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诺尔康文摘

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耳蜗畸形与诺尔康人工耳蜗植入

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文献摘要

儿童内耳共同腔畸形耳聋患者单侧植入人工耳蜗的临床效果观察

【摘要】

目的：观察单侧植入诺尔康人工耳蜗的儿童内耳共同腔畸形（Common Cavity Deformity，CCD）耳聋患者的术参考译文：后开机、调试情况以及临床听力言语康复效果。

方法：临床选取 10 例单侧植入诺尔康人工耳蜗的儿童 CCD 耳聋患者作为实验组，同时选取性别、手术年龄、植入侧别、人工耳蜗植入时长、助听器佩戴时长均配对的 10 例耳蜗结构正常的儿童作为对照组，并对两组的术后调试情况（行为反应 T 值（Threshold Level, T-level）、最大舒适阈 C 值（Comfortable Level, C-level）及言语康复情况（听觉行为分级标准（CAP）和言语可懂度分级标准（SIR）分级）进行比较。

结果：两组植入者的临床资料无显著性差异（$P > 0.05$），且术后无严重发症；在开机 3 个月、开机 6 个月时，CCD 组的各电极的 T 值、C 值均高于对照组（$P< 0.05$），动态范围无差异（$P> 0.05$）；CCD 组开机 6 个月的 T 值、C 值均高于开机时（$P < 0.05$），动态范围无差异（$P > 0.05$）；对照组开机 6 个月的 C 值、动态范围均高于开机时（$P < 0.05$），T 值无差异（$P > 0.05$）。开机 6 个月时，CCD 组的 CAP 分级低于对照组（$P< 0.05$），SIR 分级也低于对照组，但其差值无显著性（$P = 0.296$）；同时，两组开机 6 个月时的 CAP 和 SIR 分级均高于开机时（$P< 0.05$，$P< 0.05$）。

结论：植入诺尔康人工耳蜗的儿童 CCD 患者的康复效果差于耳蜗结构发育正常儿童，术后调试需要设置更大的 T 值、C 值，但是开机 6 个月时的康复效果和听力言语能力仍有显著进步，因此诺尔康人工耳蜗适用于儿童共同腔畸形耳聋患者。

【关键词】人工耳蜗；共同腔畸形；CAP；SIR

参考译文：

Clinical effects of unilateral cochlear implantation in children with hearing loss and common cavity inner ear malformation

Abstract

Objective：The aim of this study was to observe the clinical characteristics of cochlear implant mapping and hearing rehabilitation post-operation in children with common cavity deformity (CCD) and Nurotron cochlear implants.

Methods：It was a retrospective study including a group of 10 pediatric recipients who was diagnosed as common cavity deformity through the preoperative imageological examination and a control group of 10 pediatric recipients who had similar basic clinical conditions but normal cochlear structures. Mapping parameters (threshold Level (T-level) and comfortable level (C-level)) and outcomes of hearing and speech abilities' tests (categories of auditory performance (CAP) and speech intelligibility rating (SIR)) were analyzed and compared at switch-on and 6 months later.

Results：There was no significant difference in clinical materials of children with cochlear implantation between CCD group and control group (P> 0.05). All the surgeries and switch-on of children were success and without obvious complications. T and C value of each electrode in the CCD group were higher than in the control group for the time point of 3 months after switch-on and 6 months after switch-on (P< 0.05), while the dynamic range of two group was no statistical significance (P> 0.05). At the same time, both T and C values in CCD group at the time of 6 months after switch-on were significantly higher than at switch-on (P< 0.05), while the dynamic range of the two time points was no significant difference (P> 0.05). Similarly, C value and dynamic range in control group at the time of 6 months after switch-on were higher than that at switch-on (P< 0.05), while T value of the two time points was no significant difference (P> 0.05). At the time points of 6 months after switch-on, the scores of CAP in CCD group was significant lower than that in control group(P< 0.05), the scores of SIR in CCD group was lower than that in control group, but the difference was no significant (P = 0.296). However, both CAP and SIR scores in two groups at the time point of 6 months after switch-on were significantly higher than that at switch-on (P< 0.05, P< 0.05, respectively).

Conclusion：Hearing rehabilitation of the deaf children with common cavity malformation and Nurotron cochlear implant was less effective than that of children with normal cochlear structure, and it need a larger T and C value in mapping. But there was still a significant progress in both hearing rehabilitation and hearing and speech performances in recipients with CCD at the time of 6 months after switch-on. Nurotron cochlear implant was suit for children with hearing loss and common cavity malformation. .

Key Words: Cochlear implant; Common cavity deformity; CAP; SIR

Hearing improvement after cochlear implantation in common cavity malformed cochleae: long-term follow-up results.

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1. Department of Otolaryngology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea.

Abstract

Conclusion: We suggest that cochlear implantation (CI) should be a good therapeutic modality for hearing restoration in patients with common cavity malformed ears.

Objective: To analyze hearing improvement from CI performed in common cavity malformed cochleae. **Methods:** A total of 11 patients (5 male and 6 female, mean age 4.5 ± 2.8 years) and 12 ears were enrolled in this study. During the insertion of electrodes, we used C-arm fluoroscopy to avoid intrameatal placement. We evaluated hearing improvement every 6 months and the mean follow-up period was 80.5 ± 24.1 months (53-125 months). **Results:** During the operation, there were only four cases with fully inserted electrodes. Cerebrospinal fluid gushed out in two cases during the cochleostomy and postoperative meningitis occurred in two patients. One patient had to undergo reimplantation 4 years later due to device failure and recurrent meningitis. During the 48 months follow-up hearing evaluation, the ability of hearing increased along with the age. The final average MAIS, CAP, SIR, and open set one- and two-syllable word scores were 90.3 ± 18.1%, 4.9 ± 1.6, 3.1 ± 0.9, 24.1 ± 25.9%, and 48.6 ± 38.7%, respectively.

Article source:
Ahn JH, Lim HW, Lee KS. Hearing improvement after cochlear implantation in common cavity malformed cochleae: long-term follow-up results[J]. Acta Otolaryngol, 2011, 131(9):908-13.

参考译文：

共同腔畸形耳蜗患者植入人工耳蜗后的听觉改善：长期随访结果

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【摘要】

结论：我们建议人工耳蜗植入术（CI）作为共同腔畸形耳蜗患者听力重建的有效治疗手段。

目的：分析共同腔畸形耳蜗患者植入CI后的听觉改善。

方法：共11例患者（男5例，女6例，平均年龄4.5±2.8岁），共计12例术耳参与此项研究。电极植入中行c臂x线透视检查，以避免植入到内听道。术后每6个月评估听觉改善情况，平均随访周期为80.5±24.1月（53-125月）。

结果：在术中，仅四例患者电极全部植入。2例耳蜗造口术中出现脑脊髓井喷，2例术后出现脑膜炎。1例因设备故障和复发性脑膜炎，4年后行二次植入。随访评估48个月，患者听觉能力随年龄提高。植入者MAIS，CAP，SIR，开放式单音节词，双音节词测试得分，分别均为90.3 ± 18.1%，4.9 ± 1.6，3.1 ± 0.9，24.1 ± 25.9%和48.6 ± 38.7%。

【文献来源】

Ahn JH, Lim HW, Lee KS. Hearing improvement after cochlear implantation in common cavity malformed cochleae: long-term follow-up results[J]. Acta Otolaryngol, 2011, 131(9):908-13.

Cochlear implantation in 21 patients with common cavity malformation.

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1. Department of Otorhinolaryngology Head and Neck Surgery, Beijing Friendship Hospital, Capital Medical University, Beijing, China.

Abstract

Conclusion*:* The facial recess approach is preferred in common cavity (CC) malformation with an incomplete basal turn, and the transmastoid single-slit labyrinthotomy approach in classic CC malformation. Patients with CC benefit from cochlear implantation (CI) over time, but the audiological and speech development is poorer than in cases with normal cochleas.

Objectives: To discuss the surgical aspects and performance of CI in 21 patients with CC malformation.

Methods: Twenty-one CC malformations were classified into 2 types: classic CC malformation and CC malformation with an incomplete basal turn. Twenty-one patients without inner ear malformation were set as the control group. Thus, data for 42 patients were analyzed.

Results: The facial recess approach was used in 3 patients with CC malformation with an incomplete basal turn, and the transmastoid single-slit labyrinthotomy approach in 18 patients with classic CC malformation. After follow-up for 36 months, the average free-field hearing threshold was higher, and the scores for the CAP, SIR, IT-MAIS, and closed-set/open-set auditory speech perception tests were lower than in the control group ($p < 0.05$).

Key Words: CSF gusher; Inner ear malformation; behavioral audiometry; facial recess approach; performance; speech perception; transmastoid labyrinthotomy approach.

Article source:
Xia J, Wang W, Zhang D. Cochlear implantation in 21 patients with common cavity malformation[J]. Acta Otolaryngol. 2015, 135(5):459-65.

参考译文：

21例共腔畸形患者的人工耳蜗植入经验

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【摘要】

结论：可见部分耳蜗底转的共同腔畸形患者首选经面隐窝径路，而典型的共同腔畸形患者采用经乳突单缝迷路切开的方法。共同腔畸形患者随时间推移能受益于人工耳蜗植入，但其听觉和言语发展比耳蜗正常的患者较差。

目的：为了探讨21例共同腔畸形患者的人工耳蜗植入术后表现和效果。

方法：21例共同腔畸形患者分为两组：典型的共同腔畸形患者和可见部分耳蜗底转的共同腔畸形患者。对照组为21例无内耳畸形患者。分析42例患者的数据。

结果：3例可见部分耳蜗底转的共同腔畸形患者选择经面隐窝入路，18例典型的共同腔畸形患者采用经乳突单缝迷路切开方法。随访36个月，共腔畸形患者的平均声场听阈比对照组显著较差，且CAP, SIR, IT-MAIS和开放式/封闭式听觉言语感知测试得分低于对照组($p < 0.05$)。

【关键词】脑脊液自喷井;内耳畸形;行为听力测定; 面隐窝径路 ;性能;言语知觉;经乳突迷路切开术.

【文献来源】
Xia J, Wang W, Zhang D. Cochlear implantation in 21 patients with common cavity malformation[J]. Acta Otolaryngol. 2015, 135(5):459-65.

Evaluation of cochlear implantation in children with inner ear malformation.

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1. Department of Otolaryngology, Affiliated Sixth People's Hospital to Shanghai Jiao Tong University, Shanghai, China.

Abstract

Objectives*:* This study aimed to compare the outcomes of cochlear implantation (CI) in children with malformed versus normal inner ear anatomy.

Methods: We assessed 63 children with prelingual deafness, including 12 with inner ear malformations. All had undergone CI before the age of 5 y. We evaluated Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) scores before surgery and at 6, 12, and 24 months after surgery.

Results: In both groups, the CAP and SIR scores increased with time after implantation in follow-up assessments. No significant differences were found in the CAP or SIR scores between the two groups at any of the four follow-up assessments.

Conclusions: Children with inner ear malformation can benefit from CI. Although additional factors may influence the outcome of CI in children with inner ear malformations compared to children with deafness from other causes, early implantation may provide similar results.

Article source:

Zhou H, Sun X, Chen Z, et al. Evaluation of cochlear implantation in children with inner ear malformation [J]. B-ENT, 2014, 10(4):265-9.

参考译文：

评估内耳畸形患儿的人工耳蜗植入

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1. 上海交通大学附属第六人民医院耳鼻喉科，中国；

【摘要】

目的：本研究旨在比较内耳发育畸形患儿和内耳发育正常患儿的人工耳蜗植入效果。

方法：评估5岁前植入人工耳蜗的语前聋患儿63例，其中12例内耳畸形。评估患儿术前和术后6月，12月和24月的听觉行为分级标准（CAP）和言语可懂度分级标准（SIR）得分。

结果：随访评估中，两组CAP和SIR得分在术后均有提高，且CAP和SIR得分在4次随访评估中无明显差异。

结论：人工耳蜗适用于内耳畸形患儿。尽管相比于其他原因造成耳聋的患儿，内耳畸形患儿可能因附加因素影响人工耳蜗植入效果，但早期植入可能达到相似效果。

【文献来源】

Zhou H, Sun X, Chen Z, et al. Evaluation of cochlear implantation in children with inner ear malformation [J]. B-ENT, 2014, 10(4):265-9.

Speech perception and production in children with inner ear malformations after cochlear implantation.

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Abstract

Objectives: The aim of this study was to assess the speech perception and speech intelligibility outcome after cochlear implantation in children with malformed inner ear and to compare them with a group of congenitally deaf children implantees without inner ear malformation.

Methods: Six deaf children (five boys and one girl) with inner ear malformations who were implanted and followed in our clinic were included. These children were matched with six implanted children with normal cochlea for age at implantation and duration of cochlear implant use. All subjects were tested with the internationally used battery tests of listening progress profile (LiP), capacity of auditory performance (CAP), and speech intelligibility rating (SIR). A closed and open set word perception test adapted to the Modern Greek language was also used. In the dysplastic group, two children suffered from CHARGE syndrome, another two from mental retardation, and two children grew up in bilingual homes.

Results: At least two years after switch-on, the dysplastic group scored mean LiP 62%, CAP 3.8, SIR 2.1, closed-set 61%, and open-set 49%. The children without inner ear dysplasia achieved significantly better scores, except for CAP which this difference was marginally statistically significant ($p = 0.009$ for LiP, $p = 0.080$ for CAP, $p = 0.041$ for SIR, $p = 0.011$ for closed-set, and $p = 0.006$ for open-set tests).

Conclusions: All of the implanted children with malformed inner ear showed benefit of auditory perception and speech production. However, the children with inner ear malformation performed less well compared with the children without inner ear dysplasia. This was possibly due to the high proportion of disabilities detected in the dysplastic group, such as CHARGE syndrome and mental retardation. Bilingualism could also be considered as a factor which possibly affects the outcome of implanted children. Therefore, children with malformed inner ear should be preoperatively evaluated for cognitive and developmental delay. In this case, counseling for the parents is mandatory in order to explain the possible impact of the diagnosed disabilities on performance and habilitation.

Key Words: Cochlear implantation, Inner ear malformation, Speech perception, Speech intelligibility, CHARGE syndrome, Mental retardation.

Article source:
Rachovitsas D, Psillas G, Chatzigiannakidou V,et al. Speech perception and production in children with inner ear malformations after cochlear implantation[J]. Int J Pediatr Otorhinolaryngol. 2012 ,76(9):1370-4.

参考译文：

内耳发育畸形患儿人工耳蜗术后的言语识别和重建

Rachovitsas D[1], Psillas G, Chatzigiannakidou V, Triaridis S, Constantinidis J, Vital V.

1.塞萨洛尼基亚里斯多德大学耳鼻喉系，AHEPA医院，塞萨洛尼基，希腊；

【摘要】

目标：本研究旨在评估内耳发育畸形患儿在人工耳蜗术后的言语识别效果和言语可懂度，并与内耳发育正常的先天性耳聋患儿进行比较。

方法：观察在我们中心植入人工耳蜗的6例内耳发育畸形患儿（男5例，女1例），并选择植入年龄及人工耳蜗使用时间相匹配的6例耳蜗结构正常的人工耳蜗植入患儿。本次研究采用国际通用的组合测试：听觉发育情况（LIP），听觉行为分级标准（CAP）和言语可懂度分级标准（SIR），且采用适用于现代希腊语言的开放式和封闭式单词识别测试。其中畸形组患儿，2例患有CHARGE综合证，2例有精神发育迟滞，2例生长在双语家庭。

结果：开机两年以后，内耳发育畸形组测试平均得分为LiP 62%, CAP 3.8, SIR 2.1，开放式得分61%，封闭式得分49%。内耳发育正常组得分明显高于内耳发育畸形组，除CAP只有细微统计学差异(LiP：$p = 0.009$，CAP：$p = 0.080$，SIR：$p = 0.041$，封闭式：$p = 0.011$，开放式$p = 0.006$)。

结论：所有内耳发育畸形的人工耳蜗植入患儿，听觉言语能力均有提高。然而，内耳发育畸形组的表现不如内耳发育正常组。这可能跟内耳发育畸形组高比例的残疾有关，如CHARGE综合症和精神发育迟滞。另外，双语也被视为可能影响患儿人工耳蜗植入效果的一个因素。因此，内耳畸形患儿需在术前评估认知能力和发育延迟情况。在这种情况下，患者父母必须接受咨询以告知其被诊断的残疾状况对术后表现和康复方面可能造成的影响。

【关键词】人工耳蜗植入；内耳畸形；言语识别；言语可懂；CHARGE综合症；精神发育迟滞.

【文献来源】

Rachovitsas D, Psillas G, Chatzigiannakidou V,et al. Speech perception and production in children with inner ear malformations after cochlear implantation[J]. Int J Pediatr Otorhinolaryngol. 2012, 76(9):1370-4.

Cochlear Implant Electrode Choice in Challenging Surgical Cases: Malformation, Residual Hearing, Ossification, or Reimplantation.

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Abstract

Purpose of Review: The limits of cochlear implantation candidacy have been expanding over the last decade and recent studies show that patients with inner ear anomalies, significant residual hearing, ossified cochlea, or far advanced otosclerosis can benefit from implant. The cochlear implant companies are coming up with various electrode designs. The purpose of this study is to review the factors that can affect the choice of electrode array in the expanding indications of cochlear implantation and suggest some surgical tips.

Recent Findings:A comprehensive preoperative evaluation is crucial. Detailed audiologic examination and radiologic assessment of inner ear structures with high-resolution computed tomography and/or magnetic resonance imaging is necessary. The choice of electrode array should be made regarding the type of cochlea in the presence of inner ear anomalies. If the patient has residual hearing in low-frequencies, electrode array and surgical insertion technique should be as atraumatic as possible to protect apical part of the cochlea. Appropriate selection of electrodes and surgical techniques are necessary if the cochlea is obstructed by fibrosis or ossified. The surgeon also should consider the possibility of reimplantation in the future and select the initial electrode after comprehensive evaluation.

Summary: There is a diversity of electrode arrays for different indications. The selection of the most accurate electrode depends on the audiological tests, etiology of hearing loss, and cochlear anatomy. Surgeon must be prepared preoperatively for various clinical situations and unexpected surgical circumstances. One should keep in mind that making the right electrode choice will impact the outcomes of unusual or challenging cases.

Key Words: Cochlear implant，electrode，Reimplantation，Cochlear malformation，Cochlear ossification，Residual hearing

Article source:
Eshraghi AA, E Ocak. Cochlear Implant Electrode Choice in Challenging Surgical Cases: Malformation, Residual Hearing, Ossification, or Reimplantatio[J]. Current Otorhinolaryngology Reports,2017,5(4):315-322.

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疑难病例中人工耳蜗植入电极的选择：畸形，残余听力，骨化，或再植入

Eshraghi AA[1], E Ocak[2].

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【摘要】

目的：在过去十年中，人工耳蜗的植入范围界限逐渐放宽，最近的研究表明，内耳畸形、有显著残余听力、耳蜗骨化、或者严重的耳硬化症患者都可以从人工耳蜗中获益。人工耳蜗公司也在设计出不同的电极。本研究旨在探讨，在人工耳蜗植入术中，影响电极阵列类型选择的因素，并提出一些手术方面的意见。

目前进展：全面的术前评估至关重要。详细的听力检查和内耳构造高分辨率断层CT扫描评估和/或者磁共振成像都是必要的评估手段。

在内耳异常的情况下，应选择适合的电极阵列类型。如果患者在低频有残余听力,电极阵列和手术插入技术应尽可能保护耳蜗顶端，防止损伤残余听力。如果耳蜗发生纤维化或骨化，合理选择电极和手术技术十分必要。手术医生也应考虑未来再植入的可能性，并在全面评估后选择电极。

结论：不同的适应症应选择不同的电极阵列。最准确的电极的选择取决于听力学测试，听力损失的病因，耳蜗解剖结构。手术医生必须在术前做好准备以应付各种临床情况和意想不到的手术情况。并谨记电极放置准确性将影响到异常和特殊病例的植入效果。

【关键词】人工耳蜗；电极；再植入；耳蜗畸形；耳蜗骨化；残余听力.

【文献来源】
Eshraghi AA, E Ocak. Cochlear Implant Electrode Choice in Challenging Surgical Cases: Malformation, Residual Hearing, Ossification, or Reimplantation [J]. Current Otorhinolaryngology Reports, 2017, 5(4):315-322.

Clinical outcomes following cochlear implantation in children with inner ear anomalies.

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Abstract

Objectives: A significant proportion of children with congenital hearing loss who are candidates for cochlear implants (CIs) may have inner ear malformations (IEMs). Surgical and speech outcomes following CI in these children have not been widely reported.

Methods: The charts of children who were evaluated for a CI between 1/1/1986 and 12/31/2014 at a university-based tertiary level pediatric cochlear implant center were reviewed. Principal inclusion criteria included (i) age 1-18 years, (ii) history of bilateral severe to profound sensorineural hearing loss, and (iii) limited benefit from binaural amplification. Exclusion criteria included (i) underlying diagnosis of neurodevelopmental disorder and (ii) lack of follow up for speech assessment if a CI was performed. The following outcome measures were reviewed: (i) imaging findings with magnetic resonance imaging or high resolution computed tomography, (ii) intraoperative complications, and (iii) speech perception categorized as the ability to perceive closed set, open set, or none.

Results: The prevalence of IEMs was 27% (102 of 381), of which 79% were bilateral. Cochlear dysplasia accounted for 30% (40 of 136) of the anomalies. Seventy-eight of the 102 patients received a CI (78%). Surgery was noted to be challenging in 24% (19 of 78), with a perilymphatic gusher being the most common intraoperative finding. Cochlear dysplasia, vestibular dysplasia and cochlear nerve hypoplasia were associated with poor speech perception (open OR closed set speech recognition scores, 0-23%), although the outcomes in children with enlarged vestibular aqueduct were similar to those of children with normal inner ear anatomy (65%).

Conclusions: Cochlear implantation is safe in children with IEMs. However, the speech perception outcomes are notably below those of patients with normal anatomy, with the exception of when an enlarged vestibular aqueduct is present.

Key Words: Cochlear implants; Hearing restoration; Inner ear malformations; Speech recognition

Article source:
Isaiah A, Lee D, Lenes-Voit F, et al. Clinical outcomes following cochlear implantation in children with inner ear anomalies[J]. Int J Pediatr Otorhinolaryngol. 2017,93:1-6.

参考译文：

内耳畸形患儿人工耳蜗植入的临床效果

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【摘要】

目的：很大一部分的先天性听力损失儿童人工耳蜗植入候选人可能患有内耳畸形(IEMs)。此类人工耳蜗植入患儿的手术和言语效果报道还不广泛。

方法：分析1986年1月1日到2014年12月31日之间中心接受人工耳蜗植入评估的患儿结果进行汇总。主要入选标准包括：(i)年龄1-18岁。(ii) 双侧重度和极重度感音神经性听力损失史。(iii) 双耳助听器佩戴无效。排除标准包括：(i)潜在的神经发育障碍。(ii) 如进行CI手术，术后无法配合随访及言语评估。回顾了以下检查结果：(i)影像学检查结合核磁共振成像或高分辨率CT。(ii) 术中并发症。(iii) 言语识别分为开放式言语识别能力，封闭式言语识别能力，部分未测试该项内容。

结果：IEMs患病率达27%（381例中有102例），其中79%是双侧。耳蜗发育异常在所有畸形中占比30%（136例中有40例）。102例患者中78例（78%）接受了人工耳蜗植入。24%（78例中19例）的手术具有一定的难度，术中常见的并发症为外淋巴液“井喷”。耳蜗发育异常、前庭发育异常和蜗神经发育不全导致言语识别能力不良（开放式或封闭式言语识别能力得分，0-23%），但前庭导水管扩大患儿效果与内耳发育正常患儿植入效果相似（65%）。

结论：IEMs患儿进行人工耳蜗植入手术是安全的。除了前庭导水管扩大患儿，内耳畸形患儿的言语识别效果要明显低于正常患儿。

【关键词】人工耳蜗；听觉恢复；内耳畸形；言语识别.

【文献来源】
Isaiah A, Lee D, Lenes-Voit F, et al. Clinical outcomes following cochlear implantation in children with inner ear anomalies[J]. Int J Pediatr Otorhinolaryngol. 2017,93:1-6.

Outcome of cochlear implantation in children with cochlear malformations.

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Abstract

The objective of the study was the evaluation of outcomes of cochlear implantation (CI) in children with cochlear malformations. A retrospective case–control study was conducted in a tertiary referral centre. The patients were children with inner ear malformation judged by high-resolution computed tomography and magnetic resonance imaging treated with uni- or bilateral CI and a follow-up period of at least 3years. They were matched with a control group of children operated for other reasons. The patients were operated by one of two surgeons using similar techniques including a standard perimodiolar electrode in all cases. The intervention was therapeutic and rehabilitative. The main outcome measures were category of auditory performance (CAP) and speech intelligibility rating (SIR). Eighteen children were diagnosed with cochlear malformations (12% of children receiving CI). No statistical differences regarding CAP and SIR scores were found between the two groups. Only one child was diagnosed with a common cavity and performed below average. Children with auditory neuropathy performed beyond average. Children with cochlear malformations performed equally to children without malformation in the long term. Standard perimodiolar electrodes can be used despite cochlear malformations. The most important factors determining the outcome is the age of the child at the time of implantation and duration of hearing loss before CI. Awareness towards an increased risk of complications in case of inner ear malformations is recommended.

Key Words: Cochlear implantation， Inner ear malformation， Category of auditory performance (CAP)，Speech intelligibility rating (SIR) ，Complication， Control group.

Article source:
Bille J, Fink-Jensen V, Ovesen T. Outcome of cochlear implantation in children with cochlear malformations[J]. Eur Arch Otorhinolaryngol. 2015,272(3):583-9.

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耳蜗畸形患儿人工耳蜗植入效果分析

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【摘要】

本研究旨在评估内耳畸形患儿人工耳蜗植入的效果。回顾中心病例进行研究。经过高分辨率CT和磁共振成像诊断为内耳畸形的患儿并植入单侧或双侧人工耳蜗，随访3年以上。并匹配设置了因其他原因植入人工耳蜗的对照组。患者均由两名手术医生主刀，采用相似技术植入标准近轴状电极。术后实施言语治疗和言语康复进行干预。主要检测方式采取听觉行为分级标准（CAP）和言语可懂度分级标准（SIR）。18例患儿被诊断为耳蜗畸形（占人工耳蜗手术的12%）。耳蜗畸形组和对照组的CAP 和SIR 得分无统计学差异。仅一例患儿诊断为共同腔畸形且表现低于平均水平。听神经病患儿表现超出平均水平。长远看，耳蜗畸形患儿与耳蜗正常患儿表现没有显著差异。标准近蜗轴电极可适用于耳蜗畸形患儿。影响康复效果最重要的因素是患儿植入人工耳蜗的年龄和听力损失的时长。建议提高对内耳畸形患儿手术并发症的风险意识。

【关键词】人工耳蜗植入；内耳畸形；听觉行为分级标准（CAP）；言语可懂度分级标准(SIR)，并发症；对照组.

【文献来源】

Bille J, Fink-Jensen V, Ovesen T. Outcome of cochlear implantation in children with cochlear malformations[J]. Eur Arch Otorhinolaryngol. 2015,272(3):583-9.

Cochlear Implantation in Cochlear Ossification: Retrospective Review of Etiologies, Surgical Considerations, and Auditory Outcomes.

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Abstract

Objectives: 1) To review the surgical and auditory outcomes and complications of cochlear implantation in cases with cochlear ossification. 2) To evaluate association between the extent and etiology of ossification to outcomes.

Study Design: Retrospective study.

Setting: Otology and skull base surgery center.

Subjects and Methods: Charts of 40 patients (42 ears) with cochlear ossification undergoing cochlear implantation were reviewed. Demographic features, operative findings, auditory outcomes, and complications were analyzed. Operative findings included extent of cochlear ossification, extent of drilling required to obtain patent cochlear lumen, approach (posterior tympanotomy/subtotal petrosectomy), electrode insertion (partial/complete, scala tympani/vestibuli), and complications. Auditory outcomes were assessed over a 4-year follow-up period using vowel, word, sentence, and comprehension scores. Patients were divided into groups (otosclerotic/non-otosclerotic and round window/basal turn ossification) for comparison of auditory outcomes. Outcomes were compared with 60 randomly identified controls (adults with postlingual deafness) who underwent implantation with no cochlear ossification.

Results: The median age and duration of deafness of patients was 54.39 and 27.15 years, respectively. Etiology of cochlear ossification was otosclerosis in 23 of 42 ears and mixed in 19 of 42 ears (chronic otitis media, temporal bone fractures, idiopathic, meningitis, Cogan's syndrome) with exclusive round window involvement in 54.7% of cases and the rest having partial or complete basal turn ossification. 59.5% ears underwent subtotal petrosectomy for implantation. Three patients underwent scala vestibuli insertion and five had incomplete electrode insertion. Auditory outcomes were comparable in otosclerotic and non-otosclerotic cases and in round window and basal turn ossification cases. No significant differences were observed in auditory scores when compared with controls with no ossification.

Conclusions: Cochlear implantation in cochlear ossification is feasible despite surgical challenges and modifications. Auditory outcomes in basal turn ossification appear to be comparable to cases with no ossification with extent of ossification having no significant association with outcomes.

Key Words: Cochlear implantation; Cochlear ossification; Partial insertion; Scala vestibule; Subtotal petrosectomy.

Article source:

Vashishth A, Fulcheri A, Prasad SC, et al. Cochlear Implantation in Cochlear Ossification: Retrospective Review of Etiologies, Surgical Considerations, and Auditory Outcomes[J].Otol Neurotol, 2018, 39(1):17-28.

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耳蜗骨化患者的人工耳蜗植入：回顾性分析病因、手术考虑和听觉效果

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【摘要】

目的：（1）分析耳蜗骨化状态下人工耳蜗植入的手术和听觉效果以及并发症；（2）评估耳蜗骨化的程度和病因与植入效果之间的关系。

研究设计：回顾性研究 设置: 耳科学和颅底外科中心

病例与方法：分析40例（42例术耳）耳蜗骨化患者人工耳蜗植入效果。对人口学特征、手术结果、听觉效果和并发症进行分析。手术观察指标包括耳蜗骨化程度、暴露耳蜗内腔需要磨骨的程度、手术方法（后鼓室切开术/次全切除术）、电极插入（部分/完全，骨阶/前庭窗）和并发症。听觉效果评估随访4年以上，采用元音、单词、句子理解得分。将耳蜗骨化的患者分为几组（耳硬化症/非耳硬化症和圆窗/耳蜗底转骨化）以比较听觉效果。耳蜗骨化组的效果与随机选取的60例（成人语后聋）无耳蜗骨化的人工耳蜗植入患者比较。

结果：耳聋患者的平均年龄和耳聋持续时间分别为54.39和27.15年。

42例耳蜗骨化术耳的病因：23例为硬化症，19例为多种原因（慢性中耳炎、颞骨骨折、先天性、脑膜炎、Cogan’s综合症），其中54.7%存在圆窗骨化，其余可见部分或者全部耳蜗底转骨化。59.5%的术耳在人工耳蜗植入中行次全切除术。3例患者行前庭阶插入，5例患者电极不完全植入。比较了耳硬化症病例和非耳硬化症病例，与圆窗骨化病例和耳蜗底转骨化病例的听觉效果。对照组与耳蜗骨化组的听觉得分比较无显著差别。

结论：尽管手术有一定难度和限制，人工耳蜗植入手术在耳蜗骨化患者中是可行的。耳蜗底转骨化患者的听觉效果与无骨化的病例相似，骨化程度与效果无明显相关性。

【关键词】人工耳蜗植入；耳蜗骨化；部分插入；前庭阶；次全切除术。

【文献来源】
Vashishth A, Fulcheri A, Prasad SC, et al. Cochlear Implantation in Cochlear Ossification: Retrospective Review of Etiologies, Surgical Considerations, and Auditory Outcomes[J].Otol Neurotol, 2018, 39(1):17-28.

Cochlear implantation in patients with inner ear bone malformations with posterior labyrinth involvement: an exploratory study.

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Abstract

Inner ear bone malformations are one cause of profound sensorineural hearing loss. This investigation focused on those affecting the posterior labyrinth, especially enlarged vestibular aqueduct syndrome, which is associated with fluctuating and progressive hearing loss. The objectives of this study were to analyze the behavior of the electrical stimulation, auditory functionality and linguistic development in patients with inner ear malformations involving the posterior labyrinth. The study included ten patients undergoing cochlear implantation (cases: five with enlarged vestibular aqueduct, two with vestibular aqueduct stenosis/aplasia, and three with semicircular canal disorders). Post-implantation, data were gathered on the electrical stimulation threshold and maximum comfort levels and on the number of functioning electrodes. Evaluation of Auditory Responses to Speech (EARS) subtests were used to assess auditory functionality and language acquisition at 6, 12, and 24 months post-implantation. Results were compared with findings in a control group of 28 cochlear implantation patients without these malformations. No significant differences were found between case and control groups in electrical stimulation parameters; auditory functionality subtest scores were lower in cases than controls, although the difference was only statistically significant for some subtests. In conclusion, cochlear implantation patients with posterior labyrinth bone malformations and profound hearing loss, including those with enlarged vestibular aqueduct syndrome, showed no significant difference in electrical stimulation threshold with controls. Although some auditory functionality test results were lower in cases than in controls, cochlear implantation appears to be beneficial for all patients with these malformations.

Key Words: Cochlear implantation; Enlarged vestibular aqueduct syndrome; Hearing loss; Posterior labyrinth bone malformations.

Article source:

Palomeque Vera JM, Platero Sánchez-Escribano M, Gómez Hervás J, et al. Cochlear implantation in patients with inner ear bone malformations with posterior labyrinth involvement: an exploratory study[J]. Eur Arch Otorhinolaryngol, 2016, 273(4):893-8.

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内耳骨迷路畸形患者的人工耳蜗植入效果：探索性研究

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【摘要】

内耳骨迷路畸形是极重度感音神经性耳聋的原因之一。本研究着重观察影响迷路的病症，尤其是伴有波动性和进行性听力损失的前庭导水管扩大综合症。 本研究的目的是分析内耳后部迷路畸形患者的电刺激、听觉功能和语言发展。研究包括10例人工耳蜗植入患者（病例：5例前庭导水管扩大，2例前庭导水管狭窄/发育不全，3例半规管发育障碍）。术后数据采集包括T值和C值以及功能作电极的数量。在人工耳蜗术后6、12、24个月采用言语听觉反应评估（EARS）测验评估患者听觉功能和言语习得。测试结果与对照组28例无此类畸形的人工耳蜗患者进行比较。两组电刺激数据无显著差别；而听觉功能测试得分显著低于对照组，（尽管两组听觉能力的部分子测试之间并没有显著差异）。总之，后部骨迷路畸形的极重度听力损失人工耳蜗植入患者（包括前庭导水管扩大患者），与对照组电刺激阈值无显著差别。尽管一些听觉功能测试结果在某些情况下低于对照组，人工耳蜗植入手术对所有该类畸形患者有益。

【关键词】人工耳蜗植入；大前庭导水管综合症；听力损失；骨迷路畸形.

【文献来源】
Palomeque Vera JM, Platero Sánchez-Escribano M, Gómez Hervás J, et al. Cochlear implantation in patients with inner ear bone malformations with posterior labyrinth involvement: an exploratory study[J]. Eur Arch Otorhinolaryngol, 2016, 273(4):893-8.

Surgical Outcomes After Cochlear Implantation in Children With Incomplete Partition Type I: Comparison With Deaf Children With a Normal Inner Ear Structure.

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Abstract

Objective: To compare audiologic performance after cochlear implantation (CI) in children with incomplete partition (IP) Type I and age-matched children with normal cochleae.

Study design: Retrospective chart review.

Setting: Academic center.

Patients: Twenty-three children (25 ears) with IP Type I and 230 age-matched deaf children (230 ears) with nonsyndromic normal inner ears who underwent CI between January 2000 and June 2013.

Intervention CI Main outcome measure: The Categories of Auditory Performance (CAP) Scale score and the Meaningful Auditory Integration Scale (MAIS) score.Results The mean age of IP Type I patients at the time of CI was 5.3 years (standard deviation, 5.4 yr; range, 0.9–17.7 yr). The mean duration of follow-up was 4.7 years (standard deviation, 3.5 yr; range, 1.1–11.2 yr). Fourteen of the 25 IP Type I ears (56%) had cerebrospinal fluid gusher during the cochleostomy. In the IP type ears, the number of inserted electrodes was 16.3 ± 3.2 (range, 11–22), and the insertion angle was 236.5 ± 41.2 degrees (range, 180–305 degrees). The cochlear nerve was assessed in 17 of the 25 IP type ears, and hypoplasia was present in nine (53%). Facial nerve stimulation occurred in 15 of the 25 IP type ears. IP Type I patients younger than 3 years at CI had significantly lower CAP Scale and MAIS scores than age-matched controls at 12 and 24 months after CI, but similar CAP Scale and MAIS scores as age-matched controls at 42 and 72 months after CI. IP Type I patients aged 3 to 18 years at CI had similar CAP Scale and MAIS scores as age-matched controls at all post-CI time points.

Conclusion: Children with IP Type I who underwent CI performed as well as children with normal cochlea in the long-term.

Key Words: Cochlear implantation; Incomplete partition type; Inner ear anomaly.

Article source:
Suk Y, Lee JH, Lee KS. Surgical outcomes after cochlear implantation in children with incomplete partition type I: comparison with deaf children with a normal inner ear structure[J]. Otol Neurotol, 2015, 36(1):e11-7.

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不完全分隔I型患儿人工耳蜗植入术后效果：与内耳结构正常的聋儿比较

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【摘要】

目的：比较并分析不完全分隔（IP）I型患儿和年龄匹配的耳蜗正常的患儿人工耳蜗植入术后的听觉能力比较。

研究设计：回顾性图表分析

场地：学术中心

病例：回顾分析从2000年1月到2013年6月之间植入人工耳蜗的23例（25术耳）不完全分隔I型患儿与230例（230术耳）年龄匹配的内耳正常患儿。

人工耳蜗干预主要观察指标：听觉行为分级量表(CAP)得分和有意义听觉整合量表（MAIS）得分。

结果：不完全分隔I型患者CI平均植入年龄为5.3岁（标准偏差 5.4岁，范围0.9－17.7岁）。平均随访周期为4.7年（标准偏差3.5年；范围1.1－11.2年）。25例不完全分隔I型患儿中14例（56%）在耳蜗开孔时出现脑脊液“井喷”。不完全分隔 I型患耳，植入电极数为16.3 ± 3.2（范围11－22），植入深度为236.5 ± 41.2度（范围180－305度）。对25例不完全分隔型患耳中的17例进行了蜗神经检查，其中9例发育不全（53%）。25例不完全分隔型患儿中15例出现面抽。人工耳蜗植入年龄小于3岁的不完全分隔 I型患者在植入人工耳蜗后12个月和24个月的CAP和MAIS得分明显低于年龄匹配的对照组，但是在植入人工耳蜗后42个月和72个月的CAP和MAIS得分与年龄匹配的对照组相近。人工耳蜗植入年龄3到18岁的不完全分隔 I型患者与年龄匹配对照组的患者人工耳蜗植入者术后各时间点的CAP和MAIS得分相近。

结论：从长远看，不完全分隔I型人工耳蜗植入患儿与耳蜗正常的人工耳蜗植入患儿表现无显著差异。

【关键词】人工耳蜗植入；内耳不完全分隔I型；内耳畸形.

【文献来源】

Suk Y, Lee JH, Lee KS. Surgical outcomes after cochlear implantation in children with incomplete partition type I: comparison with deaf children with a normal inner ear structure[J]. Otol Neurotol, 2015, 36(1):e11-7.

诺尔康畸形耳蜗植入数据

截止2018年6月30日，已知的特殊案例共有1530例，其中：内耳畸形共有384例，其开机1个月，6个月，1年的CAP（$\bar{x}$ ±s）

特殊案例	数目	开机 1 个月 CAP	开机 6 个月 CAP	开机 1 年 CAP
耳蜗畸形	69	1.83±0.90	3.73±1.42	4.33±1.3
不完全分割 II 型（Mondini）	44	2.32±0.82	5.05±1.65	6.28±1.79
共同腔畸形	20	2.17±0.72	3.83±0.94	6.00±1.04
IP3 型	1	2.00±0.00	4.00±0.00	6.00±0.00
耳蜗发育不全	21	2.00±0.47	4.80±0.92	7.33±1.03
前庭导水管畸形	183	2.17±0.61	4.52±0.85	6.62±1.21
内听道畸形	43	2.24±0.66	3.75±1.34	6.62±1.56
前庭和半规管畸形	3	1.67±0.58	3.33±0.58	5.00±1.00

词汇

词汇	翻译
Cochlear malformation	耳蜗畸形
Ossification	骨化
Enlarged vestibular aqueduct syndrome	前庭导水管扩大综合症
Labyrinth bone malformation	骨迷路畸形
Incomplete partition type I	内耳不完全分隔I型
Inner ear malformation	内耳畸形
Common cavity malformation	共同腔畸形

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