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Classification of Facial Nerve Aberration in Congenital Malformation of Middle Ear: Implications for Surgery of Hearing Restoration

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Description

A retrospective case review study was conducted. Electronic charts from January 2000 until December 2017 in the department of otolaryngology, Beijing Tongren hospital of capital medical university, were reviewed. Diagnosis of "congenital malformation of middle ear" of the cases included in the study was confirmed by operation records and videos. During chart searching, "admission diagnosis" was screened for "middle ear malformation", "congenital middle ear anomaly", "conductive deafness", "conductive hearing loss", "otosclerosis", "ossicular chain abnormality" and "ossicular chain". Charts with admission diagnosis of "otosclerosis" were also browsed for possible mis-diagnosis in order to minimize unnecessary omissions. Inclusion criteria: Hearing loss since birth; tympanic membrane intact under otoscopy; conductive hearing loss by Pure Tone Audiometry (PTA); if mixed hearing loss, dominated by conductive loss; ossicular chain abnormality confirmed on operation records and videos. Exclusion criteria: Chronic suppurative otitis media, otitis media with effusion, traumatic ossicular fracture, microtia, aural atresia, stenosis of External Auditory Canal (EAC) with or without cholesteatoma and otosclerosis. Cases with malformation of the inner ear with remarkable bone conduction threshold elevation were also excluded.

Effective with Variable Results

Two investigators reviewed operation records independently. When there was any disagreement, discussion was undertaken until an agreement was reached. Based on the inclusion and exclusion criteria, 227 cases (256 ears) of congenital malformation of middle ear were drawn from the electronic chart system after duplication removal. The median ± SD age at surgery was 20.4±10.7 years. The male to female ratio was 1.55:1. Among the 256 ears, the leading cause of operation was hearing loss (n=248, 96.9%), suppurative discharge (n=6, 2.3%), tinnitus (n=1, 0.4%) and vertigo (n=1, 0.4%). Facial paresis was found in one case before surgery. Involvement of tympanic segment and genu of facial nerve by cholesteatoma was indicated by CT scanning before surgery and confirmed in the operation.

Since the leading cause of operation was hearing loss (248/256 cases, 96.9%), hearing improvement procedures were performed in the majority of cases after middle ear exploration. Cholesteatoma hindered primary stage hearing restoration in 4 ears and secondary stage hearing improvement surgery was expected when clearance of cholesteatoma could be assured. After releasing adhesion, the ossicular chain was found to be intact with good mobility in 14 ears, allowing immediate tympanoplasty. In a case report from Trautwein et al. evident Electrical Auditory Brainstem Responses (EABRs) were recorded after implantation in a child with AN, with significant improvement in speech perception. Shallop et al. and Peterson et al reported the preliminary results from a group of AN children implanted at Mayo clinic in Rochester, Minnesota. These children showed significant postoperative improvement in sound detection, speech perception and communication, with no difference in CI benefits compared with other CI recipients. Buss et al reported 4 children with AN who had failed to benefit from amplification trials before implantation. All subjects' performance data were comparable with other pediatric implant patients using the Paden-Brown test at the 1-year follow-up, with two out of the four better than control subjects, while all subjects showed robust contralateral reflex and EABR wave V, suggesting synchronous neural response to stimulation delivered through the implant. In a study implemented by Zeng et al. speech intelligibility was compared in 7 AN patients between CI and acoustic HA in both quiet and in noise conditions. The former produced significantly higher intelligibility than the latter and reached the level of SNHL controls. Raveh et al. Reported better speech recognition performance in 12 AN patients with CI than those with HA. They thereby advocated CI as often a good solution for failures of conventional rehabilitation. In the evaluation on speech production in some children, Meaningful Use of Speech Scale (MUSS) results demonstrated an improvement from 3% preoperatively to 29% postoperatively. Jeong et al. Reported optimistic implantation results in 9 children with AN whose performance outcomes are as good as those children implanted for sensor neural hearing loss. No statistically significant differences were found between AN implanted children and matched SNHL implanted children in performance outcomes including Categories of Auditory Performance scale (CAP, P=0.3337), Monosyllabic Word test for phonemes (MW, P=0.5768) and common phrases test (P = 0.3337). Besides, slopes of electrically evoked compound action potentials (ECAPs) amplitude growth functions were also similar (P=0.970) in the two groups, demonstrating comparable spiral ganglion populations. Rance et al. evaluated speech perception skills in children with AN fitted with cochlear implants by open-set CNC monosyllabic words. In 10 implanted AN children, 9 demonstrated significant speech discrimination improvement in phoneme recognition (P=0.006) with scores no less than 55%, which were similar with the results in the amplification aided AN group although poorer than those in the matched control group implanted for sensor neural hearing loss.

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