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Preoperative evaluation and surgical outcomes of congenital aural atresia

Doğuştan aural atrezinin ameliyat öncesi değerlendirilmesi ve cerrahi sonuçları

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ABSTRACT

Objectives: In this study we aimed to investigate the effect of preoperative evaluation on functional surgical outcomes in congenital aural atresia surgery which is one of the most challenging procedures of otolaryngology.

Patients and Methods: Twelve patients (8 males, 4 females; mean age 12.9±6.8 years; range 6 to 32 years) who underwent surgery for unilateral congenital aural atresia in our clinic between October 2007 and April 2013 were included in this study. All patients were performed pre- and postoperative physical examination, and audiologic and radiologic evaluation. Patients were preoperatively evaluated according to Altmann's classification and Jahrsdorfer's surgery indication classification. All patients were operated under general anesthesia using transatretic (anterior) approach.

Results: According to preoperative evaluation, three patients had Altmann type 1 minor malformation and nine patients had type 2 moderate malformation. No patient had type 3 major malformation. Congenital cholesteatoma was detected in two patients. According to Jahrsdorfer's surgery indication classification, three patients were excellent, four patients were very good, four patients were good and one patient was fair candidates for functional surgery. Mean air-bone gap was 43±4.1 (range 38-50) dB at postoperative sixth week, 36.3±3.4 (range 30-42) dB at postoperative third month, 30.4±2.6 (range 28-35) dB at postoperative sixth month, and 30.2±2.1 (range 25-35) at postoperative 12th month. One patient (9%) developed postoperative infection, and two patients (18%) developed restenosis.

Conclusion: Preoperative evaluation of patients in congenital aural atresia surgery is of vital importance in terms of surgical success. Obtaining successful surgical and functional outcomes is strongly related to preoperative evaluation and postoperative follow-up along with surgical technique and surgeon's experience.

Keywords: Atresia; external ear canal; preoperative evaluation.

ÖZ

Amaç: Bu çalışmada otolarengolojinin en zor işlemlerinden biri olan doğuştan aural atrezi cerrahisinde ameliyat öncesi değerlendirmenin fonksiyonel cerrahi sonuçlar üzerine etkisi araştırıldı.

Hastalar ve Yöntemler: Bu çalışmaya Ekim 2007 - Nisan 2013 tarihleri arasında kliniğimizde tek taraflı doğuştan aural atrezi nedeni ile ameliyat edilen 12 hasta (8 erkek, 4 kız; ort. yaş 12.9±6.8 yıl; dağılım 6-32 yıl) dahil edildi. Tüm hastalara ameliyat öncesi ve sonrası fizik muayene ile odyolojik ve radyolojik değerlendirme yapıldı. Hastalar ameliyat öncesi Altmann sınıflaması ve Jahrsdorfer cerrahi endikasyon sınıflamasına göre değerlendirildi. Hastaların tümü transatrezik (anterior) yaklaşım kullanılarak genel anestezi altında ameliyat edildi.

Bulgular: Ameliyat öncesi değerlendirmeye göre, üç hastada Altmann tip 1 minör malformasyon ve dokuz hastada tip 2 orta malformasyon vardı. Hiçbir hastada tip 3 majör malformasyon yoktu. İki hastada doğuştan kolesteotom saptandı. Jahrsdorfer'in cerrahi endikasyon sınıflamasına göre, fonksiyonel cerrahi için üç hasta mükemmel, dört hasta çok iyi, dört hasta iyi ve bir hasta orta adaylardı. Ortalama hava-kemik aralığı ameliyat sonrası altıncı haftada 43±4.1 (dağılım 38-50) dB, üçüncü ayda 36.3±3.4 (dağılım 30-42) dB, altıncı ayda 30.4±2.6 (dağılım 28-35) dB ve 12. ayda 30.2±2.1 (dağılım 25-35) dB idi. Bir hastada (%9) ameliyat sonrası enfeksiyon, iki hastada (%18) restenoz gelişti.

Sonuç: Doğuştan aural atrezi cerrahisinde hastaların ameliyat öncesi değerlendirilmesi cerrahi başarı açısından büyük önem taşır. Cerrahi ve fonksiyonel açıdan başarılı sonuçlar elde etmek cerrahi tekniği ve cerrahın deneyiminin yanı sıra ameliyat öncesi değerlendirme ve ameliyat sonrası takibe bağlıdır.

Anahtar sözcükler: Atrezi; dış kulak kanalı; ameliyat öncesi değerlendirme.

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Congenital aural atresia (CAA) is a term describing external and middle ear malformations that are encountered one in 10,000 to 20,000 births. Congenital aural atresia is mostly one-sided (70-90%) with a high incidence of involvement of the right ear (58-61%), and occurs twice more frequently in male patients.[1,2] External ear atresia or stenosis occurs due to lack of recanalization of the outer ear plug. In addition to external auditory canal atresia and stenosis, CAA often involves the middle ear, ossicular anomalies and microtia. Congenital aural atresia is accompanied by a syndrome in 10% of patients and as many as 6-10% of patients have concomitant inner ear malformations.[2] The main choice of treatment in CAA is surgery which is often performed for both functional and cosmetic purposes. Aside from application of modern tympanoplasty techniques and mastery of fenestration, knowledge of possible anatomic deviations of the malformed ear, middle ear structures, and facial nerve are essential.[2,3] Therefore, patients with CAA constitute a group that should be evaluated properly prior to surgery and should also be followed-up carefully in the postoperative period.

PATIENTS AND METHODS

In this study, 12 patients (8 males, 4 females; mean age 12.9±6.8 years; range 6 to 32 years) who were operated for one-sided CAA between October 2007 and April 2013 in the Ear Nose and Throat Department of Necmettin Erbakan University were included. This study was conducted in accordance with the principles of the Helsinki Declaration. The study protocol was approved by the local institutional review board. Parents of each patient signed an informed consent form. The right side was affected in six patients (50%) and the left side was affected in six patients also (50%). A detailed medical history and examination of the ear, nose and throat (Figure 1) and audiometric assessment was performed preoperatively for both ears and bone conduction was recorded. All patients were evaluated with preoperative and postoperative high-resolution temporal computed tomography (CT). The early postoperative (six-week), three-month, and long-term (6- and 12-month) hearing was measured and recorded including air conduction, bone conduction and air-bone gap levels. Middle ear ossicles, course of facial nerve, inner ear, temporal bone pneumatization degree and location of the oval window, presence of base of the stapes were evaluated preoperatively (Figure 2). In addition, the layer thickness and the shape of atretic bone on high-resolution CT, relationship between soft tissue and atresia and presence of congenital cholesteatoma were investigated. All of the patients were classified before the operation according to the classification system of Altmann and Jahrsdorfer, with indications for surgery on a 10-point scale based on radiological findings (Table 1). We evaluated all of the patients before surgery. Based on mastoid pneumatization, middle and inner ear condition, cartilage development, psycho-social problems engendered by pathology, and taking into account the child's cooperation, surgery was performed at the most appropriate time.

All patients were operated on using a transatretic (anterior) approach with facial nerve monitoring under general anesthesia via orotracheal intubation. Following postauricular incision, keeping the linea temporalis superior, the root of the zygomatic arch and mandibular condyle anterior, and the mastoid cells posterior, the atretic plates were drilled anteromedialy to the epitympanum identifying the middle fossa plate to form the new external auditory canal. The atretic bone plate was removed without any damage to the inner ear. In all cases, after the separation of the ossicular chain and after obtaining sufficient width of the newly created external auditory canal, type 1 tympanoplasty was performed using a temporalis muscle fascial graft. A split-thickness dermal graft was placed on top, circumferentially sutured to the skin of the newly created external ear canal and supported by Merocel ear pack (Medtronic Xomed, Inc., FL, USA).

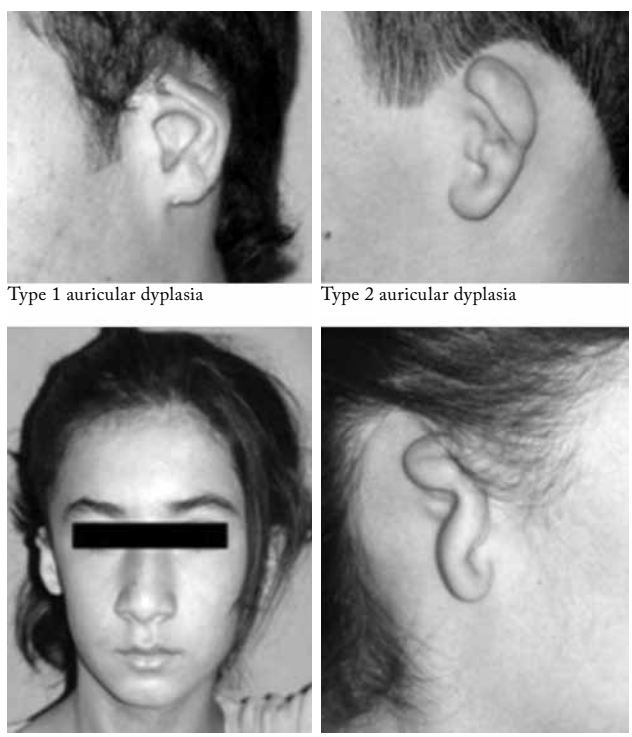
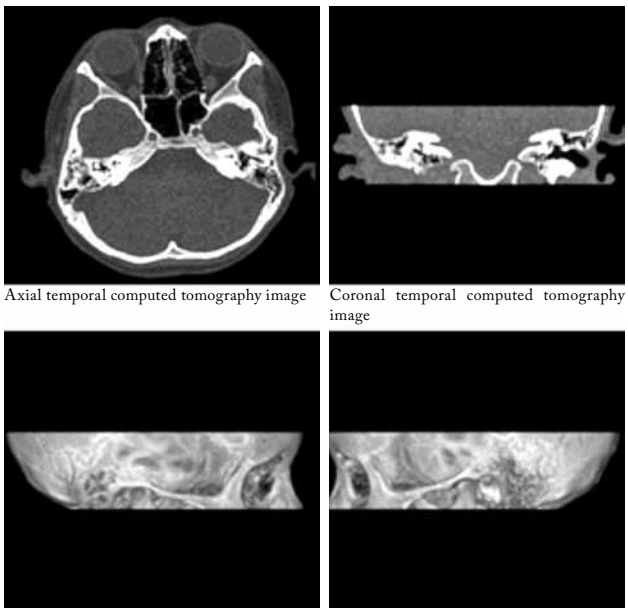


Figure 1. Preoperative pictures of patients.



Axial temporal computed tomography image | Coronal temporal computed tomography image

Three dimensional temporal computed tomography image (right atretic ear) | Three dimensional temporal computed tomography image (normal left ear)

Figure 2. Temporal computed tomography and three-dimensional images of patients.

RESULTS

According to Jahrsdorfer surgical indication staging, three patients were excellent, four patients were very good, four patients were good and one patient was a fair candidate(s) for functional surgery. According to Altmann staging, three patients had type 1 minor malformations and nine patients had type 2 moderate malformations. No type 3 major malformations were found in any patient. Audiological evaluation showed all patients had preoperative normal bone hearing thresholds on the pathological side, air and bone hearing levels in the contralateral ear were within normal limits. According to data obtained during the operation, five patients had malleus and incus fusion blocks and seven patients had near-normal anatomy. In two patients the facial nerve was turning with narrow-angle and was uncovered at the tympanic segment. There was a congenital middle ear cholesteatoma in two patients. There were no early iatrogenic complications like facial paralysis and sensorineural hearing loss encountered during surgery. One patient (9%) developed postoperative infection, two patients (18%) developed restenosis in the fifth postoperative month and 12th postoperative month, respectively. Both patients who developed restenosis were male and eight years old. The average postoperative air-bone gap was 43±4.1 dB (range 38-50) at six weeks, 36.3±3.4 dB (range 30-42) at three months, 30.4±2.6 dB (range 28-35) at six months, and 30.2±2.1 dB (range 25-35) at one year (Table 2).

Table 1
Jahrsdorfer's grading scale

Anatomical structure	Score
Stapes bone	2
Oval window open	1
Middle ear space	1
Facial nerve	1
Malleus and incus	1
Mastoid pneumatization	1
Incudo-stapedial connection	1
Round window	1
External ear	1
Total possible score	10

Table 2
Patient's scores according to Jahrsdoerfer's preoperative surgical indication classification

		Patients											
		1	2	3	4	5	6	7	8	9	10	11	12
1	Stapes bone	2	2	2	2	2	2	2	2	2	2	2	2
2	Oval window open	1	1	1	1	1	1	1	1	1	1	1	1
3	Middle ear space	1	1	1	1	1	1	1	1	1	1	1	1
4	Facial nerve	0	1	1	1	0	1	1	0	1	1	1	1
5	Malleus and incus	1	0	1	0	0	1	1	1	1	0	1	1
6	Mastoid pneumatization	0	0	0	1	0	1	1	0	0	0	1	0
7	Incudo-stapedial connection	1	1	1	1	1	1	1	1	1	1	1	1
8	Round window	1	1	1	1	1	1	1	1	1	1	1	1
9	External ear	0	0	0	0	0	0	0	0	0	0	0	0
	Total	7	7	8	8	6	9	9	7	8	7	9	8

Table 3
Air-bone gap (dB) on the side of the pathology

Postoperative period	1	2	3	4	5	6	7	8	9	10	11	12
Sixth week	42	45	46	38	50	45	35	40	45	42	44	46
Third month	38	34	40	32	35	42	30	36	38	34	37	38
Sixth month	35	25	32	32	30	25	28	32	29	30	30	32
One year	32	25	32	30	32	25	30	30	32	35	30	25

DISCUSSION

The main purpose of surgery is to provide a useful-hearing ear and external auditory canal without infection. Useful hearing levels can be obtained when the difference of the speech reception threshold is ≤30 dB and the difference of speech discrimination score is ≤15% between the pathologic and healthy ear.[4,5] According to recent studies children with unilateral hearing loss are at risk in terms of speech, language development, school performance, and attention disorder.[6,7] The main purpose of surgery in patients with CAA is providing binaural hearing levels. Binaural hearing is important to determine direction of sounds and detect speech.[8,9] Atresia surgery can be applied in bilateral cases which cannot benefit from hearing aids. However, these cases usually have more severe anomalies and surgical success rates are low.[10]

The presence of malformations like facial paralysis, anotia and severe mandibular hypoplasia are contraindications for functional surgery. As a general rule, in congenital ear malformations, the severity of deformity in the auricle and external auditory canal is parallel to degree of abnormality of the middle ear. Development of the mastoid apex is a good clinical determinant of mastoid pneumatization and development of the middle ear.[11,12] Several classifications exist for preoperative evaluation of CAA. Tanzer divided auricular anomalies into five groups. Jahrsdorfer divided them into three degrees. Nagata divided them into four groups, considering surgical planning. Today, Altmann's classification based on the anatomical malformation is frequently used. According to this classification malformations are divided into three types: type 1 - minor malformations, type 2 - moderate degree malformations (most common type of congenital aural atresia and the most suitable for the functional surgery), type 3 - severe malformations (contraindication for functional surgery). The transatretic and transmastoid approach can be used for the surgical treatment of EEC (external ear canal) atresia.

Today, the anterior approach is the more preferred technique in treatment of CAA.[13] In this study, all patients were operated on via the transatretic approach. The most common causes of postoperative failures are recurrent infections. Conductive hearing loss reoccurs as a result of refixation of ossicles, canal stenosis and lateralization of the tympanic membrane.[4,14] In our study, one patient developed postoperative infection and improved with antibiotic therapy. Two patients developed postoperative restenosis. Both patients were male, and postoperative restenosis developed 5 and 12 months after surgery respectively. Each case was operated on under general anesthesia, and a Pezzer 22FR catheter was placed in the external ear canal in order to maintain it open. The catheter was removed six weeks later. Depending on the age of the first operation, postoperative meatal stenosis is more common in younger children because canaloplasty often stimulates the proliferation of granulation and bone tissue in response to surgical trauma.[15] In our study, both patients who developed restenosis were eight years old. According to studies of postoperative meatal stenosis our cases seen at 12 and 18 months are consistent with the physiology of scarring and time of contracture.[7] Although it is difficult to predict postoperative results, restenosis rates ranged between 8% and 29%[1] while in our study, this ratio was 18%. Battelino et al.[16] reported that mitomycin-C application during the operation reduces the rates of postoperative stenosis According to Jahrsdorfer, a full-thickness skin flap from the anterior conchal region of the reconstructed auricle can be used to create the anterior part of the external canal. This provides adequate meatal opening by removing the circular suture line of a split thickness skin graft and the incidence of stenosis decreases.[17] According to the study carried out by De La Cruss et al.[1] 1985 a significant reduction in postoperative canal stenosis is achieved by using split thickness skin graft instead of full-thickness. Regardless of whether split-thickness or full-thickness graft is used, formation of scar tissue and epithelial debris narrows the external ear canal in the initial postoperative period.[18] In order to cover the surface of the bone two pedicled chondrocutanous grafts were used in some cases.[19] An anterior-inferior based periosteal flap significantly decreases the incidence of meatal stenosis, $p<0.05$.[20]

Out of 60-70% patients who underwent surgery, less than 30 dB air-bone gap can be obtained in the early period. But in the long term as many as 30% of patients with useful hearing levels can be achieved only after primary and revision surgeries.[14,21,22] In our study, air-bone gap levels obtained at postoperative sixth week, third month and sixth month were 43±4.1 dB (range 38-50), 36.3±3.4 dB (range 30-42) and 30.4±2.6 dB (range 28-35) and 30.2±2.1 dB (range 25-35) respectively and air-bone gap levels decreased over time. According to Hall and Jahrsdorfer the postoperative speech perception threshold in patients with CAA ranged from 15-25 dB but other otologists could not achieve these results.[23]

Conclusion

Preoperative evaluation of CAA cannot be ignored when it is considered that it has greater contribution to the success of surgery. To achieve successful results in terms of surgical and functional aspects depends on the surgical technique of operation and the surgeon's experience, as well as preoperative evaluation and postoperative follow-up. In cases where restenosis developed after surgery, it may be a good treatment option to place a catheter for 6-8 weeks in order to obtain an open external auditory canal.

Declaration of conflicting interests

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