

诺尔康文摘

NUROTRON DIGEST

2022年第1期

多重残疾与人工耳蜗效果专题

前言

根据研究数据显示，美国约有40-50%的听障儿童同时有其他残疾^[1]，同时英国国家聋人儿童协会（NDCS）的统计显示约有4-57%听障者患有视觉残疾，2-14%听障者患有神经发育障碍，而2-4.2%自闭症患儿及2-13%脑性瘫痪患儿同时有听力损失^[2]，因此在进行人工耳蜗干预的过程，难免会遇到多重残疾的植入者，深入探究此类患者的人工耳蜗效果及康复过程，对于术前期期望值调整、开机调试流程、言语康复过程等都有重要的指导作用。本文摘针对五类多重残疾人群，分别为视力障碍、智力障碍、脑性瘫痪、自闭症谱系障碍及唐氏综合征进行文献检索，探讨多重残疾对于人工耳蜗效果的影响，最终收录并翻译20篇近期的国内外相关文献。

对于这五类多重残疾植入者，均可通过人工耳蜗获得听声感知及言语发展，但是康复程度按照残疾类别及严重程度会有所差异。对于视力障碍患者，植入康复效果和无视力障碍的植入患者相比无显著差异，但康复需要更多技巧。对于智力障碍患者，患儿植入人工耳蜗后的听觉感知和言语发展逐渐改善，有机会发展口语交流技能。研究亦显示脑性瘫痪患者尤其是轻度或中度患儿，在人工耳蜗植入后效果可以正常同龄患者的水平。对于自闭症谱系障碍（ASD）患者，人工耳蜗可以带来好处，然而人工耳蜗的使用和调试程序都受ASD的严重程度影响，74%的自闭症人工耳蜗植入儿童为非口头交流者，与非ASD患儿相比听力结果明显较差^[3]。针对患有感音神经性耳聋的唐氏综合征（DS）患者，人工耳蜗效果与非DS患者相符，对于有严重认知障碍或其他额外需求的儿童来说，即使效果不如正常患者，依然可能提升其社会心理发展和生活质量。

欢迎各位阅读本文摘了解详细的文献内容，让我们一起为植入者带来更专业的康复，帮助更多患者重获听声！

1. Nelson, C. and S.M.J.E.S. Bruce, Children who are deaf/hard of hearing with disabilities: Paths to language and literacy. 2019. 9(2): p. 134.

2. Foundation, E., Prevalence of Additional Disabilities with Deafness: A Review of the Literature. 2012.

3. Mathew, R., et al., Cochlear Implantation in Children with Autism Spectrum Disorder: A Systematic Review and Pooled Analysis. 2022. 43(1): p. e1-e13.

目录

（一）视障与人工耳蜗的关系

Multichannel Cochlear Implantation in Visually Impaired Patients-视障患者的多通道人工耳蜗植入 1

Outcomes for Children with Deaf-Blindness with Cochlear Implants: A Multisite Observational Study-耳蜗植入治疗聋盲儿童的疗效:一项多地点观察研究 3

Cochlear Implantation in Children with Usher's Syndrome: A South Asian Experience- Usher综合征患儿的人工耳蜗植入: 南亚患儿经验 5

Outcomes of visually impaired patients who received cochlear implantations-视障患者的人工耳蜗植入效果 7

（二）智障与人工耳蜗的关系

The auditory and speech performance of children with intellectual disability after cochlear implantation-智力障碍儿童人工耳蜗植入后的听觉言语效果 9

Developmental delays assessed using the Enjoji Scale in children with cochlear implants who have intellectual disability with or without autism spectrum disorder-使用Enjoji量表评估伴随或不伴随自闭症谱系障碍的智力障碍人工耳蜗植入患儿的发育迟缓 11

Is There Any Association Between Language Acquisition and Cognitive Development in Cochlear-Implanted Children-人工耳蜗植入患儿的语言习得和认知发展的相关性 13

（三）脑瘫与人工耳蜗的关系

Cochlear implantation in children with cerebral palsy. A preliminary report-脑瘫儿童人工耳蜗植入的初步报告 15

Cochlear implantation in children with cerebral palsy-脑瘫儿童人工耳蜗植入 17

Performance after timely cochlear implantation in prelingually deaf children with cerebral palsy-语前聋脑瘫患儿适时植入人工耳蜗后的表现 19

Hearing rehabilitation in cerebral palsy: development of language and hearing after cochlear implantation-脑性瘫痪的听力康复:人工耳蜗植入后语言和听力发展 .. 21

Outcomes of cochlear implanted children with cerebral palsy: A holistic approach-脑性瘫痪患儿人工耳蜗植入的整体治疗效果 23

（四）自闭症与人工耳蜗的关系

Cochlear implant in patients with autistic spectrum disorder—a systematic review-自闭症谱系障碍患者的人工耳蜗植入——系统评价 25

Compliance with cochlear implantation in children subsequently diagnosed with autism spectrum disorder-人工耳蜗植入儿童随后诊断为自闭症谱系障碍的佩戴依从性 27

诺尔康文摘
NUROTRON DIGEST
2022年第1期
出版日期: 2022年3月1日

主办单位:
浙江诺尔康神经电子科技股份有限公司

编辑委员会:
诺尔康科学委员会和技术委员会

主编: 银力
编辑: 彭惠融
翻译: 苏珍、耿瓔映
校对: 彭惠融、章佳棋、叶影
排版: 耿瓔映

地址:
浙江省杭州市余杭区龙潭路17号
邮编: 311121
电话: 4006 222 571
传真: 0571-88179905
邮箱: service@nurotron.com
网址: <http://www.nurotron.com>

Cochlear implantation in children with Autism Spectrum Disorder (ASD): Outcomes and implant fitting characteristics-自闭症谱系障碍(ASD)儿童人工耳蜗植入:结果和人工耳蜗调试特征.....	29
Cochlear Implantation in Children with Autism Spectrum Disorder: A Systematic Review and Pooled Analysis-自闭症谱系障碍儿童人工耳蜗植入:系统回顾与综合分析	31
(五) 唐氏综合征与人工耳蜗的关系	
UK and Ireland experience of cochlear implants in children with Down Syndrome-英国和爱尔兰对唐氏综合征患儿进行人工耳蜗植入的经验.....	33
The management of children with Down syndrome and profound hearing loss-唐氏综合征伴极重度听力损失患儿的治疗.....	35
Cochlear implants in eight children with Down Syndrome - Auditory performance and challenges in assessment-八名唐氏综合征人工耳蜗儿童植入者——听觉表现和评估挑战.....	37
Long-term Outcomes in Down Syndrome Children After Cochlear Implantation: Particular Issues and Considerations-唐氏综合征儿童人工耳蜗植入后的长期结果: 特殊问题和注意事项	39

文献摘要

(一) 视障与人工耳蜗的关系

Multichannel Cochlear Implantation in Visually Impaired Patients

El-Kashlan, Hussam K.Boerst, AngeliqueTelian, Steven A.

Department of Otolaryngology-Head and Neck Surgery, University of Michigan, Ann Arbor, Michigan, U.S.A

Abstract

Objectives: To evaluate the outcome of cochlear implantation in patients with severe to profound hearing loss and visual impairment.

Study Design: Retrospective case review.

Setting: Tertiary referral center with a large cochlear implant program.

Patients: Six adults and two children with severe or profound hearing loss and significant visual impairment underwent multichannel cochlear implantation. Follow-up period ranged from 6 months to 9 years. Case history, etiology of visual and hearing loss, and benefit from cochlear implant were evaluated.

Interventions: Cochlear implantation and subsequent rehabilitation.

Main Outcome Measures: Speech perception measures were selected based on the patient age and cognitive abilities. Identical measures were used in each patient before and after implantation.

Results: As a group, patients did well after cochlear implantation. There was significant improvement in speech perception when compared with the score before implantation.

Conclusion: Cochlear implants can play a significant rehabilitative role in patients with severe visual and auditory impairment. Additional skills are required by the implant team for rehabilitation of patients with multiple sensory deficits.

Key Words: Cochlear implants; Blindness; Communication abilities

参考译文:

视障患者的多通道人工耳蜗植入

El-Kashlan, Hussam K.Boerst, AngeliqueTelian, Steven A.

美国密西根大学安阿伯耳鼻咽喉头颈外科

【摘要】

目的: 探讨人工耳蜗在重度或极重度听力和视觉障碍患者中的应用效果。

研究设计: 回顾性病例评论。

设置: 三级转诊中心, 有大型人工耳蜗植入项目。

患者: 患有重度或极重度听力损失和严重视觉障碍的6例成人和2例儿童接受了多通道人工耳蜗植入手术。随访时间6个月~9年。评估病史、视力和听力损失的病因及人工耳蜗植入的益处。

干预: 人工耳蜗植入及后续康复。

主要观察指标: 根据患者年龄和认知能力选择语音感知指标。每个患者在植入前后都采用相同的措施。

结果: 人工耳蜗植入术后, 患者整体表现良好。与植入前的评分相比, 言语识别有明显改善。

结论: 人工耳蜗可以在严重视听障碍患者中发挥重要的康复作用。在严重的视觉和听觉障碍的患者中, 人工耳蜗可以发挥重要的康复作用。植入团队需要更多的技能来帮助有多种感官障碍的患者康复。

【关键词】 人工耳蜗; 失明; 沟通

Outcomes for Children with Deaf-Blindness with Cochlear Implants: A Multisite Observational Study

Wiley, SusanMeinzen-Derr, JareenStremel-Thomas, KathleenSchalock, MarkBashinski, Susan M.Ruder, Charlotte

*Division of Developmental and Behavioral Pediatrics, Division of Pediatric Otolaryngology;

†Division of Biostatistics and Epidemiology, Division of Pediatric Otolaryngology, Cincinnati Children's Hospital Medical Center, University of Cincinnati, Cincinnati, Ohio;

‡The Teaching Research Institute, Western Oregon University, Monmouth, Oregon;

§Missouri Western State University, St. Joseph, Missouri; and Division of Speech Pathology, Cincinnati Children's Hospital Medical Center, University of Cincinnati, Cincinnati, Ohio, U.S.A.

Abstract

Introduction: Children with dual sensory impairments are receiving cochlear implants; however, little is known regarding their language outcomes.

Materials and Methods: Children between the ages of 6 months and 8 years with dual sensory impairment and cochlear implant(s) were recruited from across the United States to participate in an evaluation of language skills using the Reynell-Zinkin Developmental Scales, a tool validated on children with vision impairment and adapted for children with hearing loss. Basic demographic information was also collected from care givers.

Results: Ninety-one children completed assessments after implantation. For receptive language abilities, 32% of children obtained a level of sound detection, 15% obtained the ability to understand simple words, 21% could identify words, 5% could follow simple directions, and 22% could follow directions related to the functional use of objects. Four children had no response to sound after cochlear implantation. For expressive language abilities, 49% only had sound production skills, 9% could jargon, 18% could communicate with some words, 12% could communicate with simple sentences, and 12% could communicate with complex sentences. Children with lower developmental ages (or quotients) tended to obtain lower level expressive language skills such as sound production and jargon. Developmental abilities, rather than age at implant, were the most robust predictor associated with outcomes.

Discussion: This information can guide cochlear implant centers when discussing outcomes with families in the cochlear implant candidacy process. There is great heterogeneity in outcomes and caution should be used in discussing possible language outcomes for children with dual sensory impairments.

Key Words: Additional disabilities, Deaf-Blind, Pediatric cochlear implants

参考译文:

耳蜗植入治疗聋盲儿童的疗效: 一项多地点观察研究

Wiley, SusanMeinzen-Derr, JareenStremel-Thomas, KathleenSchalock, MarkBashinski, Susan M.Ruder, Charlotte

*儿童耳鼻喉科部门发育与行为儿科处;

†俄亥俄州辛辛那提市辛辛那提大学辛辛那提儿童医院医学中心儿童耳鼻喉科部门生物统计和流行病学处;

‡美国俄勒冈州蒙茅斯市西俄勒冈大学教学研究;

§美国密苏里州圣约瑟夫市密苏里西部州立大学; 美国俄亥俄州辛辛那提市辛辛那提大学辛辛那提儿童医院医学中心语音病理科

【摘要】

简介: 双感官障碍儿童在接受人工耳蜗植入; 然而, 关于他们的语言成果却知之甚少。

材料和方法: 从美国各地招募6个月到8岁之间患有双感官障碍和人工耳蜗植入的儿童, 使用Reynell-Zinkin发展量表对他们的语言能力进行评估。Reynell-Zinkin发展量表是一种对视力受损儿童进行验证并适用于听力损失儿童的工具。此外, 还收集了照顾者的基本信息。

结果: 91例患儿在植入后完成评估。在语言接受能力方面, 32%的儿童获得了一定程度的声音察觉能力, 15%的儿童明白简单词语, 21%的儿童能够识别单词, 5%的儿童能够遵循简单的指示, 22%的儿童能够遵循与物品功能使用相关的指示。4名儿童人工耳蜗植入后对声音无反应。在语言表达能力方面, 49%的人只具备发音能力, 9%的人会婴儿话, 18%的人会用一些单词交流, 12%的人会用简单的句子交流, 12%的人会用复杂的句子交流。发展年龄(或商)较低的儿童倾向于获得较低水平的语言表达能力, 如发声和婴儿话。发育能力, 而非植入时的年龄, 是最有力的预测结果。

讨论: 这些信息可以指导人工耳蜗中心在进行人工耳蜗术前检测过程中与家庭讨论效果。结果有很大的异质性, 在讨论双重感官障碍儿童可能的语言结果时应谨慎使用。

【关键词】 多重残疾, 失聪及失明, 儿童人工耳蜗植入

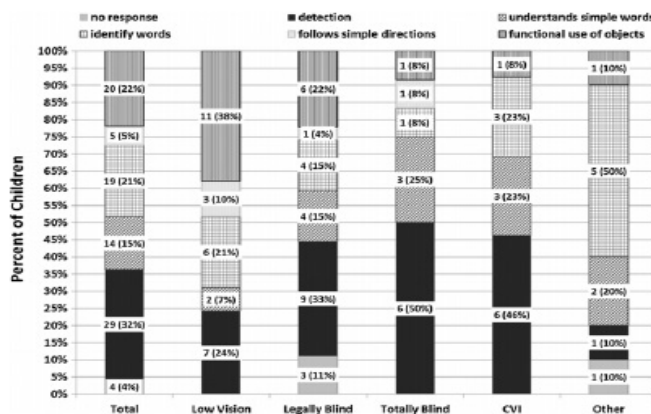


FIG. 1. Highest receptive language level on the Reynell-Zinkin scales achieved by type of vision loss. Number of children achieving each skill shown in the white boxes with percentages in parentheses. Percentages may add up to greater than 100 because of rounding. Categories of vision loss on the x-axis. Category of "Totally Blind" includes those with absolutely no vision and light perception only. CVI = cortical vision impairment.

Cochlear Implantation in Children with Usher's Syndrome: A South Asian Experience

Geetha Nair¹, Ruchima Dham², Arpana Sekhar², Raghunandhan Sampath Kumar², and Mohan Kameswaran²

Abstract

Usher's syndrome is an autosomal recessive disorder characterized by dual sensory impairment involving both the ears and eyes. Cochlear implantation paves a way to restore hearing loss in such individuals but poor vision among these patients poses additional challenges for the habilitationists. This study aimed to compare the habilitation outcomes and hearing-related quality of life scores of cochlear implantees having Usher syndrome with age-matched cochlear implantees with no such syndromic association. 27 patients aged 1–6 years with Usher syndrome underwent cochlear Implantation over a period of 10 years from 2006 to 2016 and were included in this study along with an age-matched cohort of 30 implantees with no additional disabilities. Category of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) scores were compared at 3, 6, 9 and 12 months respectively. Glasgow Benefit Inventory and Health Utility Index (HUI 3.0) questionnaires were used to assess the hearing-related quality of life in both groups at 1 year post implantation. There was significant difference in CAP and SIR scores between children with Usher's syndrome and the control group ($p < 0.05$). The overall scores in terms of quality of life as well were statistically different ($p < 0.05$). Though there was improvement in speech and language acquisition after cochlear Implantation this was found to be of lesser extent than the normative cohort. These children with additional visual disabilities required intensive, individualized therapy catering to their complex needs. Their family's perception of expected benefit from cochlear Implantation was guarded and needed to be appraised in detail prior to surgery. This experience helped prepare an institutional protocol for counseling such implantees in future.

Key Words: Usher syndrome, Cochlear implant (CI), Category of Auditory Performance (CAP), Speech Intelligibility Rating (SIR), Hearing Related Quality of Life (HR-QOL), Glasgow Benefit Inventory (GBI), Health Utility Index version 3 (HUI 3.0)

参考译文:

Usher综合征患儿的人工耳蜗植入：南亚患儿经验

Geetha Nair¹, Ruchima Dham², Arpana Sekhar², Raghunandhan Sampath Kumar², and Mohan Kameswaran²

【摘要】

Usher's综合征是一种常染色体隐性遗传病，其特征是听觉和视觉的双重感官障碍。人工耳蜗植入可为该类患者重建听力，但其视障给小儿康复专家带来了额外的挑战。本研究旨在比较患有Usher综合征的人工耳蜗植入者与年龄匹配的无Usher综合征的人工耳蜗植入者的康复效果和听力相关生命质量量表。从2006年到2016年的10年间，共有27例年龄在1-6岁的Usher综合征患者接受了人工耳蜗植入，并纳入30例年龄匹配的无其他残疾的人工耳蜗植入者队列。分表比较3个月、6个月、9个月和12个月的听觉行为表现(CAP)和言语可懂度(SIR)。采用格拉斯哥受益量表和健康效用指数量表(HUI 3.0)评估两组植入后1年的听力相关生命质量。Usher综合征患儿CAP和SIR评分与对照组有显著差异($p < 0.05$)。两组在生命质量方面的总分也有统计学差异($p < 0.05$)。尽管人工耳蜗植入后在言语和语言习得方面有改善，但其改善程度低于对照组群。这些有额外视觉障碍的患儿需要强化的、个性化的治疗来满足其复杂需求。需在术前指导患儿家人对人工耳蜗植入的期望值，并进行详细评估。这一经验帮助制定机构以后针对此类植入者的咨询辅导指引。

【关键词】 Usher综合征，人工耳蜗(CI)，听觉行为表现(CAP)，言语可懂度(SIR)，听力相关生命质量(HR-QOL)，格拉斯哥受益量表(GBI)，健康效用指数3版(HUI 3.0)

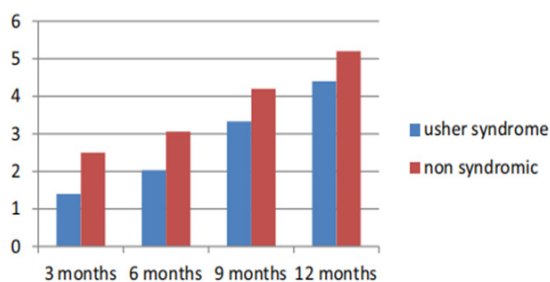


Fig. 1 Mean CAP score of ushers versus non-syndromic children

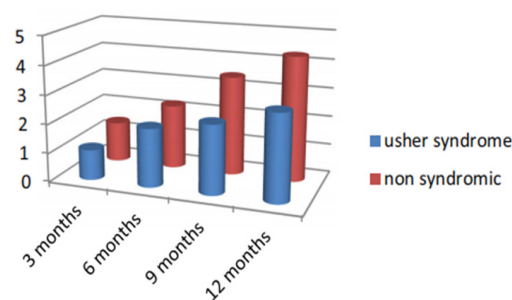


Fig. 2 Mean SIR scores of ushers versus non-syndromic children

Outcomes of visually impaired patients who received cochlear implantations

Kenichi Takano*, Aya Kaizaki, Etsuko Saikawa, Ayami Konno, Noriko Ogasawara, Tetsuo Himi

Department of Otolaryngology, Sapporo Medical University School of Medicine, Japan

Abstract

Objective: Patients with multiple sensory deficits, including hearing loss and visual impairment, present a unique problem. We evaluated the clinical outcomes of cochlear implantation in patients with severe to profound sensorineural hearing loss and visual impairment.

Methods: We retrospectively reviewed eight patients with severe sensorineural hearing loss and visual impairment who underwent cochlear implantation at our institution between 1993 and 2014. The follow-up period was between 2 and 20 years. We evaluated the case histories, etiologies of hearing loss and visual impairment, pre- and postoperative pure-tone thresholds, speech perception rates after CI using the Japanese CD speech discrimination scoring system (CI-2004 test) for words and sentences, and pre- and postoperative communication means. Postoperative speech discrimination scores were compared between patients with and without visual impairment who underwent cochlear implantation.

Results: The outcomes of cochlear implantation were good in all patients, with seven showing the ability to hold a conversation with others. The average proportion of correct answers for words and sentences in the CI-2004 test was $72.3 \pm 19.1\%$ and $86.0 \pm 16.1\%$, respectively, for the patients with visual impairment and $62.1 \pm 21.7\%$ and $78.5 \pm 20.9\%$, respectively, for those without visual impairment (based on auditory senses only). There were no significant differences in results between the patients with and without visual impairment.

Conclusion: Cochlear implantation is important for the rehabilitation of patients with severe auditory loss and visual impairment. Medical staff members require additional skills to perform auditory evaluations and rehabilitate patients with multiple sensory deficits.

参考译文：

视障患者的人工耳蜗植入效果

Kenichi Takano*, Aya Kaizaki, Etsuko Saikawa, Ayami Konno, Noriko Ogasawara, Tetsuo Himi

日本札幌医科大学医学院耳鼻喉科

【摘要】

目的：患有多重感官缺陷（包括听力损失和视力障碍）的患者存在独特的问题。评估重度极重度感音神经性听力损失和视觉障碍患者人工耳蜗植入的临床效果。

方法：回顾性分析1993年至2014年间在本机构接受人工耳蜗植入的8例重度感音神经性听力损失和视力障碍患者，随访时间为2~20年。使用日语CD语音识别评分系统（单词和短句）（CI-2004测试），以及术前和术后沟通方法，评估病史、听力损失和视觉障碍的病因、术前和术后的纯音阈值、CI后的言语感知率。比较人工耳蜗植入术后有视力障碍和无视力障碍患者的言语识别得分。

结果：所有患者的人工耳蜗植入效果均良好，其中7例患者表现出与他人对话的能力。在CI-2004测试中，有视力障碍的患者的单词和句子的平均正确率分别为 $72.3 \pm 19.1\%$ 和 $86.0 \pm 16.1\%$ ，无视力障碍患者（仅有听力障碍）分别为 $72.1 \pm 21.7\%$ 和 $78.5 \pm 20.9\%$ 。有视力障碍和无视力障碍的患者之间的结果无显著差异。

结论：人工耳蜗植入对重度听力障碍和视力障碍患者的康复具有重要意义。医务人员需要更多技能对多重感官障碍患者进行听觉评估和康复。

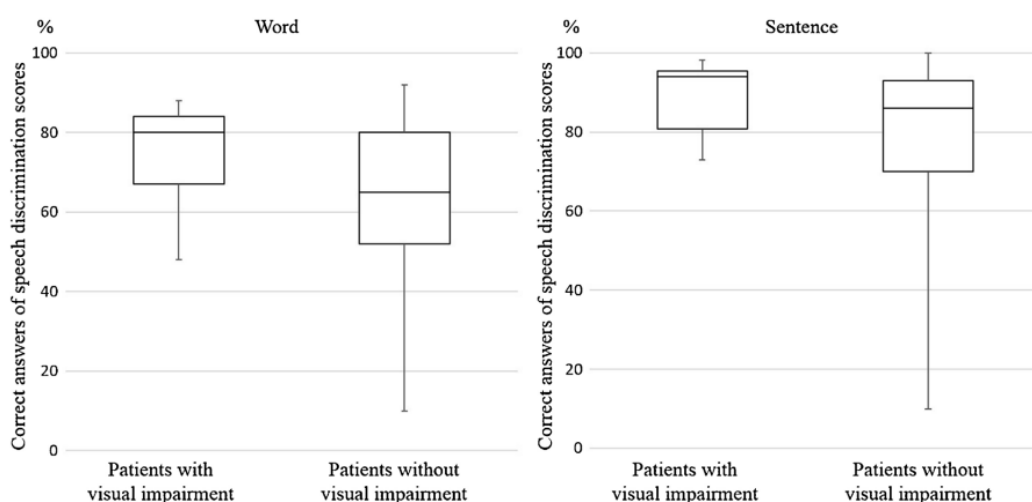


Fig. 2. Proportions of accurate answers (speech discrimination scores) for words (70 dB) and sentences (70 dB) in the Japanese CI-2004 test (based on auditory senses only) for patients with and without visual impairment. There is no significant difference in the proportions between the two groups.

(二) 智障与人工耳蜗的关系

The auditory and speech performance of children with intellectual disability after cochlear implantation

HYE-YOUN YOUM*, IL JOON MOON*, EUN YEON KIM, BO YOUNG KIM, YANG-SUN CHO, WON-HO CHUNG & SUNG HWA HONG

Department of Otorhinolaryngology-Head and Neck Surgery, Sungkyunkwan University School of Medicine, Samsung Medical Center, Seoul, Republic of Korea

Abstract

Objective: The purpose of this study was to assess the auditory and speech performance of 14 young deaf children with ID after CI.

Methods: Fourteen children with ID who underwent CI between December 2002 and February 2010 were included. Improvement in auditory perception and speech production over time was evaluated longitudinally with the Categories of Auditory Performance (CAP) score and Korean version of Ling's stages (K-Ling). The results were compared with those of age- and gender-matched implanted controls without additional disabilities. All tests were performed four times in each patient: before implantation and at 3, 6, and 12 months after implantation. Preoperative and postoperative communication modes were also assessed and compared between the two groups.

Results: Auditory perception and speech production of deaf children with an ID improved consistently after CI. In addition, the communication mode also took a favorable turn from nonverbal to vocalizations or oral communication or from vocalizations to oral communication.

Conclusions: The results revealed that children with intellectual disability (ID) who underwent cochlear implantation (CI) showed gradual progress in their auditory perception and speech development. ID in children should not be considered a contraindication for CI, because they are able to obtain a chance to develop oral communication skills following CI.

Key Words: Speech development, auditory perception, communication mode, language acquisition

参考译文:

智力障碍儿童人工耳蜗植入后的听觉言语效果

HYE-YOUN YOUM*, IL JOON MOON*, EUN YEON KIM, BO YOUNG KIM, YANG-SUN CHO, WON-HO CHUNG & SUNG HWA HONG

韩国成均馆大学医学院耳鼻咽喉头颈外科, 韩国首尔三星医疗中心

【摘要】

目的: 本研究旨在评估14例患有智力障碍 (ID) 的聋儿CI后的听觉言语效果。

方法: 纳入2002年12月至2010年2月期间接受CI的14例ID患儿。采用听觉行为表现(CAP)量表和韩语版Ling's stages (K-Ling)纵向评估听觉感知和言语产生的改善。与年龄和性别匹配并且无其他残疾的人工耳蜗植入者对照组进行比较。每例患者进行4次测试:植入前, 植入后3个月, 6个月和12个月。对两组患者术前、术后沟通方式进行评估和比较。

结果: 患有ID的聋儿的听觉感知和言语产出在CI后均持续改善。此外, 交流模式也发生了由非言语交流到发声交流或口语交流或由发声交流到口语交流的有利转变。

结论: 结果显示, 智障患儿植入人工耳蜗后的听觉感知和言语发展逐渐改善。ID患儿不应视为是CI的禁忌症, 因为在CI之后, 该类患儿有机会发展口语交流技能。

【关键词】言语发展, 听觉感知, 交流模式, 语言习得

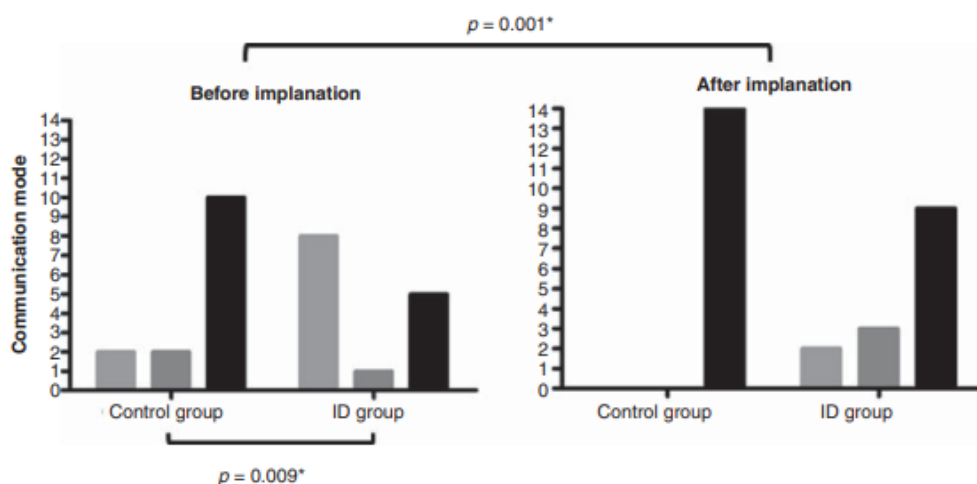


Figure 7. Communication mode before and after cochlear implantation (CI) in the intellectual disability (ID) and control groups. Communication mode in the two groups revealed a significant difference ($*p = 0.009$) and according to the time after CI ($*p = 0.001$). Light gray, non-verbal; deep gray, vocalization; black bar, oral communication.
*Statistical significance.

Developmental delays assessed using the Enjoji Scale in children with cochlear implants who have intellectual disability with or without autism spectrum disorder

Masaomi Motegi ^{ab}, Akira Inagaki ^{a*}, Toshiya Minakata ^a, Shinji Sekiya ^a, Mariko Takahashi ^a, Yoshimasa Sekiya ^c, Shingo Murakami ^a

^a Department of Otolaryngology, Nagoya City University Graduate School of Medical Sciences and Medical School, Aichi, Japan

^b Department of Otorhinolaryngology, Jikei University School of Medicine, Tokyo, Japan

^c Sekiya Otorhinolaryngology Clinic, Nagoya, Aichi, Japan

Abstract

Objective: Intellectual disability (ID) and autism spectrum disorder (ASD) are common among children who are candidates for cochlear implants. However, the implications of these comorbidities for cochlear implant placement have been not fully established. This study sought to identify these implications by comparing developmental delays among children with these conditions.

Methods: Participants were children who were followed up at least every 6 months for 24 months after cochlear implant surgery. Developmental delays were assessed using the Enjoji Scale of Infant Analytical Development (Enjoji Scale) and compared in three groups with hearing loss: those with ID (ID group, n = 4); those with ASD and ID (ASD + ID group, n = 4); and those with typical development (control group, n = 5). Developmental delay was evaluated longitudinally before and after cochlear implant placement for 18 months.

Results: Among the six subscales that make up the Enjoji Scale, language development and intelligence development were significantly delayed in all three groups and were exacerbated over time except for language development in the control group. Emotional development and social behavior were significantly delayed only in the ASD + ID group. Comparison of intergroup differences revealed delays in language development in the ID and ASD + ID groups compared with the control group.

Conclusions: The Enjoji Scale successfully demonstrated developmental delays characteristic to the underlying comorbidities of ID with or without ASD in children with cochlear implants. The Enjoji Scale can be a useful diagnostic tool for screening children with cochlear implants for ID with or without ASD.

参考译文:

使用Enjoji量表评估伴随或不伴随自闭症谱系障碍的智力障碍人工耳蜗植入患儿的发育迟缓

Masaomi Motegi ^{a,b}, Akira Inagaki ^{a*}, Toshiya Minakata ^a, Shinji Sekiya ^a, Mariko Takahashi ^a, Yoshimasa Sekiya ^c, Shingo Murakami ^a

^a日本爱知县名古屋城市大学医学研究生院耳鼻喉科

^b日本东京直系大学医学院耳鼻喉科

^c日本爱知县名古屋关谷耳鼻喉科诊所

【摘要】

目的: 智力障碍 (ID) 和自闭症谱系障碍 (ASD) 在适合植入人工耳蜗的患儿中很常见。然而, 这些合并症对人工耳蜗植入的影响尚未完全确定。本研究试图通过比较患有这些疾病的儿童的发育迟缓来确定这些影响。

方法: 受试者为人工耳蜗植入术后24个月的患儿, 至少每6个月进行一次随访。采用婴儿分析性发育Enjoji量表 (Enjoji量表) 评估发育迟缓, 并比较三组听力损失: ID组 (ID组, n = 4); ASD + ID组 (ASD + ID组, n = 4); 发育正常组 (对照组, n = 5)。纵向评估人工耳蜗植入前后18个月的发育迟缓。

结果: Enjoji量表的六个分量表中, 除对照组的语言发育外, 三组的语言发育和智力发育均显著迟缓, 并随着时间推移加剧。仅ASD + ID组的情绪发育和社会行为明显延迟。组间差异的比较显示, 与对照组相比, ID组和ASD + ID组的语言发育迟缓。

结论: Enjoji量表成功证明了伴随或不伴随自闭症谱系障碍的ID人工耳蜗植入患儿的发育迟缓。Enjoji量表可以作为一种有用的诊断工具, 用于筛查伴随或不伴随自闭症谱系障碍的ID人工耳蜗植入患儿。

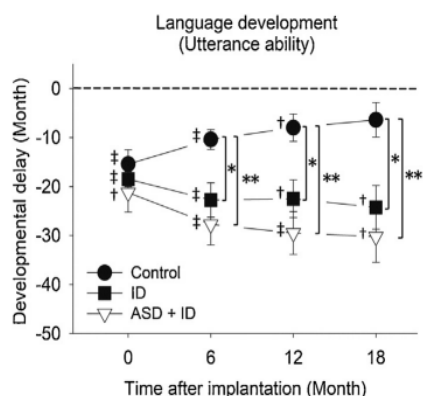


Fig. 1. Temporal changes in language development (utterance ability).

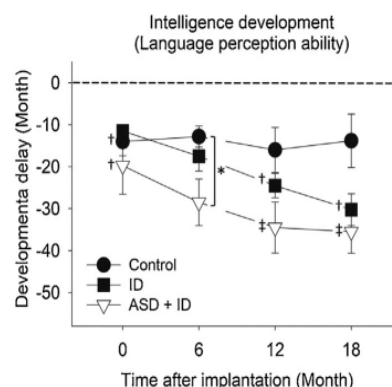


Fig. 2. Temporal changes in intelligence development (language perception ability).

There Any Association Between Language Acquisition and Cognitive Development in Cochlear-Implanted Children

Leila Monshizadeh¹, Roshanak Vameghi², Mehdi Rahimi³, Firoozeh Sajedi², Seyed Basir Hashemi¹, Fariba Yadegari⁴, Fatemeh Kasbi⁵

1.Otolaryngology Research Centre, Shiraz University of Medical Sciences, Shiraz- Iran

2.Pediatric Neurorehabilitation Research Centre, University of Social Welfare and Rehabilitation Sciences, Tehran- Iran

3.Department of Educational Psychology, Yazd University, Yazd- Iran

4.Department of Speech and Language, University of Social Welfare and Rehabilitation Sciences, Tehran, Iran

5.Neuromuscular Rehabilitation Research Centre, Rehabilitation College, Semnan University of Medical Sciences, Semnan, Iran

Abstract

Objective: Different studies on normal children and children with a sensory or intellectual disability indicate a strong correlation between the child's vocabulary domain and his cognitive abilities. Based on this, the main focus of the present study was to investigate the cognitive performance of cochlear-implanted children after a cognition-based language intervention program.

Methods: In this experimental study, 60 cochlear-implanted children were selected and randomly allocated into case and control groups. The control group received auditory verbal therapy (AVT), while the intervention group was trained by using both AVT and a language intervention protocol that was recently developed by the authors. Finally, the participants' communication abilities were assessed through the adapted version of the language subtest of Bayley Scales of Infant and Toddler Development – Third Edition (BSID 3). Five months later, the cognitive subtest was carried out. The data gathered were then analyzed using SPSS software.

Results: The study was performed on 2 groups of 20- to 24-month-old cochlear implant users, and our results confirmed a high correlation between language acquisition and cognitive development ($r = 0.76$). In addition, the cognitive and language performance of the participants who were trained by the new and specifically designed language intervention protocol as well as AVT was significantly higher than that of the control group ($P \leq .001$).

Conclusions: The new and specifically designed language intervention protocol that was mainly established based on cognitive factors such as attention and semantic memory enhancement in cochlear-implanted children improved not only their language acquisition but also their cognitive development.

Key Words: Cochlear implant, language intervention, cognitive development, vocabulary, children

参考译文:

人工耳蜗植入患儿的语言习得和认知发展的相关性

Leila Monshizadeh¹, Roshanak Vameghi², Mehdi Rahimi³, Firoozeh Sajedi², Seyyed Basir Hashemi¹, Fariba Yadegari⁴, Fatemeh Kasbi⁵

1. 伊朗设拉子医科大学耳鼻喉科学研究中心
2. 伊朗德黑兰社会福利和康复科学大学儿童神经康复研究中心
3. 伊朗Yazd大学教育心理学系
4. 伊朗德黑兰社会福利和康复科学大学语言学系
5. 伊朗塞姆南医科大学康复学院神经肌肉康复研究中心

【摘要】

目的: 对正常患儿和感官或智力障碍患儿的不同研究表明, 患儿的词汇领域与其认知能力之间存在很强的相关性。基于此, 本研究主要探讨人工耳蜗植入患儿接受认知语言干预后的认知表现。

方法: 本实验选取60例人工耳蜗植入患儿, 随机分为病例组和对照组。对照组接受听觉语言治疗(AVT), 而干预组同时使用AVT和作者最新开发的语言干预方案进行训练。最后采用Bayley婴幼儿发育量表第三版(BSID 3)语言子测试的改编版对被试者进行沟通能力评估。5个月后进行认知子测试。使用SPSS软件分析收集到的数据。

结果: 研究对象为2组20~24个月的人工耳蜗使用者, 结果证实语言习得与认知发展具有高度相关性($r = 0.76$)。此外, 接受新专门设计的语言干预方案和AVT训练的受试者的认知和语言表现显著高于对照组($P \leq .001$)。

结论: 新的专门设计的语言干预方案主要基于人工耳蜗植入患儿的注意力和语义记忆增强等认知因素而建立, 不仅提高了患儿的语言习得, 还提高其认知发展。

【关键词】 人工耳蜗, 语言干预, 认知发展, 词汇, 儿童

Table 5. The Correlation Between Language Acquisition and Cognitive Development

	Composite language	Composite cognitive
Composite language scores		
Pearson correlation	1	0.76**
Sig.		0.001
N	51	
Composite cognitive scores		
Pearson correlation	0.76**	1
Sig.	0.001	
N	51	51

**Correlation is significant at .01 (2-tailed)

(三) 脑瘫与人工耳蜗的关系

Cochlear implantation in children with cerebral palsy. A preliminary report

Andrea Bacciu^a, Enrico Pasanisi^a, Vincenzo Vincenti^a, Francesca Ormitti^b, Filippo Di Lella^a, Maurizio Guida^a, Mariateresa Berghenti^a, Salvatore Bacciu^a

^aDepartment of Otolaryngology, University of Parma, Via Gramsci, 14, 43100 Parma, Italy

^bDepartment of Neuroradiology, University of Parma, Parma, Italy

Abstract

Objectives: The aim of this study is to assess the post-implantation speech perception and intelligibility of speech produced by five profoundly deaf children with cerebral palsy.

Methods: This study is derived by a review of a prospectively maintained data collection on all patients entering the cochlear implant program. Five children with cerebral palsy who underwent cochlear implantation participated in this study. Functional outcome was assessed using the Speech Perception Categories and the Speech Intelligibility Rating scale. The follow-up of the series ranged from 12 to 45 months.

Results: At the last follow-up, two children who were placed into speech perception category 1 (detection of a speech signal) preoperatively progressed to category 6 (open-set word recognition with familiar words) postoperatively. Two children moved from preoperative category 2 (pattern perception) to postoperative category 6. One child placed into category 0 (no detection of speech) preoperatively progressed to category 4 (word identification) postoperatively. Before implantation, three children had connected speech unintelligible, and two subjects had connected speech intelligible to a listener who concentrates and lip-reads. At the last follow-up, one child had connected speech unintelligible, two children had connected speech intelligible to a listener who concentrate and lip-reads, one child had connected speech intelligible to a listener who has little experience of a deaf person's speech, and one child had connected speech intelligible to all listeners.

Conclusions: Cochlear implantation allowed these patients to dramatically improve their quality of life, increasing their self-confidence, independence and social integration.

Key Words: Cerebral palsy; Cochlear implant; Deaf; Children; Hearing loss; Outcome

参考译文:

脑瘫儿童人工耳蜗植入的初步报告

Andrea Bacciu^{a*}, Enrico Pasanisi^a, Vincenzo Vincenti^a, Francesca Ormitti^b, Filippo Di Lella^a, Maurizio Guida^a, Mariateresa Berghenti^a, Salvatore Bacciu^a

^a帕尔马大学耳鼻喉科, 格拉西, 1443100帕尔马, 意大利;

^b帕尔马大学神经放射学系, 意大利帕尔马

【摘要】

目的: 本研究旨在评估5例极重度听力损失及脑瘫儿童植入后的言语感知和言语可懂度。

方法: 本研究回顾所有参与人工耳蜗植入计划患者的前瞻性收集数据。共5例进行人工耳蜗植入的脑瘫儿童参与本研究。使用言语感知类别和言语可懂度评分量表评估功能结果。随访12~45个月。

结果: 在最后一次随访中, 两名患儿从术前言语感知类别1(察觉言语声), 术后发展至类别6(开放式熟悉词语识别)。两名患儿从术前类别2(声音规律感知)进展至术后类别6。一位儿童术前被列入类别0(没有言语察觉), 术后发展到类别4(词语辨别)。在植入前, 三位受试者连续性语言无法被理解, 两个受试者的连续性语言在专注及结合唇读情况下听者能理解。在最后的随访中, 一位儿童的连续性语言无法被理解, 两位儿童的连续性语言在专注及结合唇读情况下听者能理解, 一位儿童的连续性语言在听者缺少接触聋人口音经验下能听懂, 一位儿童的连续性语言可被所有听者听懂。

结论: 人工耳蜗植入可显著提高患者的生活质量, 增强自信心、独立性和社会融合能力。

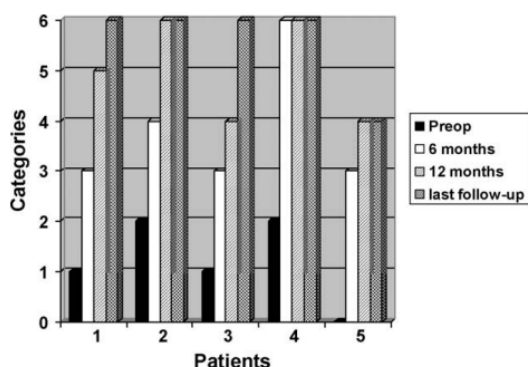


Fig. 2. Preimplantation (with hearing aids) and post-implantation speech perception categories (SPCs) according to Geer and Moog [14-16].

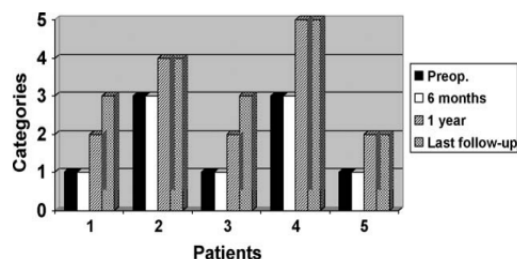


Fig. 3. Pre- and post-implantation individual Speech Intelligibility Rating scores according to Allen et al. [17] scale.

Cochlear implantation in children with cerebral palsy

Richard A. Steven^{a*}, Kevin M.J. Green^b, Stephen J. Broomfield^b, Lise A. Henderson^b, Richard T. Ramsden^b, Iain A. Bruce^b

^aManchester Auditory Implant Centre, University of Manchester, Oxford Road, Manchester M13 9PL, UK

^bManchester Auditory Implant Centre, University of Manchester, UK

Abstract

Objective: Few studies have looked at the outcomes of children with complex needs following cochlear implantation. Increasing evidence supports the case for implantation in these children. To date there is very little evidence available evaluating the role of cochlear implantation in children with cerebral palsy. In this paper we look at the Manchester Cochlear Implant Programme's experience of implantation in 36 children with cerebral palsy.

Method: A retrospective review of prospectively collected data for all children with cerebral palsy was undertaken. Cognitive and physical disability was scored by members of the cochlear implant team. A modified version of Geers and Moogs 1987 Speech Reception Score was used to assess outcome. Data was analysed looking at the relationship between cognitive and physical impairment, age at implantation and the SRS outcomes.

Results: This study demonstrated that children with cerebral palsy and a mild cognitive impairment do significantly better following implantation than those with a severe impairment ($p=0.008$). Children with mild physical impairment did not appear to do significantly better than those with moderate or severe impairments (mild versus severe $p=0.13$). Age at implantation was not a significant prognostic factor in this study group.

Conclusion: Children with complex needs are increasingly being referred for consideration of cochlear implantation. Further research is required to help guide candidacy, but each case must be considered individually. Higher functioning does appear to be the most important prognostic indicator regarding outcome but the effect of modest improvement in sound perception should not be underestimated.

Key Words: Cochlear implant; Cerebral palsy; Deaf; Complex needs; Hearing loss

参考译文:

脑瘫儿童人工耳蜗植入

Richard A. Steven^a, Kevin M.J. Green^b, Stephen J. Broomfield^b, Lise A. Henderson^b, Richard T. Ramsden^b, Iain A. Bruce^b

^a曼彻斯特大学曼彻斯特听觉植入中心, 牛津路, 曼彻斯特 M13 9PL, 英国

^b曼彻斯特听觉植入中心, 曼彻斯特大学, 英国

【摘要】

目的: 少量研究探讨特殊需求儿童人工耳蜗植入效果, 越来越多的证据支持在这类儿童身上进行植入。而到目前为止, 很少证据表明耳蜗植入在脑瘫患儿中的作用。本文介绍了曼彻斯特人工耳蜗植入计划在36例脑瘫患儿中的应用。

方法: 对所有脑瘫患儿的前瞻性资料进行回顾性分析。人工耳蜗植入组的成员对认知和身体残疾进行评分。采用改良版Geers和Moogs 1987言语感知评分 (SRS) 来评估结果。研究人员分析了认知和生理障碍、植入年龄和SRS结果之间的关系。

结果: 本研究表明, 轻度认知障碍的脑瘫患儿在植入后的表现明显好于重度认知障碍患儿($p=0.008$)。轻度身体障碍的儿童似乎并不比中度或重度障碍的儿童表现得更好(轻度与重度障碍 $=0.13$)。在这个研究组中, 植入术时的年龄并不是一个重要的预后因素。

结论: 越来越多特殊需求儿童被推荐考虑人工耳蜗植入。需要进一步的研究来帮助指导候选资格, 每个案例都必须单独考量。功能程度似乎是最重要的预后指标, 但声音感知的适度改善效果不应低估。

【关键词】 人工耳蜗; 脑瘫; 耳聋; 特殊需求; 听力损失

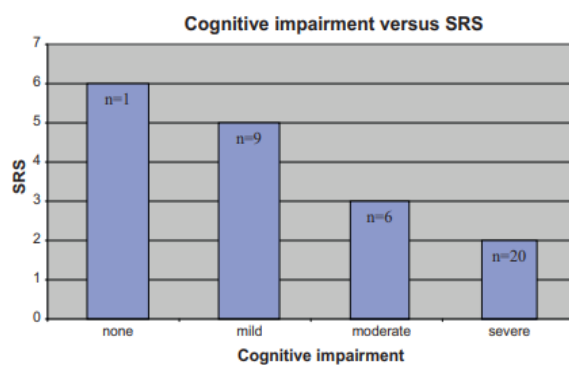


Fig. 1. Cognitive impairment alone versus Speech Reception Score (n = number in group).

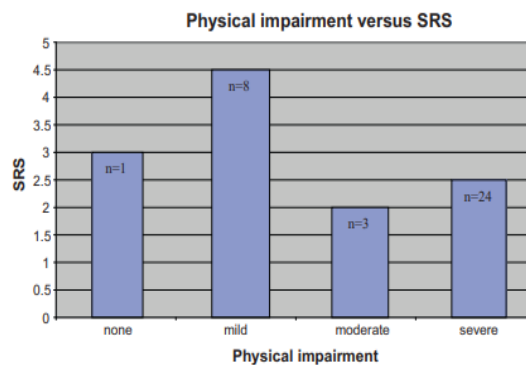


Fig. 2. Physical impairment alone versus Speech Reception Score (n = number in group).

Performance after timely cochlear implantation in prelingually deaf children with cerebral palsy

Hayoung Byun¹, Il Joon Moon¹, Eun Yeon Kim, Junoh Park, See Youn Kwon, Hyo Dam Han, Won-Ho Chung, Yang-Sun Cho, Sung Hwa Hong*

Department of Otorhinolaryngology-Head and Neck Surgery, Samsung Medical Center, Sungkyunkwan University, School of Medicine, Seoul, Republic of Korea

Abstract

Objective: To investigate auditory perception, speech production, and language ability of prelingually deaf toddlers with cerebral palsy (CP) who were implanted within a sensitive period and who received proper speech therapy. Comparison of their outcomes with age- and sex-matched CI recipients without additional disabilities was also performed.

Methods: We retrospectively reviewed a cohort of pediatric CI in Samsung Medical Center. Eight CP subjects who received CI before 3 years of age and age–sex matched control recipients who had no additional disabilities except idiopathic sensorineural hearing loss (SNHL) were included for the analysis. Preoperative evaluation included the Categories of Auditory Performance (CAP) score, Korean Version of the Ling’s Stage (K-Ling), Sequenced Language Scale for Infants (SELSI), Bailey Scales of Infant Development II assessment, Social Maturity Scale test, and grading of CP severity using severity level and Gross Motor Function Classification System for CP (GMFCS). To measure the outcome, the CAP scores, K-Ling, and SELSI were performed at 3, 6, 12, and 24 months after implantation.

Result: Four CP children with outstanding performances showed comparable achievement with matched control recipients. These patients had less severe motor disabilities (mild–moderate severity, GMFCS level 1–3), better social quotient, and better cognitive abilities. Although the others showed poor language abilities and hardly produced meaningful speech, their CAP scores reached 1 or 2 in 24 months after implantation.

Conclusion: Deaf children with CP could have various ranges of benefits up to the levels of normal peers whose only disability was hearing loss, when CI was performed within a critical period. Especially, children with mild or moderate CP had a favorable outcome after CI, equivalent to that of normal peers.

Key words: Cochlear implant; Cerebral palsy; Korean Version of the Ling’s Stage; Sequenced Language Scale for Infants; Bailey Scales of Infant Development; Social Maturity Scale

参考译文:

语前聋脑瘫患儿适时植入人工耳蜗后的表现

Hayoung Byun¹, Il Joon Moon¹, Eun Yeon Kim, Junoh Park, See Youn Kwon, Hyo Dam Han, Won-Ho Chung, Yang-Sun Cho, Sung Hwa Hong*

韩国首尔韩国成均馆大学医学院三星医院耳鼻咽喉头颈外科

【摘要】

目的: 探讨脑性瘫痪(CP)语前聋患儿在敏感期进行人工耳蜗植入并接受适当言语治疗后的听觉感知、言语产生和语言能力。与年龄和性别匹配无其他残疾的CI受试者的结果进行了比较。

方法: 我们对三星医疗中心的CI儿童组别进行回顾性分析。8名在3岁前接受CI的CP受试者和年龄性别匹配的对照组受试者被纳入分析, 这些受试者除自发性感音神经性听力损失(SNHL)外没有其他残疾。术前评估包括听觉行为表现(CAP)评分、韩文版林氏阶段(K-Ling)评分、婴儿语言发育筛查量表(SELSI)评分、贝利婴儿发展量表II评分、社会成熟度量表测试、使用严重程度等级和对于脑瘫患者的大肌肉运动功能分类系统(GMFCS)严重程度进行分级。为了测量结果, 分别在植入后3、6、12和24个月进行CAP、K-Ling和SELSI评分。

结果: 4例表现突出的CP患儿与对照组患儿的成绩相当。这些患者的运动障碍较轻(轻度至中度, GMFCS 1-3级), 社交商较好, 认知能力较强。虽然其他患者的语言能力较差, 几乎不能产生有意义的语言, 但在植入后的24个月内, 他们的CAP得分达到1或2分。

结论: 在关键时期进行CI时, 伴随CP的听障儿童可以获得不同程度的好处, 达到正常同龄仅有听力损失儿童的水平。特别是轻度或中度CP患儿在CI后的预后较好, 与正常同龄患儿相当。

【关键词】 人工耳蜗; 脑瘫; 韩文版林氏阶段; 婴儿语言发育筛查量表; 社会成熟度量表

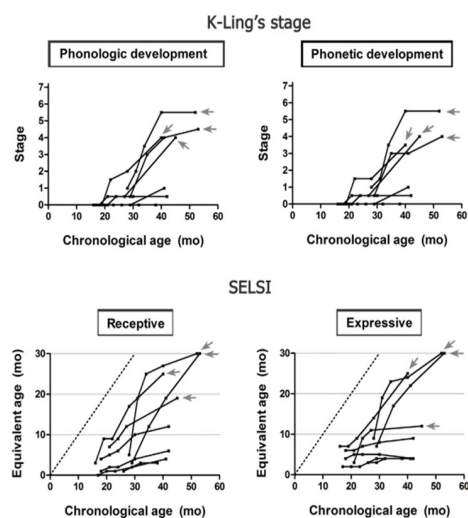


Fig. 1. Plots of performance results for individual subjects. Each connected-line means one patient. Evaluations were performed preoperatively and at 3, 6, 12, and 24 months after implantation. Note that four out of 8 patients showed outstanding speech production than the others according to the K-Ling's stage (arrows). Same patients are arrowed at SELSI plotting.

Hearing rehabilitation in cerebral palsy: development of language and hearing after cochlear implantation

Anacléia Melo da Silva Hilgenberg^a, Carolina Costa Cardoso^a, Fernanda Ferreira Caldas^a, Renata de Sousa Tschiedel^{ab}, Tatiana Medeiros Dep-eron^c, Fayez Bahmad Jr.^{a,*},

^aDepartment of Health Sciences, Universidade de Brasília (UnB), Brasília, DF, Brazil

^bDepartment of Psychology, Centro Universitário Planalto do Distrito Federal (UNIPLAN), Brasília, DF, Brazil

^cDivision of Audiology, University College of London (UCL), London, England, United Kingdom

Abstract

Introduction: Auditory rehabilitation in children with bilateral severe-to-profound sensorineural hearing loss with cochlear implant has been developed in recent decades; however, the rehabilitation of children with cerebral palsy still remains a challenge to otolaryngology and speech therapy professionals.

Objective: To verify the effectiveness of cochlear implants in the development of auditory and language skills in children with cerebral palsy.

Methods: A prospective analytical study. The evaluation of auditory responses to speech test was applied to the children in this study at regular intervals following implantation. Standardized tests that assess and quantify the development of auditory and language skills were administered and speech therapy video records and speech therapy files were analyzed. All children went through individually tailored intensive audiological rehabilitation programs following cochlear implantation.

Results: Two participants had gradual auditory and language development when compared to other participants who reached advanced levels in hearing and oral language classifications.

Key words: Cerebral palsy; Cochlear implants; Hearing loss

参考译文:

脑性瘫痪的听力康复:人工耳蜗植入后语言和听力发展

Anacléia Melo da Silva Hilgenberg^a, Carolina Costa Cardoso^a, Fernanda Ferreira Caldas^a, Renata de Sousa T schiedel^{a,b}, Tatiana Medeiros Dep-eron^c, Fayez Bahmad Jr .^{a,*},

^a巴西DF巴西利亚利亚大学健康科学系(UnB);

^b巴西DF巴西利亚联邦区普兰阿尔托大学中心 (UNIPLAN) 心理学系;

^c英国伦敦伦敦大学学院(UCL)听力学科

【摘要】

简介: 近几十年来,人工耳蜗对双侧重度至极重度感音神经性听力损失儿童的听觉康复已经取得了一定的进展;然而,儿童脑性瘫痪的康复对耳鼻喉科和言语治疗专业人员来说仍然是一个挑战。

目的: 验证人工耳蜗在脑性瘫痪患儿听力和语言能力发展中的作用。

方法: 前瞻性分析研究。本研究针对儿童研究对象进行言语测试,在植入后定期进行听觉反应评估。进行标准化测试,评估和量化听觉和语言技能的发展,并分析语音治疗视频记录和语音治疗文件。所有儿童在人工耳蜗植入后都接受了量身定制的强化听力康复治疗。

结果: 与听力和口语分类达到高级水平的其他参与者相比,两名参与者的听觉和语言发展缓慢。

【关键词】 脑瘫;耳蜗植入;听力损失

Table 4 Results of IT-MAIS and CDI tests and classifications of hearing and language of participant A.

Auditory age with CI	IT-MAIS	CDI (comprehension)	CDI (linguistic repertoire)	Classification of hearing	Classification of language
1 month	25%	-	-	1	1
6 months	32.5%	20 words	-	1	1
10 months	60%	25 words	13 words	2	1
14 months	62.5%	31 words	16 words	3	1
24 months	62.5%	44 words	20 words	3	1

IT-MAIS, Infant-Toddler Meaningful Auditory Integration Scale; CDI, MacArthur-Bates Communicative Development Inventory.

Outcomes of cochlear implanted children with cerebral palsy: A holistic approach

Maria Jaqueline Dias dos Santos^{a,*}, Dionísia Aparecida Cusin Lamoínica^a, Maria Valeriana Leme de Moura Ribeiro^b, Wendy McCracken^c, Leandra Tabanez do Nascimento Silva^d, Orozimbo Alves Costa^a

^a Department of Audiology and Speech Language Pathology, University of Sao Paulo, Al. Octavio Pinheiro Brisola, 9-75, Vila Universitaria, Bauru, Sao Paulo, Brazil

^b Department of Neurology, University of Campinas, R. Tessaíia Vieira de Camargo, 126, Cidade Universitaíria “Zeferino Vaz”, Campinas, Sao Paulo, Brazil

^c School of Psychological Sciences, University of Manchester, Ellen Wilkinson Building, Devas Street, Manchester, United Kingdom

^d Audiological Research Center, Hospital of Rehabilitation of Craniofacial Anomalies, University of Sao Paulo, Bauru, Sao Paulo, Brazil

Abstract

Objective: Analyze the progress of hearing and language in a group of children with cerebral palsy (CP) who have received cochlear implants (CI) and compare their progress in the clinical and functional domains.

Methods: This is a prospective transdisciplinary study developed within a tertiary referral center, with a group of nine cochlear-implanted children with CP, two- to seven-year-old. The assessments undertaken included audiological, language, and communication assessments complemented by the assessment of functional abilities and the level of independence as evaluated by the Pediatric Evaluation of Disability Inventory (PEDI) and Gross Motor Function Classification System (GMFCS).

Results: The outcomes varied, as two children achieved hearing comprehension in open-set evaluations. These children presented the same type of CP, athetosis, but with different functional skills and GMFCS levels. Only one of the subjects had any spoken language at the single-word level.

Conclusions: A holistic view of change and development is central to understanding progress made in children with CP who received cochlear implants (CI). The functional evaluation of these children with CP is a useful tool for monitoring their progress and measuring their outcomes with CI.

Key words: Cochlear implant; Cerebral palsy; Communication; Social function; Functional measures

参考译文:

脑性瘫痪患儿人工耳蜗植入的整体治疗效果

Maria Jaqueline Dias dos Santos^{a*}, Dioni'sia Aparecida Cusin Lamo'nica^a, Maria Valeriana Leme de Moura Ribeiro^b, Wendy McCracken^c, Leandra Tabanez do Nascimento Silva^d, Orozimbo Alves Costa^a

^a 巴西圣保罗巴烏魯圣保罗大学听力和言语语言病理学部门 Octavio Pinheiro Brisola, 9-75, Vila Universitaria, Bauru, Sao Paulo, Brazil

^b 巴西圣保罗坎皮纳斯坎皮纳斯大学神经学系 R. Tessa'lia Vieira de Camargo, 126, Cidade Universita'ria ' 'Zeferino Vaz' ', Campinas, Sao Paulo, Brazil

^c 英国曼彻斯特曼彻斯特大学心理科学学院

^d 巴西圣保罗巴烏魯圣保罗大学颅面异常康复医院听力研究中心

【摘要】

目的: 分析一组接受人工耳蜗(CI)治疗的脑瘫(CP)患儿的听力和语言功能进展, 比较其临床和功能方面的进展。

方法: 这是一项前瞻性跨学科研究, 在一个三级转诊中心进行, 研究对象是9名2 - 7岁的人工耳蜗植入性脑瘫患儿。所进行的评估包括听力、语言和沟通评估, 辅以儿童残疾评估量表(PEDI)和大肌肉运动功能分类系统(GMFCS)评估的功能能力和独立性水平。

结果: 结果各不相同, 两位儿童在开放式评价中能达到听声理解。这些患儿表现为相同类型的CP、手足徐动症, 但功能技能和GMFCS水平不同。其中只有一名受试者有单字水平的口语。

结论: 对CP患儿接受人工耳蜗植入(CI)后的变化和发展有一个整体的观点是理解进展的重心。对这些患有CP的儿童进行功能评估是监测其进展和测量其CI结果的有用工具。

【关键词】 人工耳蜗, 脑瘫, 沟通, 社会功能, 功能测量

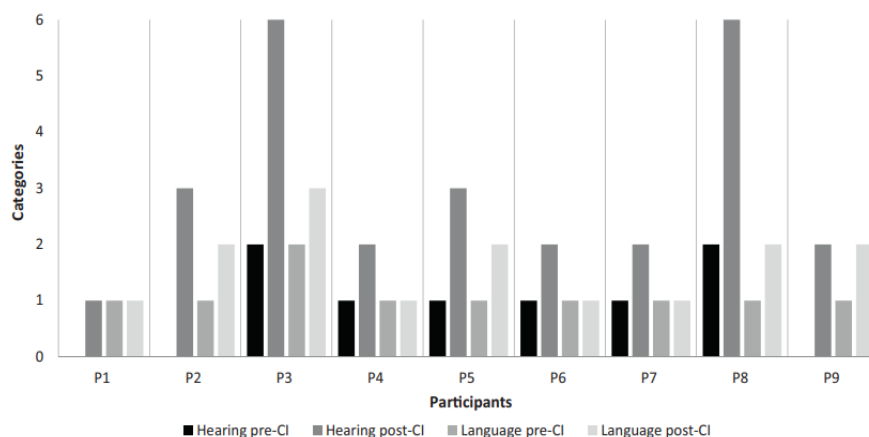


Fig. 1. Hearing and expressive language categories of each participant before and after CI.

(四) 自闭症与人工耳蜗的关系

Cochlear implant in patients with autistic spectrum disorder—a systematic review

Flávia da Silv^a, Tavares Yaná Jinkings Azevedo^a, Luísa da Matta Machado Fernandes^b, Alice Takeuti^a, Larissa Vilela Pereira^c, Aleluia Lima Lino Ledesma^a, Favez Bahmad Jr^c

^aUniversidade de Brasília (UnB), Programa de Pós-Graduação em Ciências da Saúde, Brasília, DF, Brazil

^bInstituto René Rachou – Fiocruz Minas, FESF-TECH Bahia e Grupo de pesquisa em Políticas de Saúde e Proteção Social, Belo Horizonte, MG, Brazil

^cUniversidade de Brasília (UnB), Faculdade de Ciências da Saúde, Programa de Pós-Graduação em Ciências da Saúde, Brasília, DF, Brazil

Abstract

Introduction: In cases of autism spectrum disorders with severe to profound hearing loss, cochlear implant is a therapeutic option.

Objective: To identify evidence in the scientific literature that the cochlear implant brings benefits to people with autism spectrum disorders with associated hearing loss.

Methods: Systematic review of the literature based on the criteria recommended by PRISMA. The population, intervention, comparison, outcomes, study design, PICOS strategy, was used to define the eligibility criteria. The studies that met the inclusion criteria for this second stage were included in a qualitative synthesis. Each type of study was analyzed according to the Joanna Briggs Institute's risk of bias assessment through the critical checklist for cohort studies, prevalence studies and critical criteria and case reports.

Results: Four hundred and eighty-four articles were found in eight databases and 100 in the gray literature, mentioning the relationship between cochlear implants in patients with autism spectrum disorder and hearing loss. Twelve articles were read in full and 7 were selected for qualitative analysis in this systematic review. All seven articles were analyzed on the critical evaluation checklist. Four articles had a low risk of bias and three articles had a moderate risk of bias. In this study, were included 66 patients with autism spectrum disorder and hearing loss who received cochlear implant..

Conclusions: This systematic review indicates that a cochlear implant can bring benefits to autism spectrum disorder patients with associated deafness.

Key Words: Autism spectrum disorder; Autistic disorder; Cochlear implant; Cochlear implantations; Systematic review auditory

参考译文:

自闭症谱系障碍患者的人工耳蜗植入——系统评价

Flávia da Silv^a, Tavaresa Yaná Jinkings Azevedo^a, Luísa da Matta Machado Fernandes^b, Alice Takeuti^a, Larissa Vilela Pereira^c, Alelluia Lima Loseno Ledesma^a, Favez Bahmad Jr^c

^a 巴西DF巴西利亚大学 (UnB), 卫生科学研究生课程

^b 巴西MG贝洛奥里藏特Ren é Rachou 研究所 - Fiocruz Minas, FESP-TECH Bahia 和卫生政策和社会保护研究小组

^c 巴西DF巴西利亚 (UnB)大学健康科学学院, 健康科学研究生课程

【摘要】

简介: 对于自闭症谱系障碍伴有重度至极重度听力损失的病例, 人工耳蜗是一种治疗选择。

目的: 在科学文献中找出证据, 证明人工耳蜗为伴有听力损失的自闭症谱系障碍患者带来好处。

方法: 根据PRISMA推荐的标准对文献进行系统回顾。使用人口、干预、比较、结果、研究设计、PICOS策略来定义资格标准。符合第二阶段纳入标准的研究被纳入定性综述中。根据Joanna Briggs研究所的偏倚风险评估, 通过队列研究、流行病学研究和关键标准以及病例报告的关键检查表, 对每种类型的研究进行分析。

结果: 在八个数据库中发现了48篇文章, 在灰色文献中发现了100篇文章, 提到了自闭症谱系障碍患者的人工耳蜗和听力损失之间的关系。在本系统综述中, 有12篇文章被全文阅读, 7篇被选作定性分析。所有七篇文章都在批判性评价核对表上进行了分析。四篇文章的偏倚风险较低, 三篇文章的偏倚风险为中度。在这项研究中, 纳入了66名接受人工耳蜗的自闭症谱系障碍和听力损失的患者。

结论: 该系统综述表明, 人工耳蜗可以为伴有耳聋的自闭症谱系障碍患者带来好处。

【关键词】 自闭症谱系障碍; 自闭症; 人工耳蜗; 人工耳蜗植入; 系统听觉回顾

Table 2 JBI critical appraisal checklist for cohort studies.

	Donaldson et al. ¹⁹	Eshraghi et al. ²¹	Mikic et al. ²²
Were the two groups similar and recruited from the same population?	Y	Y	Y
Were the exposures measured similarly to assign people to both exposed and unexposed groups?	U	Y	Y
Was the exposure measured in a valid and reliable way?	Y	Y	Y
Were confounding factors identified?	N	Y	Y
Were strategies to deal with confounding factors stated?	NA	Y	U
Were the groups/participants free of the outcome at the start of the study (or at the moment of exposure)?	U	Y	Y
Were the outcomes measured in a valid and reliable way?	Y	Y	Y
Was the follow-up time reported and sufficient to be long enough for outcomes to occur?	Y	Y	Y
Was follow up complete, and if not, were the reasons to loss to follow up described and explored?	Y	Y	Y
Were strategies to address incomplete follow up utilized?	U	Y	U
Was an appropriate statistical analysis used?	U	Y	Y
Bias risk (%)	50%	100%	81.81%

Y, yes; N, no; U, unclear; NA, not applicable.

Compliance with cochlear implantation in children subsequently diagnosed with autism spectrum disorder

Monica Rodriguez Valero¹, Mira Sadacharam², Lise Henderson¹, Simon R. Freeman¹, Simon Lloyd¹, Kevin M. Green¹, Iain A. Bruce^{1,2,3}

¹The Richard Ramsden Centre for Auditory Implants, Manchester Royal Infirmary, Central Manchester University Hospitals NHS Foundation Trust, Manchester Academic Health Science Centre, UK,

²Paediatric ENT Department, Royal Manchester Children's Hospital, Central Manchester University Hospitals NHS Foundation Trust, Manchester Academic Health Science Centre, UK,

³Respiratory and Allergy Centre, Institute of Inflammation and Repair, Faculty of Medical and Human Sciences, University of Manchester, UK

Abstract

Objectives: To assess the compliance with cochlear implantation (CI) in children subsequently diagnosed with autism spectrum disorder (ASD).

Methods: This was a retrospective case review and survey performed at a tertiary referral centre. Children meeting the criteria for CI who were implanted between 1989 and 2015 and who subsequently received a diagnosis of ASD were included. The primary outcome measure was to assess compliance with CI in children subsequently diagnosed with ASD. Secondary outcome measures included assessment of pre-CI risk factors that may have identified children at higher risk of a subsequent diagnosis of ASD, as well as the benefit obtained by these children following CI.

Results: 1050 children were implanted between 1989 and 2015. Of these, 22 children were diagnosed with ASD after receiving their CI. The average age at implantation was 2.6 years (median 3, range 1-8 years). The average age for diagnosis of ASD was 5 years, approximately 2 years (median 22 months, range 2-85 months) following CI. Of these, 16/22 (712.7%) regularly use their CI. 6/22 (27.2%) children became non-users of their implant. Some degree of verbal communication was used by 13/22 (59%) of our studied group.

Conclusions: There is a range of level of disabilities in ASD, with some relatively minor social communication difficulties through to severe language, cognitive, and behavioural difficulties. Compliance with CI is variable and appears to correlate with the severity of the ASD. Preoperative counselling should include information about the possible impact of later diagnosed disabilities such as ASD on performance.

Key Words: Cochlear implant, Autistic spectrum disorder, Asperger's, Compliance, Children, Hearing loss, Outcomes

参考译文:

人工耳蜗植入儿童随后诊断为自闭症谱系障碍的佩戴依从性

Monica Rodriguez Valero¹, Mira Sadadcharam², Lise Henderson¹, Simon R. Freeman¹, Simon Lloyd¹, Kevin M. Green¹, Iain A. Bruce^{1,2,3}

¹英国曼彻斯特皇家医院理查德·拉姆斯登听觉植入中心, 曼彻斯特大学中央医院NHS基金会信托, 曼彻斯特学术健康科学中心;

²英国皇家曼彻斯特儿童医院儿科耳鼻喉科, 曼彻斯特大学中央医院NHS基金会信托, 英国曼彻斯特学术健康科学中心,

³英国曼彻斯特大学医学和人文科学学院炎症和修复研究所呼吸和过敏中心

【摘要】

目的: 评价自闭症谱系障碍(ASD)患儿人工耳蜗植入的依从性。

方法: 这是一个在三级转诊中心进行的回顾性病例回顾和调查。研究纳入1989年至2015年期间符合CI标准进行植入, 随后被诊断为ASD的儿童。主要结果测量是评估后来诊断为自闭症谱系障碍的人工耳蜗儿童的依从性。次要结果测量包括评估CI术前风险因素, 这些因素可能识别后续有较高ASD风险的儿童, 以及这些儿童在CI后获得的好处。

结果: 1989年至2015年共有1050例儿童植入术。其中22名儿童在接受CI后被诊断为ASD。植入时的平均年龄为2.6岁(中位数3岁, 范围1-8岁)。ASD的平均诊断年龄为5年, 大约在植入后2年(中位数22个月, 范围2 - 85个月)。其中, 16/22(712.7%)常规使用CI。6/22(27.2%)儿童不再使用人工耳蜗。在我们的研究小组中, 有13/22(59%)的人使用了一定程度的语言交流。

结论: 自闭症谱系障碍存在多种程度的残疾, 从一些相对较轻微的社交困难到严重的语言、认知和行为障碍。对CI的依从性是可变的, 似乎与ASD的严重程度相关。术前咨询应包括后续诊断残疾的影响, 如ASD对植入效果的影响。

【关键词】 人工耳蜗, 自闭症谱系障碍, 阿斯伯格症, 依从性, 儿童, 听力损失, 效果

Table 2 Clinical characteristics of the patients with ASD

Patient	Gender	Age at implantation (months)	Autism spectrum (DSM-IV)	Co-morbidities	Disabilities	Years with implant	Usage
1	F	34	AD	—	—	20	No
2	F	20	AD	—	Learning disabilities	8	No
3	M	38	AD	Prematurity	Learning disabilities	16	Yes
4	M	20	AD	Meningitis	Learning disabilities	15	Yes
5	M	22	AD	AN/AD	—	4	Yes
6	M	45	AD	Prematurity	—	2	No
7	M	22	AD	—	Communication disabilities	2	Yes
8	M	44	AD	—	—	8	Yes
9	M	28	AD	—	—	7	Yes
10	M	60	AD	Meningitis, ADHD	Communication disabilities	3	Yes
11	M	12	AD	—	—	6	Yes
12	M	83	AD	Prematurity, ADHD	—	11	Yes
13	M	36	AD	Smith-Lemli-Opitz syndrome	Global developmental delay	6	(unilateral) Yes
14	M	37	AD	—	Communication disabilities	4	Yes
15	M	62	AD	ADHD	Learning disabilities	13	Yes
16	M	15	PDD-NOS	Waardenburg syndrome	Learning disabilities	17	No
17	M	22	PDD-NOS	—	—	7	No
18	M	54	PDD-NOS	—	Learning disabilities	11	No
19	M	32	PDD-NOS	Prematurity	Global developmental delay	6	Yes
20	M	98	Asperger's disorder	CMV	—	9	Yes
21	M	32	Asperger's disorder	Waardenburg syndrome, ADHD	—	7	Yes
22	M	34	AD	Prematurity	Learning and communication difficulties	1	Yes

Cochlear implantation in children with Autism Spectrum Disorder (ASD): Outcomes and implant fitting characteristics

Patrizia Mancinia, Laura Mariani^{a,*}, Maria Nicastrì^a, Sara Cavicchiolo^b, Ilaria Giallini^a, Pietro Scimemi^c, Diego Zanetti^d, Silvia Montino^e, Elisa Lovo^e, Federica Di Bernardino^{d,1}, Patrizia Trevisi^{e,1}, Rosamaria Santarelli^{e,1}

^aDepartment of Sense Organs, University Sapienza of Rome, Italy

^bAudiology Unit, Department of Clinical Sciences and Community Health, University of Milan, Italy

^cDepartment of Neuroscience, University of Padua, Padua - UOSD Otolaryngology and Audiology, Santi Giovanni e Paolo Hospital, Venice, Italy

^dDepartment of Specialistic Surgical Sciences, Foundation IRCCS Ca' Granda Maggiore Hospital, Milan, Italy

^eDepartment of Neuroscience, University of Padua, UOC Otolaryngology, Padua Hospital, Italy

Abstract

Background: Little is known regarding fitting parameters and receptive and expressive language development in cochlear-implanted children (CCI) with profound sensorineural hearing loss (SHL) who are diagnosed with Autism Spectrum Disorder (ASD). The aim of the study was to evaluate a group of ASD CCI users in order to describe their ASD clinical features and CCI outcomes; report on the average electrical charge requirements; and evaluate the possible correlations between electrical and psychophysical outcomes with ASD characteristics.

Materials and methods: A multicentre observational study of 22 ASD children implanted in four cochlear implant (CI) centers. Data concerning profound SHL diagnosis, ASD diagnosis, CI timing and CI compliance were collected. Sound Field (SF) was assessed through repeated behavioural measurements. Categories of Auditory Perception (CAP) and Categories of Language (CL) were used to evaluate speech perception and language skills at short (≤ 2 yrs), medium (5 yrs) and long term (>10 yrs) follow-up. Fitting parameters such as comfortable thresholds, pulse-width (pw, μsec) and clinical units converted into units of charge/phase were collected. The diagnosis of ASD was acquired by the referral neuropsychiatric department and severity was assessed through the Diagnostic and Statistical Manual of Mental Disorders (DSM-V) and the Childhood Autism Rating Scale (CARS).

Results: At the final follow-up session the median SF threshold for CI outcomes was 30 dB HL (min 15 – max 60). CAP score was extremely variable: 45.5% showed no improvement over time and only 22% of children reached CAP scores of 5–7. CL 45.5% showed no improvement over time and score was 1–2 in the majority of ASD children (72.7%), while only 18.2% reached the highest level of language skills. There were no statistically significant differences at each follow-up between subjects with or without comorbidities. CAP and CL were inversely correlated with DSM-V A and B domains, corresponding to lower speech and language scores in children with more severe ASD symptoms, and maintained their correlation at mid and long follow-ups whilst controlling for age at CI. Electrical charge requirements did not correlate with SF or age at implant but did inversely correlate with ASD severity. With regards to CI compliance: only 13.6% children (3) with severe DSM-V A/B levels and CARS score were partial/intermittent users.

Conclusion: The present study is a targeted contribution to the current literature to support clinical procedures for CI fitting and audiological follow-up in children with ASD. The findings indicate that the outcomes of CI use and the fitting procedures are both influenced by the severity of the ASD symptoms rather than the demographic variables or associated disorders.

Key Words : Cochlear implantation, Children, Autism spectrum disorders, Outcomes, Fitting

参考译文:

自闭症谱系障碍 (ASD) 儿童人工耳蜗植入: 结果和人工耳蜗调试特征

Patrizia Mancinia, Laura Mariani^{a*}, Maria Nicastrì^a, Sara Cavicchiolo^b, Ilaria Giallini^a, Pietro Scimemi^c, Diego Zanetti^d, Silvia Montino^e, Elisa Lovo^e, Federica Di Berardino^{d,1}, Patrizia Trevisi^{e,1}, Rosamaria Santarelli^{e,1}

^a意大利罗马萨皮恩扎大学感觉器官系

^b意大利米兰大学临床科学和社区卫生系听觉学组

^c意大利威尼斯帕多瓦大学神经科学系, 圣吉奥瓦尼圣保罗医院帕多瓦- uosd耳鼻喉科和听力学

^d意大利米兰马格奥雷医院基金会专业外科科学部

^e意大利帕多瓦大学神经科学系, 意大利帕多瓦医院耳鼻喉科

【摘要】

背景: 对于被诊断为自闭症谱系障碍(ASD)的极重度感音神经性听力损失(SHL)的人工耳蜗植入儿童(CCI)的调试参数和语言的理解与表达能力发展尚不清楚。本研究的目的是评估一组ASD CCI用户, 以描述他们的ASD临床特征和CCI效果; 报告平均电荷要求; 并评估自闭症谱系障碍的电性和心理物理结果之间可能的相关性。

材料与方法: 对22例ASD患儿在4个人工耳蜗植入中心进行多中心观察研究。收集深度SHL诊断、ASD诊断、CI植入时间及CI依从性等资料。通过重复的行为测量来评估声场(SF)。在短期(≤2年)、中期(5年)和长期(>10年)随访中, 用听觉表现类别(CAP)和语言类别(CL)评估言语知觉和语言技能。采集舒适阈值、脉宽(pw, μ sec)和转换为电荷/相位单位的临床单位等调试参数。ASD由转诊神经精神科确诊, 通过《精神障碍诊断与统计手册》(DSM-V)和《儿童自闭症量表》(CARS)评估其严重程度。

结果: 在最后一次随访中, CI结果的中位SF阈值为30 dB HL (最小15 - 最大60)。CAP得分变化很大: 45.5%的儿童随时间的推移没有改善, 只有22%的儿童CAP得分达到5-7分。CL得分中, 45.5%的自闭症谱系障碍儿童表现为随时间的推移没有改善, 大部分(72.7%)自闭症谱系障碍儿童的语言能力得分为1-2分, 只有18.2%的自闭症谱系障碍儿童达到了语言能力的最高水平。在每次随访中, 有或没有合并症的受试者之间没有统计学上的显著差异。CAP和CL与DSM-V A和B域呈负相关, 对应于较严重ASD症状儿童较低的言语和语言得分, 并在控制CI年龄的中长期随访中保持相关性。电荷需求与SF或植入时年龄无关, 但与ASD严重程度呈负相关。关于CI依从性: 只有13.6%(3)患有严重的DSM-V A/B水平和CARS评分的儿童是部分或间歇性使用CI。

结论: 本研究是对现有文献的有针对性的贡献, 以支持自闭症儿童CI调试和听力学随访。结果表明, CI的使用和调试程序都受ASD症状的严重程度影响, 而不是受人口统计学变量或相关疾病的影响。

【关键词】 人工耳蜗植入, 儿童, 自闭症谱系障碍, 结果, 调试

Cochlear Implantation in Children with Autism Spectrum Disorder: A Systematic Review and Pooled Analysis

Mathew, Rajeev*; Bryan, James*; Chaudhry, Daoud*; Chaudhry, Abdullah*; Kuhn, Isla**; Tysome, James*; Donnelly, Neil*; Axon, Patrick*; Bance, Manohar*,**

*University of Cambridge Foundation Trust, Addenbrooke's Hospital, Cambridge, UK

†University of Birmingham, Medical School, Birmingham, UK

**University of Cambridge, Cambridge, UK

Abstract

Objective: To determine outcomes following cochlear implantation (CI) in children with autism spectrum disorder (ASD).

Databases Reviewed: MEDLINE, Embase, Web of science, Cochrane Library, and Clinicaltrial.gov.

Methods: The review was performed according to the PRISMA statement. Primary outcomes measures were changes in speech perception and speech production scores. Secondary outcome measures included communication mode, device use, parental recommendation of implant, postoperative hyperacusis, and quality of life measures. Pooled analysis of outcomes was performed if possible.

Results: Twenty-four studies reported on 159 children with ASD. There were improvements in speech perception in 78% of cases and in speech expression in 63% of cases, though the extent of this improvement was variable. Seventy-four percent of children with ASD and CI are nonoral communicators. Intermittent/nonuse rate was 31%. Hearing outcomes are worse compared to children with other disabilities. The vast majority of parents would recommend CI based on their experiences.

Conclusion: Outcome in children with ASD and CI are highly variable and significantly poorer compared to non-ASD children. Despite this, most parents report positive experiences and the evidence supports the use of CI in children with ASD.

Key Words: Autism spectrum disorder; Children; Cochlear implants; Deafness; Developmental disability; Hearing loss; Speech; Systematic review.

参考译文:

自闭症谱系障碍儿童人工耳蜗植入:系统回顾与综合分析

Mathew, Rajeev*; Bryan, James*; Chaudhry, Daoud*; Chaudhry, Abdullah*; Kuhn, Isla⁺⁺; Tysome, James*; Donnelly, Neil*; Axon, Patrick*; Bance, Manohar*,⁺⁺

*英国剑桥大学基金会信托基金, 阿登布鲁克医院

⁺英国伯明翰大学医学院

⁺⁺英国剑桥大学

【摘要】

目的: 探讨自闭症谱系障碍(ASD)儿童人工耳蜗植入(CI)的疗效。

数据库回顾: MEDLINE, Embase, Web of science, Cochrane Library和Clinicaltrial.gov。

方法: 按照PRISMA标准进行审查。主要的结果测量是语音感知和语音产生得分的变化。次要结果测量包括沟通方式, 设备使用, 父母对人工耳蜗推荐程度, 术后听觉敏感, 生活质量测量。如果可能, 对结果进行汇总分析。

结果: 24项研究报告了159名ASD儿童。78%的患者的言语感知和63%的患者的言语表达均有改善, 尽管改善的程度各不相同。74%的自闭症人工耳蜗植入儿童是非口头交流者。间歇/不使用率为31%。与其他残疾儿童相比, 听力结果更差。绝大多数家长会根据自己的经验推荐CI。

结论: ASD和CI患儿的预后差异很大, 与非ASD患儿相比明显较差。尽管如此, 大多数家长报告了积极的经验和证据支持在自闭症儿童中使用CI。

【关键词】 自闭症谱系障碍; 儿童; 人工耳蜗; 耳聋; 发育性残疾; 听力丧失; 语言; 系统综述

TABLE 4. *Communication mode in children with ASD following CI*

Study and Year	Number	Oral	Oral/Sign/Behavior	Nonoral
Berritini et al. (2008)	2	0	0	2
Datta et al. (2019)	9	0	0	9
Donaldson et al. (2004)	7	0	1	6
Hayman et al. (2005)	1	0	0	1
Jennifer Robertson (2013)	10	1	3	6
Lachowska et al. (2016)	6	0	1	5
Meinzen Derr et al. (2014)	14	2	2	10
Messalam et al. (2018)	9	1	0	8
Nasralla et al. (2018)	4	0	1	3
Valero et al. (2016)	22	4	6	12
Waltzmann et al. (2000)	1	0	0	1
Yamazaki et al. (2012)	2	0	0	2
Zaidman Zait et al. (2018)	9	2	1	6
Total	96	10	15	71
Percentage	100	10.4	15.6	74.0

(五) 唐氏综合征与人工耳蜗的关系

UK and Ireland experience of cochlear implants in children with Down Syndrome

P.S. Hans, R. England¹, S. Prowse, E. Young, P.Z. Sheehan

ENT and Hearing Clinic for Children with Down Syndrome, Department of Paediatric Otorhinolaryngology, Royal Manchester Children's Hospital, Oxford Road, Manchester M13 9WL, United Kingdom

Abstract

Objective: Down Syndrome (DS) is associated with a high incidence of hearing loss. The majority of hearing loss is conductive, but between 4 and 20% is sensorineural, which in the main is mild or moderate and is managed with conventional behind-the-ear hearing aids. Cochlear implantation is an elective invasive procedure, performed to provide some form of hearing rehabilitation in individuals with severe to profound sensorineural hearing loss, and initially candidacy criteria were strict—excluding patients with additional disabilities. With good results and expanding experience, more candidates with additional disabilities are now being implanted. A survey of UK and Ireland Cochlear Implant Programmes sought to identify the number of individuals with DS who have been implanted with a cochlear implant (CI) and to provide relevant information on outcomes of implantation in these individuals.

Methods: E-mail survey of all programmes within the British Cochlear Implant Group (BCIG). Postal questionnaire to programmes identified to have implanted a child with Down Syndrome, with data collection on pre-operative assessment, surgical and post-operative outcomes. Case series review.

Results: Three of 23 BCIG programmes have implanted a child with Down Syndrome. Four children have received implants. No intraoperative or post-operative surgical complications were encountered. All children had middle ear disease, but no problems with implantation were associated with their middle ear condition. All children remain implant users, 12 months to 4 years post-implantation.

Conclusions: Cochlear implantation is an option for a child with Down Syndrome and associated severe to profound sensorineural hearing loss. Clinicians caring for these children and their families should consider referral for assessment by a Cochlear Implant Programme.

参考译文:

英国和爱尔兰对唐氏综合征患儿进行人工耳蜗植入的经验

P.S. Hans, R. England¹, S. Prowse, E. Young, P.Z. Sheehan

英国皇家曼彻斯特儿童医院儿科耳鼻喉科, 唐氏综合征儿童耳鼻喉科, 曼彻斯特牛津路M139WL

【摘要】

目标: 唐氏综合征(DS)与听力损失的高发病率有关。大多数听力损失是传导性的,但4%到20%是感音神经性的,主要是轻度或中度的,可以通过传统的耳后助听器进行干预。人工耳蜗植入是一种选择性侵入性手术,旨在为重度至极重度感音神经性听力损失的个体提供某种形式的听力康复,最初的候选标准很严格——排除有其他残疾的患者。凭借良好的结果和不断扩大的经验,更多有其他残疾的患者也陆续植入。一项针对英国和爱尔兰人工耳蜗计划的调查项目试图确定植入人工耳蜗(CI)的DS患者的数量,并提供有关这些人植入结果的相关信息。

方法: 对英国人工耳蜗集团(BCIG)内所有项目的电子邮件调查。对已植入的唐氏综合征儿童项目进行邮寄问卷调查,收集有关术前评估、手术和术后结果的数据,案例系列审查。

结果: 23个BCIG项目中有3个已植入唐氏综合征的儿童项目。四个儿童接受了植入,未发生术中或术后手术并发症。所有儿童都患有中耳疾病,但无中耳状况相关的植入问题。植入后12个月至4年,所有儿童仍为植入用户。

结论: 人工耳蜗植入是患有唐氏综合征并伴有重度至极重度感音神经性听力损失的儿童的一种选择。照顾这些儿童的临床医生及其家人应考虑转诊由人工耳蜗计划进行评估。

Table 3
Characteristics and outcomes for the four children implanted.

	Child			
	A	B	C	D
Age at implantation	46 months	44 months	25 months	39 months
Duration of implant use	12 months	20 months	36 months	50 months
Additional disabilities	Prematurity, cardiac anomaly requiring surgery	None reported	Neo-natal jaundice, gastro-intestinal abnormalities	None reported
Middle ear disease	Two previous ventilation tube insertions	Previous ventilation tube insertion	Middle ear effusions	Acute otitis media during assessment process
Pre-implantation non-language cognitive skills	Severely delayed	Severely delayed	Severely delayed	Delayed
Pre-implantation language	Total communication. Very limited sign language, no oral communication	Very limited sign language (Makaton), no oral communication	Total communication, no oral communication	Simple sign language, no oral communication
Imaging abnormalities	CT—middle and inner ear characteristic of DS	None	Absence CN VIII on R on MRI	None
Side implanted	R	L	L	R
Post-implantation audiological measurements	Consistent audiogram (Table 4)	First response to sound field audiometry at 20 months (Table 5)	Not able to measure sound field audiometry	Not able to measure sound field audiometry
CAP score	2	1	2-3	4-5
SIR score	1	1	1	1

The management of children with Down syndrome and profound hearing loss

E. Phelan¹, R. Pal¹, L. Henderson², K. M. J. Green², I. A. Bruce^{1,2,3}

¹Paediatric ENT Department, Royal Manchester Children's Hospital, Central Manchester University Hospitals NHS Foundation Trust, Manchester Academic Health Science Centre, UK,

²The Richard Ramsden Centre for Auditory Implants, Peter Mount Building, Manchester Royal Infirmary, UK,

³Respiratory and Allergy Centre, Institute of Inflammation and Repair, Faculty of Medical and Human Sciences, University of Manchester, UK

Abstract

Introduction: Although, the association between Down syndrome (DS) and conductive hearing loss is well recognized, the fact that a small proportion of these children may have a severe to profound sensorineural hearing loss that could benefit from cochlear implantation (CI) is less well understood. The management of significant co-morbidities in children with DS can delay initial diagnosis of hearing impairment and assessment of suitability for CI can likewise be challenging, due to difficulties conditioning to behavioural hearing tests.

Methods: We performed a retrospective case note review of three children with DS referred to the Manchester Cochlear Implant Programme.

Results: Three illustrative cases are described including CI in a 4 years old. Using conventional outcome measurement instruments, the outcome could be considered to be suboptimal with a Categories of Auditory Performance score of 4 at 6 months post-op and at last follow up. In part, this is likely to reflect the delay in implantation, but the role of cognitive impairment must be considered. The cases described emphasize the importance of comprehensive radiological and audiological assessment in children with DS being considered for CI.

Conclusion: The influence of cognitive impairment upon outcome of CI must be taken into account, but should not be considered a contra-indication to implantation in children with DS. Benefit that might be considered limited when quantified using existing general outcome measurement instruments, may have a significant impact upon psychosocial development and quality of life in children with significant cognitive impairment, or other additional needs.

Key Words: Cochlear implantation; Down syndrome; Sensorineural hearing loss

参考译文：

唐氏综合征伴极重度听力损失患儿的治疗

E. Phelan¹, R. Pal¹, L. Henderson², K. M. J. Green², I. A. Bruce^{1,2,3}

¹英国曼彻斯特皇家儿童医院儿科耳鼻喉科、曼彻斯特中央大学医院 NHS 基金会信托基金、曼彻斯特学术健康科学中心

²英国曼彻斯特皇家医院理查德拉姆斯登听觉植入中心、彼得·蒙特大楼

³英国曼彻斯特大学医学与人文科学学院炎症与修复研究所呼吸和过敏中心

【摘要】

简介：虽然唐氏综合征 (DS) 与传导性听力损失之间的关联已得到广泛认可。事实上，这些儿童中的小部分可能患有重度至极重度感音神经性听力损失，可以从人工耳蜗植入 (CI) 中收益，但仍未全面理解。由于难以对行为听力测试进行条件反射，患有 DS 的儿童的重重并发症的管理可能会延迟听力障碍的初步诊断，对评估 CI 的适用性同样具有挑战性。

方法：我们对曼彻斯特耳蜗植入计划中提到的三名 DS 儿童进行了回顾性病例报告审查。

结果：描述了三个说明性案例，包括一位 4 岁接受 CI 的儿童。使用常规的结果测量工具，在术后 6 个月和最后一次随访时，听觉性能分类评分为 4 分，可以认为结果是不理想的。在某种程度上，这可能反映了植入的延迟，但必须考虑认知障碍的作用。所描述的案例强调了对考虑进行 CI 的 DS 儿童进行综合放射学和听力学评估的重要性。

结论：必须考虑认知障碍对 CI 效果的影响，但不应将其视为 DS 儿童植入的禁忌症。使用现有的一般结果测量工具量化时可能认为益处有限，但对于有严重认知障碍或其他额外需求的儿童来说，可能对其社会心理发展和生活质量有重大影响。

【关键词】人工耳蜗；唐氏综合征；感音神经性听力损失

Table 1 Common ear abnormalities in children with Down syndrome

Ear	Abnormality	Effect
Outer ear	Stenotic EAC (Shott <i>et al.</i> , 2001)	Impacted cerumen CHL Difficult access for VT insertion
Middle ear	Eustachian tube dysfunction (Igarashi <i>et al.</i> , 1977)	HA mould fitting may be problematic
	OME (Brooks <i>et al.</i> , 1972; Balkany <i>et al.</i> , 1979a, 1979b)	Increased risk of persistent OME
	Ossicular chain abnormalities (Brooks <i>et al.</i> , 1972)	CHL – limits usefulness of OAEs in hearing threshold assessment
Inner ear	Dehiscence of facial nerve (Bilgin <i>et al.</i> , 1996; Harada and Sando, 1981)	CHL
	Abnormal facial nerve anatomy (Bilgin <i>et al.</i> , 1996; Harada and Sando, 1981)	Increased risk of inadvertent damage during ME surgery
	Hypoplastic mastoid (Blaser <i>et al.</i> , 2006)	Increased risk of inadvertent damage during ME surgery
	Absent cochlear nerve (Blaser <i>et al.</i> , 2006; Hans <i>et al.</i> , 2010)	Limited access for posterior tympanotomy in CI
	Hypoplastic nerve (Blaser <i>et al.</i> , 2006)	SNHL – CI not appropriate
	Short cochlear duct length (Bilgin <i>et al.</i> , 1996)	SNHL – May limit the outcome of CI
Dysplastic cochlea (Blaser <i>et al.</i> , 2006)		Must be considered when selecting the most appropriate CI electrode array length
		SNHL – Must be considered when selecting the most appropriate CI electrode array length
		SNHL – Must be considered when selecting the most appropriate CI electrode array length
Mondini defect (Bilgin <i>et al.</i> , 1996; Igarashi <i>et al.</i> , 1977)		Increased risk of balance problems. Increased risk of post-CI imbalance
Abnormal vestibule, semicircular canals, vestibular aqueduct (Blaser <i>et al.</i> , 2006)		

Conductive hearing loss (CHL); ventilation tubes (VT); otitis media with effusion (OME); middle ear (ME); sensorineural hearing loss (SNHL); cochlear implant (CI).

Cochlear implants in eight children with Down Syndrome – Auditory performance and challenges in assessment

Mariann Gjervik Heldahl, Beth Eksveen, Marie Bunne

Department of Otorhinolaryngology, Oslo University Hospital – Rikshospitalet, Oslo, Norway

Abstract

Objectives: A small proportion of children with Down Syndrome (DS) have severe to profound hearing loss and may potentially benefit from a cochlear implant (CI). Evidence on outcomes in DS is very limited, and there is a need for further investigation to provide a basis for clinical evaluation of candidates and outcomes. This study aims to explore outcomes of CI in children with DS in Norway.

Methods: Data on all children with DS and CI in Norway were collected from the CI registry and patients' records at the national pediatric CI center. Main outcome measures were: use of CI, Category of Auditory Performance (CAP), Speech Intelligibility Rate (SIR), and parents' and caregivers' views of the benefits of CI.

Results: Eight children with DS have received CI in Norway, all bilaterally. The outcomes varied greatly. All children used their CIs, and all of them responded to environmental sounds. Four children reached CAP score 5 after several years of use, (i.e., they understand phrases without lip reading). All children scored at least 2, (i.e., responds to speech sounds). One child reached a SIR score of 3, (i.e. connected speech is intelligible to experienced listeners). The rest of the children reached SIR scores of 1 or 1–2, (i.e., connected speech is unintelligible). Without exception, parents had an entirely positive attitude to their children using a CI. Co-morbidity and middle ear disease frequently delayed implantation.

Conclusion: Our experience with CI in children with DS is positive. However, CI cannot replace other types of communication for these children, and it is important to give parents realistic expectations prior to surgery. Outcomes might be considered limited when evaluated with instruments for testing auditory performance and speech intelligibility constructed for children without additional disabilities. We do not believe that such outcomes reflect the benefit in real life.

Key Words: Cochlear implant, Down syndrome, Outcome

参考译文:

八名唐氏综合征人工耳蜗儿童植入者——听觉表现和评估挑战

Mariann Gjervik Heldahl, Beth Eksveen, Marie Bunne

挪威奥斯陆大学医院耳鼻喉科

【摘要】

目标: 一小部分唐氏综合征 (DS) 儿童患有重度至极重度的听力损失, 可能会从人工耳蜗(CI) 中受益。DS 结果的证据非常有限, 需要进一步调查为候选者和临床评估提供基础。本研究旨在探讨挪威 DS 儿童 CI 的效果。

方法: 挪威所有 DS 和 CI 儿童的数据均来自 CI 登记处和国家儿科 CI 中心的患者记录。主要结果指标是: CI 的使用、听觉行为分级标准 (CAP)、言语可懂度分级标准 (SIR) 以及父母和照顾者对 CI 益处的看法。

结果: 挪威有8例DS患儿接受了CI治疗, 均为双侧, 结果差异很大。所有儿童都使用CI, 并且他们都对环境声音做出了反应。四个儿童在使用几年后CAP 分数达到5 (即不借助唇读, 即可理解常用的短句)。所有儿童得分至少为 2 (即对语言声能够做出反应)。一个儿童的 SIR 分数达到 3, (即有经验的听众可以理解连贯的言语)。其余儿童的 SIR 分数为 1 或 1-2, (即, 连贯的言语不易懂)。毫无例外, 父母对使用 CI 的孩子持完全积极的态度。合并症和中耳疾病经常导致延缓植入。

结论: 我们对 DS 儿童 CI 的经验是正面的。然而, CI 不能取代这些儿童的其他沟通方式, 在手术前给父母实际期望值非常重要。当使用为没有额外残疾的儿童构建的测试听觉表现和言语清晰度的工具进行评估时, 结果可能被认为是有限的。我们不认为这样的结果反映了现实生活中的好处。

【关键词】人工耳蜗, 唐氏综合征, 结果

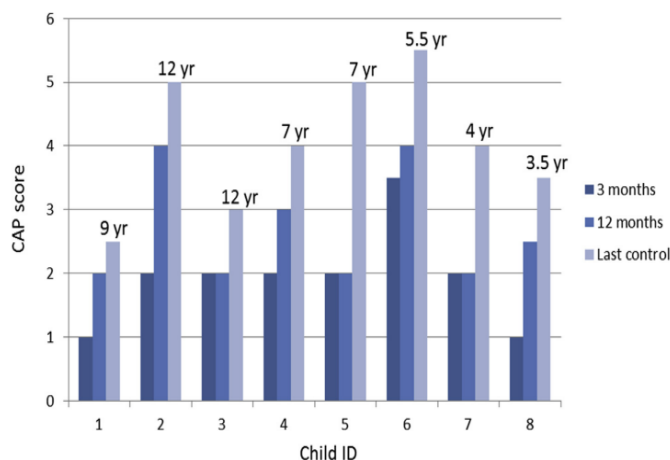


Fig. 1. Categories of Auditory Performance (CAP) scores at 3 months, 12 months and at last follow-up (number of years between switch-on of the first implant and last follow-up noted).

Long-term Outcomes in Down Syndrome Children After Cochlear Implantation: Particular Issues and Considerations

Clarós, Pedro^{*}; Remjasz, Agnieszka[‡]; Clarós-Pujol, Astrid^{*}; Pujol, Carmen^{*}; Clarós, Andrés^{*}; Wiatrow, Andrzej[§]

^{*}Clarós Clinic, Cochlear Implant Center

[†]Department of Otorhinolaryngology at Stefan Zeromski Specialist Hospital, Cracow

[‡]Scholarship in Clarós Clinic, Barcelona, Spain

[§]Institute of Psychology Polish Academy of Sciences, Warsaw, Poland

Abstract

Objectives: The aim of the study was to analyze the long-term outcomes after cochlear implantation in deaf children with Down syndrome (DS) regarding age at the first implantation and refer the results to preoperative radiological findings as well as postoperative auditory and speech performance. Additionally, the influence of the age at implantation and duration of CI use on postoperative hearing and language skills were closely analyzed in children with DS.

Study Design: Retrospective analysis.

Setting: Referral center (Cochlear Implant Center).

Materials and Methods: Nine children with Down syndrome were compared with 220 pediatric patients without additional mental disorders or genetic mutations. Patients were divided into four categories depending on the age of the first implantation: CAT1 (0–3 yr), CAT2 (4–5 yr), CAT3 (6–7 yr), and CAT4 (8–17 yr). The auditory performance was assessed with the meaningful auditory integration scales (MAIS) and categories of auditory performance (CAP) scales. The speech and language development were further evaluated with meaningful use of speech scale (MUSS) and speech intelligibility rating (SIR). The postoperative speech skills were analyzed and compared between the study group and the reference group by using nonparametric statistical tests. Anatomic abnormalities of the inner ear were examined using magnetic resonance imaging (MRI) and high-resolution computed tomography of the temporal bones (HRCT).

Results: The mean follow-up time was 14.9 years (range, 13.1–18.3 yr). Patients with DS received a multichannel implant at a mean age of 75.3 months (SD 27.9; ranging from 21 to 127 mo) and 220 non-syndromic children from reference group at a mean age of 51.4 months (SD 34.2; ranging from 9 to 167 mo). The intraoperative neural response was present in all cases. The auditory and speech performance improved in each DS child. The postoperative mean CAP and SIR scores were 4.4 (SD 0.8) and 3.2 (SD 0.6), respectively. The average of scores in MUSS and MAIS/IT-MAIS scales was 59.8% (SD 0.1) and 76.9% (SD 0.1), respectively. Gathered data indicates that children with DS implanted with CI at a younger age (<6 years of age) benefited from the CI more than children implanted later in life, similarly in a control group. There were additional anomalies of the temporal bone, external, middle, or inner ear observed in 90% of DS children, basing on MRI or HRCT.

Conclusion: The early cochlear implantation in children with DS is a similarly useful method in treating severe to profound sensorineural hearing loss (SNHL) as in non-syndromic patients, although the development of speech skills present differently. Due to a higher prevalence of ear and temporal bone malformations, detailed diagnostic imaging should be taken into account before the CI qualification. Better postoperative outcomes may be achieved through comprehensive care from parents/guardians and speech therapists thanks to intensive and systematic rehabilitation.

Key Words: Additional disabilities; Auditory performance; Cochlear implantation; Down syndrome; Outcomes cochlear implant; Speech perception; Speech recognition.

参考译文：

唐氏综合征儿童人工耳蜗植入后的长期结果：特殊问题和注意事项

Clarós, Pedro*； Remjasz, Agnieszka†； Clarós-Pujol, Astrid*； Pujol, Carmen*； Clarós, Andrés*； Wiatrow, Andrzej§

*克拉罗诊所，人工耳蜗中心

†克拉科夫斯特凡·泽罗姆斯基专科医院耳鼻喉科

‡西班牙巴塞罗那克拉罗诊所的奖学金

§波兰华沙波兰科学院心理研究所

【摘要】

目的：本研究的目的是分析唐氏综合征（DS）耳聋儿童首次植入人工耳蜗后的长期结果，包括首次植入术时的年龄，并将结果与术前放射学结果以及术后听觉和言语表现相参照。此外，还密切分析了植入年龄和CI使用时间对DS儿童术后听力和言语能力的影响。

学习规划：回顾性分析。

环境：转诊中心（人工耳蜗中心）。

材料和方法：研究人员将9名患有唐氏综合征的儿童与220名没有其他精神障碍或基因突变的儿科患者进行了比较。根据首次植入的年龄将患者分为四类：CAT1（0-3岁）、CAT2（4-5岁）、CAT3（6-7岁）和CAT4（8-17岁）。听觉表现通过有意义听觉整合量表（MAIS）和听觉行为分级标准（CAP）量表进行评估。言语和语言的发展是通过有意义言语使用量表（MUSS）和言语可懂度分级标准（SIR）进一步评估的。通过非参数统计测试，对研究组和参照组的术后言语能力进行了分析和比较。使用磁共振成像（MRI）和颞骨高分辨计算机断层扫描（HRCT）来检查内耳解剖异常。

结果：平均随访时间为14.9年（范围为13.1-18.3年）。DS患者接受多通道植入的平均年龄为75.3个月（SD 27.9；范围为21至127月），参照组的220名非综合征儿童的平均年龄为51.4个月（SD 34.2；范围为9至167月）。所有病例术中皆有神经反应。每个DS儿童的听觉和言语表现都有所改善。术后CAP和SIR的平均得分分别为4.4（SD 0.8）和3.2（SD 0.6）。MUSS和MAIS/IT-MAIS量表的平均得分分别为59.8%（SD 0.1）和76.9%（SD 0.1）。收集到的数据表明，在较小的年龄（<6岁）植入CI的DS儿童比晚期植入的儿童更受益于CI，与参照组类似。根据MRI或HRCT，90%的DS儿童有颞骨、外耳、中耳或内耳的其他异常情况。。

结论：在患有DS的儿童中，尽管言语技能的发展表现不同，早期的人工耳蜗植入在治疗重度至极重度感音神经性听力损失（SNHL）方面是一种有效方法，与非综合征患者类似。由于耳部和颞骨畸形的发病率较高，在进行CI诊断前应考虑到详细的影像诊断。通过父母/监护人和言语治疗师的全面护理，并通过强化和系统的康复治疗，可以取得更好的术后效果。

【关键词】其他残疾 听觉表现 人工耳蜗植入 唐氏综合征 人工耳蜗 言语感知 言语识别

杭州总部

地址：浙江省杭州市余杭区龙潭路17号

邮编：311121

电话：4006 222 571

传真：0571-88179905

邮箱：service@nurotron.com

网址：<http://www.nurotron.com>