

Original Article

Acoustic Reflex After Surgical Repair in Patients with Congenital Aural Atresia

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Cite this article as: Kim MB, Cho Y. Acoustic reflex after surgical repair in patients with congenital aural atresia. *J Int Adv Otol.* 2022;18(6):482-487.**BACKGROUND:** This study aimed to examine the plasticity of nerves indirectly by acoustic reflex after surgical repair of unilateral congenital aural atresia.**METHODS:** This study is a retrospective study including 80 patients who had undergone surgical repairs of congenital aural atresia before 18 years old and for whom acoustic reflex testing was performed postoperatively. Several variables correlated with acoustic reflex were analyzed to find factors affecting the presence of acoustic reflex.**RESULTS:** Among 80 patients, 44 were positive for acoustic reflex. As a result of multivariate analysis, good postoperative hearing ($P = .017$), younger age at surgery ($P = .028$), and the longer time interval between surgery and acoustic reflex test ($P = .040$) were revealed as factors affecting the acoustic reflex.**CONCLUSION:** Acoustic reflex was present in 55% of surgically managed patients with congenital aural atresia. The results of this study imply that the reflex arc of acoustic reflex may be restored after proper surgical reconstruction and prolonged use of reconstructed ear in patients with unilateral congenital aural atresia.**KEYWORDS:** Congenital aural atresia, canalplasty, acoustic reflex

INTRODUCTION

Congenital aural atresia (CAA) is a disorder that is characterized by varying degrees of congenital hypoplasia of the external auditory canal (EAC). Congenital aural atresia is often accompanied by malformations of the auricle, middle ear, or inner ear. Congenital aural atresia occurs in about 1 per 10 000-20 000 live births, of which about 70% are unilaterally involved.¹ In addition to cosmetic problems, the patients suffer from severe conductive hearing impairment on the side of the lesion from birth.

Management options that can resolve these conductive hearing losses (CHL) include bone conduction hearing devices, middle ear implants, and surgical reconstruction. Among these options, canalplasty has the advantage of rebuilding the natural sound conduction pathway and reducing the need for additional hearing devices.² In previous studies, it has been reported that there is an improvement of hearing of about 20-30 dB and, in about half of patients, the air-bone gap (ABG) is corrected within 30 dB after canalplasty.^{2,3} Therefore, recent consensus has recommended canalplasty as a desirable treatment method for CAA, if the Jahrsdoerfer score on preoperative computed tomography (CT) is 7 or higher.⁴

In case of congenital sensorineural hearing loss, deterioration of the central auditory pathway always occurs, and this is the same in unilateral hearing loss.⁵ In addition, it has been reported that restoring hearing at an early age is important for the proper development of the central auditory system.^{6,7} However, the status of the central auditory system in cases with CAA has not been sufficiently investigated, and the effect of hearing restoration on the development of the central auditory system is also unknown.⁸

Acoustic reflex (AR), the reflexive contraction of the intratympanic muscles resulting from high-intensity sound stimulation, is a non-invasive physiologic test that can evaluate the auditory brainstem function.^{9,10} A recent study reported that the appearance of postoperative AR was associated with postoperative hearing thresholds in CAA patients, but it is not known what other factors affect it.⁸

This study aimed to investigate the plasticity of the central auditory pathway indirectly using AR after surgical repair of unilateral CAA and to identify the factors that influence the presence of AR.

METHODS

Subjects

A total of 96 patients who had undergone canalplasty by a single surgeon between 2009 and 2019 to treat unilateral CAA and tested for AR after surgery were enrolled in the study. Canalplasty was performed only on patients with Jahrsdoefer scores of 7 or more in the preoperative temporal bone CT scan. Reconstruction of sound conduction was attempted using intact native chain reconstruction (INCR), a partial ossicular replacement prosthesis (PORP), and a total ossicular replacement prosthesis (TORP) according to the status of the ossicular chain. Patients with central auditory disorder, middle ear disease, or facial nerve disorder, or who were over 18 years of age at the time of surgery were excluded, and 85 patients were eventually included. The research was conducted under common ethical rules and was approved by the Institutional Review Board of a tertiary referral center (IRB No: 2020-03-054).

Surgical Technique

We performed the surgery in the same way as previously reported.³ The anteriorly based skin flap was elevated and cartilage was removed with soft tissue. Periosteum on the mastoid cortex was preserved and the anteriorly, superiorly, and inferiorly based periosteal flaps were elevated to expose the mastoid cortex (Figure 1). The new ear canal was made at the junction of the linea temporalis and the superior aspect of the posterior rim of the glenoid fossa by drilling, which proceeded medially through the atretic plate and into the epitympanum. The atretic plate was removed with diamond burrs, curettes,

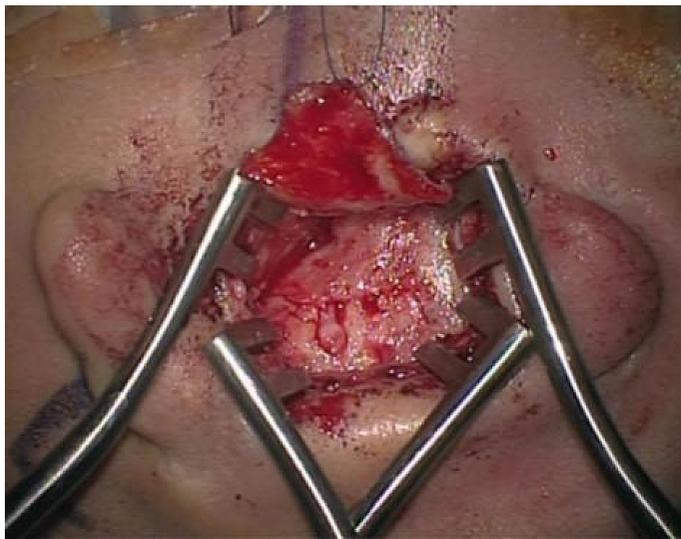


Figure 1. Exposure of the mastoid cortex.

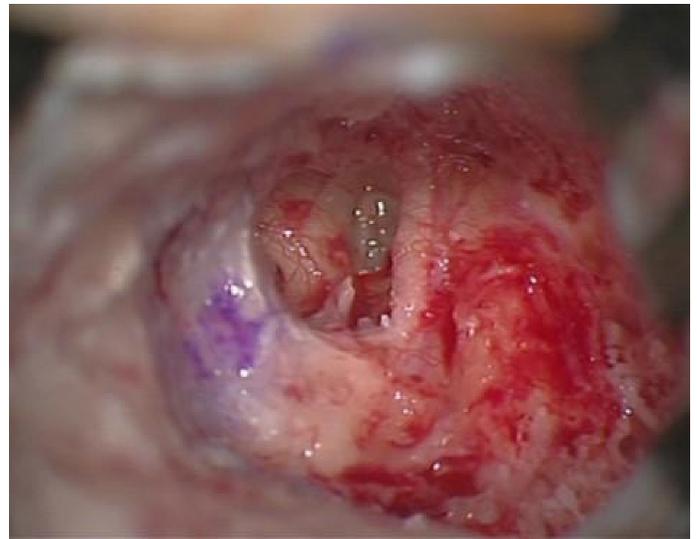


Figure 2. Middle ear structures were assessed. Violet dye indicates the course of the facial nerve.

or laser, and the mobility of ossicular chain was assessed (Figure 2). If necessary, the ossicular reconstruction was performed (Figure 3). Exposed air cells were covered with soft tissue and sliced cartilage. We made the tympanic membrane using an onlay graft of the temporalis fascia (Figure 4). A split-thickness skin graft was obtained at 0.008-0.010 inch thick in 6 × 5 cm size from the inner thigh using a dermatome and covered the newly made EAC (Figure 5). The outer end of skin graft was sutured to the skin and pieces of Merocele® wick (Medtronic Xomed Surgical Products, Jacksonville, Fla, USA) were placed in the EAC and hydrated to expand (Figure 6).

Acoustic Reflex Test and Audiometric Evaluation

Postoperative AR was evaluated at regular follow-up visits from February 2017 to January 2020. Ipsilateral AR (IAR) was checked in the normal ear with sound stimulation to the same ear, while contralateral AR (CAR) was checked in the normal side with stimulation to the operated ear. All AR was recorded with pure tone stimuli at 500, 1000, 2000, and 4000 Hz using a GSI TympStar middle ear analyzer (Grason-Stadler, Eden Prairie, Minn, USA). Stimuli were initially

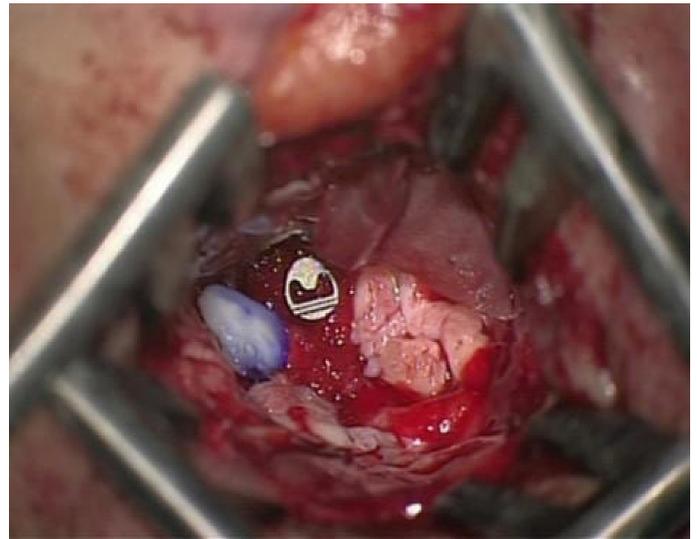


Figure 3. Placement of ossicular prosthesis and cartilage slice.

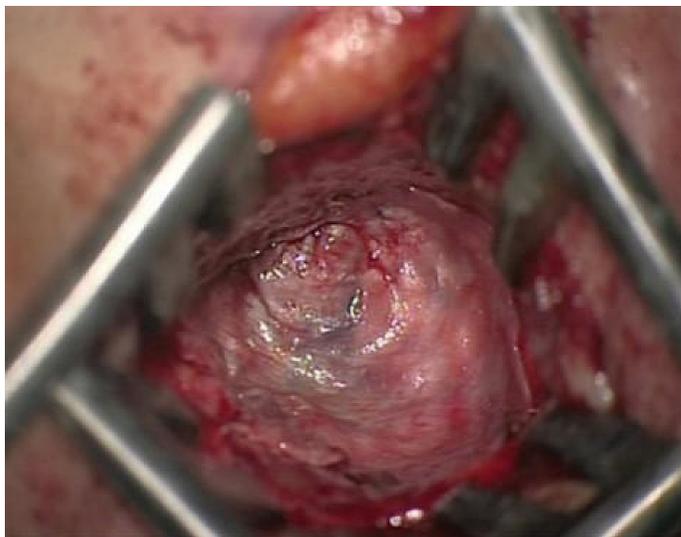


Figure 4. Tympanic membrane was made using onlay graft of the temporalis fascia.

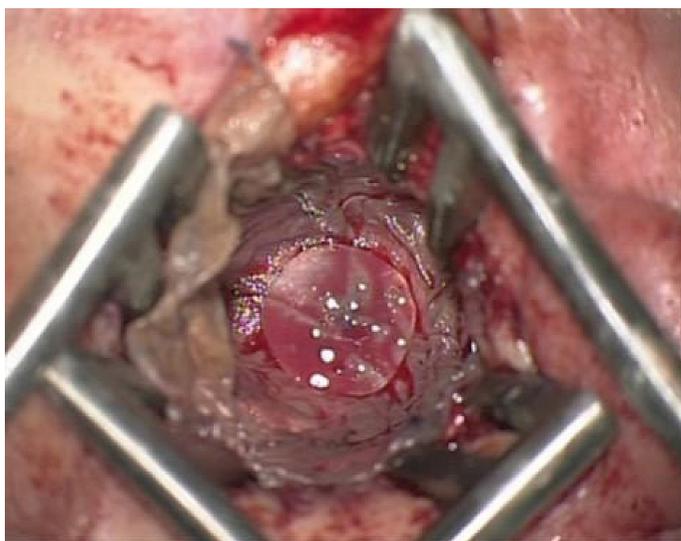


Figure 5. Split-thickness skin graft was draped with a silastic sheet.



Figure 6. Skin graft was sutured and Merocel® packing was placed.

presented at 80 dB and increased to 5 dB and step up to 120 dB until an AR was measured. The AR was regarded as absent if not detected at 120 dB stimuli.

Pure tone audiometry was also performed at the same time, and the hearing thresholds and 4-frequency pure tone average (PTA) were obtained at 500, 1000, 2000, and 4000 Hz.

Analysis of Factors Influencing the Presence of Acoustic Reflex

Patients were grouped according to the presence of CAR, and the mean value of pure tone threshold was compared between groups for each frequency as well as the PTA.

Other factors that might be associated with the presence of AR were also investigated. The factors analyzed included sex, age at surgery, side of operation, severity of aural atresia according to the Schuknecht classification,¹ procedures for hearing restoration, and time interval between the operation and AR test.

Statistical Analysis

All analysis was performed using the International Business Machines Statistical Packages for the Social Sciences Statistics Data Editor Version 26.0 (IBM SPSS Corp.; Armonk, NY, USA). For group comparisons of continuous variables, independent *t*-tests were applied, and for those of categorical variables, chi-square tests and Fisher’s exact tests were used, as appropriate. Variables in multivariate analysis were selected based on a *P*-value <.10 from univariable analysis results. *P*-values below .05 were considered statistically significant.

RESULTS

Demographics

This study included 65 male (76.5%) and 20 female participants. Fifty-one patients (60%) had right-sided aural atresia and the others had a left-sided anomaly. The average age at surgery was 11.47 ± 2.93 years, and the mean time interval between surgery and the AR test was 40.49 months (ranging from 1 to 119 months). According to the Schuknecht classification, type C was most common, being

Table 1. Demographic Information of Enrolled Patients

	n (%)	
Sex	Male	65 (76.5)
	Female	20 (23.5)
Affected side	Right	51 (60)
	Left	34 (40)
Schuknecht type	Type B	16 (18.8)
	Type C	69 (81.2)
Method of hearing restoration	INCR	27 (31.8)
	PORP	46 (54.1)
	TORP	10 (11.8)
	Others	2 (2.3)
Age at surgery, mean ± SD	11.47 ± 2.93	
Time interval between surgery and AR test, mean (range)	40.49 months (1-119)	

INCR, intact native chain reconstruction; PORP, partial ossicular replacement prosthesis; TORP, total ossicular replacement prosthesis; AR, acoustic reflex.

observed in 69 ears (81.2%) followed by type B in 16 ears (18.8%). The procedure for hearing restoration was INCR in 27 patients, PORP in 46 patients, and TORP in 10 patients. Tympanoplasty without ossicular chain reconstruction was performed in 2 patients due to an invisible stapes. (Table 1).

Presence of the Acoustic Reflex

Out of 85 total patients, IAR was absent in 5 patients and CAR was absent in these subjects. Among the 80 patients who demonstrated positive IAR, 44 (55%) showed CAR at 1 or more frequencies. Contralateral acoustic reflex was most frequently collected at 1000 and 2000 Hz (38 patients, 44.7% for each frequency), followed by 500 Hz (21 patients, 24.7%) and 4000 Hz (eight patients, 9.4%). Eight patients who tested negative for CAR underwent an AR test again after an average of 2 years, and 5 of them showed positive CAR on the second AR test.

The postoperative mean air conduction (AC) PTA of 80 patients who had IAR was 44.14 ± 15.43 dB and the mean ABG was 34.56 ± 11.44 dB. The mean AC threshold of 44 patients with CAR was better (41.08 ± 14.16 dB) than that of patients without CAR (47.88 ± 16.26 dB) (*P* = .049). This difference is also observed when analyzed by frequencies, except for 500 Hz. When ABG was analyzed, no difference was found between the patients with and without CAR (*P* = .26). The average age at surgery was 10.77 ± 2.66 years old in patients in whom CAR was present and 12.44 ± 2.84 in patient in whom CAR was absent

Table 2. Comparison Between Patients With or Without Contralateral Acoustic Reflex

Variable	Overall	Contralateral Acoustic Reflex		<i>P</i>
		Absent (n = 36)	Present (n = 44)	
Gender*				
Male	61 (76.3)	26 (72.2)	35 (79.6)	.44
Female	19 (23.7)	10 (27.8)	9 (20.4)	
Age#	11.5 (2.8)	12.4 (2.8)	10.8 (2.6)	<.001
Side of operation*				
Right	47 (58.8)	20 (55.6)	27 (61.4)	.60
Left	33 (41.2)	16 (44.4)	17 (38.6)	
Schuknecht*				
Type B	13 (16.3)	5 (13.9)	8 (18.2)	.60
Type C	67 (83.7)	31 (86.1)	36 (81.8)	
Type (n = 78)*				
INCR	25 (32.1)	15 (44.1)	10 (22.7)	.12
PORP	43 (55.1)	16 (47.1)	27 (61.4)	
TORP	10 (12.8)	3 (8.8)	7 (15.9)	
PTA_AC#	44.1 (15.4)	47.9 (16.3)	41.1 (14.2)	.049
PTA_ABG#	34.6 (11.4)	36.1 (11.0)	33.3 (11.7)	.26
Duration#	38.0 (33.2)	31.1 (24.8)	43.7 (38.1)	.08

*Number (%); #Mean (standard deviation); The bold values indicate statistically significant results.

PTA, pure tone audiometry; AC, air conduction; ABG, air-bone gap; AR, acoustic reflex; INCR, intact native chain reconstruction; PORP, partial ossicular replacement prosthesis; TORP, total ossicular replacement prosthesis.

(*P* = .008). The average time interval between operation and AR test (*P* = .081), gender (*P* = .598), affected side (*P* = .652), Schuknecht type (*P* = .872), and method of hearing restoration (*P* = .111) was not different between groups. The results of group comparisons are presented in Table 2.

Factors Associated with the Presence of Contralateral Acoustic Reflex

To find out which factors affect the presence of AR, univariable and multivariate logistic regression analyses were performed. The variables were postoperative AC threshold, ABG, age at surgery, time interval between operation and AR test, method of hearing restoration, gender, affected side, and the Schuknecht type of CAA. Results from univariable analysis revealed that age at surgery was significantly associated with the presence of AR during longitudinal follow-up (*P* = .01, odds ratio [OR]: 0.80, and 95% CI: 0.67-0.95). Variables with a *P*-value <.10 from univariable analysis results were selected in multivariate analysis. After multivariate analysis, good postoperative hearing (*P* = .03, OR: 0.96, CI: 0.93-0.996), younger age at surgery (*P* = .01, OR: 0.79, CI: 0.65-0.95), and longer time interval between surgery and AR test (*P* = .04, OR: 1.02, CI: 1.001-1.03) were revealed as factors that positively affected the presence of AR. The results of logistic analysis are presented in Table 3.

DISCUSSION

Congenital aural atresia is a condition that causes severe congenital unilateral auditory deprivation, and it is thought that some parts of the AR arc may be affected due to possible deterioration of auditory stimulation since birth.

Table 3. Results of Uni- and Multivariable Regression Analysis

Variable	Univariable Analysis		Multivariate Analysis	
	OR (95% CI)	<i>P</i>	OR	<i>P</i>
Gender				
Male	1.5 (0.53-4.21)	.45		
Female	Reference			
Age	0.80 (0.67-0.95)	.01	0.79 (0.65-0.95)	.01
Side of operation				
Right	1.27 (0.52-3.11)	.60		
Left	Reference			
Schuknecht				
Type B	1.38 (0.41-4.65)	.61		
Type C	Reference			
Type		.13		
INCR	Reference			
PORP	2.53 (0.92-6.96)	.55		
TORP	3.50 (0.73-16.84)	.28		
PTA_AC	0.97 (0.94-1.00)	.06	0.96 (0.93-0.996)	.03
PTA_ABG	0.98 (0.94-1.02)	.26		
Duration	1.01 (1.00-1.03)	.10	1.02 (1.001-1.03)	.04

The bold values indicate statistically significant results.

PTA, pure tone audiometry; AC, air conduction; ABG, air-bone gap; AR, acoustic reflex; INCR, intact native chain reconstruction; PORP, partial ossicular replacement prosthesis; TORP, total ossicular replacement prosthesis.

There are several studies that reported AR restoration after cochlear implantation in patients with sensorineural hearing loss. When the hearing threshold is recovered to almost a normal level after cochlear implantation, it is reported that CAR is restored at about 60%-70%.¹¹⁻¹³ However, the appearance of AR after surgical correction in congenital CHL has not been sufficiently reported.

Sharma et al⁷ reported that it is important to perform cochlear implantation surgery at a younger age to achieve better development of the central auditory system in deaf children. Moreover, Vanderauwera et al⁶ reported that unilateral hearing loss also affects the neural network and that early hearing restoration is important to achieve proper neuroplasticity.

In the case of CHL, however, the effect of corrective surgery on neuroplasticity has not been investigated enough. According to Byun et al.⁵ hearing in noise significantly recovered in teenagers after surgical correction of unilateral CAA, because patients with CAA had less auditory deprivation than those with severe to profound sensorineural hearing loss due to the presence of continuous internal sound stimulation.

The AR refers to the reflexive contraction of the intratympanic muscles, especially the stapedius muscle, resulting from intense sound stimulation. The main physiologic role of AR is to protect the cochlea from high-intensity sound by making the ossicular chain stiffer and reducing the penetration of sound energy reaching the inner ear. The reflex is activated bilaterally to protect both inner ears, and the cochlear nerve and the facial nerve are involved in this reflex arc. The auditory stimulation is delivered to the cochlear nerve and reaches the ventral cochlear nucleus and trapezoid body, where it is transmitted to the ipsilateral and contralateral superior olivary complexes of the brain stem. From there, neurons proceed to the facial nerve nucleus, from which motor neurons of the facial nerve contract the stapedius muscle.¹⁴ If there is CAR, it is evident that the reflex arc across the brainstem is intact. For this reason, impairment of AR is reported in cases with some central auditory lesions^{9,10} and CAR could be used to monitor the recovery of brainstem function.¹⁵

It has been established that the AR is present with about 85-90 dB HL of pure tone stimulus in a normal hearing human.¹⁶ In people with sensorineural hearing loss, the AR threshold is at the same level as that in normal subjects when the hearing threshold is less than 55 dB HL, and it increases with more deterioration of hearing.¹⁷ In the case of CHL, AR does not appear in the affected ear due to the pathology that causes sound conduction disturbance. When stimulation is delivered to the affected side with CHL and CAR is measured, the threshold rises or no reflex occurs according to the severity of hearing loss.

In the present study, CAR appeared in 55% of patients with CAA after canalplasty. This result is similar to that of sensorineural hearing loss after cochlear implantation, which recovers hearing to an almost normal level. We analyzed factors affecting the presence of CAR postoperatively. In multivariate analysis, 3 factors have been shown to independently affect the presence of CAR; postoperative hearing, age at surgery, and time interval after surgery.

A previous study reported that the worse the hearing level, the higher the chance of AR absence in CAA patients, and this is a predictable

finding, as in our study.⁸ In patients with worse postoperative hearing, a higher level of stimulation sound is necessary to trigger AR. Acoustic reflex does not easily appear when hearing is poor because stimulation sounds of higher than 120 dB HL cannot be used in the AR test. While AR appeared in nearly half of the patients with 90 dB HL of sensorineural hearing loss,^{18,19} the subjects in the current study had an average hearing level of 44dB HL and showed AR in 55%. Previously, Gelfand et al¹⁴ also reported that there was a 50% chance of absent reflexes for CHL of 42 dB HL, and our results are largely consistent with that study. In the case of sensorineural hearing loss, due to the recruitment phenomena, AR may present with a stimulus below the maximum stimulus even when the hearing loss is severe. But in the case of a CHL, the AR threshold is considered to increase parallel to the hearing threshold.

Other than hearing level, CAR was more likely to be observed when canalplasty was performed at an earlier age and when the AR test was performed for a long time after surgery. Acoustic reflex is more likely to appear when a patient with unilateral CAA is operated on at a younger age. It is well known that neuroplasticity declines with aging.²⁰ Thus, auditory input after early canalplasty might have affected the restoration of the reflex arc of AR. Also, the longer the auditory stimulus is delivered after reconstructive surgery, the higher the likelihood of reflex arc restoration due to the longer actuation of neuroplasticity. Although hearing was best 3 months after surgery, AR continued to appear more frequently over time, indicating that the period after surgery, independent of hearing, affected AR recovery. Of note, in 5 out of 8 patients who did not show CAR at the first test, CAR appeared at retest after an average of 24 months.

This study has a limitation that the follow-up periods and time intervals between surgery and AR test were not consistent between subjects. Nevertheless, it is meaningful that this study investigated the restoration of AR after reconstructive surgery in a large cohort of patients with congenital CHL.

In conclusion, this study revealed that CAR was present in 55% of patients with CAA after canalplasty and appeared more frequently with better postoperative hearing, when the surgery was performed at a younger age and when tested longer after surgery. These findings suggest that early and prolonged provision of appropriate auditory stimulation to patients with congenital CHL may lead to better restoration of the acoustic reflex arc.

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Informed Consent: N/A.

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