bone grafts harvested from the iliac crest, which could be used instead of plates and with which the fixation must be performed with screws.[12] However, this method has not been evaluated specifically for the management of atrophic mandibular fracture.

In conclusion, it is suggested that until a high level of evidence is available, treatment decisions should continue to be based on the clinician's experience. Thus, we agree with Madsen et al,[8] who reported that patients with comorbid medical conditions, as the most patients with atrophic mandibular fractures, necessitate treatment using methods known to be successful, not methods hoped to be successful. Therefore, a load-bearing plate, and sometimes associated with nonvascular autogenous bone grafting (if the mandibular fractures are with a vertical height of ≤10 mm), should be performed for management of atrophic mandibular fracture because this treatment method gets the most predictable outcomes.

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Congenital Aural Atresia

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Abstract: Congenital aural atresia is a spectrum of ear deformities present at birth that involves some degree of failure of the development of the external auditory canal. This malformation may be associated with other congenital anomalies; it occurs as a result of abnormal development of the first and second branchial arches and the first branchial cleft and most often occurs sporadically, although the disease may be manifested in different syndromes. Congenital aural atresia is considered one of the most difficult and challenging surgeries for the otologic surgeon. The goals of atresia surgery are to restore functional hearing, preferably without the requirement of a hearing aid, and to reconstruct a patent, infection-free external auditory canal. The repair is usually done at the age of 6 years, so children with bilateral atresia may need hearing amplification in the first few weeks of life until the age at surgery. To optimize the surgical outcome, careful audiological and radiological evaluation of the patient should be performed preoperatively. Also, postoperative frequent packing and regular follow-up are mandatory to avoid restenosis and infection of the newly created canal. With careful intraoperative dissection and regular follow-up, complications of surgery can be avoided.

Key Words: Congenital aural atresia, hearing loss, pediatric otology, microtia, ear malformation

Congenital aural atresia (CAA) is an ear deformity present at birth involving some degree of failure of the development of the external auditory canal (EAC). Often, the malformation involves the tympanic membrane, ossicles, and middle ear space to varying degrees. Although associated abnormalities of the auricle are common, the inner ear development of those cases is most often normal.[1,2]

Incidence of the disease is about 1 in 10,000 to 20,000 live births, unilateral atresia occurs 3 to 5 times more commonly than bilateral atresia. Males are more often affected than females, and in unilateral cases, the right ear is more commonly involved. This anomaly most often occurs sporadically, although patients of autosomal dominant or recessive inheritance have been reported. The disease has been reported in association with hydrocephalus, posterior cranial hypoplasia, hemifacial microsomia, cleft palate, and genitourinary abnormalities. Congenital aural atresia may be a part of different syndromal abnormalities such as Treacher Collins, Goldenhar, Crouzon, Mobius, Klippel-Feil, Fanconi, DiGeorge, and Pierre Robin syndromes.[3–5]

The challenge to the ear surgeon is to restore the sound conduction pathway through the atretic EAC and malformed middle ear to the normal cochlea.[1,4] Congenital aural atresia occurs as a result

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TABLE 1. De la Cruz and Teufert's[1] Classification of CAA

Minor malformations	Normal mastoid pneumatization
	Normal oval window footplate
	Good facial nerve footplate relationship
	Normal inner ear
Major malformations	Poor mastoid pneumatization
	Abnormality or absence of oval window/footplate
	Abnormal course of the facial nerve
	Abnormalities of the inner ear

of abnormal development of the first and second branchial arches and first branchial cleft; therefore, for good understanding of the disease, an otologist should be familiar with the development of the ear and its associated structures.

EAR DEVELOPMENT

Development of the external ear begins during the fourth week of gestation as 6 mesenchymal proliferations that enlarge to form ridges known as the hillocks of His. These hillocks, which surround the first branchial cleft or primitive meatus, fuse to form the primitive auricle by the third month of gestation.[6] The formation of EAC starts with invagination of the first branchial cleft to form the primitive meatus. This area is located between the first and the second branchial arches. The first branchial cleft invaginates and advances medially as an epithelial plate as early as the second month of fetal life. Its ingrowth temporarily meets the lateral growth of the first branchial pouch. The cleft is an ectodermal structure, whereas the pouch is an endoderm, so the external ear develops from the former, and the middle ear develops from the latter. The union between the first branchial cleft and the branchial pouch forms the meatal plate, which is the precursor of the tympanic membrane. At sixth month of gestation, the epithelial plate starts to canalize from medial to lateral direction to meet the primitive meatus.[3] At birth, the EAC comprises the bony tympanic ring medially and a membranous cartilaginous part laterally. Postnatally, the bony tympanic ring lengthens and transforms from a ring into a bony cylinder. Thus, the EAC increases in length and reaches its adult proportion by the age 4 to 5 years.[1]

Development of the ossicles begins in the fourth week of gestation. First, the malleus and incus appear as a fused mass; separation into 2 distinct ossicles typically occurs by the eighth week of gestation. The first branchial arch (Meckel cartilage) forms the head and neck of the malleus, and the body and short process of the incus. The second branchial arch (Reichert cartilage) develops the manubrium of the malleus, the long process of the incus, and the stapes superstructure. By the 16th week of gestation, the ossicles reach their adult size.[3,6]

The inner ear develops from the otic placode that appears in the third week of gestation. Invagination of the otic placode forming the otic vesicle is apparent by the fourth week; the semicircular canals have taken shape by the sixth week, and the utricle and saccule have formed by the eighth week. Development of the cochlea begins in the seventh week, and by week 12, the complete 2½ cochlear turns have formed. The membranous labyrinth is entirely developed by 15 weeks' gestation, and ossification of the surrounding otic capsule is complete by 23 weeks' gestation.[7]

The facial nerve is the nerve of the second branchial arch. Its development starts with the differentiation of neuroblasts from the acoustic-facial primordium between the fourth and the fifth week of gestation. The course of the nerve has completely formed by 17 weeks. However, at this time, the nerve is located in a more anterior-superior position, and the eventual migration to its normal adult position is dependent on the normal development of the tympanic ring and mastoid.[6,8]

From this basic knowledge of ear embryology, we can detect that completion of the external ear development occurs rather early in gestation, whereas the recanalization of the EAC occurs later. Thus, a severely deformed auricle may indicate an arrest of development at an early stage; therefore, it is more likely to be associated with EAC, middle ear, facial nerve, and possibly inner ear anomalies. In contrast, CAA with normally developed auricle is most likely representative of a later arrest in development and has a higher likelihood normal middle and inner ear structures.

EVALUATION OF PATIENTS

First, CAA may be associated with other congenital anomalies that should be excluded. Early hearing evaluation in the first few days of life is very important, either in unilateral or in bilateral cases, because ipsilateral and contralateral inner ear abnormalities may be associated with CAA.[1] Auditory brain stem response audiometry is used to assess the integrity of the auditory pathway as early as possible; this is to ensure speech and language development by hearing amplification for children born with hearing impairment.[9] Early enrollment in special education enhances speech and language development. Radiological evaluation in the form of high-resolution computed tomography (CT) is done at the age of 5 to 6 years; it can show the thickness of atretic bone, conditions of the middle ear space and ossicles, cochlear morphology, and position of the facial nerve. Also, high-resolution CT could show the presence of associated temporal bone anomalies that may be present in cases with CAA such as congenital cholesteatoma that may necessitate early intervention.[10,11]

As noted, 2 requirements are needed before atresia repair, which are (1) an audiometric evidence of cochlear function and (2) a radiological evidence of normal cochlear morphology.[4] In an effort to estimate the surgical outcome of CAA, different classification systems have been adopted. De la Cruz et al[3] divide the disease into minor and major categories (Table 1); they stated that cases of minor malformations have a high possibility of postoperative good hearing, whereas cases of major malformations are best treated with hearing amplification device in the form of bone-anchored hearing aid system. Jahrsdoerfer et al[12] addressed another classification system based on different anatomical findings: mastoid pneumatization, oval window, round window, facial nerve course, status of the ossicles, middle ear space, stapes, and appearance of the auricle (Table 2). The authors assigned 10 points for these anatomical findings; a score of 8 or higher

TABLE 2. Jahrsdoerfer's[14] Grading System of Candidacy for Atresiaplasty

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

Type of candidate: score of 10 = excellent, 9 = very good, 8 = good, 7 = fair, 6 = marginal, ≤5 = poor.

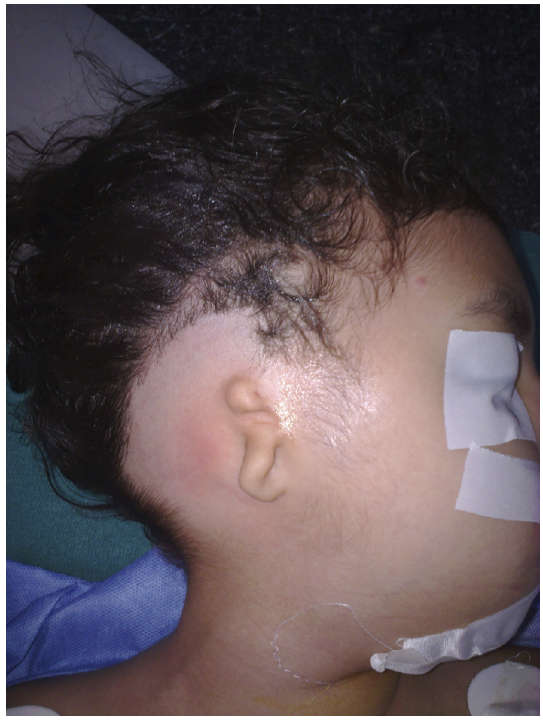

FIGURE 1. A 6-year-old patient with right CAA and microtia.

predicts the best chance of surgical success, a score of 7 implies a fair results, and a score of 6 is expected to have marginal results, whereas a score less than 6 indicates that the patient would have poor results from atresia surgery.

PATIENT COUNSELING

Congenital aural atresia may be present as either an isolated deformity, that is, sporadic, nonsyndromic, or syndromic that associated with other congenital anomalies. Parents of a child with sporadic CAA are counseled that the possibility of the disease in their subsequent siblings is no greater than that for the general population.[4,13] Counseling the parents about the expected surgical results should be performed before surgery. Patients who have minor malformations or a score of 8 or better on the Jahrsdoerfer grading scale are given a greater than 80% chance of significant hearing improvement.[14] Postoperative deterioration of hearing over time due to graft lateralization may occur in some patients.[4,15] The risk of high-tone sensorineural hearing loss (SNHL) caused by inner ear trauma may reach up to 7%, whereas the risk of facial nerve paralysis is less than 1%.[16] Other surgical risks are comparable to those for other middle ear surgeries.[1] Parents should be declared that the newly created EAC is lined by skin graft that is taken from the thigh or lower abdomen, and there will be a wound other than the one in the ear region, and both wounds necessitate a special postoperative care.[4] Patients and parents should be reminded that frequent postoperative visits are critically important especially in the early postoperative period.[1,4,13,14]

TIMING OF REPAIR

Auricular reconstruction should precede atresia repair; this is because atresia surgery may jeopardize the blood supply of the soft tissues surrounding the newly created meatus.[17] A harvested sculptured costal cartilage—used for auriculoplasty—needs good vascularity to survive. By the age of 6 years, the costal cartilage may be sufficiently developed enough to enable the surgeon to start with the first step of auricular reconstruction. However, recently, some authors prefer the use of synthetic materials for microtia repair such as a porous high-density polyethylene (Medpor) as it is more tolerated, can be used at an earlier age, and can be performed before or after atresia repair as it is not dependent on blood supply.[1] In bilateral CAA, hearing amplification should be started as early as possible with bone-anchored hearing device that can be applied in the first few weeks of life unit the age at surgery.[18]

However, hearing amplification is not necessary for patients with unilateral atresia if hearing is normal in the contralateral ear. Also, it was stated that there is no urgency for early surgical intervention in unilateral cases; the operation can be deferred until the age of adolescence at which the patient can share in the decision.[14] On the other hand, some otologists prefer to operate on unilateral atresia at the age of 6 years, like in bilateral cases, as they suggested that binaural hearing is essential for normal speech and language development.[1,4]

METHODS OF REPAIR

Under general anesthesia with oral endotracheal intubation, the patient is placed supine with the head turned away. The postauricular area is shaved, and the auricle and its surrounding area are sterilized and draped (Fig. 1). Computed tomography of the temporal bone (Fig. 2) should be available in the operating theater, and it should be evaluated thoroughly before starting the dissection. Facial nerve monitoring electrodes are placed, and muscle relaxants are avoided. The thigh or the lower abdomen is similarly prepared for harvesting skin graft. A postauricular incision is performed, and care is taken not to expose the grafted costal cartilage in patients with prior auricular reconstruction. Harvesting temporalis fascia graft is performed and left to dry for later use in tympanic membrane creation. A T-shaped incision is made in the periosteum, and the skin, subcutaneous tissue, and periosteum are elevated anteriorly until identification of glenoid fossa with exposure of the temporomandibular joint. Palpation of the condyle, mastoid tip, posterior border of mastoid process, and supramastoid crest (if present) is helpful for the surgeon to identify the position of the new meatus.[4]

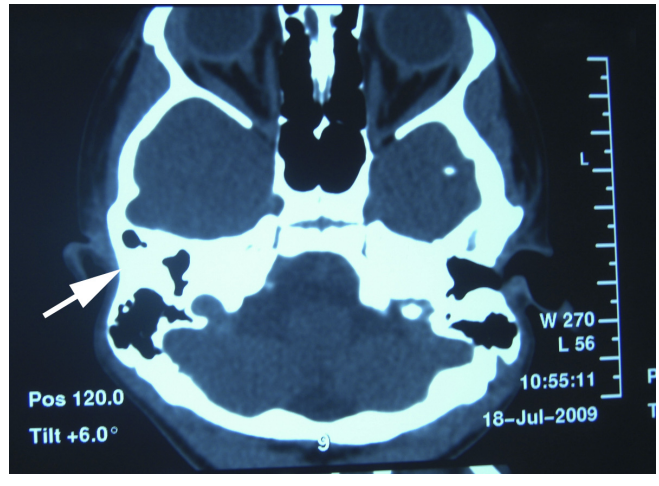

FIGURE 2. An axial CT shows right CAA with a well-developed middle ear space and pneumatized mastoid; the arrow points to the atretic plate.

Two basic approaches have been described for canaloplasty. An anterior approach starts with drilling in the cribriform area of the mastoid posterior to the glenoid fossa; drilling should be close to the suprameatal crest (if present) or high up close to the tegmen (if the crest is absent). Dissection proceeds medially until identification of the epitympanum, where the fused malleus-incus complex is found. Exposure of the middle ear space is done through widening the new ear canal to about 12-mm diameter without opening an excessive number of mastoid air cells to avoid mastoid cavity problem.[1] A posterior approach starts with drilling of the mastoid bone like in cortical mastoidectomy, with identification of the sinodural angle, which is followed anteriorly to the antrum. The lateral semicircular canal is identified and used as a landmark. Atticoantrostomy is performed, and the atretic plate is carefully removed with gentle removal of bone lateral to the middle ear space. This atretic bone is removed by diamond burr and curettes to completely expose the middle ear.[4] In both approaches, care is taken not to enter the temporomandibular joint, not to touch the ossicular mass on drilling, and not to drill posteroinferior to the middle ear space, as the facial nerve may be encountered at this point.[19]

Tympanoplasty comprises reconstruction of the ossicular chain and creation of tympanic membrane. Removal of the deformed ossicular mass is done; restoration of conductive hearing mechanism is performed by repositioning the ossicular mass on the mobile stapes or its footplate, when this is not possible, total or partial ossicular reconstruction prosthesis is used.[4] If the ossicular chain is found to be intact, it is left in place. The dried temporalis fascia graft that has been harvested early is used for creation of the new tympanic membrane. The graft is placed medial to the malleus (if present) or lateral to a piece of cartilage if prosthesis is used.[12] Packing the newly created EAC with dry cotton piece could support the temporalis fascia and ossicular mass in place until creation of a new meatus.[4]

Meatoplasty is finally performed. The auricle is turned back, and the periosteum is tacked posterosuperior to the new ear canal with an absorbable suture; this is to ensure that the new meatus will be at the same level to the newly created canal. Skin, subcutaneous tissue, and cartilage are excised in a 12-mm diameter cuff over the new canal. The new meatus like the new canal is created wider than normal by one and third as it will diminish through normal healing by the effect of fibrosis.[14]

A split-thickness skin graft is harvested with a dermatome from the lower abdomen or thigh on the same side of the operated ear. The donor site is covered with sterile gauze soaked in 1% lidocaine to decrease the postoperative pain. The skin graft is used to line the newly created EAC after removal of the temporary pack; care is taken to overlap the peripheral edge of the temporalis fascia graft with skin and also to cover all the bone circumferentially, as uncovered raw bone may lead to granulation tissue formation. The lateral edge of the skin graft is sutured to the margin of the new meatus. Packing the newly created EAC with Merocel sponge is performed, and the postauricular incision is closed in the usual fashion with posterosuperior tacking sutures to maintain the new meatus open and to pull the auricle posterior and superior to the meatus.[1,4,19]

POSTOPERATIVE CARE

Patients can be discharged from hospital on the third postoperative day. Oral antibiotics are given for 2 weeks. By the end of the second postoperative week, the pack of the ear canal is removed, with reminding the parents to come every week for removal of crusts and repeated repacking for at least 6 weeks. Packing is done with sterile gauze impregnated with antibiotic and steroid cream. Dressing of the donor site is removed by the end of the third postoperative week. Those cases need follow-up for life for cleaning of the ear canal and removal of crusts, debris, and wax.[4,14]

COMPLICATIONS OF SURGERY

Patients may suffer from deterioration of hearing after they have experienced satisfactory hearing level; it is caused by tympanic membrane lateralization. Its incidence is about 5%.[4] Various attempts have been paid to avoid this problem. A tab is performed in the temporalis fascia graft to be tucked in the anteroinferior part of middle ear space.[1] A tunnel is performed at the anterior part of the medial end of EAC, in which the temporalis fascia graft is inserted.[4] A silastic disc is used by some surgeons to support the temporalis fascia graft in place.[1] Also, the anesthesiologist is instructed to decrease the expired oxygen to 25% or less; it is preferable to maintain the patient on room air (if possible), as expired oxygen concentrations greater than 30% may cause ballooning of the graft.[12]

Stenosis of the EAC may occur in the bony part of the EAC due to granulation tissue formation, which can be avoided by complete coverage of the raw bone with skin graft. Also, the cartilaginous part may stenose secondary to inadequate removal of auricular cartilage during creation of the external meatus. Its incidence is variable in the literature, ranging between 3% and 30%. Postoperative long-term stenting and repeated repacking have been used to prevent this complication.[4,20]

High-tone SNHL may be encountered in up to 7%.[1] It is presumably caused by drilling on the ossicular chain when dissecting it away from the atretic bone.[21]

A major devastating complication is the facial nerve paralysis. It is due to trauma to facial nerve, which may take an aberrant course in some cases. Fortunately, it is rare (1%), and it can be minimized with careful dissection and intraoperative facial nerve monitoring.[14,16]

Chronic infection of the newly constructed EAC may occur as a result of the lack of normal keratin migration in the skin-grafted canal and the lack of production of protective cerumen. It may occur in 17% of patients; however, this problem can be minimized by creating a widely patent meatus, and the patients should be counseled on aural hygiene and recommended to return regularly for microscopic debridement.[4,21]

CONCLUSIONS

Congenital aural atresia is a congenital disorder that can affect the child's communication and consequently seriously affect the social life of his/her family, especially in bilateral cases. It can occur as an isolated deformity or may be part of various syndromes. In bilateral cases, hearing amplification is mandatory in the early few years of life for normal speech and language development. Counseling of the parents is a part of the management; it includes discussion with parents about the disease, its associated anomalies (if present) and operative and postoperative complications. Evaluation of patients with auditory brain stem response audiometry for measuring the cochlear function and CT for delineation of temporal bone anatomy should be performed as a preoperative requisite. Operative intervention is a challenge to the otologic surgeon. It comprises 3 important steps: canaloplasty, which is a creation of new EAC; tympanoplasty, which includes restoration of the conductive hearing mechanism of the middle ear; and meatoplasty, which entails opening of the external skin to make an external meatus. Complications of surgery that can be minimized or even avoided may include tympanic

membrane lateralization, restenosis of the EAC, SNHL, facial nerve palsy, and repeated infection.

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Frontonasal and Fibrous Dysplasia in a Patient With Unilateral Cleft Lip and Palate

William M. Weathers, MD, Erik M. Wolfswinkel, BS, Steven B. Albright, MD, Larry H. Hollier Jr, MD, Edward P. Buchanan, MD

Abstract: Frontonasal dysplasia is a rare entity. It has characteristic physical deformities: hypertelorism, broad nasal root, median facial cleft of the upper lip or palate, clefting of the nasal alae, poorly formed nasal tip, cranium bifidum occultum, and a widow's peak hairline. Fibrous dysplasia is a benign bone tumor in which normal bone is replaced by fibrous, poorly formed osseus tissues. We present a patient with frontonasal dysplasia who desired correction of her hypertelorism. Incidentally, fibrous dysplasia was found in her left orbit complicating surgical correction. In addition, the patient has velopharyngeal insufficiency and a class III malocclusion. The interplay of all these craniofacial defects makes the sequencing and timing of surgery important in this unique patient.

Key Words: Fibrous dysplasia, frontonasal dysplasia, box osteotomies

Frontonasal dysplasia is a rare condition characterized by hypertelorism and nasal abnormalities. It was first described 1970 but has since been refined to encompass a constellation of physical findings. These include 2 or more of the following: hypertelorism, broad nasal root, median facial cleft of the upper lip or palate, clefting of the nasal alae, poorly formed nasal tip, cranium bifidum occultum, and a widow's peak hairline.[1,2] Other case series and reports have shown associations with heart anomalies, mental delay, optic disc anomalies, and central nervous system anomalies demonstrating a wide spectrum of the disease.[3–6] Fibrous dysplasia is a benign bone tumor in which normal bone is replaced by fibrous, poorly formed osseus tissues.[7] It can be a solitary lesion or include multiple bony sites. The involved bone is fragile and predisposed to pathologic fractures.[8] In this case report, we present a patient with frontonasal dysplasia and fibrous dysplasia of the frontal, sphenoidal, and ethmoidal bones.

CLINICAL REPORT

An 18-year-old female patient presented to our craniofacial clinic with a chief complaint of hypernasality. Her medical history was significant for frontonasal dysplasia and left unilateral cleft lip and

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