatization, mastoid pneumatization, position of the tegmen, and position of the facial nerve at the oval window for more accurate risk stratification and surgical planning.

IMAGING FEATURES THAT AFFECT SURGERY

EAC

There may be stenosis or membranous versus bony atresia of the EAC.^{3,19} Narrowed ear canals are often angulated caudolateral to craniomedial (Fig 1).¹⁷

Mastoid Pneumatization

Mastoid aeration correlates with the degree of external ear anomaly. Reduced pneumatization results in decreased drilling space, downward sloping of the middle cranial fossa, and low tegmen position.^{2,3,22} The mastoid may be described as pneumatized, opacified, or ebonized (Fig 2). A pneumatized mastoid is present when there are an aerated antrum and aerated cells. The aerated cells extend lateral to the inner atresia plate and labyrinth, and into the mastoid. A fluid or soft-tissue-filled mastoid is considered an opacified mastoid. The mastoid is considered ebonized if it is sclerotic or ossified.²¹

Middle Ear Space

The middle ear is qualitatively assessed for size and aeration (Fig 3). Size measurements were previously proposed,^{22,23} however, absolute measurements have yet to be described or uniformly used.²¹ A nonaerated or opacified middle ear cavity may predict poorer hearing outcome after surgery. This can be due to difficulty in identifying the tympanic cavity and ossicular chain during drilling, postoperative ossicular chain re-fixation, and postoperative stenosis with a small middle ear space. Identification of the bony Eustachian tube is important because it may often be abnormal.^{1,2,9,22} Fat attenuation within middle ear opacification may signify the presence of a cholesteatoma or an epidermoid cyst. MR imaging in these patients is useful to differ-

Table 3:— Siegert scoring system for surgical candidacy for congenital aural atresia

Structures	Configuration	Points
EAC	Normal/fibrotic atresia/bony atresia	2/1/0
Aeration of the mastoid	Distinct/moderate/absent	2/1/0
Size of the tympanic cavity	Large/moderate/eburnated	2/1/0
Aeration of the tympanic cavity	Distinct/moderate/absent	2/1/0
Facial nerve	Normal/slightly aberrant/strongly aberrant	4/2/0
Course of arteries and veins	Normal/slightly aberrant/strongly aberrant	2/1/0
Malleus and incus	Normal/dysplastic/absent	2/1/0
Stapes	Normal/dysplastic/absent	4/2/0
Oval window	Open/closed	4/0
Round window	Open/closed	4/0
Maximum score		28

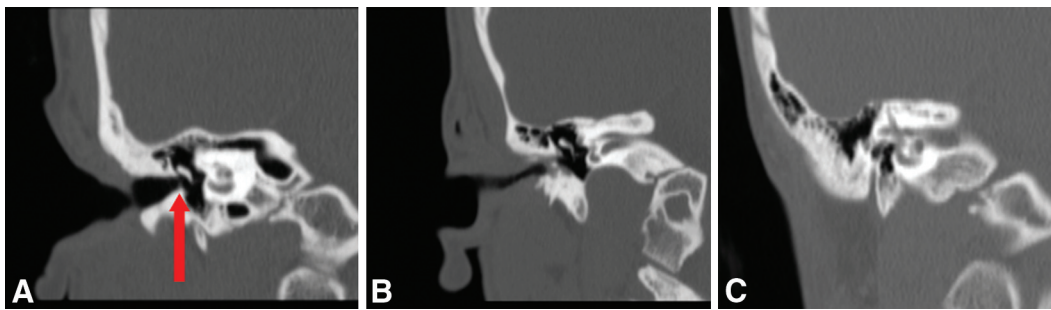


Fig 1. Appearances of EAC abnormalities. (A) Coronal CT in a patient with a hypoplastic EAC and a hypoplastic tympanic membrane (*red arrow*). **(B)** Coronal CT in a patient with a hypoplastic canal and a medial bony atretic plate. **(C)** Coronal CT in a patient with complete bony atresia and a thick atretic plate.

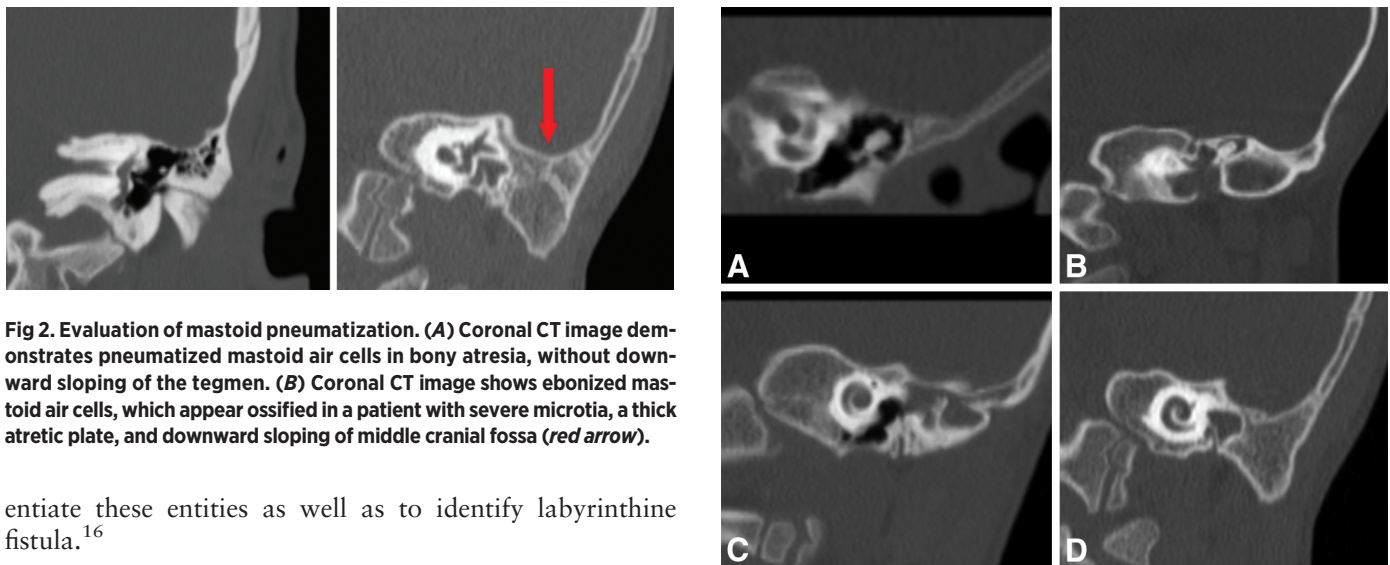


Fig 2. Evaluation of mastoid pneumatization. (A) Coronal CT image demonstrates pneumatized mastoid air cells in bony atresia, without downward sloping of the tegmen. **(B)** Coronal CT image shows eburnated mastoid air cells, which appear ossified in a patient with severe microtia, a thick atretic plate, and downward sloping of middle cranial fossa (*red arrow*).

entiate these entities as well as to identify labyrinthine fistula.¹⁶

Ossicular Chain

The ossicles are frequently dysplastic, rotated, adherent to the middle ear wall, absent, or display arthrodesis. Oblique coronal planes along the long axes of the malleus and incus allows depiction of the ossicular anatomy and may uncover incudostapedial joint abnormalities. The incudomalleolar joint can be assessed on oblique sagittal and axial planes.¹¹

Fig 3. Middle ear size and aeration. (A) Coronal CT demonstrates a large and aerated tympanic cavity. **(B)** Coronal CT shows moderately sized opacified middle ear, with downward sloping of the tegmen and an eburnated mastoid. **(C)** Coronal CT displays a small moderately aerated tympanic cavity. **(d)** Coronal CT reveals a small and opacified middle ear.

Malleus-Incus Complex. EAC atresia may be associated with abnormal fusion of the malleus to the incus, fixation of the malleus neck to the atresia plate, and/or hypoplasia of

the manubrium (Fig 4).²⁰ Note should be made of a deformity of the malleus-incus complex, a lack of the incus long process, or an ossicular dissociation.^{21,23}

Incus-Stapes Joint. The joint between the lenticular process of the incus and head of the stapes resembles a “hockey stick” near the oval windows on oblique coronal sections.¹¹ Incudostapedial joint dysplasia often coexists with abnormal malleoincudal articulation. The long process of the incus may be thin and hypoplastic (Figs 5 and 6).²

Stapes. The stapes is so important that the presence of the stapes is assigned 2 points on the Jahrsdoerfer scale and 4 points on the Siegert score. Without a stapes, surgical reconstruction is much more complex, with results that are less predictable.¹ With the current multidetector high-resolution CT technique, oblique axial reformation as described by Henrot et al¹⁴ could identify minor stapes deformity that may otherwise be subtle but not uncommon.⁵

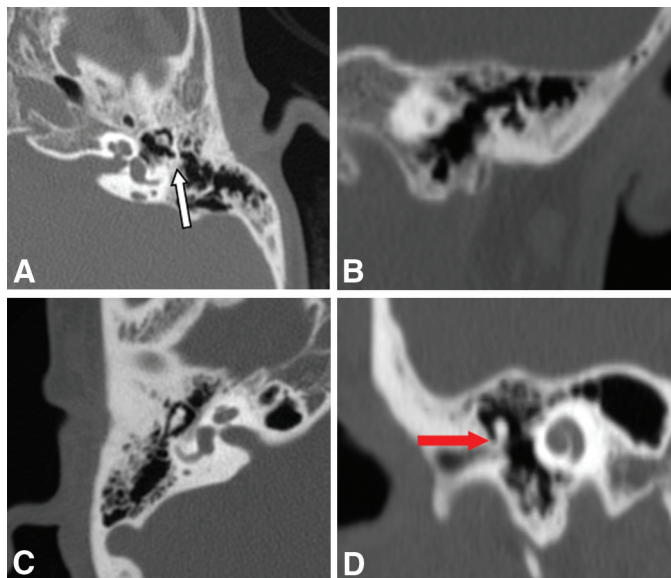


Fig 4. Abnormalities of the malleus-incus complex. Axial (A) and coronal (B) CT images exhibit fusion at the malleoincudal joint. There is fusion of the dysmorphic ossicles to the atretic plate (white arrow); axial (C) and coronal (D) CT images demonstrate a normal malleoincudal joint, but the malleus is fused to the lateral tympanic wall (red arrow).

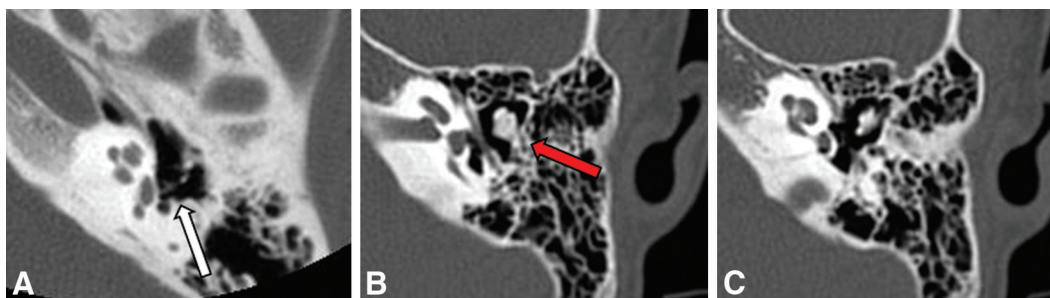


Fig 5. Configuration of the stapes. (A) Axial CT in a patient without EAC atresia shows an intact stapedial superstructure (white arrow). (B) Axial CT images from a patient with atresia show that the malleus and incus are fused to the lateral wall (red arrow) and lack of a normal stapes (b, c).

Oval Window

The vertical oval window diameter on coronal imaging should measure at least 1 mm,²² with a normal size that approaches 2 mm in diameter. A window of only 1 mm in diameter, for example, is considered stenotic and does not receive a point on the Jahrsdoerfer rating scale. This is further specified by stapes corridor and oval window corridor measurements by using the Active Middle Ear Implant Score. Nondefinition of the oval window, overlying bone attenuation, or small oval window corridor receives a low rating (Fig 7).^{15,17,21,23}

Round Window

The round window identified posterior and inferior to the oval window and medial to the tympanic sinus, is seldom

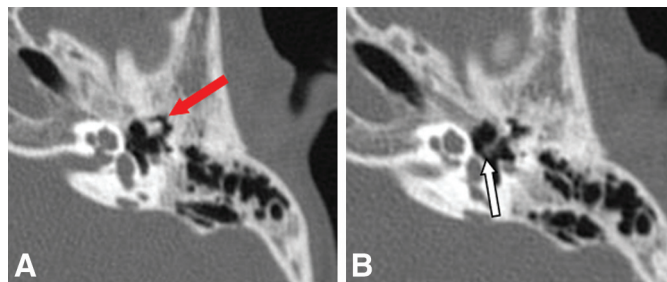


Fig 6. Configuration of the incus-stapes joint. (A) Axial CT shows an abnormal bony mass that represents the malleus and incus (red arrow) and appears ankylosed to the anterior and posterior walls of the hypoplastic middle ear. (B) The stapedial superstructure (white arrow) is not well separated from the bony mass and likely represents a combination of the incus and the stapes capitellum.



Fig 7. Configuration of the oval window. Coronal CT image shows that the oval window in this patient is covered by bone (red arrow). The vertical diameter on coronal imaging should measure at least 1 mm.

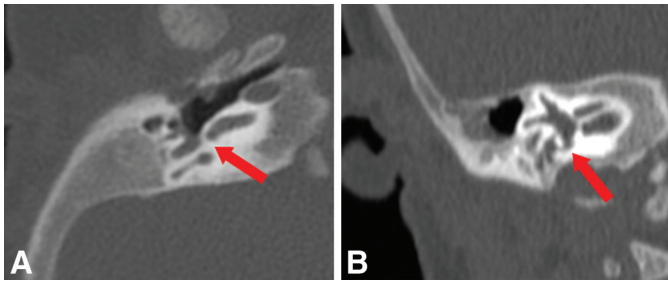


Fig 8. Configuration of the round window. Axial CT (A) and coronal CT (B) demonstrate round window stenosis (red arrows). The round window should measure at least 1 mm.



Fig 9. Course of the facial nerve. Coronal CT in a patient with atresia shows that the facial nerve overlies the oval window (white arrow), which may preclude surgical intervention.

absent in EAC atresia.²² It is located at the base of the basal turn of the cochlea and must be patent for effective cochlear function. The window is considered stenotic when it is ≤ 1 mm. If the window is stenotic, then the Active Middle Ear Implant Score proposes additional anterior and posterior round window corridor measurements to further characterize the surgical field (Fig 8).^{21,23}

Facial Nerve

Facial nerve injury represents a serious risk of atresia repair.^{1,9} The course of the facial nerve is often altered and may be difficult to follow up in patients with EAC atresia. To plan the surgical approach, tympanic and mastoid segment anomalies are particularly important to describe because surgical repair may not be entertained if there is a poor nerve position.⁹ Common abnormalities include dehiscence or caudal displacement of the tympanic segment to overlie the oval window with narrowing of the oval window niche, and anterior and lateral displacement of the mastoid segment (Fig 9).^{1,5,19,22,23}

The second genu and descending mastoid portion of the facial nerve is often displaced anteriorly; this factor is very important in determining surgical candidacy.²³ Extracranial facial nerve injury may be higher risk if there is anterolateral displacement of the stylomastoid foramen.²² Other facial nerve abnormalities include displacement of the intratympanic portion between the round and oval window niche and duplications of the mastoid segment.³

Table 4:— Checklist for evaluation of imaging features that affect surgery

Structure	Commentary
External auditory canal	Membranous or bony atresia; atresia plate thickness
Mastoid pneumatization	Pneumatized, opacified or ebonized
Middle ear space	Large or small, aerated or opacified
Malleus-incus complex	Normal or dysplastic
Incus-stapes joint	Normal or dysplastic
Stapes	Normal or dysplastic
Oval window	≤ 1 mm is stenotic, 2 mm is normal
Round window	≤ 1 mm is stenotic
Facial nerve course	Tympanic segment position relative to the oval window; mastoid segment anterior or lateral displacement

Other Abnormalities

Mandibular condyle, glenoid fossa, and zygomatic arch abnormalities can be associated with aural atresia, particularly in patients with craniofacial syndromes. Aural atresia may be seen in certain syndromes, such as Goldenhar, Treacher-Collins, Pierre Robin, Klippel Feil, Apert, Wilder-vanck, and Crouzon syndromes.^{2,20-22}

A checklist for the radiologist who is interpreting a CT study for a patient with congenital aural atresia is provided in Table 4.

CONCLUSION

EAC atresia is difficult to repair surgically, with significant risks and complications. Recent advances in auditory rehabilitation, including the use of bone-conducting and bone-anchored hearing aids, have presented alternatives for traditional repair. Thus, presurgical evaluation with diagnostic imaging is essential for determining good surgical candidates for EAC reconstruction. Imaging assessment of patients with congenital aural atresia may expand and become even more rigorous over time, and the radiologist's understanding and precise evaluation of important anatomic structures is critical in the assessment of surgical candidacy. To optimize patient care, a close collaborative relationship between the surgeon and the radiologist is critical.

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